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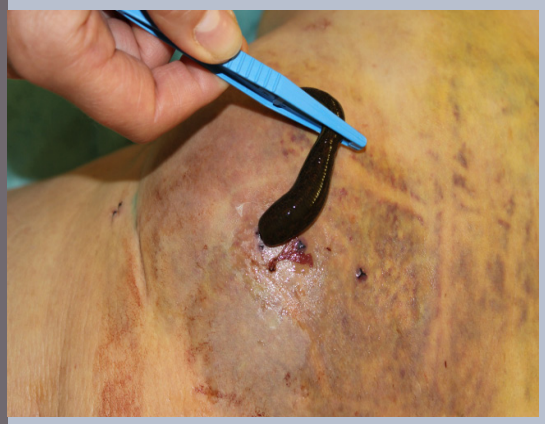

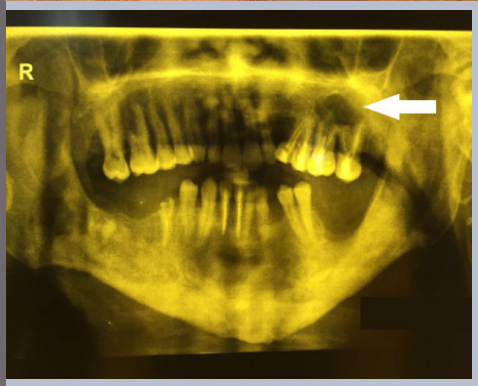

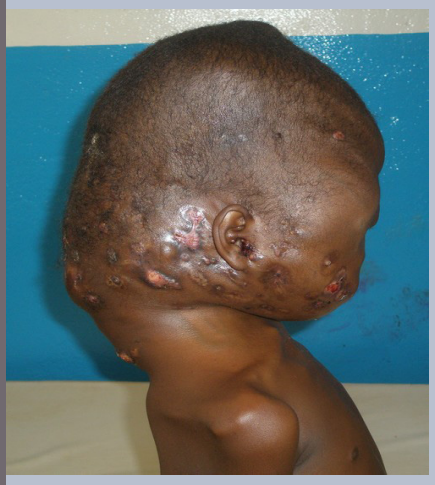

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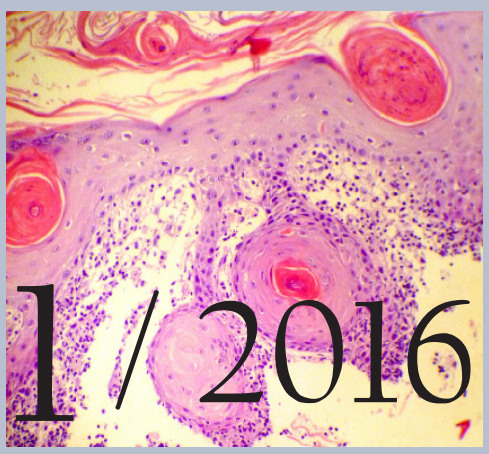

Editorial Pages

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Our Dermatology Online

Original Article

Impact of hand eczema severity on quality of life: a hospital based cross-sectional study

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ABSTRACT

Introduction: Hands are important organs of expression, communication, and are necessary for household and work-related activity. Thus, hand eczema can deteriorate quality of life. This study aims to find impact of hand eczema severity on quality of life. **Methods:** A cross-sectional study was done in a tertiary care hospital in Punjab from January to July, 2014. A total of 69 hand eczema patients of either gender aged ≥16 years were enrolled after taking an informed consent. Disease severity was assessed by hand eczema severity index (HECSI) score; and quality of life by dermatology life quality index (DLQI) questionnaire. The data was evaluated using statistical tests like frequency, chi-square, oneway ANOVA, t-test etc. **Results:** Out of 69 patients, 63.8% were males and 36.2% females. The commonest age group affected was 21-40 years (55.1% cases). Aggravating factors were reported by 76.8% patients, the commonest trigger being summer season (47.8%) followed by soaps and detergents (21.7%). The mean±S.D. for DLQI was 6.22±5.42 and for HECSI was 18.54±17.05. There was no statistically significant impact of age, occupation and duration of disease on DLQI or disease severity except gender (p-value being 0.028 for DLQI; 0.035 for HECSI). There was no significant correlation between HECSI score and DLQI. **Conclusion:** Majority of the patients with hand eczema had a significant impairment of their quality of life. There was a statistically significant impact of gender on hand eczema severity; although no correlation was found between DLQI and HECSI score in this study.

Key words: Hand eczema; Hand eczema severity index; Quality of life

INTRODUCTION

Hands are important organs of expression, communication, and are necessary for household and work-related activity. Hand eczema is a common and chronic dermatological condition. Though the exact prevalence of hand eczema is difficult to estimate as it is not a reportable disease and many patients even do not seek treatment. An estimated 2-10% of population is likely to develop hand eczema at some point of time during life. It appears to be the most common occupational skin disease, constituting up to 80% or more of all occupational contact dermatitides [1]. The disease has onset before 20 years of age in one-third of the patients [2]. Due to its high prevalence, early onset with chronic course and relation to occupation, it can have enormous socioeconomic consequences and a massive impact on patient's quality of life. For the assessment of its impact on quality of life (QoL), generic and dermatology-specific questionnaires can be used as disease-specific questionnaire is missing presently [3]. This study assessed QoL in hand eczema patients presenting in our dermatology outpatient clinic, related QoL to disease severity and morphological subgroup of hand eczema, and identifies various modifying factors influencing QoL.

MATERIALS AND METHODS

A cross-sectional study was done in the dermatology outpatient department at a tertiary care center in North India from January to July 2014.

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Inclusion Criteria

All patients of either gender and aged 16 years or more who presented with hand lesions suggestive of eczema were included in the study after informed consent.

Exclusion Criteria

- Patients less than 16 years of age.
- Patients whose skin scraping for fungus was positive on potassium hydroxide (KOH) mount.
- Patients who had palmar psoriasis (biopsy proved or with other psoriatic skin lesions and/or nail involvement).
- Patients who did not give consent to be part of the study.

Demographic profile, symptoms, details of occupation, duration, and aggravating factors were recorded. Examination included sites of involvement, morphology of the lesions. Morphological diagnosis was categorized as pompholyx, fissured hand eczema, hyperkeratotic hand eczema, nummular hand eczema, fingertip eczema and interdigital eczema.

Data on QoL was obtained from a self-administered questionnaire using the Dermatology Life Quality Index (DLQI). This is a dermatology-specific questionnaire which has been proven useful for assessment of QoL in hand eczema patients [4]. The DLQI is a 10-item questionnaire, which covers six aspects of daily life experienced during the past week: (i) symptoms and feelings, (ii) daily activities, (iii) leisure items, (iv) work and school, (v) personal relationship items, and (vi) treatment. The DLQI score is calculated by summing the score of each question, with a maximum score of 30 and a minimum score of 0. The higher the score, the greater the impairment of life.

Hongbo, *et al*. in his study looked at the relationship between DLQI and the patients' views of the overall impairment of their skin-related quality of life. He proposed the following classification of DLQI score: 0-1: No effect on patient's life, 2-5: Small effect on patient's life; 6-10: Moderate effect on patient's life; 11-20: Very large effect on patient's life; 21-30: Extremely large effect on patient's life. He stated that classifying DLQI will aid in the clinical interpretation of an individual's DLQI score, thus help in making clinical decisions [5]. The severity of hand eczema was assessed using a scoring system (Hand Eczema Severity Index, HECSI) [6]. It includes scoring of morphological signs such as erythema, infiltration, vesicles, fissures, scaling, and oedema as well as scoring of the affected area on the hands (fingertips, fingers, palms, back of hands, and wrists). The final score varies from 0-360.

Statistical Analysis

The data was evaluated using statistical tests like frequency, chi-square, oneway ANOVA, t-test etc.

RESULTS

A total of 69 patients were included in the study. Out of these, 63.8% (44/69) were males and 36.2% (25/69) were females with M:F ratio being 1.76:1. The commonest age group affected was 21-40 years (55.1% cases) followed by 41-60 years (23.2%), less than 20 years (17.4%) and >60 years (4.3%). The commonest age of onset of hand eczema was 21-40 years (49.3%) followed by less than 20 years of age (34.8%). However, males had an earlier onset of disease when compared to females (Table 1).

In our study, 30.4% were housekeepers followed by construction workers including masons, manual labourers (30.4%), farmers (13.0%), and others including doctor, engineer, student, driver (21.7%). About 23.2% (16/69) patients were illiterate while 34.8% (24/69) had passed primary school and 42.0% (29/69) had read up to secondary school or higher. Around 42% (29/69) patients had disease for more than 5 years; 36.2% cases had disease duration between 1-5 years and only 21.7% (15/69) had symptoms for less than 1 year. Aggravating factors were reported by 76.8% patients, the commonest trigger being summer season (47.8%) followed by soaps and detergents (21.7%), cement (11.6%), metals (10.1%) and cutting vegetables (7.2%). Out of total 5 patients who reported exacerbation of hand eczema with vegetables, all were housewives. Again, 80% of cases reporting worsening with soaps and detergents were housewives. Based on clinical findings, 44.9% patients were diagnosed with

Table 1: Age of onset of hand eczema

Age at onset of disease (years)	Males		Females		Total	
	N	% of all males	n	% of all females	n	% of all cases
≤20	18	40.9	6	24	24	34.8
21-40	20	45.5	14	56	34	49.3
41-60	5	11.4	5	20	10	14.5
>60	1	2.3	0	0	1	1.4
Total	44	100	25	100	69	100

pompholyx, 27.5% had fissured hand eczema, 23.2% had hyperkeratotic hand eczema, 2.9% had nummular eczema, and fingertip eczema was seen in 1.4% cases (Fig. 1).

The mean±S.D. for DLQI was 6.22±5.42 and for HECSI was 18.54±17.05. The median values for DLQI and HECSI were 4.00 and 12.00 respectively. Dermatology life quality index ranged from 1 to 20 while corresponding range for HECSI was 2 to 84. Statistically significant higher HECSI scores were found in males as compared to females, while no significant difference was found for DLQI values (Table 2).

Age, duration of disease, educational status, and occupation did not significantly affect the quality of life (*P* values = 0.261, 0.386, 0.698, 0.378) (Table 3). Disease severity did not show any significant correlation with the above parameters (*P* values = 0.597, 0.782, 0.645, 0.324) (Table 3).

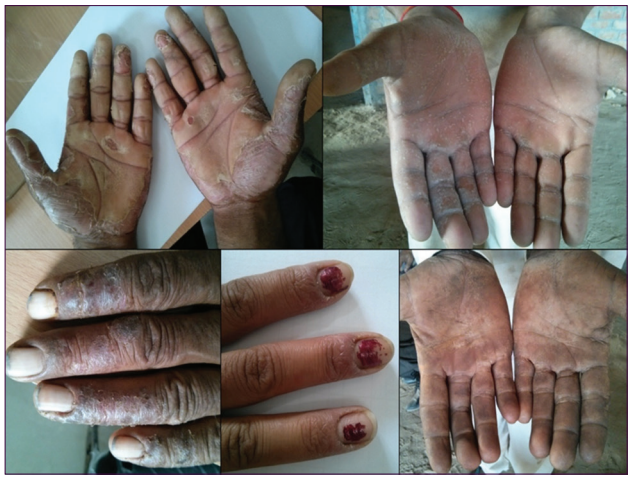

Figure 1: Various morphological patterns of hand eczema seen in our patients.

Table 2: Mean values for DLQI and HECSI for males and females

Scoring system	Total	Males	Females	*p*-value
DLQI	6.22	5.32	7.80	0.067
HECSI	18.54	21.77	12.84	0.035

Table 3: DLQI and HECSI in terms of gender, age, duration, education status and occupation

Variables	*p*-value	
	DLQI	HECSI
Gender	0.067	0.035
Age	0.261	0.597
Duration	0.386	0.782
Education	0.698	0.645
Occupation	0.378	0.324

There was no significant correlation between HECSI and DLQI in this study

DISCUSSION

Hand eczema is a common occupational dermatoses requiring dermatological care. Studies published in the past have shown significant negative impact of severity of hand eczema on the quality of life of an individual.

Men were almost twice as commonly affected than females which correlate well with increasing trend of hand eczema seen in males as stated in some of the recent studies [7,8]. According to literature, in one-third of patients, the disease occurs before the age of 20 years [9] which is quite similar to what was observed in our study. Also, it was seen in our study that males had earlier onset of disease than females. Higher prevalence and earlier age of onset in males may be attributed to the fact that men are employed in occupations such as construction, farming etc. where they get exposed to numerous allergens which may contribute enormously to hand eczema. Also such exposure starts at a younger age amongst males so cumulative exposure in men is higher than females.

The risk of hand eczema is occupation-related as well, being higher in industrial workers and masons due to exposure to various chemicals [10]. In our study, 34.8% were housekeepers, 30.4% were involved in construction work (including masons, construction site labourers) and 13% were farmers, thereby, predominantly affecting population that is regularly exposed to diverse types of chemical allergens.

The mean DLQI in the study was 6.22, underlining that hand eczema has a significant negative impact on the quality of life. This finding is similar to the observation in other similar studies [11,12].

Similar to the study by Agner *et* al, it was observed that although females had less severe hand eczema than males, QoL was equally affected [13]. This may stem out of greater cosmetic concern in females when compared to males. However in contrast to their study, age, occupation, and duration of disease did not significantly affect the quality of life or disease severity in our patients. There was no significant correlation between disease severity assessed by HECSI score and quality of life, however patients with even low HECSI score had a significant negative impact on their quality of life.

CONCLUSION

Majority of the patients with hand eczema had a significant impairment of their quality of life. There was a statistically significant impact of gender on hand eczema severity; although no correlation was found between DLQI and HECSI score in this study.

Statement of Human and Animal Rights

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Statement of Informed Consent

Informed consent was obtained from all patients for being included in the study.

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Our Dermatology Online

Original Article

A clinical study of the cutaneous manifestations of hyperthyroidism in Kashmir valley - India

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ABSTRACT

Introduction: Thyroid hormones are instrumental in regulating the health and appearance of skin and when the thyroid gland becomes underactive or overactive, a variety of skin problems result. These dermatologic manifestations may occur secondary to the abnormal thyroid hormone levels or due to the presence of thyroid autoantibodies that interact with skin components. **Aims:** The present study was designed to ascertain the varied cutaneous manifestations of hyperthyroidism. **Methods:** This was a hospital based cross sectional study conducted over a period of one year. A total of forty diagnosed cases of hyperthyroidism constituted the subject material for the study and were evaluated for the presence of any cutaneous manifestation. **Results:** In our study group of 40 patients, the predominant cutaneous symptom was increased sweating (80%), followed by heat intolerance (42.5%). The predominant cutaneous sign in hyperthyroid patients was increased skin temperature, noticed in 47.5% of patients. This was followed by soft, smooth and velvety skin (37.5%), palmar erythema (35%), fine thin hair (22.5%) and hyperpigmentation (10%). **Conclusions:** The interaction between thyroid gland and skin is very complex. So, dermatologists need to be cognizant of the ways in which these two organs interact.

Key words: Thyroid hormones; Cutaneous; Hyperthyroidism

INTRODUCTION

Endocrine disorders may occasionally present with cutaneous manifestations. Thyroid disorders have a high prevalence in medical practice and are associated with a wide range of diseases with which they may or may not share the etiological factors. One of the organs which best shows this wide range of clinical signs is the skin [1]. In thyroid diseases, many symptoms arise on the skin and most of these symptoms disappear with the treatment of thyroid disease.

Some dermatological skin findings and diseases may be the first symptoms of thyroid diseases. Since most of these cutaneous manifestations of thyroid disorders are nonspecific, these do not allow diagnosis without the estimation of endocrine function [2].

There is a very limited data available in literature regarding the cutaneous changes associated with hyperthyroidism. So, the present study was designed to ascertain the varied cutaneous manifestations of hyperthyroidism.

METHODS

This study was a hospital based cross-sectional clinical study conducted in collaboration with the endocrinology division of SMHS Hospital (associated teaching hospital of Government Medical College Srinagar). The present study was conducted over a period of six months from February 2010 to July 2010. A total of forty consecutively diagnosed cases of hyperthyroidism were included in the present study. There was no age limit for inclusion in the study. The

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diagnostic criteria for hyperthyroidism were:
a) Clinical manifestations of hyperthyroidism
b) Depressed or negligible TSH levels
c) Elevated serum T3 and T4 levels.

Diagnosis of hyperthyroidism was made by a suppressive TSH level in blood. Levels of T3 and T4 were also measured in blood and if one of both were elevated, the diagnosis was confirmed.

These patients were evaluated for the presence of any cutaneous manifestation. A detailed medical history pertaining to hyperthyroidism was elicited in each case with particular reference to the cutaneous complaints including duration, history of evolution and progression. An informed consent was taken from each patient, after which a general physical examination, systemic examination and a detailed dermatological examination was carried out and the relevant details recorded and tabulated. Apart from routine laboratory investigations, thyroid function tests (TSH, T3 and T4) were done by electro-chemiluminesense assay (ECLIA). Statistical analysis of the data was performed by appropriate statistical methods using Statistical Package for Social Sciences (SPSS Version 17) and inferences were drawn.

Ethics

This study was performed on human subjects; thus, all patients were aware of the presence of the study and they were fully informed about the drug and its side-effects.

RESULTS

A total of 40 hyperthyroid patients were included in the study. Age range, mean age, male/female ratio and type of hyperthyroidism is depicted in (Table 1). The medical complaints in the history in our study group are depicted in (Table 2). The most common cutaneous symptom was increased sweating, complained by a total of 32 (80%) patients. This was followed by heat intolerance, reported by 17 (42.5%) patients. Diffuse alopecia was complained by a total of 14 (35%) patients and hyperpigmentation by 6 (15%) patients. Fast nail growth and fine thin hair were reported as a cutaneous symptom by 5 (12.5%) patients each. Soft and friable nails were noticed by just 1 (2.5%) patient. Alopecia areata was reported by 2 (5%) patients. Most of these patients had more than one cutaneous symptom (Table 3). We noticed 5 (12.5%) hyperthyroid patients, who had no cutaneous symptom as such.

The predominant cutaneous sign in hyperthyroid patients was increased skin temperature, noticed in 19 (47.5%) patients. This was followed by soft, smooth velvety skin, which was seen in 15 (37.5%) patients. Palmar erythema (Fig. 1), was noticed as a cutaneous sign in 14 (35%) patients, while as hyperpigmentation in 4 (10%) hyperthyroid patients. The least common cutaneous sign was facial flushing, seen in only 2 (5%) patients. We did not observe pretibial myxedema in any patient. There were 9 (22.5%) hyperthyroid patients with no cutaneous signs on examination. The cutaneous signs in our patients are tabulated in (Table 4).

Table 1: Age range, mean age, male/female ratio and type of hyperthyroidism

Age range	Mean age	Male/ female ratio	Type of hyperthyroidism (%)
16 to 50 years	31.9±7.43 years	1:4	Graves disease: 31 patients (77.5) Toxic adenoma: 5 patients (12.5) Toxic multinodular goiter: 4 patients (10)

Table 2: Distribution of hyperthyroid patients as per clinical history (N=40)

Clinical history*	No. of patients	Percentage
Hyperactivity and irritability	5	12.50
Heat Intolerance and sweating	23	57.50
Palpitations	21	52.50
Weight loss	28	70.0
Increased appetite	12	30.0
Weakness and fatigue	13	32.5
Diarrhea	3	7.50
Insomnia and decreased concentration	3	7.50

*Some patients had more than one symptom in clinical history

Table 3: Cutaneous symptoms in hyperthyroid patients (N=40)

Cutaneous symptom*	No. of patients	Percentage
Heat intolerance	17	42.50
Increased sweating	32	80.0
Hyperpigmentation	6	15.0
Fast nail growth	5	12.50
Soft nails	1	2.50
Fine thin hair	5	12.50
Diffuse alopecia	14	35.0
Alopecia areata	2	5.0

*Some patients had more than one cutaneous symptom

Table 4: Cutaneous signs in hyperthyroid patients (N=40)

Cutaneous sign*	No. of patients	Percentage
Increased skin temperature	19	47.50
Palmar erythema	14	35.0
Facial flushing	2	5.0
Pretibial myxedema	0	0.0
Soft, smooth skin	15	37.5
Hyperpigmentation	4	10.0

*Some patients had more than one cutaneous sign

The predominant finding on examination of hair was fine thin hair with diffuse non-scarring alopecia (Fig. 2), seen in a total of 9 (22.5%) patients. Alopecia areata (Fig. 3) was noticed in just 2 (5%) patients. No hair changes were noticed in 29 (72.5%) of our hyperthyroid patients. Hair changes are depicted in (Table 5).

In our study group, the predominant nail change was fast nail growth, seen in 3 (7.5%) patients. Distal onycholysis (Plummer's nail) was not noticed in any of our patients. The other associated cutaneous diseases which we noticed in the patients in the hyperthyroid group were vitiligo, acne vulgaris, dermatitis herpetiformis, acropachy (Fig. 4) and ephelides. The associated cutaneous diseases are depicted in (Table 6).

Mean and median values of TSH, T3 and T4 in our patients are shown in (Table 7).

In our study group, majority of the patients were on antithyroid drugs, few of them were receiving radioactive iodine and 2 patients were scheduled for surgery for toxic thyroid nodules. Most of these patients were lost to follow up, so the effect of various treatment modalities over various cutaneous manifestations of hyperthyroidism could not be ascertained.

DISCUSSION

The skin in hyperthyroidism is warm, moist and smooth, bearing a resemblance to infantile skin. Warmth can be attributed to increased cutaneous blood flow and peripheral vasodilatation, which may also lead to the commonly noticed facial flushing and palmar erythema seen in hyperthyroid patients. Generalized hyperhidrosis may be noted with a predilection for palms and soles.

Table 5: Hair changes in hyperthyroid patients (N=40)

Hair changes	No. of patients	Percentage
Fine thin hair	9	22.5
Alopecia areata	2	5.0

Table 6: Associated cutaneous diseases in hyperthyroid patients (N=40)

Associated cutaneous disease	No. of patients
Vitiligo	1
Acne vulgaris	1
Dermatitis herpetiformis	1
Acropachy	1
Ephelides	1
Total	5

Table 7: Thyroid function status

Thyroid function status	Mean	Median
T3 (ng/ml)	2.89±1.10	2.77
T4 (µg/dl)	16.61±4.06	15.86
TSH (mIU/ml)	0.13±0.92	0.09

Thyroid function status

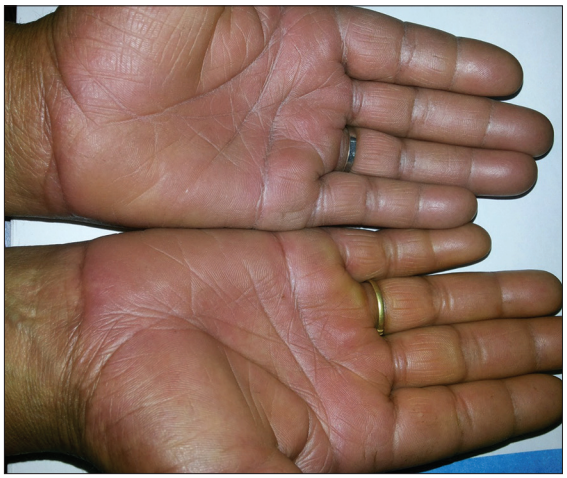

Figure 1: Palmar erythema in a patient of hyperthyroidism.

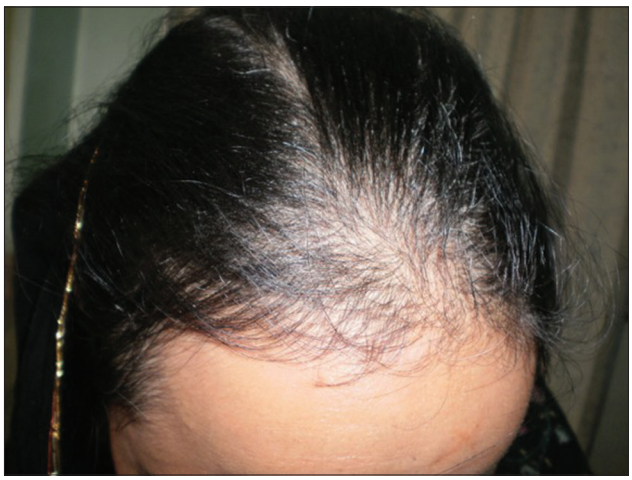

Figure 2: Fine thin hair in a hyperthyroid female.

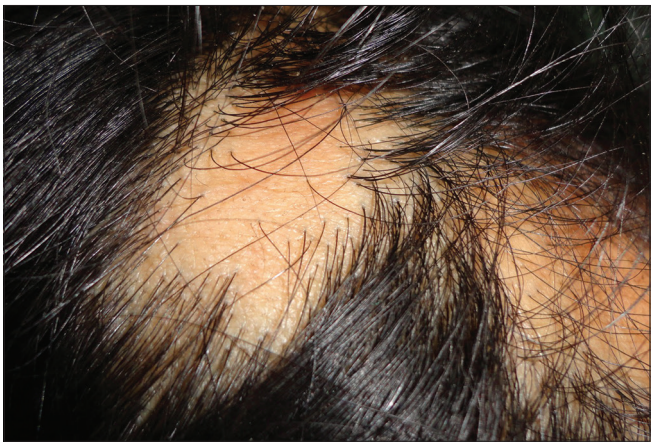

Figure 3: Alopecia areata in a female with hyperthyroidism.

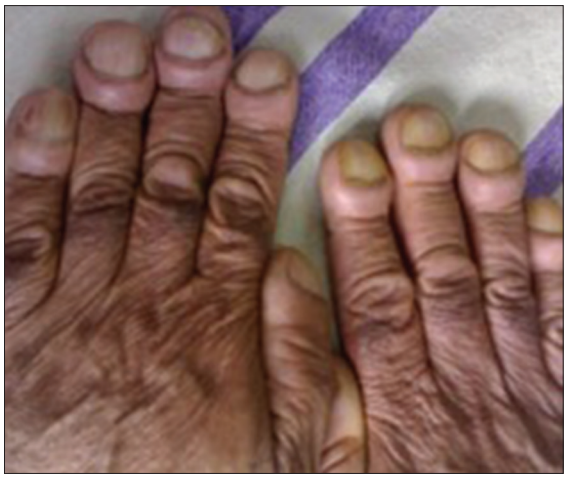

Figure 4: Acropachy in a male with hyperthyroidism.

Scalp hair is soft and fine and sometimes accompanied by diffuse, non-scarring alopecia [3]. Approximately 5% of patients may present with nail findings. Characteristic, though not pathognomonic, is the "Plummer's nail" with a concave contour and distal onycholysis. Hyperpigmentation may be seen in a distribution resembling that seen in Addison's disease, and is particularly pronounced in darker skin types [4]. Hyperpigmented eyelids have been described (Jellinek's sign).

Grave's disease is characterized by the cutaneous findings of hyperthyroidism in addition to distinctive cutaneous features including pretibial myxedema or dermopathy (0.5 to 4% of patients) and acropachy (1%). Clinical presentation may vary from a "peau d orange" appearance to the extensive infiltration resembling elephantiasis verrucosa nostra. Most often, lesions appear as bilateral asymmetric, raised, firm plaques or nodules varying in color from pink to purple-brown and sometimes accompanied by woody induration [5].

Grave's dermopathy occurs less frequently than ophthalmopathy, and although it is usually seen to occur with ophthalmopathy, it may occur alone [5]. The vast majority of patients with dermopathy have Grave's disease. Histologically, there is an accumulation of hyaluronic acid in the dermis more so than in the subcutis.

Thyroid acropachy is a triad consisting of distal clubbing, soft tissue swelling of hands and feet, and periosteal new bone formation. The first, second, and fifth metacarpals, the proximal phalanges of hands, and first metatarsal and proximal phalanges of feet are the most commonly affected. Pathognomic radiographic osseous changes are comprised of periosteal reaction of a lamellar type paralleling the diaphysis and has been described as "feathery". The vast majority of cases are associated with Grave's disease.

In our study group of 40 patients, there were 32 (80 %) females and just 8 (20 %) males. This was in concordance with the observations made by Rai D et al in their study, in which the percentage of females was found to be 83% [6]. This observation of female preponderance may be due to an increased association of autoimmune disorders in females, autoimmunity being an important cause of hyperthyroidism.

The most common cutaneous symptom in our hyperthyroid patients was increased sweating, seen in 80% of cases, followed by heat intolerance(42.5%). Comparable results were obtained by Rai D et al in their study. Hyperpigmentation was a cutaneous symptom in 15% of patients. Hair changes reported by our patients included fine thin hair (12.5%), diffuse alopecia (35%) and alopecia areata (5%). These findings were in contrast to the study by Rai D et al, in which hair changes were reported by 64% of cases [6]. Nail changes reported by our patients included fast nail growth (12.5%) and soft nails (2.5%).

We observed that the most common cutaneous sign in our hyperthyroid group of patients was increased skin temperature, seen in 47.5% of cases, followed by soft smooth skin (37.5%) and palmar erythema (35%). Ideally skin temperature can be assessed by either a wired skin electrode or a wireless skin temperature data logger or a more sophisticated thermal imaging system. Due to the non-availability of either of these in our institution, we could not record the skin temperature of the patients.

Hyperpigmentation was observed in 10% of hyperthyroid patients. None of our patients had pretibial myxedema, while as Leonhardt JM et al reported pretibial myxedema to be present in 0.5-4% of patients of Grave's disease [7]. In our study, we observed that the predominant finding on examination of hair in hyperthyroid patients was fine thin hair (22.5%) and alopecia areata, noticed in 5% of cases. In other study, they noticed hair changes in just 2.6% of their cases [8]. In our hyperthyroid group, we observed that the predominant nail change was fast nail growth, seen in 7.5% of our cases. Distal onycholysis (Plummer's nail) was not noticed in any of our patients whereas in some

studies, nail changes were noticed in 5% of the cases [9]. We noticed acropachy in 1 patient (2.5%), while as some studies have reported acropachy to be present in 0.1-1% of cases [7].

Associated cutaneous manifestations in our hyperthyroid patients included vitiligo, seen in 1 patient similar to other studies [10]. We found one patient of hyperthyroidism with dermatitis herpetiformis as an associated cutaneous diseases similar to that in other studies [11,12].

We also found 1 hyperthyroid patient each with acne vulgaris and ephelides. We believe that these findings in our patients were coincidental.

CONCLUSIONS

We conclude that there definitely exists an association between cutaneous signs and symptoms with hyperthyroidism but this interrelationship is complex. Advances in molecular and immunological studies have heightened our understanding of the pathogenesis of some of the aspects of these disorders, although innumerable questions remain.

Limitations

- Our sample size was not large
- The complaints of the patients were subjective
- There was no follow up of our patients in order to ascertain the effects of various antithyroid treatment modalities on the cutaneous manifestations of hyperthyroidism.

Suggestions

Cutaneous signs suggestive of hyperthyroid state should be followed up by routine thyroid function studies. Should the thyroid disease be classified as autoimmune, the clinician should be vigilant for any of the potential associated disorders for which the patient may be at risk throughout the patient's entire life. Optimal management of the cutaneous manifestations of hyperthyroidism relies on an understanding of their pathophysiology, early recognition and treatment.

Statement of Human and Animal Rights

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Statement of Informed Consent

Informed consent was obtained from all patients for being included in the study.

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Our Dermatology Online

Original Article

The impact of psoriasis on the lifequality: a cohort of 140 Moroccan patients

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ABSTRACT

Introduction: The alteration of the life quality in psoriasis is currently proved. **Aim:** To evaluate the particularity of this impact in Moroccan psoriatic population. **Methods:** It was a prospective cohort of 140 psoriatic patients who filled the 16-Skindex questionnary to evaluate this alteration of the life quality. **Results:** The life quality was significantly affected in patients having severe or old psoriasis and in young females, also it was related to the low Socioeconomic level and the living in rural areas, to the presence of psoriatic arthritis, to the scalp, the nails and mucosal involvement and to the use of systemic treatments. However, the emotional life, theimpact on daily activities and the sleep quality were not affected in our population. **Conclusions:** we had a low negative impact on the sleep quality and the emotional life which may be explained by the role of the family support in our society.

Keys words: Psoriasis; quality of life; Prospective cohort study

INTRODUCTION

The Psoriasis is an inflammatory, systemic and multifactorial skin disease affecting about 2% of the Moroccan population [1]. It is considered as an autoimmune disease with abnormality of mitosis and differentiation of keratinocytes where the cyclic nucleotide and lipid mediators play a key role [2,3].

This systemic disease is also known linked to a great number of comorbidities especially metabolic [4] and psychiatric [5] ones.

The alteration of the life quality of psoriatic patients has been shown in many epidemiological studies in adults and in children [6-14]. So the aim of our study was to evaluate this psycho-social impact of psoriasis in Moroccan psoriatic patients.

MATERIALS AND METHODS

It was a descriptive, analytical, unicentric and prospective study of 140 psoriatic patients treated in the Department of Dermatology ofthe Hospital Hassan II of Fez,during a period of 1 year: 2013/2014. Epidemiological andclinical data were collected in the psoriasis consultation by our doctors of the Department of Dermatology.

Psoriasis severity was calculated based on the Psoriasis Area Severity Index (PASI): Mild psoriasis: PASI <7, Moderate psoriasis: PASI between 8 and 12 and Severe psoriasis: PASI> 12.

we used the 16-Skindex as a questionnary for the evaluation of the impact of psoriasis on the life quality of psoriatic patients which include several items: pruritus, psychological impact: "No improving and recidivism, fear of worsening or persisting lesions or scars, frustration, shame, depression and anger ", relationships with others and the integrity in society, Emotional life, impact on daily activities and hobbies.

We estimated that a low impact on the life quality is a score of Skindex<10, a moderate impact on the

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life quality if the Skindexis between 10 and 50,and a significant impact on the life quality if the Skindex> 50.

Data analysis was performed using the SPSS 20 software, 2 kinds of analysis were carried out: Descriptive and univariate analysis.

In the univariate analysis: we analyzed the epidemiological and clinical data significantly related to each item of the Skindex.

RESULTS

We collected 140 patients with psoriasis for this study, the mean age was 30.5 years.

58.9 % of our patients were young (between 15 years and 45 years old) and we had a slight male predominance (51 %). Our patients had a moderate socioeconomic level in 55.5 % of cases and 50,4 % of them had a recent psoriasis.

Plaque psoriasis was the most frequent form (80,5%) and we had a scalp involvement in 74.1 % of cases. 51,8% of patients had a mild psoriasis while 31.4 % had a severe one.

Besides, severe itching was present in 31,9% of our patients and 26,8% of patients had moderate pruritus.

Furthermore, 50% of our patients had an important impact on the life quality, 35 % had a moderate impact and 15 % of patients had a low impact on the life quality.

Concerning the treatments used in our patients: 74.5 % of patients used topical treatments, 24.8 % used systemic treatments (30 patients: methotrexate, 2 patients: Infliximab (Remicade) for Psoriatic arthritis, 2 patients: Retinoids), and 26.6% of patients used phototherapy UVB TL01 (Table. I).

In the Univariate Analysis

The characteristics of psoriasis significantly related to the Pruritus in our population study were: the low SEL (p= 0,05), the Mild psoriasis (p=0,011) and the living in rural areas (p=0,05) while the No improving and recidivism was significantly associated with Young age (p=0,000), Female gender (p=0,000), Psoriatic arthritis (p= 0,004) and Psoriasis of the scalp,the nails and mucosal psoriasis (p=0,000).

Table 1: Descriptive analysis

Number of patients=140	N (%)
Demographic data	
Age (years)	
≤15	4 (2,9)
15-45	82 (58,9)
> 45	52 (37,4)
Gender	
Male	71 (51)
Female	67 (48,2)
Socio economiclevel	
Low	51 (37,2)
Moderate	76 (55,5)
High	10 (7,3)
Cultural level	
None	17 (12,4)
Primary	33 (24,1)
Secondary	52 (38)
Academic	35 (25,5)
Origin	
Rural	42 (30,7)
Urban	95 (69,3)
Duration of the disease (years)	
<5	69 (50,4)
5-10	36 (26,3)
>10	32 (23,4)
Clinical Data	
Type of psoriasis	
Plaque	112 (80,5)
Guttate psoriasis	12 (8,6)
Psoriaticarthritis	5 (3,6)
Pustular psoriasis	3 (2,1)
Erythrodermic psoriasis	6 (4,3)
Particular locations of psoriasis	
Scalp	103 (74,1)
mucosal psoriasis	7 (5)
Inverse psoriasis	1 (0,7)
Nail psoriasis	55 (39,5)
Palmoplantar psoriasis	13 (9,3)
Body surface area (BSA) (%)	
<10	71 (51,8)
Between 10 and 30	23 (16,8)
Between 30 and 50	29 (21,2)
BSA >50	14 (10,2)
PASI	
Mild psoriasis	87 (68,5)
Moderate psoriasis	16 (12,6)
Severe psoriasis	24 (18,9)
The impact on the life quality	
Pruritus	
Intense	44 (31,9)
Moderate	37 (26,8)
Psychological impact	
No improving and recidivism	65 (46,7)
Fear of worsening or persisting lesions or scars	73 (53,3)
Frustration	41 (41,3)
Shame	51 (37,2)
Depression	59 (42,8)
Anger	64 (46,4)
Emotionnal life	23 (16,7)
Integrity in the society	
Relationships with others	30 (21,7)

Cond..

Table 1: *(Continued...)*

Number of patients=140	N (%)
Desire to beaccompanied	31 (22,6)
Impact on daily activities and hobbies	
Daily activities and quality of sleep	38 (27,7)
Hobbies	31 (22,5)
Total skindex	
Low impact on the life quality	21 (15)
Moderate impact on the life quality	49 (35)
Important impact on the life quality	70 (50)
Treatements	
Topicaltreatment	102 (74,5)
Systemictreatment	34 (24,8)
Phototherapy	37 (26,6)

The Fear of worsening or persisting lesions or scars was more frequent in women (p=0,05) and the Shame was significantly related to the Young age (p=0,02) and Severe psoriasis (p=0,006). Anger was significantly related to severe psoriasis (p=0,013), old one (0,07) and the use of systemic treatments (p=0,046).

Besides, Relationships with others was altered in female patients (p=0,041),young patients (p=0,019) and patients having Severe psoriasis (p=0,056), and generally, the quality of life was significantly altered in patients having severe psoriasis (p=0,016) and old psoriasis (p=0,009).

However, the following items were not significantly affected or related to a particular characteristics of psoriasis: the frustration, the emotional life, the desire to be accompanied, the impact on daily activities, the sleep quality and Hobbies (Table. II).

Table 2: Univariate analysis

The skindex items	Characteristics of psoriatic patients significantly associated with the item affected	*P* value
Pruritus	Low SEL	0,05
	Mild psoriasis	0,011
	Originfrom rural regions	0,05
No improving and recidivism	Young age	0,000
	Female	0,000
	Psoriaticarthritis	0,004
	Psoriasis of the scalp, the nails and mucosal psoriasis	0,000
Fear of worsening or persisting lesions or scars	Female	0,005
Shame	Young age	0,02
	Severe psoriasis	0,006
Depression	BSA >30%	0,026
Anger	Severe psoriasis and BSA >30%	0,013
	Old psoriasis	0,007
	Systemictreatment	0,046
Relationshipswithothers	Young age	0,019
	Female	0,041
	BSA >30%	0,056
Total skindex	Severe psoriasis and BSA >30%	0,01
	Old psoriasis	0,009

DISCUSSION

Psoriasis is a complex multifactorial skin disease which is known linked to manymetabolic [15], autoimmune and psychiatric [10] comorbiditiesthat must be taken into account in the management of this disease [16].

The psychological impact of psoriasis and the alteration of the life quality iscurrently proven in many studies,There are even some series that have proven that this negative impact is stronger than other chronic dermatitis such as atopic dermatitis [17], and that this impact of psoriasis is similar to other dangerous diseases such as breast cancer and certain serious heart diseases [18]. Other studies proposed a theory that this alteration of the life quality is compounded by the other comorbidities of psoriasis. Furthermore, a recent study proved that this alteration of the quality of life affects not only psoriatic patients but also their families [19].

In our study, we aimed to evaluate this alteration of the life quality in Moroccan patients, we didn't study the life quality on psoriatic patients families, or the relationship of this impact and comorbidities. So we noticed that the Severity and theduration of psoriasis are the two characteristics significantly related to the general alteration of the life quality which is almost the same for other studies (resumed in Table III).

This impact is also increased by the scalp and nailsinvolvement which affects the general health, emotional life, and increase the severity of psoriasis [20-22]. In our psoriatic patients,the scalp and nail involvement were significantly related to the fear of recurrence and persistence of lesions which may be explained by the fact that these areas of the body are the symbol of beauty especially in women.

Besides, there is currently a great interest in the psoriatic arthritis (PSA) and his negative impact on the life quality,especially if it complicates skin psoriasis, So several scales for assessing the quality of life of these patients were validated [23,24].

Furthermore, many studies proved the alteration of the life quality in patients with PSA than patients

Table 3: Review of literature

Study	Year	Number of patients	Method of measurement	Results
Taiwanese[28]	2011	480	Dermatology life quality index (DLQI)	The psoriasis severity and the young age have a negative impact on quality of life
Chilean[29]	2011	153	Skindex 29	Important impact in males , young age , recent psoriasis, facial involvement
Japanese[30]	2012	213	Questionnary (total work and productivity impairment)	The severity of psoriasis has an impact on work and productivity
American[31]	2012	5604		The severity of psoriasis increases the feeling of anger, frustration, embarrassment, pruritus and pain
Spanish (Pso life study)[5]	2013	304	Dermatology life quality index (DLQI)	Impact parallel to the severity of psoriasis and the involvement of uncovered areas
Polish[32]	2013	100	Satisfaction with life scale	The importance of satisfaction level increases with age
Polish[33]	2013	168	Skindex 29	Important Impact parallel to the severity of psoriasis, young age, somatic symptoms and disease acceptance
Iranian[34]	2014	55	Questionnary (social functioning (SF)-36)	alteration of the well-being and the quality of work
Spanish (Arizona study)[35]	2014	1022	DLQI, Short form 36 questionnary	Alteration of quality of life in females with sleep disorders, depression, anxiety
Malaysian[36]	2013	250	(DLQI) and Version 2 of the 12-Item Short-Form Health Survey	The severity of psoriasis and the young age have a negative impact on quality of life with increased health care costs
Our study	2013/ 2014	139	Skindex 16	The severity, duration of psoriasis, young age, female gender, low CL and SEL have a negative impact on the life quality

with cutaneous psoriasis only [25,26], except one study which noticed that there was no change in the impairment of the quality of life with the presence of PSA in patients with cutaneous psoriasis using the PSAQOL questionnary [27]. In our study, we had a small sample of PSA with skin psoriasis (5 patients) because these patients are also followed by Rheumatologists especially those without skin psoriasis. Despite this fact, we proved that it affected significantly the life quality of our patients especially the fear of no improving and recidivism.

Pruritus

Psoriasis is known among the most pruritic inflammatory dermatoses according to the results of several studies [37-41]. Furthermore, this pruritus increases the negative impact of psoriasis on the life quality such as sleep disorders, sexual, appetite and concentration troubles and the alteration of the quality of work [42-44].

In our study,Pruritus was present in 58,7% of our patients and was intense in 31,9%. It was significantly related to the Lower SEL, the origin from rural regions of and it increases the severity of psoriasis.

Depression

If psychiatric comorbidity is important in psoriasis, depression is by far the most common psychiatric illness encountered [45-47]. The links between psoriasis and depression are not only psychopathological, biological factors may explain this association (elevated levels of substance P and TNF, decreased serotonin levels) [48]. There is thus a vicious circle "psoriasis- alteration of the life quality- depression" which may further complicate the management of psoriasis, because the treatment of psoriasis doesn't improve necessarily the depression. On the contrary, it is obvious that depressed psoriatic patient could not treat correctly his psoriasis [49].

A UK population-based cohort study of 146,042 patients [50] demonstrated an increased incidence of diagnoses of depression, anxiety and suicidality in psoriasis; the authors estimated that over 10400 diagnoses of depression, 7100 diagnoses of anxiety, and 350 diagnoses of suicidality were attributable to psoriasis each year, while Gupta [51] found that 5.5% of patients with psoriasis had active suicidal ideation and that 9.5% expressed a death wish and that this depression is increased by the pruritus. Another study demonstrated an increased use of antidepressant drugs in psoriasis [52].

In our study, Depression was significantly observed in patients with severe psoriasis and fortunately we had no cases of suicidality.

Sleep Quality

the sleep quality is among the domains the most affected in patients with psoriasis and this Sleep disturbances can cause significant quality of life impairment,which

was proved in many studies (koo and al [53]; Delfino and al [54]; Hu and al [55]. This sleep impairment in psoriasis is known linked to many reasons such as the pruritus, the psychological burden and the obstructive sleep apnea which is a common and an increasingly prevalent sleep disorder that is receiving attention in terms of a potential association with psoriasis, psoriatic arthritis and rheumatoid arthritis [56].

In our study, this sleep quality was not significantly affected,maybe because of spiritual and religious reasons and the role of the family support in our society.

Relationships and Social Integrity

Human kind is known very sociable, but this sociability could be injured in some situations that makes the person want to be alone and distant from others. This situation could be in some chronic diseases like psoriasis.In the same time, psoriasis may attract attention and cause avoidance and public rejection which may cause a disturbance in the social integrity in psoriatic patients.

This social integrity disturbance is proven in some epidemiological studies [57,58] like the survey of Poot [59] that found severe family dysfunction in these patients in comparison with families without a psoriasis. Even in our study, we noticed an alteration of relationships with others especially in young female patients and patients with severe psoriasis with a significant persistence of the desire to be accompanied.

The psoriasis affects also sexual functioning. In Gupta's cross –sectional survey [60] of 120 inpatients, 40% reported a decline in sexual activity since the onset of psoriasis. Another survey of Sampogna and al [61] proved this sexual dysfunctioning.

In our study the sexual life has not been well exploited but psoriasis did not influence significantly the emotional life of our patients.

Anger

Anger is also among the most psychological troubles that we can observe in chronic diseases, unfortunately, it's association with psoriasis has not been well described.

In anIndian recent study [62] of 48 psoriatic patients,the prevalence of anger was estimated: 58.3%, and in another American study, the prevalence of anger was more important: 89% [63].

In our study, we had a low prevalence of anger in comparison with others which may be explained by the religious convictions of our patients.

However, anger was significantly related to severe and old psoriasis and the use of systemic treatments in our patients.

Othersitems: like the fear of no improving and recidivism; the fear of worsening or persisting lesions or scars and the shame were items proved related to psoriasis in our population but were not described in other publications according to our knowledge.

Besides, the frustration and the desir to be accompanied were not related to psoriasis in our population and it were not described enough in the literature since there is just one descriptive American study of 75000 patients that reported the frustration in 89% of psoriatic patients [63].

CONCLUSION

In our study, Severe and old psoriasis causes an important impact on the life quality especially in young females which leads us to insist on the psychiatric approach of these patients to complete the global management of this chronic disease by the realization of a team work containing a psychologist in the special consultation of psoriasis patients.

Statement of Human and Animal Rights

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Statement of Informed Consent

Informed consent was obtained from all patients for being included in the study.

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Our Dermatology Online

Brief Report

Chilhood leprosy: Clinical and epidemiological study in the Department of Dermatology, Clinicas Hospital, Faculty of Medical Sciences, National University of Asuncion-Paraguay, 2005-2014

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ABSTRACT

Introduction: Leprosy in childhood is not a common finding. The risk of a child to develop the disease is 4 times greater in contact with close people and 9 times higher among household contacts. The maximum risk observed is when the contact is Multibacillary (MB) and intradomicilliary. Leprosy in childhood reflects the clinical characteristics of adult, with some peculiar aspects. Non-contagious forms (IL and TT) are common during childhood. The contagious forms (BB, LB and LL) are less frequent due to higher required incubation period. **Aim:** To describe the clinical and epidemiological characteristics of childhood leprosy in the Department of Dermatology, Clinicas Hospital from January 2005 to July 2014. **Methods:** Retrospective, observational cross-sectional study with an analytical component. **Results:** The total number of leprosy patients was 369, and of these 11 were pediatric patients (2.98%) with a predominance of males (8/11) from 3 to 16 years. The BI ranged from negative to 3+. 6/11 were MB. The evolution was good in all cases and two patients developed leprorreactions. The lesions were predominant in facial location. 6/11 patients had family contacts. **Conclusions:** Leprosy in children is more common than is reported, especially in endemic areas. In <5 years, the disease is very rare. More than half of the cases of children with leprosy have a positive contact. It is considered that in <5 years the spread is always intradomiciliary; this shows the importance of monitoring contacts, which will be possible with the determination of all stakeholders in order to banish the undetected cases and prevent damage.

Key words: Hansen disease; Leprosy; Childhood hansen disease

INTRODUCTION

Hansen's disease is a contagious disease of chronic course caused by Mycobacterium leprae that has tropism for the skin, mucous membranes and peripheral nerves. Other organs may also be compromised.

Because the consequences, degree of disability and deformity that occurs, are indispensable early diagnosis and treatment, allowing an endemic disease control.

In the field of pediatric dermatology leprosy remains a little described and undervalued in daily practice, so it becomes a diagnostic challenge because of the diversity of clinical manifestations that may occur, making necessary a thorough skin and neural examination in all children presenting suspicious lesions suggestive and an infectious source [1].

Aims

General

To describe the clinical and epidemiological characteristics of childhood leprosy in the Department of Dermatology, Clinicas Hospital, in the period from January 2005 to July 2014.

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Specific

1. To set the epidemiological characteristics of the study population.
2. To determine the clinical characteristics of leprosy in childhood.
3. To describe the type of treatment given and evolution.

MATERIALS AND METHODS

Design

Retrospective, observational cross-sectional study with an analytical component. The study was conducted at the Department of Dermatology of CH, FMS-NUA, between January 2005 and July 2014.

Reference Population

Asunción is the capital of the Republic of Paraguay and is located on the right bank of the Paraguay River (which divides the country into two regions) standing in the Eastern Region. Its metropolitan area called Gran Asunción has a population of 2,870,000 inhabitants. Its area is 118 km^2 and population density 4,411 inhabitants/km^2.

Inclusion Criteria

All patients with leprosy in childhood, diagnosed clinically and with pathological confirmation, that have consulted in the Department of Dermatology of CH. FMS-NUA.

Exclusion Criteria

No leprosy patients in pediatric age or no pathological confirmation of the disease.

Sources

Medical records of patients with diagnosis of Hansen's disease in childhood.

RESULTS

- The total number of leprosy patients was 369, and of these 11 were in pediatric age (2.98%).
- The disease predominated in males.
- The age of onset was from 3 years to 16 years.
- Bacillary Index (IB) ranged from negative to 3+.
- Six of the eleven cases presented Multi bacillary forms; the TT and LL forms were 4 cases each.
- The evolution was good in all cases and two developed leproreactions.
- The predominant location of the lesions was on the face.
- Six of the eleven cases had a family contact.

The summary of findings stated in Table 1.

DISCUSSION

Pediatric patients suffering from leprosy were 11, which corresponds to 2.98% of the total infected patients (369), lower than in other series, as Terencio de las Aguas (7.7%) and Fakhouri et al (17.3%) [1-5].

Among the children affected, males predominated coinciding with a national study of another service of dermatology; other authors did not find any differences in sex [4,5].

Ages of children affected ranged from 3-16 years, with school children (6-12 years) and adolescents the hardest hit, coinciding with the literature reviewed [1].

Table 1: Summary of findings

Sex/age/orig	FMH	CC	Local	Evol	PD	BI	Treat	Reac
F/3/R	Father LL	Papules	LLL	3 months	TT	Negative	PB	No
F/8/U	Mother LL	Hipocrom macules	LL and UL	1 month	IL	Negative	PB	No
F/10/U	Father LL	Hipocrom macules	RUL	2 years	BB	Negative	MB	No
M/11/U	No	Eritem macules	Face	1 year	TT	Negative	PB	No
M/12/R	No	Eritem macules	UL	3 years	LL	2+	MB	No
M/12/U	Mother LL	Hipocrom macules	Back	1 year	BL	2+	MB	Yes
M/13/U	No	Eritem macules	Face, UL, LL	3 weeks	LL	3+	MB	Yes
M/13/U	No	Hipocrom macules	Face	2 years	TT	Negative	PB	No
M/14/R	Father LL	Eritem macules	RLL	4 months	TT	Negative	PB	No
M/16/U	No	Ulcer	LL	6 years	LL	1+	MB	No
M/16/R	Aunt	Hipocrom macules	Face/back	2 years	LL	3+	MB	No

Sex: M: Male; F: Female, Age: Years, Origen: U: Urban; R: Rural, FMH: Familiar medical history, CC: Cause of consultation, Local: Localization of lesions, Evol: Evolution, PD: Pathological diagnosis, BI: Bacillary index, Treat: Treatment, Reac: Leprosy reactions, LLL: Left lower limb, LL: Lower limb, UL: Uper limb, RUL: Right uper limb, RLL: Right lower limb, PB: Pauci bacillary, MB: Multi bacillary, IL: Indeterminate leprosy, TT: Tuberculoid leprosy, LL: Lepromatous leprosy, BB: Borderline borderline leprosy, BL: Lepromatous borderline leprosy, BT: Tuberculoid borderline leprosy

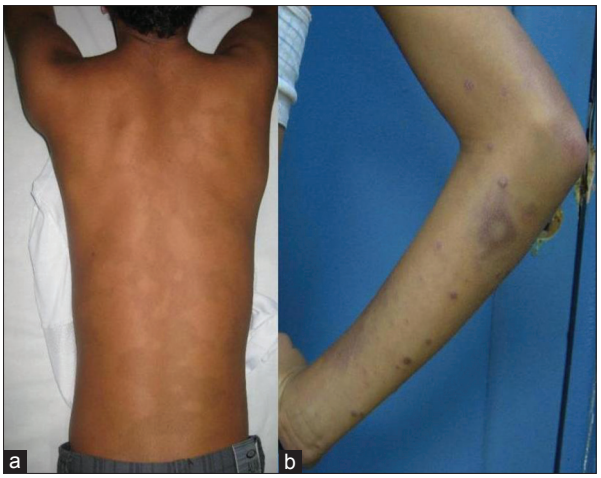

Figure 1: (a) Clinic. Hypochromic macules, between 2 and 6 cm, net limits, in the back, (b) Clinic. Erythematous plates between 0.3 and 0.8 cm, net limits, jagged edges, with the center of colored skin ("inverted dish") in arms. There are nodules, also.

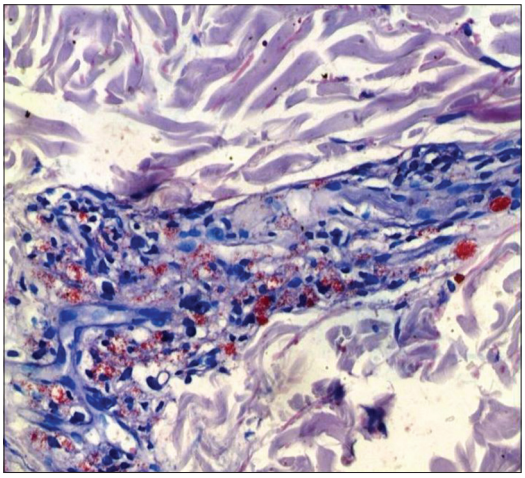

Figure 2: Histopathology. Peri adnexal chronic inflammatory infiltrate with multiple bacilli (ZiehlNeelsen 40X).

The BI ranged from negative to 3+, with 5 positive cases (45%), is inconsistent with the literature in which the positivity percentage is lower (2%). This is because the predominant number of MB cases in our study (6/11), opposite as usually PB cases described more frecquent in children [5,6].

Evolution was good in all cases and two developed leproreactions.

The elementary dermatological more frecquent lesion was a macule, with facial location, and in exposed areas, which is the most frequent site of occurrence in children with leprosy in other countries [7].

Six of eleven cases had a family contact. In most series this is the constant [1,3,8].

In developing countries, where leprosy is a public health problem, it is well accepted that children below five years are more likely to develop it than adults. About 17.13% of all cases of leprosy in India are in children under 15 years. Van Beers et al. show that the risk of a child developing leprosy is 4 times greater in contact with close people and 9 times higher among household contacts. The maximum risk is observed when the contact is Multibacillary and intradomicilliary [3].

Leprosy in childhood presents with a variety of clinical and histological manifestations, which necessitate a thorough skin examination in every child with suggestive or suspicious skin lesions and an infectious source (Figs 1ab and 2). Many skin lesions are usually asymptomatic and often mimic other dermatologic pictures [1,3].

CONCLUSIONS

1. Leprosy in children is more common than we tend to think, especially in endemic areas, as is our country [9]. In patients with less than five years, the disease is very rare [10].
2. More than half of the cases of children with leprosy have a positive contact.
3. It is consider that in patients with less than five years the spread is always intradomiciliary; this shows the importance of monitoring contacts, which will be possible with the determination of all stakeholders in order to banish the undetected cases and prevent damage.

Statement of Human and Animal Rights

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Statement of Informed Consent

Informed consent was obtained from all patients for being included in the study.

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Our Dermatology Online

Brief Report

Histopathological spectrum of benign melanocytic nevi – our experience in a tertiary care centre

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ABSTRACT

Introduction: Melanocytic lesions show great morphological diversity in their architecture and the cytomorphological appearance of their composite cells. Histological assessment of these melanocytic nevi constitutes a substantial proportion of a dermatopathologist's daily workload. The aim of our study was to observe the histological spectrum and types of benign melanocytic nevi and melanoma and also to identify the unusal/atypical histological features in these melanocytic nevi. **Results:** Intradermal nevus was the most common benign melanocytic nevi comprising 11 (62.5%) out of the total 13 cases. In ten cases lesions were located on head and neck region. Maximum number (60%) of cases were seen between 30-40 years of age. **Conclusion:** Melanocytic lesions of the skin are of notorious challengefor the pathologist. Face was the most common site and intradermal nevus was the most common lesion in our study.

Key words: Melanocytic lesions; Histological features; Intradermal nevus

INTRODUCTION

Benign Pigmented Lesions and Malignant Melanoma

Melanocytic proliferations are composed of one or more of three related types of cells: Melanocytes, nevus cells or melanoma cells, each of which may be located in the epidermis or in the dermis. Melanocytes are solitary, dendritic cells with small regular nucleus. Nevus cells are rounded or spindle shaped cells arranged in clusters with small regular nucleus. Melanoma cells are rounded or spindle shaped arranged in clusters and sheets with large irregular hyperchromatic nucleus [1].

Benign tumors of nevus cells are called melanocytic nevi, while malignant tumors are called malignant melanomas and the cells of these lesions are called as melanoma cells. Melanocytic lesions are of importance primarily because of malignant melanoma, which is the single most common potentially lethal neoplasm of skin [1].

Melanocytic Nevus

Melanocytic nevus is generally considered to be a benign neoplastic proliferation of melanocytes (Fig. 1). Melanocytic nevi are only rarely present at birth. Most nevi appear in adolescence and early adulthood. Melanocytic nevi are defined by the presence of nevus cells, which, even though they are melanocytes, differ from ordinary melanocytes by being arranged at least partially in clusters or nests, by the tendency to be round rather than having dendritic cell shape and a propensity to retain pigment in their cytoplasm [1]. Many nevi appear to be clonal [2].

There are transitional stages in the life cycle of nevi, which are believed to start out as junctional nevi, then become compound nevi and after having become intradermal nevi, undergo involution [1].

Junctional Nevi

It presents as a well circumscribed brown to black macule which can appear anywhere on the body

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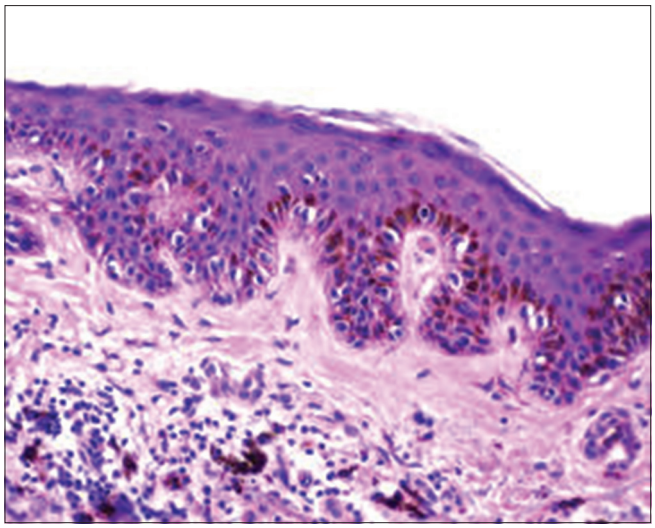

Figure 1: Melanocytic nevus Histologically Melanocytic nevi are defined by the presence of nevus cells, which, are partially in clusters or nests, by the tendency to be round rather than having dendritic cell shape and a propensity to retain pigment in their cytoplasm.

surface and appears during childhood or early adolescence [2].

Histopathology

It is composed of nevus cells that lie in well-circumscribed nests either entirely within the lower epidermis or bulging downward into the dermis but still in contact with the epidermis. The nevus cells in these nests generally have a regular, cuboidal appearance, although they are occasionally spindle-shaped.Varying amounts of melanin granules are seen in the nevus cells [1].

Compound Nevi

They occur more commonly in children and adolescents [2].Clinically, a compound nevus is a pigmented papule or a plaque [1].

Histopathology

A compound nevus possesses features of both a junctional and an intradermal nevus. Nevus cells in the upper, middle, and lower dermis may present characteristic morphologic variations called types A, B, and C, respectively. Usually, the type A nevus cells in the upper dermis are round to cuboidal and show abundant cytoplasm containing varying amounts of melanin granules. Melanophages are occasionally seen in the surrounding stroma. The cells in the mid-dermis usually are type B cells. They are smaller than the type A cells, display less cytoplasm and less melanin, and generally lie in well-defined aggregates or cords. Type C nevus cells in the lower dermis tend to resemble fibroblasts or Schwann cells, because they are usually elongated and possess a spindle-shaped nucleus. They often lie in strands and only rarely contain melanin [1].

Intradermal Nevus

It is the most common type of melanocytic nevi and vast majority are found in adults [2]. Intradermal nevi show essentially no junctional activity. Nevus cells are confined to the dermis where they are arranged in nests and cords. Multinucleate cells may be seen. In the deeper parts of the lesion nevus cells may assume a neuroid appearance [1].

Baloon cell nevus

This is a rare lesion clinically indistinguishable from ordinary nevus [2]. The ballooning is believed to develop from melanosome degeneration [2].

Histologically, nevus may be compound or dermal and the swollen nevus cells have clear cytoplasm and a central hyperchromatic nuclei. The diagnosis is restricted to lesions containing over 50% of balloon cells and not nevi showing focal balloon cell changes [2,3].

Halo Nevus

It is characterised by the presence of a depigmented halo several cms in width, around a melanocytic nevus [2]. Most persons with halo nevus are children or young adults, and the back is the most common site [1].

Histologically, there is dense lymphocytic infiltrate within the dermis and nevus cells are seen surviving in nests or singly among lymphocytes. Macrophages are also present in the infiltrate. The depigmented halo shows an absence of melanin pigment and melanocytes in the basal layer. Rarely a halo nevus is devoid of inflammatory cells [2]. At a later stage, only a few, and finally no distinct nevus cells can be identified [1].

Spitz Nevus

Synonym

Spindle cell nevus, epithelioid cell nevus, benign juvenile melanoma [2].

The Spitz nevus, named after Sophie Spitz, who first described it in 1948 [1,2]. The lesion usually is solitary and is encountered most commonly on the lower

extremities and face. In most instances, it consists of a dome-shaped, hairless, small pink nodule. In 95% of the patients, the size of the tumor is <1 cm. Its differentiation from a melanoma can often be very difficult and occasionally even impossible [1].

Histologically, most Spitz nevi are compound, 5-10% are junctional and 20% are intradermal lesions (Fig. 2).

The Major Diagnostic Criteria Include

1. Symmetrical appearance of lesion
2. Cell type- epitheloid and/or spindle cells
3. Maturation of nevus cells
4. Lack of pagetoid spread of single melanocytes
5. Coalescent, pale pink Kamino bodies.

Minor Criteria

1. Junctional cleavage
2. Pseudoepitheliomatous hyperplasia
3. Superficial dermal edema and telangiectasia
4. Giant nevus cells (multinucleate and uninucleate)
5. Absence of pleomorphism.

Congenital Melanocytic Nevus

A congenital melanocytic nevus may be defined as a lesion present at birth and containing nevus cells (Fig. 3). Congenital nevi are found in about 1% to 2% of newborn infants [1]. Majority are less than 10mm in diameter [1,2]. Giant congenital nevi have the distribution of a garment(garment nevi) [1].

Histopathology

Congenital nevi may be junctional, compound or intradermal in type, depending on the age at which they are removed. Nevi removed after neonatal period show the presence of nevus cells between collagen bundles singly or in Indian file or extension of nevus cells around nerves, vessels and adnexae [2,4].

Blue Nevus

It is a small slate blue to blue black macule or papule found in the extremities, acquired after infancy. Histologically, the common blue nevus is composed of elongated, sometimes finely branching melanocytes in the interstices of dermal collagen in upper and middermis. Laterally the lesion merges with the dermis without a clear margin and macrophages containing melanin are often found [2,4].

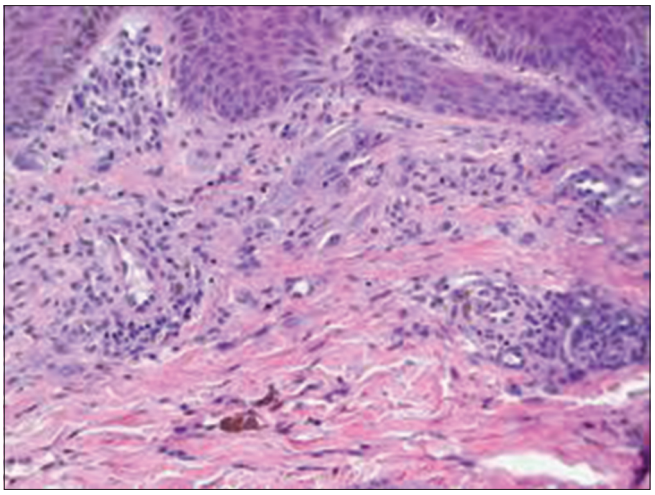

Figure 2: Spitz nevus Histologically The Spitz nevus, Histologically The lesion usually is solitary Cell type- epitheloid and/or spindle cells with orderly maturation of nevus cells.

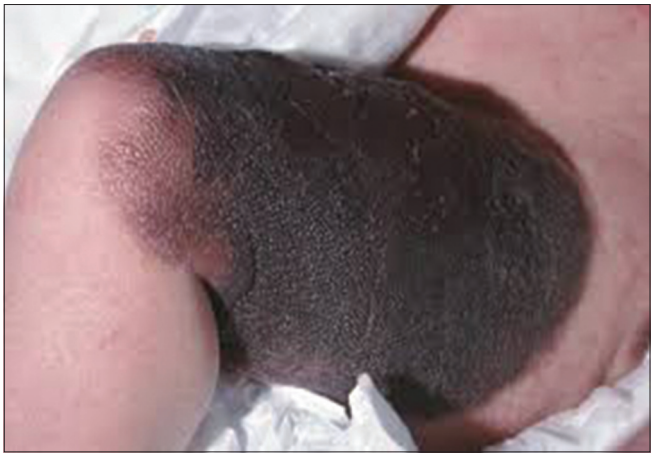

Figure 3: Congenital melanocytic nevus: A congenital melanocytic nevus lesion present at birth.

The cellular blue nevus is composed of dendritic melanocytes with islands of epitheloid and plump spindle cells with abundant pale cytoplasm and usually little pigment. Heavily pigmented varieants do occur.

MATERIALS AND METHODS

A total of 13 cases of melanocytic nevi were studied retrospectively over a period of seven years. Skin biopsies with a primary clinical diagnosis of nevus/mole/melanoma were considered in the study. Relevant data was recorded and analysed.

Mesenchymal tumours of skin, haematological tumours of skin, neural tumours of skin, nonneoplastic lesions of skin and all tumours arising from mucosal area of

Table 1: Age distribution of benign melanocytic tumours of skin

Age in years	0-9	10-19	20-29	30-39	40-49	50-59	60-69	70-79	80-89	Total
Benign tumours										
Melanocytic										
Intradermal nevus	-	1	1	7	1	-	-	-	1	11
Compound nevus	1	-	-	-	-	-	1	-	-	2
Total	3	5	7	11	10	5	6	3	3	53

mucocutaneous junction such as glans penis and eyelid margin were excluded.

The study was prospective (2years) as well as retrospective (5 years) and was done during the period of September 2004 to September 2011 i.e.7 years. Data for retrospective study was obtained from departmental records, tissue blocks and slides. Data for prospective study was obtained from clinical records, tissue specimens, tissue blocks and slides Clinical details were obtained and maintained according to the proforma.

All the biopsies and resected specimens received in the histopathology section were immediately fixed in 10% formalin for 24 hours. Gross features of the specimen were noted. Multiple sections of the specimen were taken. Then they were processed and embedded in paraffin wax. Three-five microns thick sections were prepared and then stained with Haematoxylin & Eosin.

Detailed study of the sections was performed under the light microscope and then the final diagnosis was given.

Ethical clearance

Ethical clearance has been obtained from Ethical committee of institution.

Statistical Methods Applied

Following Statistical methods were applied in the present study.

1. Number and percentage
2. Descriptive statistics

Table 1 shows age distribution of melanocytic tumours of skin.

Benign Melanocytic Tumours

Intradermal nevus

This lesion was seen in 11 cases. In all 11 cases the lesion was located in head and neck region. Maximum number (60.3%) of cases were seen between 30-40 years of age. Histologically the dermis showed nests, cords and sheets of nevus cells showing maturation. The cells showed varied amount of intracytoplasmic melanin pigment. Also few melanophages were seen in the dermis [4].

Table 2: Comparison of melanocytic tumours of skin with distribution of different benign tumours of skin

Tumours (total)	Number of cases	Percentage
Epidermal (11)		
Actinic keratosis	2	3.76
Verruca vulgaris	4	7.54
Verruca plataris	1	1.88
Seborrheic keratosis	2	3.76
Warty dyskeratoma	1	1.88
Keratocanthoma	1	1.88
Adnexal- Hair follicle (13)		
Pilomatricoma	8	15.14
Trichofolliculoma	1	1.88
Trichoepithelioma	4	7.54
Sweat gland (16)		
Chondroid syringoma	5	9.44
Hidradenoma	2	3.76
Apocrine hidrocystoma	1	1.88
Poroma	5	9.44
Spiradenoma	3	5.66
Melanocytic (13)		
Intradermal nevus	11	20.80
Compound nevus	2	3.76
Overall total	53	100

Compound nevus

This lesion was diagnosed in the two cases. One patient was a seven year old child and another was 70 year old male patient. Histologically epidermis was thinned out and showed junctional activity at dermoepidermal junction with dermal component of nevus cells showing maturation.

Table 2 shows comparison of melanocytic tumours of skin with distribution of different benign tumours of skin.

So to summarize the above table benign adnexal tumours formed the majority 29 cases (54.74%) followed by melanocytic tumours 13 (24.56%) and benign epidermal tumours 11 (20.70%). Thus indicating predominance of benign adnexal tumours of total 53 benign skin tumours [1,4].

CONCLUSION

Benign Melanocytic Nevus

In the present study there were 13 cases of benign melanocytic naevi, out of these 11 were intradermal and two were compound naevi. In ten cases lesions were located on head and neck region. Maximum number (60%) of cases were seen between 30-40 years of age. Shoko M, et al [1] has analyzed 531 cases of naevi out of which 15 (2.82%) were junctional, 134 (25.23%) cases were compound naevi, and 382 (71.9%)cases were dermal naevi. Thus we also noted maximum number of intradermal naevi in our study as that of Shoko M, et al.

Statement of Human and Animal Rights

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Statement of Informed Consent

Informed consent was obtained from all patients for being included in the study.

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Our Dermatology Online

Brief Report

Vitriols do guarantee an efficacious reduction of the human sweat when secreted from eccrine glands

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ABSTRACT

Introduction: Eccrine glands in human body are designed to secrete a salty solution containing variable percentages of urea and/or lactic acid and/or lactates. **Aim:** We want to demonstrate that it is possible to determine the preponderance of the former or of the latter in order to achieve the astringency of the same eccrine glands using vitriols, that is white or blue vitriol (zinc sulphate or copper sulphate). **Results and Conclusion:** The results are really thrilling, and it is suggestive to notice that gymnastical exercises and climate temperatures may or not influence the production of urea and/or lactic acid, nevertheless vitriols are exceptional to achieve the desired expectations, as far as astringency and anti-perspiration is concerned.

Key words: Blue vitriol, White vitriol, eccrine glands, sweat urea, sweat lactic acid

INTRODUCTION

In man, sweating is chiefly a means of thermoregulation which is achieved by the water-rich secretion from the eccrine glands, which are the major sweat glands of the human body, found in virtually all skin and produce a clear, quasi-odorless substance, consisting primarily of water and chlorides of Sodium and Potassium, urea and lactic acid. Maximum sweat rates of an adult can be up to 2–4 liters per hour or 10–14 liters per day (10–15 g/min•m^2).Evaporation of sweat from the skin surface has a cooling effect due to evaporative cooling. Hence, in hot weather, or when the individual's muscles heat up due to exertion, more sweat is produced.

We are well acquainted with the fact that 20-minute stay in a climate chamber at 40°C (e.g. an hammam) results in a 5 % reduction in body weight [1], by means of a production of a copious sweat extremely rich in urea and chlorides. The same body weight loss can be induced by the same period of time occupied by running exercise, even if during exercise human sweat is richer in lactic acid and sodium lactates than urea and chlorides [2]. Objectively it must be stressed that sodium and chloride concentrations anyway result much lower in the sweat induced by thermal exposure than that induced by the running exercise ($p < 0.01$), while urea concentrations are significantly higher after thermal exposure than after the running exercise ($p < 0.01$). Potassium concentrations do not differ significantly with either procedure. These findings suggest that sweat composition varies with the kind of induction and that more salt seems to be lost through exercise induced sweating than by just sitting in a hot environment.

On the other hand [3-5) it has been observed that 15 min of exercises (e.g.:cycling) at different temperatures give raise to elevated pools of lactic acid and sodium, meanwhile the pool of urea remains constant, at whichever temperature the gymnastic exercise is performed.

Notwithstanding these general and empirical rules, there is an avalanche of physical-chemical variables that must kept in consideration when one decides to find a way of abating the secretion of eccrine glands:

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first of all the diet that can be rich or poor in minerals and abuse of spices, the individual's skin pH, his age and the drugs he uses to take.

Anyway, exercise, like running or cycling, especially when marathoners or pedalers practice their sport during hot temperatures and wear lycra pants, may produce an enormous quantity of sweat by the eccrine glands, and the pool of lactic acid and/or urea must be anyway determined case by case, in order to find a real solution to exert an astringency with regards to the eccrine glands, when they tend to produce more urea than lactic acid or vice versa, for causes that do not pertain this dissertation.

The phenomenon of astringency is the mirror opposite of the phenomenon of inflammation: as inflammation is characterised by hyperaemia with redness, hypersensitivity connected with sensation of pain, increased and altered permeability of the cell membranes and of vasal endothelia accompanied by oedema and exudation, so astringency is characterised by its capability to coagulate the upper layers of epidermal tissues letting the permeability of cell membranes decrease with the subsequent obstacle to the exchanges between cells and extra cellular liquids and vasoconstriction of arterioles and capillaries pertaining to the same eccrine glands.

It must be even considered that a hyperproduction of lactic acid, that tends to remain onto the stratum corneum of the epidermis, may be considered a severe source of inflammation, and thus white vitriol (zinc sulphate) is advisable amongst all the types of astringent substances, since it behaves as an excellent anti-inflammatory agent [6,7].

On the other hand, when urea, hyperproduced by eccrine cells, tends to remain onto stratum corneum of the epidermis, it risks to dissolve the intercellular matrix of the cells of the same stratum corneum, promoting desquamation with simultaneous secretion of bradykinin, and therefore blue vitriol (cupric sulphate heptahydrate) seems to represent the best astringent agent owing to the fact that it is connected with a favourable mild escariothic activity,endowed with a mild capacity of denaturing the proline of the keratin of the stratum corneum itself [8].

And thus each individual which tends to hyper produce lactic acid during his phase of sweating [5] (independently from his gymnastic activities and the temperatures he performs his training) shall be treated by the use of an acqueous solution of white vitriol (2.5%) and each individual which tends to hyper produce urea (and it will be stated in this stated that women represent the majorpart, and hormonal reasons are to be placed before whichever other cause or contributory cause) shall be treated by the use of a solution of blue vitriol (2.5%).

It is supervacaneous to stress that, being human sweat a physiological solution that presents the same percentage of urea or lactic acid independently from the quantity of secreted liquid, whenever the pool of lactic or urea decreases, by artificial ways like inducted astringency, as it is possible to observe in this study, the minor is the production of lactic acid or urea, the minor too is the secretion of salty water.

MATERIALS AND METHODS

For the detection of the lactic acid in the sweat secretion, an organdy pad was rubbed onto the inner thighs and groin, and each single pad was then immerged in a solution of deionised water for the entire night.

The solution was added by 2cg of pyrocathechin and 5ml of Oleum (smocking sulphuric acid). The determination of the quantity of lactic acid (ml) is detectable thanks to the coloration that runs from weak pinkish to frank crimson. It is to stress that 0.02ml of lactic acid turns crimson, and a colorimetric scale exists for this purpose.

For the detection of urea, the same type of organdy pads rubbed on the skin of inner thighs were immerged in a solution of deionised water for all the entire night and the presence of urea was detectable by observing the coalescence (that indicates the presence of just 1 μg/100 ml) or indeed the progressive formation of brilliant white crystals, that indicated the presence of urea (50 mg/100 ml) [7] dissolving the same solution of urea in concentrated oxalic acid.

I have recruited 20 volunteers (ten male individuals 18-35 y old, designed as A,B,C,D,E,F,G,H,I,L and ten female individuals 18-55 y old, designed as M,N,O,P,Q,R,S,T,U,V) with the perspective of determining the different secretion of urea or lactic acid in each of everybody.

Table 1: Contents of lactic acid and urea before the beginning of treatment with vitriols

Case	Content of lactic acid (mmol/lt of sweat)	Content of urea (mmol/lt of sweat)
A	18.4	11.8
B	22.2	14.3
C	19.5	16.2
D	24.5	17.9
E	17.6	11.1
F	5.1	24.2
G	11.4	23.8
H	10.2	22.9
I	7.3	24.8
L	9.4	25.1
M	5.1	27.1
N	26.4	9.2
O	6.5	22.8
P	7.3	27.5
Q	3.4	25.1
R	29.1	11.0
S	7.7	22.8
T	25.2	8.9
U	3.3	27.3
V	5.2	25.9

Table 2: Contents of lactic acid and urea after three days of treatment with the solutions of white and blue vitriols

Case	Content of lactic acid (mmol/lt of sweat)	Content of urea (mmol/lt of sweat)
A	7.7.	11.0
B	8.9	13.7
C	6.6	15.4
D	11.1	11.3
E	8.7	10.8
F	5.0	7.8
G	10.9	14.1
H	10.0	9.5
I	6.9	11.2
L	9.1	10.7
M	5.1	8.9
N	12.2	8.8
O	5.5	11.6
P	6.5	10.9
Q	3.2	6.8
R	9.7	10.5
S	7.3	12.0
T	6.6	8.7
U	2.8	9.8
V	4.9	11.1

Among the 10 women 7 of those showed a strong secretion of urea and only three, being young cyclists (cases N, R and T) showed a strong secretion of lactic acid indeed.

Among the ten men 5 of those showed a strong secretion of lactic acid and the others a strong secretion of urea (independently from their attitude to practice some sport or taking saunas).

In Table I it is possible to observe the first measurements of lactic acid and urea, taken in the afternoon of a working or rest day, in a springtime day when temperature is normal (22°C) and relative humidity is regular (65%).

After the first evaluation of contents of lactic acid and urea of every single case, we have prayed all the 20 volunteers to apply the solutions, according to the following logical rule:

A,B,C,D,E,N,R,T were asked to apply the solution of white vitriol

F,G,H,I,L,M,O,P,Q,S,U,V were asked to apply the solution of blue vitriol

The applications were made in the morning before to begin the working day or rest day (that eventually involves naturally taking saunas or making whichever sport or gymnastic activity) for three consecutive days.

Final evaluations of contents of lactic acid and urea were performed in the afternoon of the third day of experiments.

In Table II are plotted the values of lactic acid and urea after the evaluation at the third day.

RESULTS

It is evident that when individuals hypersecreting lactic acid are treated with white vitriol for three days, the pool of lactic acid, measured after the complete treatment, shows a decrease of 39.51%. Comparatively when individuals hypersecreting urea are treated with blue vitriol for three days, the pool of urea, measured after the complete treatment, shows a decrease of 43.07%.

In the case of treatment with white vitriol, the pool of urea decreases very slightly and thus not significantly, likewise in the case of treatment with blue vitriol, the pool of lactic acid decreases equally very slightly.

In order to avoid the perception of malodour evoked by the presence of urea, it is advisable to add a fragrance to the formula, which is capable to camouflage the smell of ammonia, and the fragrances suggested are MENTHA SPICATA LEAF OIL and VIOLA ODORATA OIL [9].

DISCUSSIONS AND CONCLUSIONS

It is possible to determine prior the hypersecretion of lactic acid or urea in individuals when they have had a regular perspiration during 8-12 hours and thus it is possible to abate the hyperprodution of lactic acid and urea by using lotions containing white or blue vitriol.

It is very interesting to emphasize that the astringency evoked by white vitriol does not concern the decrease of urea and vice versa the astringency evoked by blue vitriol does not concern the decrease of lactic acid, so the determination of people lactic-acid secreting or urea secreting is compulsory to find the right method to choose the best dermal cosmetic remedy.

Statement of Human and Animal Rights

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Statement of Informed Consent

Informed consent was obtained from all patients for being included in the study.

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Our Dermatology Online

Case Report

Chronic hypertrophic discoid lupus erythematosus mimicking sqamous-cell neoplasia

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ABSTRACT

Introduction: Cutaneous Hypertrophic Lupus Erythematosus (CHLE) is a rare variant Of Chronic Discoid Lupus Erythematosus (CDLE) that is characterizated for the involvement of sites of the previous lesions, specially the face, with hyperkeratotic ,elevated borders and crusted and erosive centers, refractories to therapy, that simulates epidermal neoplasias. The histopathological aspect was almost confusing due to the presence of atipias, loss of polarity of keratynocites,basal membrane interruption and heavy band of basal and dermic infiltrates of lymphocytes. Local treatment with potent corticosteroids was found it made up the involution of lesions, that had been presumed neoplastic in nature. **Material and Methods:** Clinical history,follow-up and outcome of a case. **Results:** We present a 69 year-old female patient with a 16 year-history of CDLE. After systemic treatment with cloroquine remained without cutaneous lesions until development of hypertrophic CDLE lesions in the previously affected skin. These lesions become worse with the use of tachrolimus topic and exhibited the aspect actinic keratosis-like in the three biopsies performed. Thereafter local Imiquimod induced its clinical tumoral aspect. Immunohistochemic with CD123 identificates some dendritic plasmocitoid cells in the upper dermis and dermoepidermal limit, althoug not too prominent. Only after the use of local high potency corticosteroid treatment the lesions were completely resolved and the patient remained asymptomatic for one year long,until present. **Conclusion:** The relevance of topic clobetasol response in the differential diagnosis of HCDLE vs actinic keratosis/squamous neoplasia, which made up the complete resolution of cutaneous lesions and avoid aggressive surgical behaviour.

Keywords: Cutaneous hypertrophic Lupus erythematosus; Hypertrophic Discoid Lupus erythematosus

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Our Dermatology Online

Case Report

Lupus eritematoso crónico discoide hipertrófico simulando precursor de neoplasia de células escamosas

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RESUMEN

Introducción: El Lupus eritematoso crónico discorde hipertrófico (LECDH) es una rara variante del Lupus eritematoso crónico discoide (LECD) caracterizada por aparecer en sitio de lesiones previas, especialmente en la extremidad cefálica, con lesiones hiperqueratósicas, con borde elevado y centro erosivo y costroso, resistentes a la terapéutica, que simulan neoplasias epidérmicas. La histopatología contribuye a esta confusión por la existencia de hiperqueratosis, atipías, pérdida de polaridad de los queratinocitos e interrupción de la membrana basal por un denso infiltrado de linfocitos. El tratamiento con corticoides locales puede ser definitorio, ya que logra la regresión de las lesiones, presuntamente neoplásicas o preneoplásicas. **Material y Método:** Historia clínica y seguimiento de un caso clínico. **Resultados:** Se presenta una paciente femenina de 69 años,argentina, portadora de un LECDH que, tras 16 años de tratamiento sistémico con cloroquina y ya libre de lesiones cutaneas activas, desarrolla este tipo de lesiones en algunos sitios de afectación previa. Las mismas empeoran notablemente con el uso de tacrolimo tópico primero, e imiquimod después, con aspecto clínico erosivo y costroso, y diagnóstico de queratosis actínicas en las tres tomas biópsicas efectuadas. La tinción con CD123 identifica algunas células dendríticas plasmocitoides en la dermis superior y límite dermoepidérmico, sin ser prominentes. El uso de clobetasol local logra la resolución de las mismas. **Conclusiones:** Se destaca la importancia de la aplicación local de corticoides para efectuar el diagnóstico diferencial entre HCDLE y queratosis actínica/carcinoma de células escamosas, relevante para descartar una conducta quirúrgica agresiva.

Palabras Clave: Lupus eritematoso crónico discoide hipertrófico

INTRODUCCIÓN

El Lupus Eritematoso Crónico Discoide (LECD) es una forma clínica del Lupus eritematoso,caracterizada por lesiones cutáneas eritematosas, con atrofia,escama, telangiectasias y discromía. En un porcentaje variable estas lesiones se ubican sólo en cabeza, cuello y cuero cabelludo (LECD fijo), o afectan mayor superficie (LECD extendido). El riesgo de evolucionar a LES en los que se inician como LECD se ha estimado en 17 a 30% y en el LES habrían lesiones discoides en el 8 a 28% de los casos[1].

La forma Hipertrófica (LECDH) es rara,constituye aproximadamente el 2% de LECD y fue descripta por Paul Esnard Bechet en 1940 con esa denominación inicial, a la que luego denominó "*Lupus Eritematoso Hipertrófico y Profundo*" [2,3].

Años después, otros autores señalaron la característica hipertrófica, proliferativa y atípica de las lesiones epidérmicas en la microscopía, y describieron pacientes que provenían de una cirugía micrográfica de Mohs previa, sin haber hallado tumor alguno para ser extirpado [4]. Estas observaciones adquieren interés

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actual, ya que, si bien es aceptado que el LECD puede preceder y favorecer el cáncer cutáneo, según ellas el LECDH tambien puede simularlo, y lo más importante, puede requerir otra terapéutica menos agresiva, que debiera ser intentada antes de la cirugía.

Presentamos una paciente que padece LECDH con compromiso cutáneo exclusivo desde hace 16 años, con seguimiento y tratamientos locales y sistémico con cloroquina y fotoprotección que, tras permanecer sin lesiones durante años, presenta otro tipo de lesiones, clínica e histológicamente compatibles con lesiones preneoplásicas, que empeoran notablemente con tratamiento orientado hacia ese diagnóstico (queratosis actínicas) y mejoran notablemente con el uso de corticoides locales.

CASO CLINICO

MM, 69 años, médica,viuda,argentina, enviada por reumatólogo en 1998 (55 años).

Antecedentes Personales: No tuvo embarazos, actualmente menopáusica. HTA transitoria en noviembre del 2000 (trae RMN de cerebro por ACV transitorio).

Antecedentes Familiares: Prima hermana fallecida padeció LES desde los 20 años.

Examen Físico: Consulta en el 2003 (59) presentando Livedo reticular moderado y extendido y alopecia difusa. Lesiones hiperqueratósicas en labio superior, eritemato- escamosas con costra, telangiectasias y pigmentación en mejillas.

Laboratorio: Se registran fuera de lo normal Acps ANA Ho 1/320, Acps anti- fosfolípido β2 GP1 IgG 14.5 (dudoso),eritrosed. 8mm e hipercolesterolemia.

Estudios complementarios: Ecodoppler arterial de vasos cuello sp, ecografia renal s/p., doppler cardíaco: s/p. Biopsia 1998 de piel de cara: LECD (Dr.I: Calb).

Tratamientos previos: Inicialmente se indicó cloroquina 100,Deflazacort 7,5 mg/día, isotretinoina 20 mg, protector solar, vit A ácida 0.025 local que produjo eritema, erosión y ardor,interrumpido al mes por piel seca,fisuras, y escamas. No toleró talidomida.

En septiembre del 2003 prueba tacrolimo 0.03% y cloroquina logrando estar en un mes libre de lesiones.

En el 2012 consulta de control SIN LESIONES (Fig. 1) con Cloroquina 100 mg/dia.

Motivo de consulta actual (febrero 2013) aparecen nuevas lesiones de igual característica y mayor tamaño distribuídas en mejilla derecha, labio superior e inferior, (Fig. 1a) que no mejoran (empeoran) con el uso de tacrolimo 0.1% local (Fig. 1b).

Todas las determinaciones de acps resultan ser negativas,salvo ANA 1/160. Se realiza una 1er biopsia marzo 2014: "Queratosis actínica hipertrófica" (Foto 2a) Tratamiento: Imiquimod 5% crema local 5 días por semana, luego tres.Se advierte empeoramiento notable con ulceración, costra necrótica en labio, mayor tamaño, eritema y descamación (Fig. 1c).

2da Biopsia abril 2014 N°14-03424 (mejilla) Queratosis actínica hipertrófica. 1-Las secciones semiseriadas muestran en la epidermis acentuada hiperqueratosis,

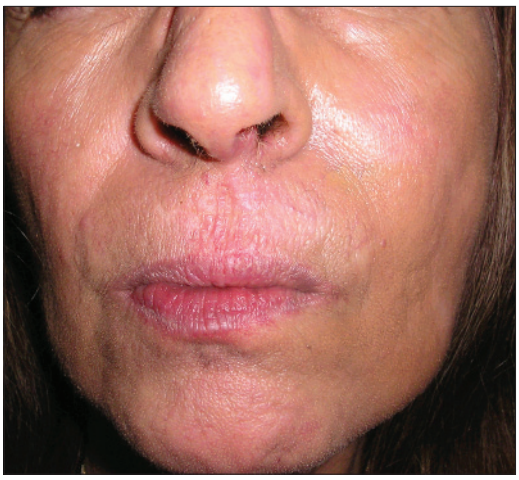

Figure 1: Sin lesiones en 2012 ,tratamiento con cloroquina.

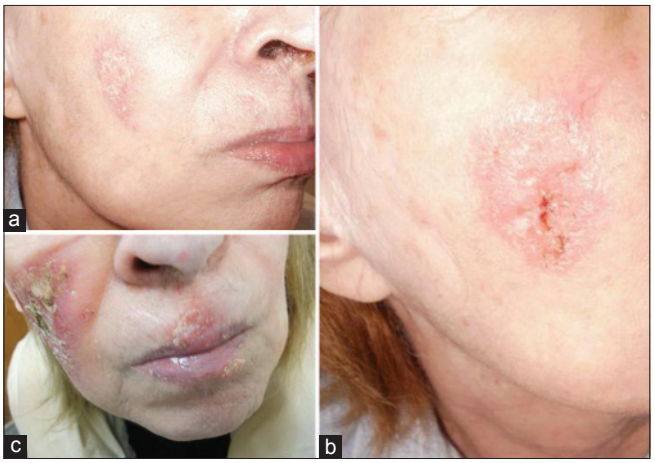

Figure 1: (a) Lesiones nuevas faciales, (b) posterior al uso de imiquimod 5%, (c) posterior a uso de tacrolimo 0.1%

paraqueratosis, acantosis, pérdida de la polaridad celular, queratinocitos atípicos,y en la dermis superficial linfocitos en banda y elastoidosis actínica (Figs. 2b - 2d).

2- labio: queratosis actínica. Las sesiones hp semiseriadas muestran epidermis con paraqueratosis, atipías de queratinocitos basales y suprabasales y en la dermis superficial linfocitos en banda subepidérmica y elastoidosis actínica.

Tratamiento: Se consulta para método de Mohs, o criocirugía en las lesiones más chicas. En el interín se intenta clobetasol (crema) local, dos veces por día. Mejoría a las dos semanas y cuatro semanas, apreciando sólo eritema y leve descamación en un sector de la periferia de la lesión de la mejilla izquierda. Labios sin costra ni erosión. Intenta nuevamente tacrolimo tópico logrando nuevo empeoramiento, por lo que reanuda clobetasol local logrando rápida mejoría y casi desaparición de las lesiones hasta la actualidad (Fig. 3).

Se obtuvo el consentimiento informado del paciente.

Antes del estudio, el paciente dio consentimiento escrito para el examen y la biopsia, tras haber sido informado sobre el mismo y el objetivo de éste.

DISCUSION

Bechet en 1940 presenta un paciente discutido en un ateneo de la Sociedad de Dermatología de Nueva York. Da el nombre de *Lupus Eritematoso hipertrófico* a la lesión localizada en labio inferior y borde bermellón, de bordes elevados, centro deprimido o cribiforme que evoluciona a costra y es diagnosticada histológicamente como Lupus eritematoso. Refiere que ha observado en otros casos que deja cicatriz importante con pérdida de sustancia y que, afortunadamente es una forma poco común[2]. En 1942 en la misma revista describe el "*LE hipertrófico y profundo*",que se inicia igual y avanza a la hipodermis (citado por Ottani en 1977) quien presenta un caso más parecido al nuestro, con lesiones en cuero cabelludo y cuello,con costras y escamas adherentes en el centro. La histología demuestra afectación inflamatoria severa de la epidermis y la interfase.,pero responde al tratamiento antimalárico [3].

Más recientemente otros autores (dermatólogos y patólogos) presentan otros casos, haciendo referencia a la descripción original y unos pocos señalan otras características como: las atipias e hiperplasia

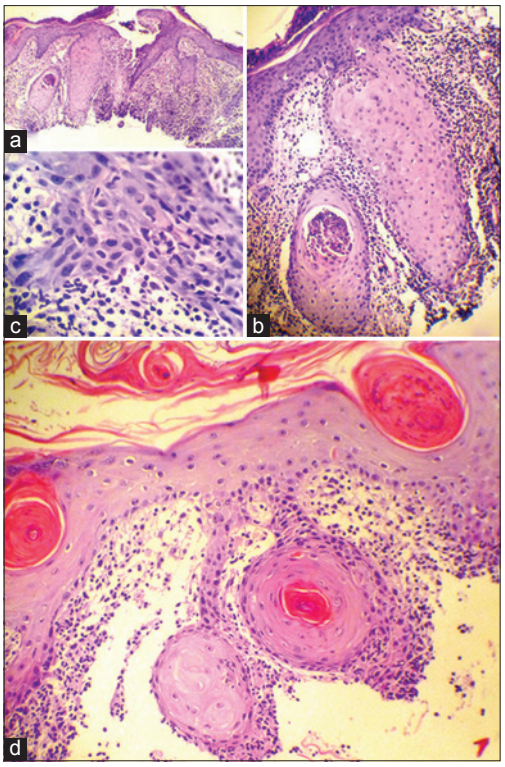

Figure 2: (a) He y e 40x ,2b 150x , 2c 400x,2d. Hiperplasia epidérmica, numerosas atipias basales, y células apoptóticas, denso infiltrado mononuclear en banda, borramiento de la MB en sectores. 2d Notar hiperqueratosis con tapones córneos en 2d.

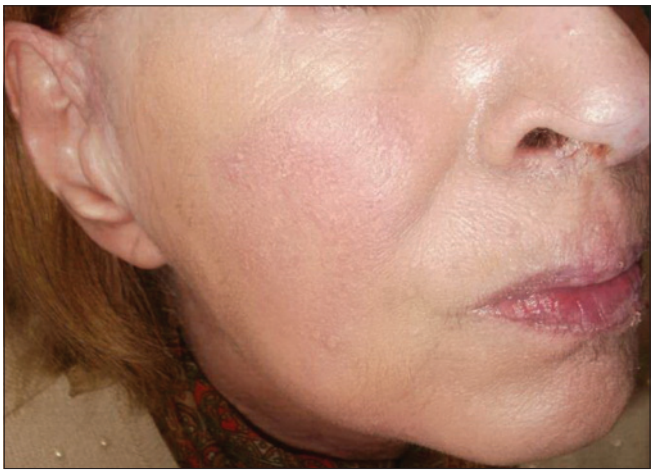

Figure 3: Mejoría y desaparición de lesiones luego de tratamiento con clobetasol tópico.

de la epidermis, la respuesta a los corticoides locales, y la falta habitual de compromiso sistémico, señalando el diagnóstico diferencial con las neoplasias escamosas [4-7]. Las lesiones son hiperqueratósicas, verrugosas,en áreas de fotodaño generalmente. A veces atróficas cribiformes en el centro con bordes indurados. Otras desarrollan pápulas hiperqueratósicas con tapones córneos en el centro,que remedan en su aspecto clínico e histopatológico el queratoacantoma.

y han sido descriptas en sitios de lesiones previas discoides. Responden a retinoides y esteroides tópicos a diferencia de las neoplasias escamosas. Generalmente este lupus no tiene compromiso sistémico. Las lesiones frecuentemente presentan atipía reactiva escamosa y pueden simular otras lesiones cutáneas escamoproliferativas. Recientemente por inmunohistoquímica se ha demostrado presencia de abundantes células CD123 +(cel dendríticas plasmocitoides) que no se hallan en las queratosis actínicas y son útiles para el diagnóstico diferencial [4].

En un estudio hp de 14 casos de Daldon, todos presentaron atipia de queratinocitos basales y elastosis solar y se acompañaron de lesiones discoides, nueve con compromiso facial, y uno desarrolló carcinoma espinocelular (CSS) en la nariz 26 años después del inicio [8].

En la microscopía se describen dos patrones: 1- la mayoría tiene infiltrados linfocitarios en banda tipo liquen plano con hiperplasia pseudoepiteliomatosa irregular y cambios vacuolares de la interfase. Numerosos queratinocitos apoptóticos y es común la atipia reactiva de los queratinocitos basales. Son claves diagnósticas de LECDH: la hiperqueratosis c/ tapones foliculares asociados a infiltrados perivasculares y anexiales, mucina dérmica distribuida en forma uniforme y engrosamiento de la MB. Se observaron células plasmáticas prominentes en dermis superficial. Hay fibras elásticas entre las células epidérmicas y el tope de los brotes papilares alongados pudiendo observarse eliminación transepitelial de las mismas. 2-Queratoacantoma-like: centro queratinoso crateriforme,con proliferación epitelial escamosa exuberante. El Diagnóstico Diferencial se establece con: la queratosis liquenoide benigna, queratosis actínica liquenoide y carcinoma epidermoide. Liquen plano hipertrófico y erupciones por drogas y, como menos probables:

halogenodermas, y micosis profundas. La correlación clínicopatológica (ubicación de lesiones, coexistencia con lesiones de LECDH) favorece el diagnóstico de LECDH. Se aconseja probar con corticoides tópicos, pero tener en cuenta que el Carcinoma epidermoide puede ser tambien una complicación tardía del LECDH [4,6].

La incidencia de SCC en LECD ha sido comunicada en 3.3% y aparecerían en forma tardía, después de dos o tres décadas [9]. El labio inferior ha sido el área más comunmente afectada, mientras que el labio superior se afectó en 2,3% de los SCC relacionados al LECD. Ocasionalmente se describió en cuero cabelludo. Los factores predisponentes serían: fotosensibilidad, cicatrices, infección crónica, e inmunosupresión. El SCC en LECD ha demostrado adoptar una conducta más agresiva. Las recurrencias, metástasis y mortalidad fueron 10% a 20% mayores que as no relacionadas al lupus eritematoso y por consiguiente se aconseja una terapéutica agresiva [9-13].

Asanalfi y Werth [7] sugieren que el diagnóstico debiera ser hecho después de repetir biopsias, y una cuidadosa reevaluación del curso de una lesión específica en relación a las lesiones lúpicas, porque diferenciar entre LECDH y SCC puede ser dificultoso clínica e histológicamente. En última instancia, Perniciaro aconseja probar el tratamiento corticoide local como elemento definitorio ante la duda [5]. Además de la descripción de esta rara forma clínica, queremos destacar en este caso:

a- el aspecto tumoral de estas lesiones que pueden inducir al error, notable en los casos publicados que provienen de cirugía de Mohs previa, sin hallazgos tumorales. b- el empeoramiento notable con imiquimod local que no encontramos referido antes (administrado como tratamiento de queratosis actínicas en una superficie amplia) que puede explicarse por el patrón de interferón que caracteriza las lesiones cutáneas del lupus eritematoso.

c- La historia previa de 16 años, sin manifestaciones sistémicas clínicas ni laboratoriales en los últimos diez años, con buena respuesta previa a la terapéutica habitual hasta entonces (cloroquina,tacrolimo local, protector solar).Livedo reticular persistente.

d- Reaparición de lesiones en los sitios afectados anteriormente (mejilla, labio inferior y superior), sin haber suspendido la terapéutica.

e- Biopsias compatibles con queratosis actínica hipertrófica (tres) en dos oportunidades. Exhiben numerosas atipias basales y suprabasales, pérdida de polaridad celular, hipertrofia epidérmica, infiltrado linfocitario prominente que borra la basal en sectores. No se advierten mitosis.

f- Mejoría notable con tratamiento corticoide local, y empeoramiento con tacrolimo local reiterado en dos oportunidades.

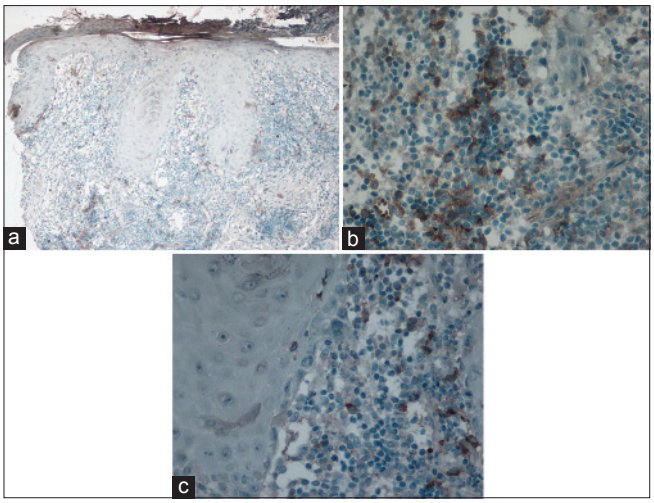

Figure 4: (a, b, c) IHQ. CD123+ . Observar acúmulos de células dendríticas plasmocitoides en dermis superior e interfase dermoepidérmica.

g-Presencia de cel dendríticas plasmocitoides CD123+ en el infiltrado (Fig. 4). Recientemente Walsh y col. analizando 27 muestras de 9 pacientes con este diagnóstico comparadas con 39 muestras de 36 pacientes portadores de neoplasia escamosa considera que las células dendríticas plasmocitoides podrían ser importantes para el diagnóstico de esta forma clínica si cumpliera tres condiciones: constituir el 10% del infiltrado, afectar el límite dermoepidérmico, y disponerse en grupos de 10 células o más [14].

Surgen preguntas acerca de la naturaleza de las lesiones informadas en estos pacientes como queratosis actínicas, que no responden al imiquimod o empeoran, mientras que, ocasionalmente, se ha referido respuesta favorable al tratamiento habitual del LECD [15] o al imiquimod, con el mismo aspecto histológico. Qué proporción de estas lesiones serán verdaderas neoplasias incipientes, ya que se habla de mejoría con corticoides locales, pero no se documenta la desaparición de las mismas, que nosotros observamos en este caso. Recientemente se aconseja el análisis dermatoscópico de toda la lesión para detectar zonas de neoplasia incipiente [16].

Efectuamos marcación con CD123 (para células dendríticas plasmocitoides) que se han referido como prominentes en las lesiones del lupus eritematoso crónico y especialmente en esta variante [17], pero no en las lesiones preneoplásicas [18], observandose la presencia de algunas células en dermis superficial (Dr. José G. Casas (Fig. 4), lo cual no parece ser concluyente.

CONCLUSIONES

Concluímos que la característica más prominente de esta forma clínica,al menos en nuestra paciente, es la respuesta sostenida al tratamiento local con corticoides de alta potencia. En cuanto a la presencia de las células dendríticas plasmocitoides, otros aportes deberán contribuir a definir los criterios de ubicación y cantidad que ayudan a avalar este diagnóstico.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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Our Dermatology Online

Case Report

Chronic cutaneous lupus erythematosus with systemic symptoms or systemic lupus erythematosus?

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ABSTRACT

Chronic cutaneous lupus erythmatosus (CCLE) and Systemic lupus erythematosus(SLE) are two separate diseases with some overlapping features. Upto 28% patients with CCLE are susceptible to develop SLE. But in some cases patients may present with CCLE with systemic features without developing SLE. Here, we present a case of Chronic cutaneous lupus erythematosus with systemic symptoms simulating SLE with no positive immunological markers to support the diagnosis of SLE.

Key words: Chronic cutaneous lupus erythematosus; Negative anti-nuclear antibody; Systemic lupus erythematosus

INTRODUCTION

Chronic cutaneous lupus erythematosus (CCLE) is a benign disorder characterised with skin lesions. But in some cases in addition to skin lesions patient may present with systemic symptoms, thus mimicking Systemic lupus erythematosus (SLE). Here, we present a case of CCLE with systemic symptoms distinguished from SLE by the absence of Anti-nuclear antibody, Anti-dsDNA and Rheumatoid factor.

CASE REPORT

A 50 year old female presented to our hospital with complaints of hypopigmented itchy plaques over back and both pinnae since 6 years. She also complained of similar lesions in the scalp which were associated with loss of hair. History dates back to 6 years when patient noted an itchy papule on the upper back which gradually increased in size along with development of similar lesions over both pinnae and scalp over a period of next 4 years. History of photosensitivity was present. She also complained of recurrent oral ulcerations, pain and swelling in both ankle and knee joints for the past 6 years. On general physical examination she was poorly built, had pallor and diffuse non-scarring alopecia of the scalp. On mucocutaneous examination the lesions on the scalp, both pinnae and upper back were in the form of hypopigmented plaques with central atrophy and well defined irregular hyperpigmented margins of size ranging from 0.5X0.6 cm to 3X4cm (Figs 1 - 2). The lesion on the upper back had yellow coloured adherent scales in the lower half (Fig. 3). On removing the adherent scale, its undersurface showed horny plugs, so called tin-tack sign. Oral mucosa and nails were normal. On being investigated, her hemoglobin was 8.5g/dl. Total leucocyte count, Differential leucocyte count, Erythrocyte sedimentation rate, total serum proteins, differential serum proteins,fasting blood sugar, VDRL, Renal profile and Liver profile were within normal range. LE cells were not seen. Tests for Anti-nuclear antibodies, Anti-dsDNA antibodies and Rheumatoid factor were negative. Xrays showed effusion in Right knee joint and right ankle joint. Histopathology of the atrophic plaque showed acanthosis, keratotic plugging, necrotic keratinocytes, thickened basement membrane, ectatic blood vessels and focal areas of pigment incontinence. Superficial and deep perivascular and periadnexal lymphocytic infiltrate and leucocytoclastic vasculitis was also noticed. Histopathology from the scaly plaque showed marked orthohyperkeratosis,

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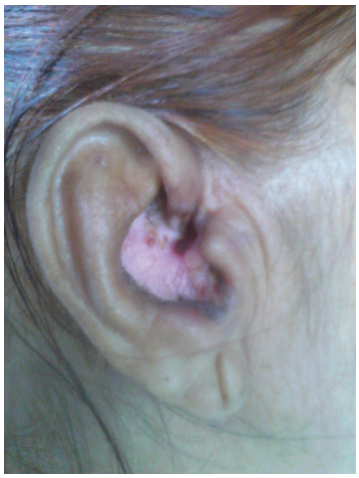

Figure 1: Hypopigmented atrophic plaque in right pinna.

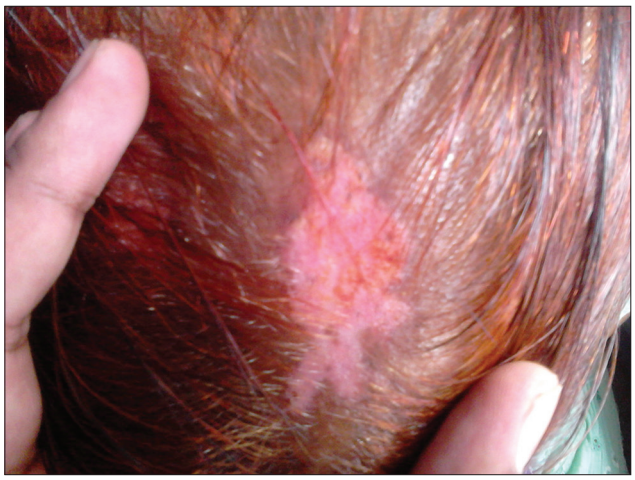

Figure 2: Hypopigmented atrophic plaque with loss of hair on the scalp.

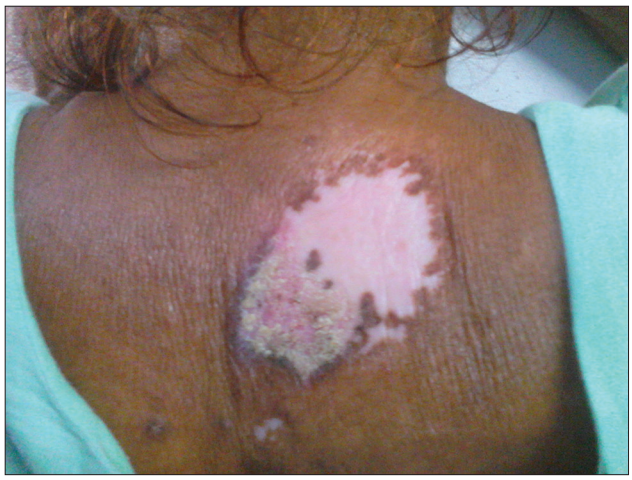

Figure 3: Hypopigmented atrophic plaque with hyperpigmented margins and yellow coloured adherent scales in the lower half of the lesion.

parakeratosis, acanthosis, necrotic keratinocytes, basal vacuolization and focal lymphocytic infiltrate at dermoepidermal junction. The findings were consistent with Chronic Cutaneous Lupus Erythematosus. Hence, based upon the clinical symptoms and histopathological findings she was diagnosed as a case of Chronic Cutaneous Lupus Erythematosus with systemic symptoms. The Diagnosis of SLE was not made as although the clinical criteria were fulfilled but no immunological criteria was present.

DISCUSSION

Discoid lupus erythematosus (DLE) also known as Chronic cutaneous lupus erythematosis (CCLE) is a benign disorder of the skin, clinically characterized by red scaly patches which heal with atrophy, scarring and pigmentary changes, and histopathologically by vacuolar degeneration of basal cell layer of epidermis and patchy dermal lymphocytic infiltrate. DLE is subdivided into a localized form in which lesions are confined to the face and neck or a disseminated form in which lesions also occur elsewhere on the body [1]. It has clinical variants like hypertrophic CCLE, lupus panniculitis, mucosal CCLE, tumid CCLE and chilblain CCLE. CCLE is more commonly seen in females between the age group of 20-40 years. Though all races are affected, the prevalence may be higher in African Americans than in Caucasians [2]. Genetic factors and somatic mutations are implicated in the pathogenesis of DLE. Three to five somatic mutations affecting autosomal genes and one mutation involving X-linked gene in lymphocytic stem cells of predisposed individuals, results in development of a forbidden clone of lymphocytes which synthesize cellular autoantibodies. Four temporally sequential phases are a prerequisite for clinical expression of lupus viz. inheritance of susceptibility genes, induction of autoimmunity by autoreactive T cells, expansion of autoimmune process with autoantibodies and immunological injury. Immunological injury is attributed to the action of autoantibodies and the immune complexes they form, which causes tissue damage [3].

Systemic features may be seen along with cutaneous lupus erythematosus. Up to 28% of patients with DLE are susceptible to develop systemic lupus erythematosus SLE. Various clinical and laboratory indicators, such as widespread DLE lesions, arthralgias/arthritis, nail changes, anaemia, leucopenia, high erythrocyte sedimentation rates (ESRs) and high titres of antinuclear antibodies (ANAs) are associated with progression to SLE in patients with DLE [4].

The Diagnosis of SLE can be made using the SLICC criteria given as under.

Requirements

≥ 4 criteria (at least 1 clinical and 1 laboratory criteria) or biopsy proven lupus nephritis with positive ANA or Anti-DNA.

Clinical Criteria

1-Acute cutaneous lupus. 2-Chronic cutaneous lupus. 3-oral or nasal ulcers. 4-Non-scarring alopecia. 5-Arthritis. 6-Serositis. 7-Renal. 8-Neurologic. 9-Hemolytic anemia. 10-Leukopenia. 11-Thrombocytopenia.

Immunologic Criteria

1-ANA. 2-Anti-DNA. 3-Anti-Sm. 4-Antiphospholipid Ab. 5-Low complement (C3,C4,CH50). 6-Direct Coombs' test [5]. Although in our case four clinical criteria could be fulfilled namely 1- Chronic cutaneous lupus, 2- oral ulcers, 3- Non-scarring alopecia, 4- Arthritis but no immunological criteria was present hence a diagnosis of CCLE with systemic sympyoms was made rather than SLE.

There have been sporadic reports of neoplastic change in DLE which range from squamous cell carcinoma and basal cell carcinoma to malignant fibrous histiocytoma and atypical fibroxanthoma [6]. The incidence of squamous cell carcinoma developing in DLE ranges from 3.3 -3.4% in various studies [7].

There are various treatment modalities available for cutaneous lupus erythematosus like topical corticosteroids, topical tacrolimus, hydroxychloroquine, azathioprine, methotrexate, mycophenolate mofetil, cyclophoaphamide, cyclosporine, high dose intravenous immunoglobulin, etc [4].

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles. Written informed consent was obtained from the patient for publication of this article and any accompanying images.

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Our Dermatology Online

Case Report

A multiple drug allergy syndrome, multiple drug intolerance syndrome and/or allergic drug reaction with multiple immune reactants

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ABSTRACT

Background: Adverse drug reactions (ADR), multiple drug allergy syndrome (MDAS) and multiple drug intolerance syndrome (MDIS), are very common in the clinical practice worldwide due to an aging population. **Case report:** Here we describe a 61 year old Caucasian female, who presented complaining of large, tense bullae on her right anterior arm; the bullae had been present for one week, with pruritus. The patient was taking multiple medications. The patient presented with multiple large, tense blisters and scattered, smaller blisters on her legs and axillae. **Materials and Methods:** Biopsies for hematoxylin and eosin (H&E) examination, direct immunofluorescence (DIF) and immunohistochemistry (IHC) analysis were performed. The H&E staining demonstrated diffuse, moderate epidermal spongiosis, with a subepidermal blister. Lymphocytes and eosinophils were present within the blister lumen and around superficial dermal blood vessels, hair follicles and sweat glands. Notably, blood vessels and lymphatics had altered shapes, many being narrowed, twisted and or dilated. DIF showed significant deposits of fibrinogen, Complement/C3c and IgA around the dermal vessels; some reactivity was also present at the basement membrane zone of the skin, as well as around dermal skin appendices. Of great interest was the reactivity seen with anti-human fibrinogen against some dermal cell junctions. IHC staining showed the presence of mainly CD4 and BCL-2 positive cells around skin appendices, with a few CD8 positive cells also present. Cyclooxygenase-2 was also very positive around the dermal blood vessels, as well as within cells of the dermal inflammatory infiltrate. The dermal lymphatics and dermal blood vessels appeared dilated, demonstrated by staining for D2-40/podoplanin and von Willembrand factor. **Conclusion:** Adverse drug reactions and multiple drug allergy syndrome have several overlapping features; the spectrum of immunopathologic and histopathologic features are not fully established. Our case contributes to our knowledge of some of these features. In the authors' experience, we have noticed that the hallmark of these reactions is the prevalence of strong deposits of fibrinogen in several vessels and dermal skin appendices. In addition, we have noted positivity on some types of dermal cell junctions. MDAS, MDIS and ADRs represent examples of how to categorize adverse, simultaneous reactions to multiple classes of chemically unrelated drugs.

Key words: Multiple drug allergy syndrome, multiple drug intolerance syndrome, adverse drug reactions, fibrinogen, vessels, intraepidermal blisters

Abbreviations: Multiple drug allergy syndrome (MDAS), multiple drug intolerance syndrome (MDIS), adverse drug reactions (ADRs), direct immunofluorescence (DIF), immunohistochemistry (IHC), basement membrane zone (BMZ), *Ulex europaeus* agglutinin 1 (UEA), hematoxylin and eosin (H&E), 4',6-diamidino-2-phenylindole (DAPI), B-cell lymphoma 2 gene (BCL-2), cyclooxygenase-2 (COX-2).

INTRODUCTION

Multiple drug allergy syndrome (MDAS) is a clinical diagnosis made in patients with adverse reactions to two or more chemically unrelated drugs, with an underlying immune-mediated mechanism causing the reaction [1]. The term multiple drug intolerance syndrome (MDIS) has been used to describe patients who express adverse

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drug reactions to three or more drugs, without a known immunological mechanism [2,3]. Thus, it is difficult in some cases to know if these nosologic entities refer to the same phenomenon. Even more confusing is to consider how single drug adverse drug reactions (ADRs) relate to a defined MDAS or MDIS case. ADRs usually affect the skin and/or mucosae (toxicoderma) [4-12], and are challenging to dermatologists, toxicologists, emergency physicians, allergists and immunologists.

A better understanding of the etiopathogenic mechanisms of drug reactions is needed, and especially when these reactions are concomitant with the presence of other sequelae such as viral infections. Such non-drug reaction forces could make selected immunologic processes more complex than previously considered [5-9]. It is also believed that allergic drug reactions may have a genetic predisposition, and that epidemiologic factors can also play a role. We describe a case that exemplifies the complexity of adverse drug reactions.

CASE REPORT

An obese, diabetic 61 year old female presented to the dermatologist with flaccid bullae and plaques, on erythematous bases. The patient was taking Doxycycline hyclate 100 mgs caps one at day orally; Micardis HCt; oral Metformin® 850 Mgs tablets; an oral multivitamin; Symlin® SubQ, Lantus® SubQ; Novolag® SubQ. The patient described no personal or family drug allergy history. Biopsies for hematoxylin and eosin (H&E) examination, direct immunofluorescence (DIF) and immunohistochemistry (IHC) analysis were performed. After receiving the biopsy results, the patient was prescribed with econazole 1% topical cream once or twice a day, and hydrocortisone topical 2.5% lotion; the dermatologist also began working to change and/or decrease her medications in consultation with her other physicians.

MATERIALS AND METHODS

Skin biopsies were taken for histology (H&E) studies, for immunohistochemistry (IHC) and for direct immunofluorescence studies (DIF); our techniques were performed as previously described [7-12].

Direct immunofluorescence (DIF)

For DIF, the skin was imbedded in optimal cutting temperature (OCT) compound for frozen-sectioning. The skin was then incubated with primary and/or secondary antibodies. We utilized FITC conjugated rabbit antisera to human IgG, IgA, IgM, IgD, IgE, Complement/C1q, Complement/C3, Complement/C4, Kappa light chains, Lambda light chains, albumin and fibrinogen. All of our antibodies were obtained from Dako(Carpinteria, California, USA) with the following details and exceptions. Our anti-human IgA antiserum (alpha chain) and anti-human IgM antiserum (mu chain) were obtained from Dako. Our anti-human-IgE antiserum (epsilon chain) was obtained from Vector Laboratories (Burlingame, California, USA). Our anti-human IgD antibodies were obtained from Southern Biotechnology (Birmingham, Alabama, USA), and utilized at dilutions of 1:20 to 1:40. The slides were counterstained with 4',6-diamidino-2-phenylindole(DAPI; Pierce, Rockford, Illinois, USA). We also used Texas Red conjugated *Ulex europaeus* agglutinin 1 (UEA) (from Vector Burlingame, CA, USA) as a vascular marker. The samples were consistently run with positive and negative controls. We classified our findings as negative (-), weakly positive (+), positive (+++) and strongly positive (++++).

IHC double staining

Our double stained IHC was performed using a double staining system; we utilized the Leica Bond Max platform autostainer with bond polymer refined Red detection DS9390, alkaline phosphatase linker polymer and fast red chromogen (red staining) as previously described [5-9]. We also used a bond polymer refined detection DS9800, horseradish peroxidase linked polymer and DAB chromogen (brown staining). The following antibodies were utilized from Dako: Mouse monoclonal anti-human CD4; CD8; CD15; CD68; B-cell lymphoma 2 gene (BCL-2) oncoprotein, clone 124; cyclooxygenase-2 (COX-2), clone CX-294; polyclonal rabbit anti-human vonWillembrand factor, and D2-40/podoplanin. The following antibodies were used from Novocastra (Chicago, Illinois, USA): anti-human CD4 clone 4B12; anti-human C8 Clone C8/144B, and von Willembrand factor. We counterstained the slides with hematoxylin. Positive and negative controls were consistently performed.

RESULTS

Microscopic description

A: Examination of the H&E tissue sections demonstrated diffuse, moderate epidermal spongiosis present.

A subepidermal blister was noted, with small numbers of lymphocytes and eosinophils noted within the blister lumen. Significant superficial papillary dermal edema was noted. The dermis also displayed a mild, superficial, perivascular infiltrate of lymphocytes and histiocytes; eosinophils and neutrophils are rare. No frank vasculitis was present; however, some fibrinoid alterations were seen in dermal blood vessel walls (**Fig. 1**).

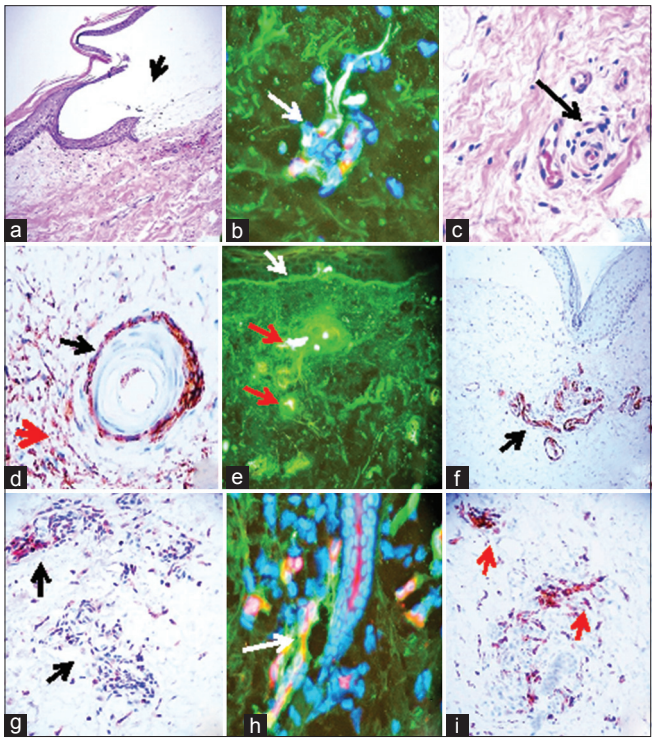

Figure 1: (a) H&E stain shows a subepidermal blister (black arrow), with dermal edema and dilation of dermal vessels. (b) DIF using FITC conjugated anti-human fibrinogen, staining positive around the upper dermal vessels (green staining; white arrow). The nuclei of endothelial cells were counterstained with DAPI (light blue staining), and the vessels were stained with *Ulex europaeus* (orange staining). (c) An H&E stain, featuring the inflammatory infiltrate around the upper and intermediate dermal vessels (black arrow; 200X). (d) IHC staining, showing positivity of the lymphatic marker D2-40/podoplanin on lymphatic vessels around a hair follicle (brown staining; black arrow), and also neo-formation of lymphatics in the adjacent dermis (brown staining; red arrow). (e) DIF using FITC conjugated anti-human IgA, and showing positive staining against the upper and intermediate vessels (green/white staining; red arrows). The white arrow highlights the linear BMZ staining. overexpressed under the blister. (f) IHC, showing von Willembrand factor expressed and grouped under the blister (brown staining; black arrow). (g) Double color IHC, showing positive staining for with BCL-2 in brown and COX-2 in red in the inflammatory infiltrate around dermal blood vessels (black arrows). (h) zDIF, using FITC conjugated anti-human fibrinogen and showing positive staining around upper dermal blood vessels (green staining; white arrow). The vessel endothelial cells were stained with Texas red conjugated *Ulex europaeus* to confirm colocalization (pink/red staining). In addition, the nuclei of endothelial cells and keratinocytes were counterstained with DAPI (light blue). (i) Double color IHC, showing positive staining with BCL-2 in brown and COX-2 in red around blood vessels supplying a dermal eccrine sweat gland duct (red arrows).

Direct immunofluorescence (DIF)

Our DIF demonstrated the following results: IgG (+, focal dermal perivascular); IgA (++, focal dermal perivascular and faint linear basement membrane zone staining) (**Fig. 1**); IgM (+, focal dermal perivascular and focal epidermal); IgD (+, focal dermal perivascular); IgE (-); Complement/C1q (+, focal dermal perivascular); Complement/C3 (++, focal dermal perivascular, and focal epidermal); Complement/C4 (-); Kappa light chains (+, focal dermal perivascular); Lambda light chains (+, focal dermal perivascular and focal epidermal); albumin (+, focal punctate dot epidermal stratum corneum) and fibrinogen (++++, focal dermal perivascular, perieccrine and +, faint linear BMZ). Of interest was the positivity of some type of dermal cells junctions seen with fibrinogen (++) (**see Fig. 1**).

Immunohistochemistry (IHC)

D2-40 seemed to be overexpressed on lymphatic vessels in areas such as under the blister, and around hair follicles and sweat glands. Von Willembrand factor staining showed that the blood vessels in the upper dermis were somehow compartmentalized under the blister (**Fig. 1**). CD15 was negative, as well as CD68. CD8 was positive only in small amounts around some upper dermal vessels. In contrast, CD4 was positive around several upper dermal blood vessels and around eccrine sweat gland ducts. BCL-2 followed the same pattern of positivity as CD4. COX-2 was very positive around the dermal blood vessels, as well as in other areas of the inflammatory infiltrate.

DISCUSSION

From the immunologic point of view, we have seen allergic skin reactions becoming more common in an aging population. These reactions involve interactions between multiple medications, and also reactions to soaps, detergents, hair dyes, shampoos, toothpastes and food preseratives [5-9]. Multiple simultaneous medications can alter how the body metabolizes the drugs, and also can produce alterations in the immune system. The ADR/MDAS/MDIS classifications are potentially more complex than the classic Gell and Coombs classification [13]. The Gell-Coombs classification divides drug hypersensitivity and other immune reactions into four categories, known as type I-IV reactions. In cases like ours, we see an immune response that features several units of the Gell-Coombs

classification, manifested by our COX-2, B and T lymphocytic and fibrinogen findings.

Our laboratory has extensive experience with skin biopsies from patients with multiple drug reactions. In many of our ADR/MDAS/MDIS cases, we note that a histologic hallmark is the presence of fibrinoid material in the superficial and deep dermal blood vessels [9-12]. The classic immune response is demonstrated by a significant anti-human-fibrinogen antibody component. In the current case, we noted that the dermal vessels demonstrated changes in their sizes and shapes; these changes could trigger deposits from the clotting cascade. We also demonstrated that both lymphatics and blood vessels were affected by these changes. We speculate that these vessels could become immunologically active by the drug reaction damage, and molecules such as inducible COX-2 could become active in these sites [14]. It also has been reported that many drug eruptions are due to T cell-mediated hypersensitivity reactions; these reactions can involve activation of multiple pro-inflammatory mechanisms, which would explain the varied manifestations. However, other allergic drug reaction immune components have also been described [15,16]. In this context, our CD68 staining was largely negative, suggesting that significant activation of the classic antigen presenting cell pathway did not occur. Moreover, some immunologic aspects of drug reactions challenge our classical understanding of antigen processing and presentation. New immunologic hypotheses have been proposed in their study, including whether complement and/or immunoglobulins can formally alter molecules. The altered molecules could then, in theory, act as new haptens [15,16].

In our case, we also found that some of the cells staining with CD4 and CD8 were also positive with BCL-2; BCL-2 is considered an important anti-apoptotic protein [17]. In addition, recent discoveries have also noted that BCL2 molecules are indispensable for activation and maturation of T lymphocytes following antigen presentation [17]. One group of authors identified patient factors that could increase the risk of MDIS, in a total of 25,695 patients with documented drug intolerance. Their findings demonstrated that MDIS was associated with female gender, multiple comorbidities, and previous hospital admissions. A documented allergy to penicillin did not increase the likelihood of MDIS [2]. The cases seen in our laboratory also had shown agreement with these findings, especially relative to comorbidities and advanced patient age.

In our experience, when a multiple drug allergy syndrome and/or allergic drug reaction is being evaluated the DIF usually demonstrates significant reactivity with fibrinogen and complement. We have noted significantly less reactivity with IgG and IgA, contrary to classic autoimmune blistering diseases that often demonstrate significant deposits of these immunoglobulins, Complement/C3 and fibrinogen at the basement membrane zone. Additionally, most linear IgA deposits at the BMZ seen by DIF in adults present in allergic drug reactions [17].

CONCLUSIONS

The approach and assessment of patients with possible ADRs/MDAS/MDIS involves taking a comprehensive drug allergy history, ruling out viral or other concomitant infections [18], and immediately contacting other physicians that are treating the patient to work together to decrease the dosage of and/or discontinue some medications.

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Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Case Report

Primary squamous cell carcinoma kidney: A rare case report

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ABSTRACT

Primary squamous cell carcinoma (SCC) of the kidney is a very rare clinical entity. Only a few cases have been reported in world literature. Here we report a case with renal SCC. The patient presented with flank pain, fever and vomiting. In ultrasonography, renal mass was detected and after nephrectomy followed by histopathological examination, it was diagnosed as SCC. It was associated with renal calculi and hydronephrosis. The lack of characteristic presentation like hematuria, pain and palpable mass causes delay in diagnosis results in locally advanced or metastatic disease at presentation.

Key words: Renal; Carcinoma; Squamous cell carcinoma

INTRODUCTION

Primary neoplasms of the renal collecting system are uncommon accounting for only 4% to 5% of all urothelial tumours. The transitional cell type is the most frequently diagnosed (85%-95%) followed by squamous cell carcinoma (6%-15%) [1]. Squamous cell carcinoma of the renal pelvis is a rare tumour. The incidence of this tumour is 1.4% of all renal malignancy [2]. Primary renal squamous cell carcinoma is a very rare tumour and few cases have been reported in world literature [3].

CASE REPORT

A 68 year old female patient presented with complaints of pain in the right flank pain for 2 month. Fever and chills for 15 days. Physical examination was unremarkable Routine haematology revealed. neutrophilic leucocytosis and moderate degree of normochromic normocytic anemia. Ultrasonography of abdomen and CT showed enlarged right kidney with loss of normal renal architecture and a large mass measuring (5×4.) cm. On gross examination, the kidney was larger in size measuring (11×5) cm.

Cut surface revealed (Fig. 1) Variegated G/W solid areas, thinned out cortex, dilated calyces and a large stone. Growth of tumour was infiltrating in nature.

Histopathological examination revealed features of Well differentiated invasive squamous cell carcinoma with urolithiasis (Fig. 2).

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

DISCUSSION

Squamous cell carcinoma of the renal collecting system is a rare malignancy with poor prognosis accounting for about 10 % of renal pelvic tumors and 0.5 % of all renal tumors [4,5]. They are frequently associated with long standing staghorn calculi, chronic kidney infection, hydronephrosis and analgesic abuse [5]. Hypercalcemia, leukocytosis and trombocytosis have been reported as a part of paraneoplastic syndromes in RSCC cases [6,7]. Although being nonspecific, a solid mass, hydronephrosis and calcifications are common

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Figure 1: Variegated G/W solid areas, thinned out cortex, dilated calyces and a large stone.

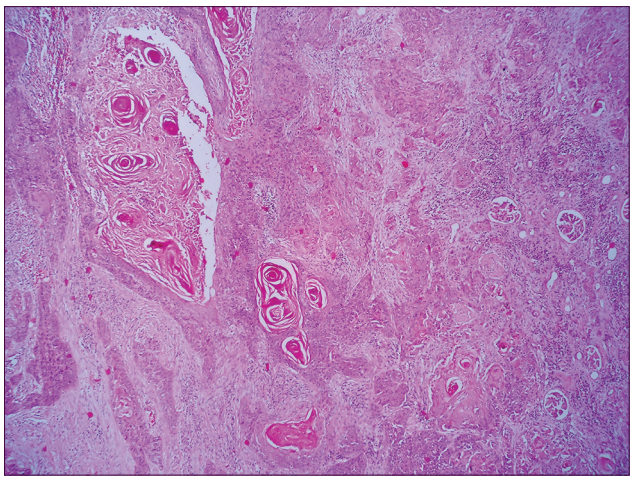

Figure 2: Shows kidney, Glomeruli and nests of squamous cell carcinoma.

radiologic findings, which may explain why the diagnosis could be missed before the histopatological examination.

In a series of 4 patients with squamous cell carcinoma of renal pelvis, mean age was 60 yrs. M: F was 3:1, right to left side ratio was 1:1 and most common presenting symptom was flank pain and haematuria. In 100% of cases, there was presence of staghorn type of renal stones [8].

Renal pelvic tumours are almost never palpable clinically, however they may block the urinary out flow and lead to palpable hydronephrosis.

Hydronephrosis is more common in renal tumour than renal pelvic ones2. In 50% of renal pelvic tumours there is precxisting or concomitant bladder urothelial tumour [9].

In general, these tumours are highly aggressive and are at high stage when detected. Most of them are histologically high grade and outcome is generally unfavorable. Extensive infiltration of the renal parenchyma and retroperitoneal soft tissues are very common [10]. In one series, 84% of the tumour were found at operation to be locally advanced or metastatic [11]. The prognosis was very poor. The current primary treatment of renal squamous cell carcinoma is nephrectomy with or without ureterectomy3 followed by radiotherapy and chemotherapy.

In the present case the patient was 68 yrs old, presented with flank pain, fever and chills. Associated etiological factors like Calculi and hydronephrosis were detected.

Grossly, infiltrative pattern of tumour mass occupied most of the renal parenchyma was seen. So the origin of the tumour was from renal parenchyma not from renal pelvis.

CONCLUSION

Squamous cell carcinoma of urothelial tract particularly renal pelvis is thought to arise through a process of metaplasia of urothelium. Various etiological factors are responsible for squamous metaplasia and subsequent carcinoma.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles

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Our Dermatology Online

Case Report

Sebaceoma of the lip originating in Fordyce's spot – A rarity

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ABSTRACT

Introduction: Sebaceomas are relatively rare benign neoplasms differentiating towards sebaceous glands. These dermal neoplasms present as solitary papule or nodule. The age of presentation is sixth to ninth decade with a female preponderance. They may be associated with underlying visceral malignancies. We present a case of sebaceoma of the lip. This case is being presented because of its rare site of occurrence. **Case report:** A seventy five year old lady presented with a single, well defined, painless and progressively enlarging flesh coloured papule over the upper lip of two months duration. No systemic signs or symptoms suggestive of visceral malignancy were present. Clinical diagnosis of papilloma was proferred. Histopathological examination of the excision biopsy revealed nests and lobules of basaloid cells and few mature sebocytes seeming arising from central hyperplastic sebaceous gland. Final diagnosis of sebaceoma was rendered. **Conclusion:** Sebaceoma is an adnexal tumour typically affecting the face and the scalp. To the best of our knowledge, this is the first case to be reported in literature in the lip. The possible origin of sebaceoma in the present case could be ectopic sebaceous glands (Fordyce's spots) which occur in increasing frequency in elderly individuals, especially in the lip.

Key word: Sebaceoma, lip, papule, Fordyce's spot, papilloma

INTRODUCTION

Sebaceomas are uncommon benign dermal lesions [1]. Troy and Ackerman coined the term sebaceoma to describe what was earlier known as sebaceous epithelioma, two decades ago [1,2]. Sebaceoma is a benign sebaceous tumour with >50% basaloid cell content. Sebaceoma is usually not suspected clinically due to its rarity [1]. Clinically, it presents as solitary papule or nodule on the face and scalp affecting elderly individuals with a female preponderance [2]. Sebaceoma may be associated with visceral malignancies. Sebaceoma is characterised by variably sized lobules composed of basaloid cells along with single or clustered mature sebaceous cells, and exhibit sebaceous ductal differentiation [3]. Fordyce's spots are ectopically located sebaceous glands, more common on the vermillion border of the upper lip [4].

We present a case of sebaceoma arising in Fordyce's spot which is a rarity.

CASE REPORT

A 75 five year old lady presented to the surgical clinic with a single asymptomatic papule over the upper lip since two months. On clinical examination, the papule was well defined, flesh coloured and painless. There was no evidence of visceral or any other malignancy. Clinical diagnosis of papilloma was given. The lesion was excised. The specimen received in the pathology department measured 0.5x0.5x0.5cm weighed <1

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gms and was dome shaped single skin covered tissue bit, cut section of which showed grey white areas. Microscopically, a well circumscribed, symmetrical dermal lesion was noted, around a central sebaceous gland which showed hyperplasia of the basaloid cells and was continuous with the tumor (Fig. 1). The tumour was composed of closely packed nests and lobules of basaloid cells with few vacuolated mature sebocytes along with duct like structures (Fig. 2). Based on all these features the final histopathological diagnosis of sebaceoma was given.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

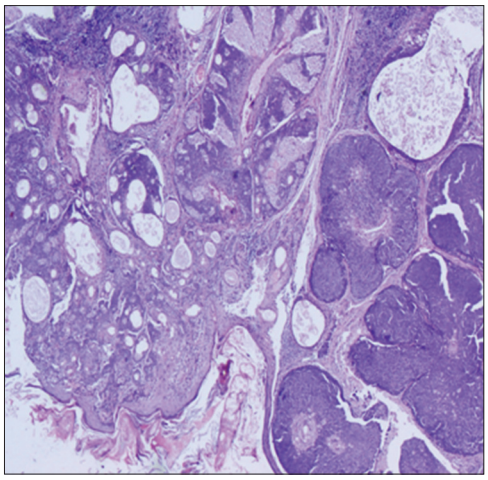

Figure 1: Nests and lobules of basaloid cells with few mature sebocytes arising from central hyperplastic sebaceous gland, (H&E, 10X).

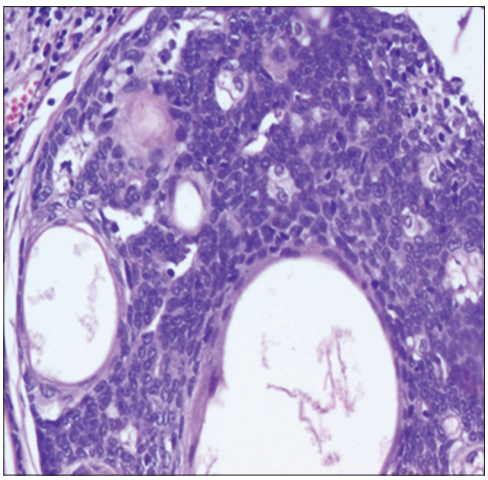

Figure 2: Basaloid cell aggregate with few vacuolated mature sebocytes around duct like structure, (H&E, 40X).

DISCUSSION

Sebaceomas are benign sebaceous neoplasms presenting as a solitary yellow to flesh-colored papule on the face and scalp of elderly females [2]. Histopathological examination is characterized by a dermal tumor consisting of multiple variably sized discrete lobules composed predominantly of basaloid cells, admixed with single or clustered mature sebaceous cells lacking an organized lobular architecture separated by dense eosinophilic connective tissue. The basaloid cells are typically small and uniform with round to oval nuclei. Absence of nuclear pleomorphism, stromal retraction, peripheral palisading and sparse mitotic activity is typical. The sebaceous cells are mature, with eosinophilic bubbly cytoplasm and scalloped nuclei. Sebaceous ductal differentiation and cyst formation are common features [2,3]. Stromal retraction and peripheral palisading are absent. Epidermal involvement is sometimes seen [2]. Foci of squamous metaplasia may rarely be seen [3].

A verrucous variant of sebaceoma has been separated from the classical sebaceoma based on the connection with a hyperplastic infundibulum in the upper portion of the lesion, prominent granular layer and basosquamous differentiation. Its architecture and cytology are however similar [3,5].

Classical sebaceoma has to be differentiated from sebaceous adenoma, sebaceous carcinoma, and basal cell carcinoma with sebaceous differentiation and trichoblastoma with sebaceous differentiation on histopathological examination [2].

Sebaceous adenomas and sebaceomas can be considered as two ends of a spectrum of benign sebaceous neoplasia, with the former being more organoid while the latter shows extensive basaloid differentiation. [1]. Histopathologically, sebaceous adenoma is a well-circumscribed dermal nodule formed of lobules of predominate central mature, bland sebaceous cells with peripherally located one or two layers of germinal basaloid epithelial cell. There is no central draining duct [3]. The central sebocytes are larger with eosinophilic bubbly cytoplasm, although indentation of the nuclei is often less prominent. There can be connection with the overlying squamous epithelium [1].

Sebaceous carcinomas show irregular lobular patterns with evidence of asymmetry, poor circumscription, and infiltrative growth pattern with preponderance of pleomorphic, basaloid cells that are arranged

in solid sheets showing cytonuclear atypia, high mitotic activity and necrosis which is not seen in sebaceoma [2,3]. Scattered sebocytes are often present within the basaloid tumour mass. Peripheral palisading and artefactual clefting are absent [3].

Some authors believe that sebaceoma is synonymous with basal cell carcinoma (BCC) with sebaceous differentiation. BCC with sebaceous differentiation is similar to classical BCC, but with a component of sebaceous differentiation [3]. Sebaceomas can be differentiated from basal cell carcinoma with sebaceous differentiation as the latter shows aggregate of follicular germinative basaloid cells composed of pleomorphic basaloid cells showing brisk mitotic activity along with distinct peripheral basal cell palisading, loose fibromucinous stroma, focal sebaceous differentiation, basaloid tumour necrosis and tumour–stroma separation artefact in formalin fixed sections [3,4].

Histopathologic features of trichoblastoma with sebaceous differentiation are those of the large nodular type of trichoblastoma characterized by large nodular aggregations composed of follicular germinative cells with palisading borders and highly fibrotic stroma, Limited differentiation toward follicular germs and rudimentary papillae is seen. In addition to these features, sebocytes and sebaceous duct-like structures are observed within the basaloid aggregations [5].

Sebaceoma can occur in association with Muir–Torre syndrome, an autosomal dominantly inherited disorder characterised by visceral malignancies (colorectal, upper gastrointestinal, endometrial and urological malignant neoplasms), tumours of sebaceous glands or keratoacanthoma [3]. But in this case no associated visceral malignancies or any other tumours were observed.

Lazar et al mentions that the common sites of sebaceoma are the head and neck area [1]. In a study by Misago et al, the most common site of sebaceoma is the head [5]. In the present case, the site of sebaceoma is the Fordyce's spot in the upper lip.

Fordyce's spots are ectopically located sebaceous glands described by Fordyce in 1896. The common sites of Fordyce's spots are lips, oral mucosa and rarely genital mucosa. JH Lee, et al reported a male predilection for Fordyce's spots, with most patients in their early or middle adulthood. Previous studies have reported female predilection for Fordyce's spots involving the elderly patients. In the present case, Fordyce's spot is involving an elderly female patient [4].

Studies have documented that the most common site of involvement of Fordyce's spots is vermilion border of the upper lip with most patients presenting with asymptomatic tiny papules and plaques. In the present case, the patient presented with asymptomatic papule over the upper lip. On histopathology examination, Fordyce's spots shows normal sebaceous gland composed of single sebaceous lobule or gland which consists of small clusters of mature sebocytes with a sebaceous duct, opening directly onto the epithelial surface which is located in the dermis or submucosa [4]. In the present case, Fordyce's spot showed features of sebaceoma which is a rarity.

CONCLUSION

- To the best of our knowledge, this is the first case of sebaceoma to be reported in literature in the lip
- The possible origin of sebaceoma in the present case could be ectopic sebaceous glands (Fordyce's spots)

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Case Report

Bilateral linear nevus comedonicus on eyelids - A rare presentation

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ABSTRACT

We describe a female child with linear comedone-like lesions over both the eyelids which histologically confirmed Naevus Comedonicus. She only partially responded to topical tretinoin cream. There was no clinically evident overt systemic association in our case.

Key words: Developmental defects; Eye lids; Hair Follicle; Nevus comedonicus; Tretinoin

INTRODUCTION

Naevus Comedonicus (NC) is an uncommon developmental defect of the pilosebaceous apparatus (or rarely, may be of the sweat duct) characterised by group of slightly elevated papules which have central, dark, firm hyperkeratosis plug resembling a comedo [1]. It was first described by Kofmann in 1895 [2]. It commonly affects scalp, face, trunk and uncommonly the genitalia [3], palms and soles [4]. Usually it is unilateral and an isolated cutaneous defect. Widespread cutaneous involvement or giant lesions are exceptional as are the systemic associations such as skeletal defects, cataract, cerebral anomalies [1,5,6] and arterio-hepatic dysplasias (Alagille syndrome) [7].

CASE REPORT

An 8-years old female child was brought by her parents for asymptomatic "string of black dots" on both of her eyelids noticed since six months which were slowly progressive.

She was otherwise healthy. There was no history of similar eruptions in the other family members. Her past health was good. There was no preceding history of trauma or eyelid surgery. There is no history of convulsions, headache or jaundice. There is no history of usage of mascara, eyeshadows or other cosmetics. We also specifically asked about the history of topical application of steroid creams.

On examination, there were multiple, firm, pin-head sized papules with a keratotic, pigmented centre with linear configuration on the upper eyelids starting 1cm supero-medially to the medial canthus, extending upto the medial third of the length of upper eyelids on each side. (Fig. 1). The lesions had more or less, a symmetrical distribution. These were non-tender and not adherent to the deeper structures. There were no lesions on other areas of skin. Her gait and posture was normal. Her complete ocular, cutaneous and systemic examinations were normal.

Skin biopsy revealed multiple dilated follicular infundibuli below the epidermis, surrounded by dense mononuclear inflammatory infiltrate. Individual dilated follicular opening was filled with eosinophilic, keratinous material (Fig. 2). Overlying epidermis was unaffected. Her baseline work up including blood counts, sugar, urinalysis and liver function tests was normal. She was prescribed topical tretinoin cream

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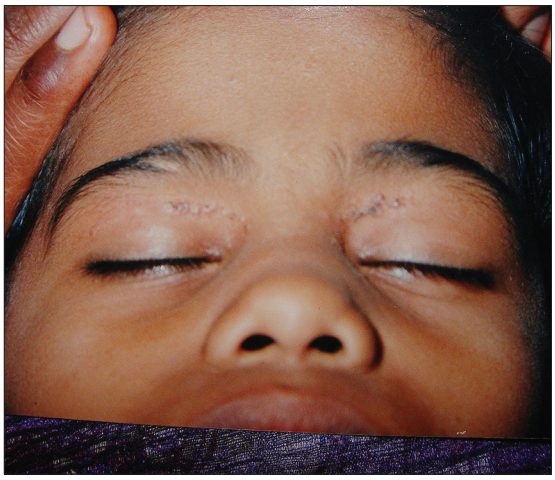

Figure 1: Bilateral, linear papules with central, black comedones on upper eyelids.

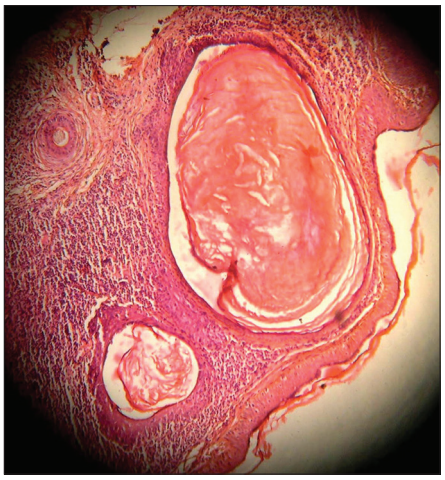

Figure 2: Skin Biopsy (X10): Individual follicular opening filled with eosinophilic keratinous material surrounded by dense mononuclear infiltrate.

0.025% twice daily, which resulted in partial flattening of comedones at the end of three weeks. She was further lost to follow-up.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

DISCUSSION

NC usually has unilateral, grouped or linear configuration. Symmetrically bilateral linear affection is rare [8]. NC may result into cosmetic disfigurement especially when the lesions are large and are on the exposed areas such as face. NC may be confused at times with the other linear eruptions such as epidermal naevus (which may actually be associated with NC), linear adnexal tumours, lichen planus, porokeratosis, lichen striatus, lipoid proteinosis or tattoo reactions. However, a distinctive clinical appearance of linear papules with central dark, keratin plug is virtually diagnostic. In atypical cases, the diagnosis is resolved on histopathogical examination. The lesions of NC may occasionally have acute inflammatory component with or without secondary infection, may then result into scarring.

NC is usually treated with topical retinoic acid. However, it may be fairly resistant to the topical therapy [8-11]. Topical treatments with keratolytics including salicylic, lactic and tartaric acids [8], pore-strip packs [10], Ammonium lactate solution [9] and Tacalcitol [8] dermabrasion; manual extraction of the comedones [11] and surgical excision [11] with or without plastic reconstruction are the other modalities of the treatment. Tissue expansion has been tried in one patient with giant NC [11].

Ophthalmologists may be the first physician to see cutaneous problems such as NC in their patients. It is, therefore, prudent to know of the conditions which are diagnostic by their morphological appearance so as to facilitate early clinical diagnosis and associated serious systemic defects, if any. The patient of NC may be informed about possible complications like inflammation, abscess formation which may result in scar formation and likely onset of cataracts in young age [12].

To our best knowledge, only one case of bilateral NC on eyelids is reported on pubmed in a 79-yrs old male patient [13]. The cause in this patient was unclear. In contrast, our case of NC in a child truely represents a developmental defect and therefore, is certainly unique.

In conclusion, bilateral linear NC on the upper eyelids is an extremely rare presentation. It causes significant cosmetic concern, which may be associated with anxiety of the parents of the affected children.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Case Report

Giant primary melanoma of the skin arising on the left foot

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ABSTRACT

Melanoma of the skin usually does not reach a greater dimension. The authors describe unique case of the patient with a huge (giant) primary cutaneous melanoma. 56-year-old man was diagnosed to have melanoma arising on the left foot. Partial surgical amputation with a complete tumor removal was done. Melanoma measured 8 x 4 cm and exhibited massive ulceration, Breslow's thickness 15 mm, Clark's level V, high mitotic and proliferative activity. A patient was classified as stage III B. After surgery, he was relegated to the oncological dispensary health care and took part additional clinical investigations. Finally, PET/CT scan revealed pulmonary and regional lymph node metastases. Therefore, paliative chemotherapy was started. Huge skin melanomas like this are very rarely diagnosed in clinical practice and they are almost always the result of long-term neglect of growing lesion. Currently, it is not answered, whether certain specific factors predispose some melanomas to grow such enormously.

Key words: Malignant melanoma; Biological behaviour; Foot

INTRODUCTION

Melanoma of the skin is one of the most aggressive and prognostically the most unfavorable malignancies in humans. Currently, it is extensively studied oncological entity, mostly due to its dramatically increasing incidence all over the world [1,2]. Cutaneous melanoma usually does not reach a greater dimension. In recent analysis, Seidenari et al. [3] found a mean (microscopically verified) horizontal diameter 1.06 cm. In contrast to skin carcinomas, pathological TNM (Tumor, Nodes, Metastasis) classification of cutaneous melanoma [4] does not include horizontal, but vertical dimension (Breslow's thickness), which is prognostically much more important. Considering an aggressive biological behaviour and high metastatic potential of melanoma there is a small chance, it could grow to the greater size. Moreover, some melanomas may also completely regress, what is usually accompanied by metastases [5]. Therefore, a presence of metastasis of malignant melanoma with unknown primary origo is not unique finding in clinical practice [6,7]. Anyway, medical literature has sporadically documented case reports of huge (so-called giant) melanomas, although their cut-off limit has not been consensually defined. Some papers [8,9] have considered giant melanomas those, diameter of which exceed 10 cm. However, several authors [10-15] have described cases of giant melanomas not reaching this size. Herein, we present a patient with a huge (giant) cutaneous melanoma arising on the foot.

CASE REPORT

A 56-year-old man presented with a huge solid tumor involving the toes and instep of the left foot. He admitted the lesion was long standing and originally started to arise on the skin between the fourth and fifth toe almost 3 years ago. Initially, it was asymptomatic and painless. During the last months, however, tumor has began to grow rapidly, progressively increased in size and was accompanied by extensive ulceration, intermittent bleeding and pain. Therefore, he had to see a medical attention and visit an ambulatory surgeon

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(March, 2015), who performed a probatory incision. After getting a histologically proven diagnosis, he reffered him to the hospital. Subsequently, a patient was admitted to the Department of Surgery, where a partial amputation of the foot was preferentially planned. Routine preoperative clinical investigations were done. Among them, none revealed an evidence of distant metastases. X-ray imaging of affected leg did not confirm a tumor infiltration of the bone tissue. A chest X-ray showed no obvious pathology in the pulmonary parenchyma. The standard laboratory parameters were (except slightly elevated serum glucose) within normal limits.

A partial surgical amputation and exarticulation of the foot was done, biopsy specimen was immediately fixed in formalin and sent for definitive histopathological examination. A received biopsy sample consisted of a part of the foot with three toes. On macroscopic examination, the toes, interdigital and metatarsal regions were massively infiltrated by gray-brownish tumor mass. The fourth toe was affected most severely, it was twisted and deformated. A tumor measured 8 x 4 cm in greatest dimension, it was irregularly shaped, protuberant and ulcerated. All nails were intact without tumor changes. There were four colored surgical stitches fixed on the basis of the sample to better indicate resection margins.

Histopathology revealed a primary malignant melanoma predominantly composed of atypical spindle-shaped cells (Figure 1). It was markedly ulcerated with a purulent detritus on the ulcer basis. Breslow's thickness was at least 15 mm and Clark's level V. Immunohistochemically, it was strongly positive for melan A (Figure 2), HMB-45, S-100 protein and negative for polyclonal cytokeratins. Disseminated deposits of melanin pigment were present in the entire sections. Mitotic activity reached 25 mitoses per 1 mm^2. Proliferative activity (Ki-67 index) was also high (cca 40%). Neither lymphovascular nor perineural tumor invasion was found in excised sections. In one region, small tumor satellite occurred 8 mm from the main tumor mass. There was scattered intratumorous lymphocyte infiltration (TIL +). Because of advanced stage of the lesion, it was not possible to clearly determine an original histological type of melanoma. However, in one histological section, a small focus of residual atypical lentiginous melanocyte proliferation was detected (Figure 3), which might suggest (even in the contex with anatomical site) an acral lentiginous

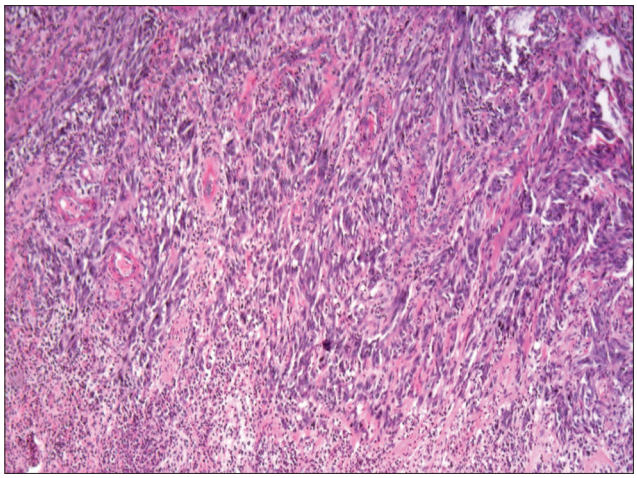

Figure 1: Melanoma predominantly composed of atypical spindle-shaped cells accompanied by secondary desmoplastic reaction and leukocyte infiltration (hematoxylin & eosin staning, original magnification, 100x).

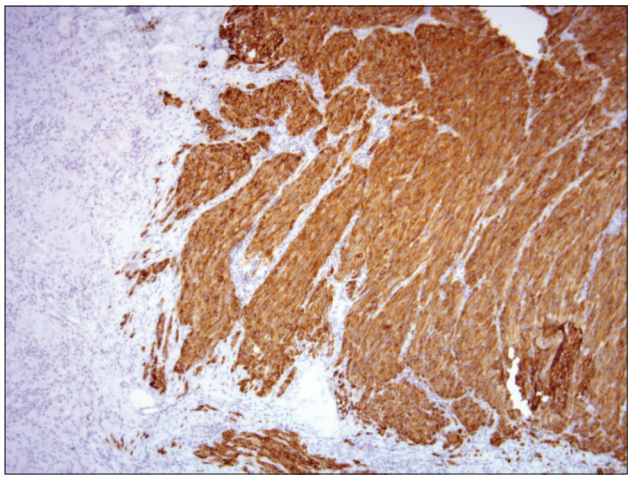

Figure 2: Strong immunopositivity for melan A. (monoclonal mouse antibody against melan A, clone A 103, DAKO, original magnification, 100x).

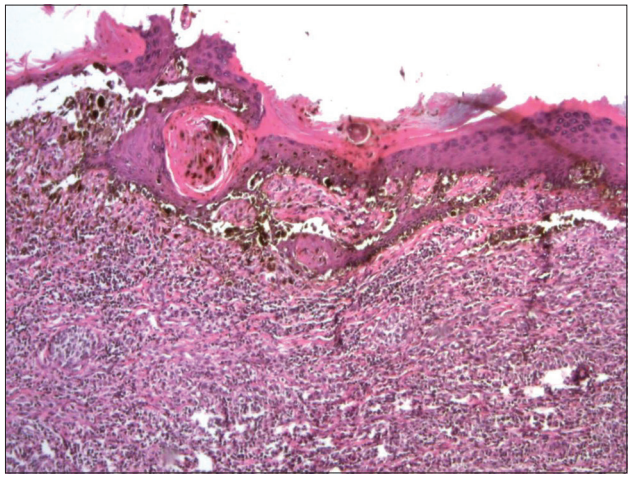

Figure 3: A focus of atypical lentiginous melanocytic proliferation at the periphery of melanoma. (hematoxylin & eosin staning, original magnification, 100x).

type. According to the UICC (Union for International Cancer Control) TNM staging system [4], the patient was classified as stage III B (pT4b,N2c,MX). All resection margins marked with surgical stitches were free of tumor and a minimum of 15-mm clearance was achieved. Surrounding skin tissue did not exhibit a solar damage.

Shortly after surgery, CT (computed tomography) scan of thoracic cavity and mediastinum was realized. It confirmed multiple intraparenchymatous nodular lesions in both lungs, some of which unsharply demarcated. The largest one measured 13 x 11 mm and grew out the pleural surface. Thus, melanoma metastases or mycotic infection were considered in differential diagnosis. Furthermore, enlarged lymph node (15 x 11 mm) in the right pulmonary hilus was visible. CT imaging of the abdomen and pelvic cavity did not reveal obvious pathological changes in the visceral organs, either intraabdominal lymphadenopathy. Similarly, both groins were without evident enlargement of the lymph nodes. A postoperative course was uneventful, the patient left the hospital and consequently, he was relegated to the oncologic dispensary health care. An oncologist recommended further examinations to elucidate the origin of pathologic nodules in the lungs. Bronchoscopy did not confirm persuasive pathological changes within the bronchial tree. An aspiration lavage was done and sent for cytological analysis, but no tumor cells nor mycotic microorganisms were found in the fluid. Therefore, the patient underwent PET/CT (positron emission tomography – computed tomography) examination, that showed an increased glucose metabolism (FDG, 2-fluoro-2-deoxy-D-glucose) within the nodules in both lungs, as well as in the left groin. Based on clinically very suspcicious evidence of metastases, a paliative chemotherapy was started. At the time this report was written, our patient had underwent monochemotherapy with dacarbazine. Additionally, there was not detected BRAF gene mutation in tumor tissue.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

DISCUSSION

Huge skin melanomas like this are very rarely diagnosed in routine clinical practice, mainly because the patients tend to come for treatment early before the tumor enlarges. Therefore, they are almost always the result of long-term neglect of growing lesion. An overview of the cases that have been published until now, including our present case report are summarized in Table 1. As may be seen, they usually occur in the middle or elderly age with approximatelly equal sex distribution. However, sometimes have been observed in young people up to 35 years of age. In the most documented cases they were localized on the trunk [8,10,13,16-18], but involved also other sites, such as scalp [9,19], arm [20,21], shoulder [22], thumb [11,12], and eyelid [14]. To the best of our knowledge, there has been published only one paper describing giant cutaneous melanoma involving the

Table 1: Summary of giant cutaneous melanomas, that have been published until now, including our present case report

Literature	Age	Sex	Location	The largest dimension	Breslow's depth	MTS
Ching & Gould [9]	70	Male	Scalp	14.5 cm	18 mm	+
Tseng et al. [20]	88	Male	Left arm	10 cm	31 mm	+
Tseng et al. [20]	63	Male	Right arm	23 cm	75 mm	+
Kim et al. [11]	56	Female	Left thumb	7 cm	> 4 mm	+
Eisen et al. [13]	47	Male	Back	9 cm	40 mm	+
Grisham [8]	45	Female	Back	13 cm	55 mm	+
Harting et al. [16]	29	Male	Back	25 cm	54 mm	+
Kruijff et al. [10]	56	Female	Back	8 cm	48 mm	+
Panajovic et al. [19]	57	Male	Scalp	12 cm	100 mm	a/n
Del Boz et al. [21]	29	Female	Left arm	20 cm	70 mm	+
Zeebregst et al. [12]	74	Female	Left thumb	7 cm	No mention	+
Pai et al. [14]	53	Male	Left eyelid	5 cm	45 mm	+
Di Meo et al. [17]	60	Female	Abdomen	18 cm	40 mm	+
De Giorgi et al. [18]	45	Female	Abdomen	16 cm	0.45 mm	a/n
Zou & Lam [15]	35	Female	Left leg	8 cm	No mention	a/n
Zou & Lam [15]	72	Male	Right leg	8 cm	No mention	+
Liu et al. [12]	60	Female	Left shoulder	15 cm	> 100 mm	+
Our case	56	Male	Left leg	8 cm	15 mm	+

MTS: Metastases in regional lymph nodes or distant organs, confirmed either clinically or histologically. +: Present. a/n: Absent or not mentioned

foot. Zou and Lam [15] briefly introduced two patients with nodular malignant melanoma located on the sole, both reaching the largest diameter of 8 cm.

According to the literature data [17], a majority of giant melanomas of the skin grew up primarily (*de novo*) without association with precursor melanocytic lesion. Occasionally, there have been described the cases arising from congenital [16] or acquired pigmented melanocytic nevus [14,21]. In our case, it was likely to develop *de novo*, because a patient did not state a previous pigmented skin lesion on the incriminated region. In advanced melanomas, it is very difficult (and usually impossible) to determine an original histological type. Therefore, several case reports of giant melanomas have not precisely classified an histological type. We suppose that our patient could originally have acral lentiginous melanoma, which is the most typical for this anatomical site. In addition, microscopic features of atypical junctional lentiginous melanocytic proliferation were detected at the periphery of the tumor mass. Anyway, in such advanced stages, an exact histological typing does not play a prognostical significance. It is not surprising, a majority of reported giant melanomas were accompanied by metastases in regional lymph nodes or distant organs, whether detected within primary tumor diagnosis, or during further investigations. Except two case reports [18,19], all others corresponded to the clinical stage III or IV and the patients were usually subsequently treated with paliative modalities (i.e. chemotherapy or α-interferon immunotherapy). However, in such cases, cure effect, as well as disease outcome and survival are hardly predictable.

In conclusion, it is not clear, whether melanomas, which are capable to progress in such large dimension exhibit different biological phenotype from that of "usual" melanoma of equivalent thickness. Further, we have insufficient knowledge, whether certain specific factors predispose some melanomas to grow such enormously. Some melanoma patients with much more smaller primary lesions behave poorly with early distant metastasis, while others develop very extensive primary tumors without or with only late metastases. In addition, some melanomas may fully regress and this phenomenon is usually accompanied by metastatic spread. More light remains to be shed on the pathobiology, malignant progression and pathways of metastasis in this neoplasia, as they are still only partially understood.

ACKNOWLEDGEMENTS

The authors wish to thank Dr. Kuchár Pavol (surgeon) for assistance concerning the clinical information and Dr. Doboszová Jana (pathologist) for technical assistance.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Case Report

An unusual misleading multiple nodules on the extremities – a case report

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ABSTRACT

Benign fibrous histiocytoma is a common benign dermal neoplasm mainly composed of a mixture of fibroblastic and histiocytic cells. It is also known as dermatofibroma, sclerosing haemangioma, or nodular subepidermal fibrosis. There are many histological variants of fibrous histiocytoma such as aneurysmal, cellular, epitheloid, atypical, keloidal and palisading subtypes. The diagnosis of cutaneous benign fibrous histiocytoma is generally easy; however, rare variants may be difficult to identify and the diagnosis can only be confirmed after histopathological examination and by immunohistochemical staining. We report a case of 52 yr old woman with asymptomatic multiple pigmented raised skin lesions over both lower limb and right arm which was histopathologically diagnosed as atypical benign fibrous histiocytoma(ABFH) involving subcutaneous tissue and the immunohistochemical staining was done and the treatment was proceeded with complete wide surgical excision due to its higher tendency to recur locally and the patient was advised for regular follow up.

Key words: Dermatofibroma, Atypical fibrous histiocytoma, fibrous histiocytoma.

INTRODUCTION

Dermatofibroma (DF), originally described by Unna [1] in 1894 [2], is a common benign dermal neoplasm formed by proliferation of histiocytes and fibroblast, so also called as benign fibrous histiocytoma) [3]. They usually present with solitary or multiple, flesh coloured to brown, firm, asymptomatic or mildly tender papule, plaque or nodule of 1cm in diameter with tethering of the overlying epidermis to the underlying lesion. On lateral compression of lesion they show a dimpling over the surface known as "dimple sign" or "button holing". Most common on extremities, especially the lower limbs and often seen in women [4]. There are numerous clinicopathological variants such as cellular, aneurysmal, atypical, epitheloid, atrophic, lichenoid, keloidal and ulcerative fibrous histiocytoma [5-7] out of which our case presented with a rare group of atypical benign fibrous histiocytoma.

CASE REPORT

A 52 year old lady came with c/o asymptomatic multiple pigmented raised skin lesions over her right arm and both lower limbs since 5 years. Initially started as a single lesion over her left leg which then gradually increased in number and started to involve the other leg & Rt arm over the past 1 year. H/o topical application of clobetasol with salicylic acid cream over the lesion for 3 weeks present but did not showed any change in lesion except the surrounding skin hypo pigmentation. The patient had no h/o trauma prior or any other significant medical problem.

On examination, multiple well defined oval hyperpigmented nodules of varying size of 2 cm× 1.5 cm present over anterior aspect of right arm and medial as well as lateral aspect of both lower limbs. On palpation, it was firm, non tender, mobile with tethering of skin to underlying structure and dimpling was present on lateral pressure of the lesion (Figs. 1 and 2).

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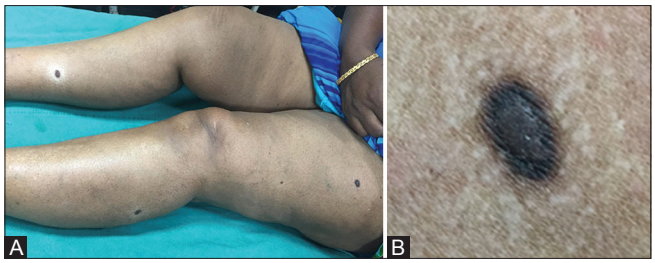

Figure 1: (a). Multiple well defined hyperpigmented oval nodules over both lower limbs (b). A closer view of nodule over left upper thigh.

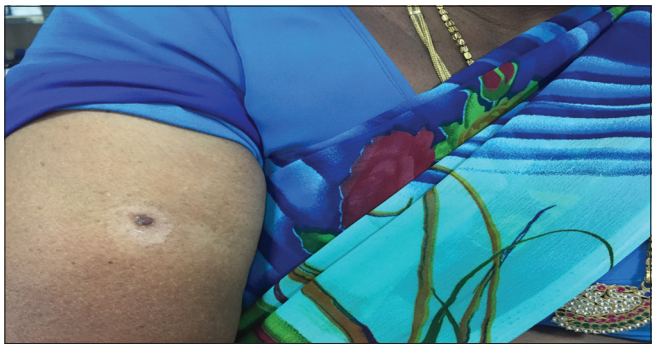

Figure 2: A well defined hyperpigmented nodule over the right arm

Routine investigations like CBC, LFT, RFT, Urinalysis, serology was within normal limit. We arrived into a differential diagnosis of dermatofibroma, malignant melanoma, atypical dysplastic nevus, angioma and fibrous xanthogranuloma.

Although the initial clinical diagnosis was dermatofibroma, establishing a conclusive diagnosis was difficult initially. A wide local excisional biopsy of single lesion was done which revealed atypical benign fibrous histiocytoma with feature of mild irregular acanthosis &dermis showed ill defined lesion comprised of spindle cells which exhibit mild atypia and giant cells. These cells were surrounded by collagen bundles and the lesions were extending upto subcutaneous tissue (Figs. 3 and 4) and later immunohistochemical staining was done which showed CD 34 negative and positive for Factor XIIIa and vimentin.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

DISCUSSION

Atypical benign fibrous histiocytoma (ABFH) was first described by Fukamizu et al in 1983. It is also known as atypical fibrous histiocytoma or dermatofibroma with monster cell [8]. It is due to proliferation of fibroblastic

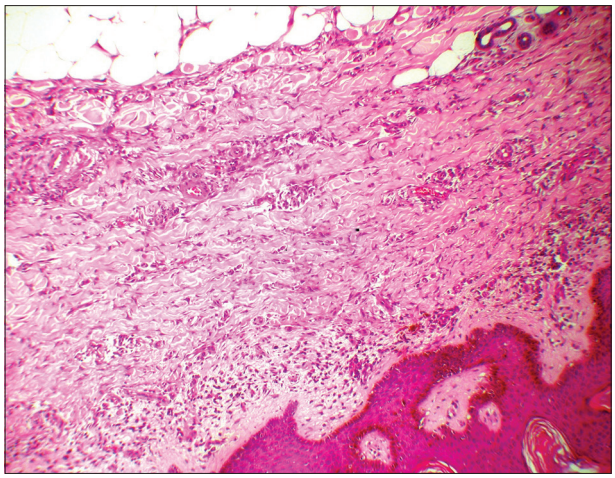

Figure 3: Tumour shows irregular acanthosis with a large area of illdefined lesion composed of spindle cells with atypia & giant cells with focal extension into subcutaneous tissue.

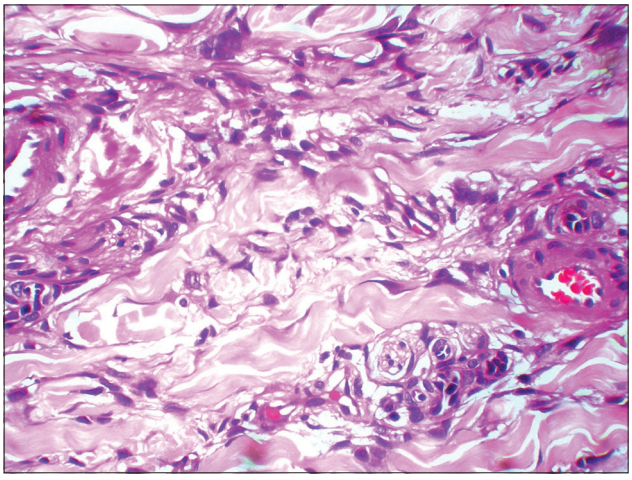

Figure 4: Dermis shows ill defined lesion comprised of spindle cells with atypia and giant cells which are surrounded by collagen bundles

and histiocytic cells, occurs frequently in the dermis. A deep penetrating type involving subcutaneous tissue is usually rare comprising less than 2% of all FH [9]. They usually occurs as a nodule on the lower extremities, especially in adults with male predominance [7,10].

Histopathological variants like cellular, atypical, aneurysmal DF as well as dermatofibroma arising on the face, subcutaneous and deep soft tissues have an increased risk for local recurrence (upto 20%) and have been reported to metastasize to the lymph node and lungs and even caused death in some patients [11-13]. Out of all these AFH alone tends to show a higher recurrence rate of 14% than ordinary fibrous histiocytoma (2-3%) and even rare metastases have been described [7,11,14,15].

Interestingly, our case reported herein was clinically characterized by the presence of multiple lesions,

involving both upper as well as lower extremities, histopathologically revealed an unusual atypical variant of DF with invasion of atypical cells into the subcutaneous tissue. Immunohistochemical staining showed positivity for factor XIIIa and vimentin and negativity for CD34 thus differentiates ABFH from dermatofibrosarcoma protuberans [9,15].

As per the treatment modality, all the tumors was surgically excised completely with clear margins and the patient was adviced for regular follow up.

CONCLUSION

ABFH is a poorly recognized variant of fibrous histiocytoma which usually lacks a clear cut predictive morphological pattern. Recognition of this variant is important because it has high potential for local recurrence and metastasis and so, a complete surgical excision and regular follow up is recommended in all cases after the final diagnosis.

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Our Dermatology Online

Case Report

Giant cervico-facial mycetoma caused by Streptomyces somaliensis in a 14-year-old girl

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ABSTRACT

Mycetomas are inflammatory pseudo-tumors in multiple locations that affect the skin, the subcutaneous tissues and, sometimes, the bones. Their treatment depends on the type of parasite. Fungal mycetomas, also called eumycetomas, are treated mainly through surgery, while actinomycotic mycetomas are treated primarily with drugs. We report here the case of a 14-year-old girl afflicted with a giant cervico-facial mycetoma. The patient was born to poor and illiterate parents in a rural area of the Diffa province, at 1500 km from the capital city of Niamey. Histological examination of a biopsy specimen allowed a diagnosis of actinomycetoma due to *Streptomyces somaliensis*. The patient showed a remarkable sensitivity to ketoconazole, but she ultimately died due to a lack of sufficient medication.

Key words: Cervico-facial mycetoma, *Streptomyces somaliensis*, ketoconazole, Niger

INTRODUCTION

The mycetoma is a pathological condition in which fungal or actinomycotic exogenous agents produce parasitic buds [1]. It is a chronic infectious disease that mostly affects the foot and, more rarely, other parts of the body [2-4]. The rural population is most exposed to the infection due to small injuries sustained in contact with thorny shrubs harboring the infectious agents [3,5]. Gill, was first to recognize mycetoma as a disease entity in 1842 when he worked at a dispensary in the southern province of Madura [6]. The treatment is primarily surgical in the case of fungal mycetomas and medicinal in the case of actinomycotic mycetomas. Both types of treatments are problematic, having rather variable results [5,6]. This paper reports on the case of a 14-year-old girl afflicted with a giant cervico-facial mycetoma due to *Streptomyces somaliensis*. Despite a favorable development was observed, the disease, unfortunately, lead to the death of the patient after the parents, because of poverty and neglect, interrupted the treatment.

CASE REPORT

The patient, a girl aged 14, was born in a rural area of Niger at 1500 km to the east from Niamey, the capital city, to poor and illiterate parents. The parents brought her to medical attention after trying during four years an unspecified traditional medication. When we examined her in April 2011, she had a giant swelling at the cervico-facial area that has deformed the right side of the face, with sores and crusted lesions on the eyelids. This swelling reached the posterior cervical area and formed a block with an occipital swelling. The whole affected area was covered with watery sores that emit yellowish buds. A yellowish purulence was oozing from the right ear (Fig. 1). The remainder of the physical examination turned out nothing remarkable. Radiography of the head made from the face, showed diffuse sclerosis and osteoporosis on the skull and the right maxillary in particular (Fig. 2). An anatomical and pathological examination was carried out on a tissue sample after a hematoxylin and eosin (H&E) staining. This examination showed in a PNN

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Figure 1: A view of the swelling before treatment: from the face (A), the left profile (B), the right profile (C) and from the back (D).

infiltrate several greyish buds of actinomycetes from the *Streptomyces somaliensis* species, thus confirming the actinomycotic mycetoma diagnosis (Fig. 3). The pre-therapeutic assessment (notably transaminases, NFS, creatinine and glucose tests) revealed nothing remarkable. In May 2011 the treatment was started based on ketoconazole at 200 mg per day. The girl's father declined hospitalization, opting for an outpatient regime. We examined the patient after three months and noticed an improvement, despite an array of malnutrition symptoms. In particular, most lesions have dried and healed (Fig. 4). The same tests as in the pre-therapeutic assessment were conducted but turned out nothing remarkable. The ketoconazole treatment was renewed at 200 mg per day and an examination appointment was set for December 2011. Unfortunately, the patient died in June 2012, after six months without medication.

DISCUSSION

Mycetomas mostly affect the feet or other parts of the lower limbs. However, the manifestations of the disease on other body parts are the result of a direct inoculation or are due to parasite metastasis [7]. People are generally ignorant of contamination risks following injuries against shrubs, and this is the case for our patient, who was aged 14, in spite of the fact that she and her parents live in an area known to have a lot of thorny shrubs [5,6]. The clinical diagnosis of mycetomas is usually easy, but other chronic conditions must be ruled out, such as other deep fungal infections, tuberculosis and benign or malignant tumors [6,8,9]. Generally, people in Niger have such confidence in traditional treatments that when these fail, they tend to think that medical treatments, too, will fail. As a result, they do not seek treatment or come in only when it is very late [3]. In the case of our patient, the parents waited four years before coming to the hospital and this led to the development of a tumorous swelling with multiple sores, as is characteristic of the actinomycotic forms of the disease [10,11]. Mycetomas are prevalent

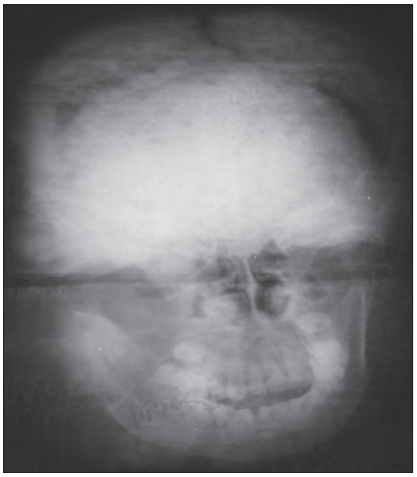

Figure 2: Diffuse sclerosis and osteoporosis (noticeable despite the poor picture quality) in the entire head skeleton (skull and jaws).

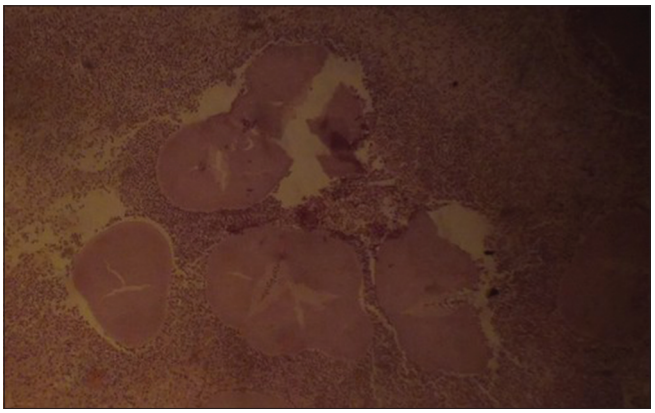

Figure 3: H&E stain (x40) of a PNN infiltrate: Numerous greyish buds *(Streptomyces somaliensis* organisms*)*.

in Niger, both in their fungal and actinomycotic forms [5]. Pathological and anatomical examinations are necessary in order to specify the pathological agent, as noted by some authors [6,12-14]. In our case, these examinations allowed the identification of *Streptomyces somaliensis* organisms. *Streptomyces Somaliensis* appears to be equally prevalent in Niger, Mauritania and Yemen, when compared with other actinomycetes [2,7], but it is absent in other areas despite the important variations in their geography [4]. Just as is the case with our patient,

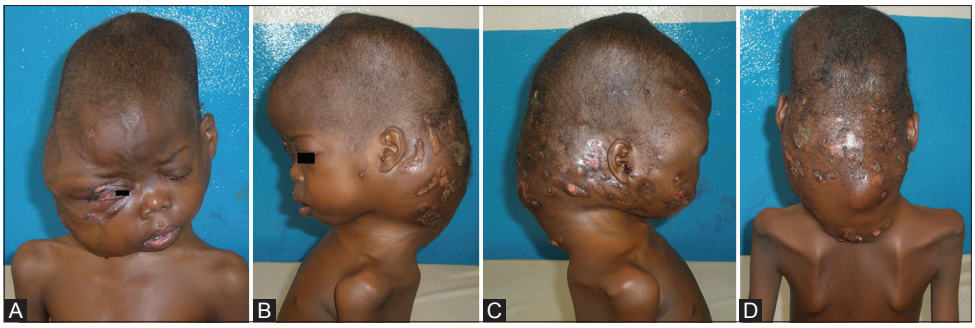

Figure 4: Improvement after a 3-month treatment based on ketoconazole (drying and healing of the lesions) from 4 perspectives (A : Face, B : Left profile, C : Right profile, D : Back).

Streptomyces Somaliensis seems to affect areas other than the foot, in particular the facial and cervical areas [15,17]. Its parasitic effects on the bone are, as is the case with our patient, sclerosis and osteocondensation [15,16]. *Streptomyces Somaliensis*, just like *Nocardia farcinita*, has become resistant to certain antibiotics, which explains the failure of many treatment protocols [18,19]. Despite the fact that the imidazole family of compounds is not indicated for the treatment of actinomycetomas, a complete remission with ketoconazole was observed on a similar case of head and neck mycetoma caused by *Streptomyces somaliensis* [15]. Similarly, a favorable development was observed in the case of a knee actinomycetoma due to the *Nocardia otitidiscaviarum* after 4 months of treatment, as reported from the Comoros [20]. We believe that in our case, the favorable course observed after 3 months of treatment could have also lead to a complete recovery with the ketoconazole, had the treatment been followed for at least one year. Unfortunately, poverty and neglect decided on the case, and the patient died after six months without drugs.

CONCLUSION

We believe that at least a one-year antifongic treatment based on ketoconazole would be effective, no matter the size of a tumor. Given the endemic nature of the pathological fungus in mycetomas, an awareness campaign is necessary so that newly affected people seek early medical treatment. The imidazole family of compounds must also be tested in further studies for minimal one year, in order to confirm their effectiveness on actinomycotic mycetomas, especially those due to *Streptomyces somaliensi* organisms.

Statement of Informed Consent

Informed consent was obtained from the patient's father for being included in the study.

Written informed consent was obtained from the patient's father for publication of this article and any accompanying images.

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Our Dermatology Online

Case Report

A case of adult onset disseminated juvenile xanthogranuloma

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ABSTRACT

Juvenile xanthogranuloma (JX) is a rare, benign, non-Langerhans histiocytic proliferative disease that etiology is unknown. It is usually seen in children and infants. JX in adult is very rare. A 41-year-old female patient was admitted to our clinic with papules on her face, torso and extremities. A few lesions had occured 3 years ago on her face, they disseminated all over her body after having a traffic accident one year ago which for she had operations and she also concurrently was diagnosed asdiabetes mellitus (DM). Based on clinical and histopathological findings, the diagnosis of JX was made. There is no systemic involvement of JX detected. JX seen in adults are very rare and usually associated with hematological malignancy. The present case is a rare adult onset disseminated JX case without malignancy association and representative for the opinion that trauma and DM may be triggering factors.

Key words: Non-Langerhans histiocytosis; Juvenile xanthogranuloma; Adult

INTRODUCTION

Juvenile xanthogranuloma (JX) is a rare, non-Langerhans histiocytic proliferative disease, generally seen in childhood [1]. JX in adult is very rare and in some of adult cases, there were reported concurrent hematological malignancies [2-4]. In the present case, a 41-year-old female patient who has disseminated JX lesions with adult onset is presented.

CASE REPORT

41 year old female patient was admitted to our clinic with papules all over her body. Lesions started 3 years ago on her face and they remained stabile until having a traffic accident 1 year ago. After accident and subsequently operation, the lesions began spreading on whole body. In her personal history, there were a surgical operation because of the accident and concurrently diagnosis of diabetes mellitus. In her family history there was nothing important. In dermatological examination, multiple yellowish-pink coloured, some dome-shaped, indurated papules were detected on her head, hairy skin, external auditory canal, pubis and upper torso (Figs 1 - 3). Routine laboratory tests were normal. Histopathological examination of the lesions showed histiocytic infiltration and multinuclear giant cells and actin, desmin, S100 and factor XIIIa stains were found negative whereas CD68 was positive (Figs 4a - d). Based on clinical and histopathological findings, patient was diagnosed as JX. There were no pathological findings in ocular and respiratory examinations of the patient. The patient was followed because the lesions has possibility of spontaneous resolution. But, 6 months later, there were no regression in control examination.

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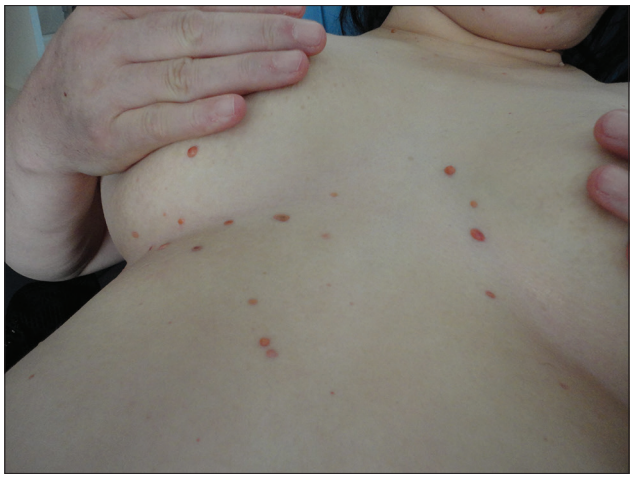

Figure 1: Multiple yellowish-pink coloured, some dome-shaped, indurated papules on the torso.

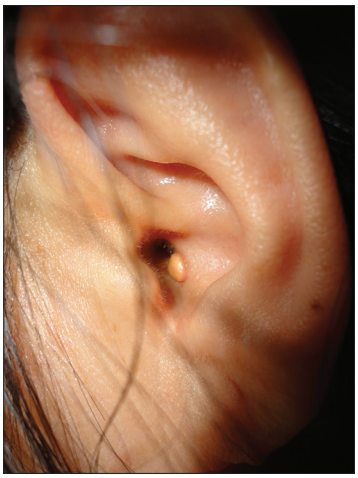

Figure 3: Papule on external auditory canal.

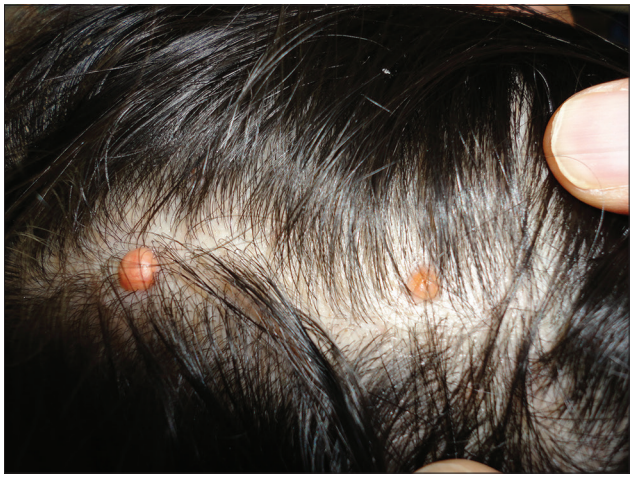

Figure 2: Multiple yellowish-pink coloured papules on the scalp.

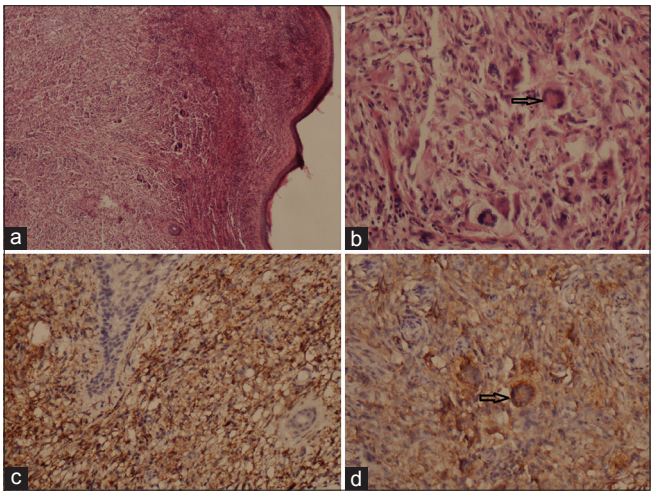

Figure 4: a) Histiocytic infiltration and multinuclear giant cells (H&E x40), b) Closer apperance arrow: Touton type giant cell (H&E x200), c) CD68 positivity of histiocyts (CD68x 40), d) Closer apperance arrow: Touton type giant cell (CD68x200).

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

DISCUSSION

JX is an uncommon non-langerhans histiocytic proliferative disease which %75 of cases were diagnosed in the first year of life. It is rarely seen in adults and the lesions generally seen as solitary and resisted to spontaneous regression [1]. Some of the adult cases with disseminated lesions were associated hematologic malignancy, mostly leukemias [2-4]. Our case is adult onset disseminated JX case without malignancy association.

The etiology of JX is still unknown however, it is suggested that it might be a reactive disorder responding to any traumatic or infectious stimulus [1]. Our case has trauma history and diabetes mellitus and dissemination of the lesions was simultaneously with trauma diagnosis of diabetes mellitus. So, we think that trauma and/or diabetes mellitus may be potential triggering factors for JX.

In many cases, lesions are limited to the skin and benign characterized [1]. Extracutaneous infiltration was reported in %5 of cases. The most common site are eye and lung. Early recognition is important to avoid complications [5]. In our case, there was no pathological findings in physical or radiological examinations that indicates any systemic involvement.

Associations between JX and neurofibromatosis type 1 (NF1), juvenile myelomonocytic leukemia (JML), urticaria pigmentosa, Niemann-Pick disease and

diabetes mellitus were reported [6]. Our case has also diabetes mellitus.

Differential diagnosis firstly includes non Langerhans-Langerhans cell histiocytosis. The other differential diagnoses are xanthomas, molluscum contagiosum, neurofibroma, spitz nevus. Diagnosis of JX is done by clinical, histopathological and immunohistochemical examinations [7,8]. In histological examination of JX lesions, characterized Touton cells with ring shaped nucleus, foamy cytoplasma are seen and S100, CD1a stains are negative whereas CD68 stain is positive similarly in our case [9].

The lesions usually regress spontaneously in 3-6 years or sometimes with hyperpigmentation, atrophy or anetoderma. For diagnosis or cosmetic concerns, they could be excised [1]. Our case has multiple lesions, so excision was not suggested.

In conclusion, JX may occur in adult and dermatologist should keep in mind in the patients with yellowish-pink coloured, indurated papules. Trauma and diabetes mellitus may be a triggering factor for dissemination of JX.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Case Report

Bullous Wells' syndrome

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ABSTRACT

Wells' syndrome (WS) is an uncommon inflammatory skin disease which typically presents single or multiple erythematous and edematous urticarial plaques similar to cellulitis. The lesions may evolve into blue-grey morphea-like lesions and may persist for weeks or months. They ultimately heal without scar. Other clinical presentations reported in literature include papular and nodular and, rarely, bullous eruptions. Previously, bullous Wells' syndrome was rarely reported in the literature. Herein, we describe a case of a female patient with bullous Wells' syndrome localized to the upper limbs without any associated disorders.

Key words: Bullous lesion; Drug therapy; Wells syndrome

INTRODUCTION

Wells syndrome is an acute, rare dermatosis characterized by painful and itchy urticarial and cellulitis-like plaques. The lesions evolve rapidly over 2-3 days into plaques that resolve spontaneously over 2-8 weeks without scarring. Definitive diagnosis is made by pathology which is rich edema and infiltration of eosinophils in the dermis. In the subacute stage, flame figures surrounding collagen bands are observed. The presence blisters on the lesion is very rare in Wells syndrome. Wells syndrome's pathophysiology has not been elucidated and this disease often shows recurrence [1,2]. We present a case of Wells syndrome characterized by blisters here.

CASE REPORT

A 44-year-old female presented with redness, fluid-filled blisters and pain on left arm. Past history at both dorsum of the feet, extensor surface of the tibia and malar region redness and blisters were complaints. The patient's resume had urticaria history and antihistamine use. On dermatological examination, an erythematous and edematous well demarcated plaque with multiple tense and flaccid bullae containing serous fluid were observed on the left forearm extensor surface (Fig. 1). The Nikolsky's sign was negative. Blood count, peripheral smear, renal and liver function tests, immunoglobulin E (IgE) level were within normal limits. Erythrocyte sedimentation rate (ESR) in the first hour was 45 mm (0-20). Chest X-ray and ultrasonography (USG) for lymph nodes and abdominal USG produced normal results. Histopathology examination from a biopsy taken from the skin showed subepidermal blister containing, eosinophils, fibrinous material, lymphocytes and occasional polymorphonuclear leukocytes (Fig. 2). Dermal edema, perivascular, interstitial and periappendducular eosinophils intermingled with mononuclear inflammatory cells were seen in the dermis (Fig. 3). Direct immunofluorescence staining was negative. Based on the clinical presentation and the histopathology, a diagnosis of bullous Wells syndrome was made. Systemic steroid (prednisolone 60 mg/day), anti-histamine therapy (cetirizine 5 mg once daily) was combined with eau borique 2% daily dressing and topical antibiotics wound care. The lesions dramatically regressed within several days and steroid dose was tapered. The patient's lesions healed without scarring.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

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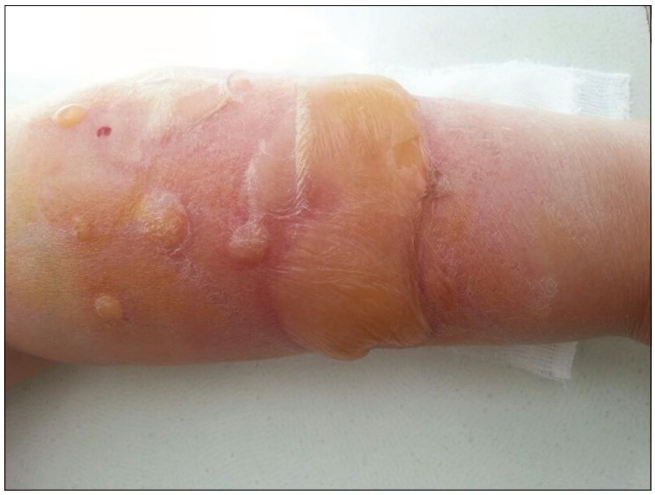

Figure 1: Erythematous and oedematous plaque with multiple tense and flaccid bullae on the forearm extensor surface.

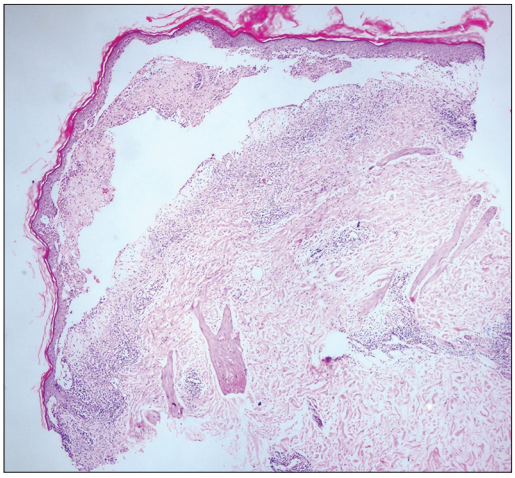

Figure 2: Subepidermal blister formation, dermal edema, both interstitial and angiocentric mononuclear inflammatory cells accompanied by eosinophils (H&E x40).

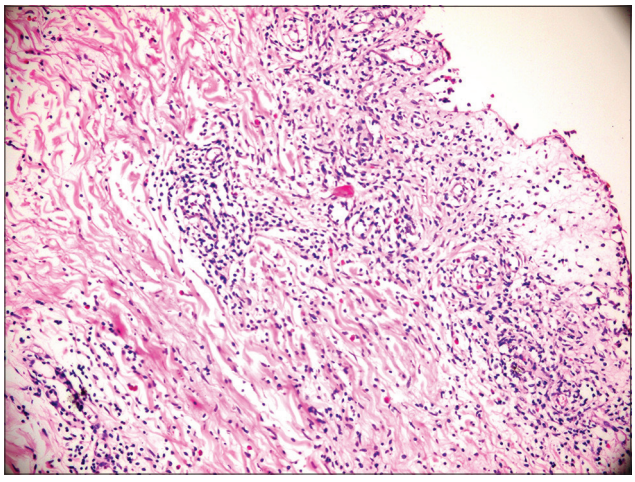

Figure 3: In the dermis, both interstitial and angiocentric mononuclear inflammatory cells accompanied by eosinophils (H&E x200).

DISCUSSION

Classical clinic presentation of Wells syndrome is painful and slightly itchy recurrent urticarial and cellulitis-like plaques. Papulonodular lesions, bluish-green, slate gray, pink or violaseous plaques have also been described [3,4]. Blisters may occur very rarely on cellulite-like plaques. To our knowledge, bullous Wells syndrome cases have been reported less than 15 in the literature to date [5-8]. Wells syndrome affects mostly adults and may involve any race and both genders [2]. Fever, fatigue, joint pain may be seen. Blood eosinophilia and increased IgE may be found [4]. In our patient, there was no accompanying symptoms, peripheral eosinophilia and increased immunoglobulin E level.

The exact etiology of Wells syndrome are unknown, however, it has been reported that insect bites, leukemia, bacterial, viral, fungal and parasitic infections, myeloproliferative disorders, drugs may be triggering factor for the disease [1,4-6]. Our patient did not have any triggering factors. Also, the pathogenesis of the disease is not known exactly, but urticarial lesions may occur with the abnormal reaction of eosinophils [4]. Aberrant and inadequate eosinophil skin homing is one of the major fact in disease expression of Wells' syndrome. An increase in Interleukin (IL)-5 levels have been observed due to Wells' syndrome. IL-5 does not only mobilize eosinophils from the bone marrow but also promotes homing of eosinophils by altering expression of adhesion molecules. In addition, increased levels of IL-5 appear to induce expression of CD25, the alfa chain of the IL-2 receptor, which increases eosinophil degranulation and following tissue destruction [9].

Histopathologically, dermal edema, rich infiltration of eosinophils in the superficial and deep dermis is observed in the acute stage. Flame figures are formed in the subacute stage when degranulating eosinophils coat basophilic collagen bundles with eosinophilic major basic protein (MBP). Phagocytic histiocytes are observed around the flame figures during the resolving stage. The flame figures are distinctive, but not pathognomonic for Wells syndrome. They may occur approximately 50% of patients with Wells syndrome [3]. Also, flame figures may be seen in other dermatosis such as bullous pemphigoid, insect bite reactions, cutaneous mastocytoma, eczema, prurigo, fungal infections and herpes gestationis [1]. Flame

figure was not detected in our patient. This condition may be associated with biopsy performed in the acute phase (3th days after onset of lesions).

There is bacterial cellulitis, erysipelas; arthropods bite reactions, allergic contact dermatitis, bullous pemphigoid, and Churg-Strauss syndrome in differential diagnosis of Wells syndrome. Detailed history, clinical examination and infection markers such as leukocyte count, sedimentation rate, C-reactive protein and fecal parasite search, serum IgE levels and looking parasite-specific antibodies, patch test, histopathological examination and direct immunofluorescence staining allow making differential diagnosis [10].

Topical steroids may be preferred in the treatment of mild cases, but oral corticosteroids are the first choice in severe cases. A few days therapy of systemic steroid achieved a dramatic response. Antihistamines such as cetirizine, minocycline, colchicine, antimalarials, dapsone, griserofulvin, interferon-alpha and cyclosporine are other treatment options [10]. In our case, the lesions dramatically regressed within several days with the oral cetirizine and oral prednisolone 60 mg/day therapies. Complete cure of the lesions was obtained within 2 weeks and gradually steroid therapy was discontinued. The patient has been in remission for 5 months.

In conclusion, presence of bullous lesions in the Wells' syndrome is a rare finding. Wells syndrome should be considered at the differential diagnosis of diseases characterized by blistering.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Case Report

Gorlin syndrome: a case report

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ABSTRACT

Gorlin syndrome is a rare autosomal dominant disorder which characterize by multi-organ abnormities such as odontogenic keratocysts in the jaw, skeletal abnormities and multiple basal cell carcinoma etc. We report a case of this syndrome in a young man with palmar pits, multiple facial BCC, clacifications of the falx cerebri and bifid rib.

Key words: Gorlin- Golts syndrome; Nevoid basal cell carcinoma syndrome; Odontogenic keratocysts; Bifid rib

INTRODUCTION

The Gorlin syndrome, also known as nevoid basal cell carcinoma, resulting from mutation in the PTCH1 gene and inherited as an autosomal dominant trait with high penetration and variable expressivity. The estimated prevalence varies from about 1/57000 to 1/256000 with a male to female ratio of 1:1 [1]. It arises in all ethnic groups, but most of reported cases are in whites [2].

In the following we describe a case of this syndrome in a 38 year old man with emphasizing on its clinical and radiologic manifestations.

CASE REPORT

A 38 year old male patient presented to our dermatology department with numerous slow growing pigmented papules on his face and scalp. The patient first begun noticing the appearance of these lesions when he was teenager (14 years old) and then they became larger in size and number over the time and some of them became erosive recently.

He gave history of a facial swelling of the right mandible 10 years ago and subsequently had been operated for a mandibular cyst few years back.

He denied any same history and lesions in his family and child. Patient's medical, personal and family history was non-contributory.

In physical examination dysmorphic features such as macrocephaly, prognatism and linear scar of operation in right side of his mandible were observed. Inspection revealed numerous (over 20) pigmented, some skin colour, papules and nodes on his cheeks, eyebrow, nose and scalp, predominantly on sun exposed areas (Fig. 1a). Some of these lesions were erosive. No regional lymph node was involved. The skin of his palms has tiny pits (Fig. 1b).

Multiple biopsies were done, the specimens were fixed in 10% formalin and submitted to histological examination and microscopically all of them showed basal cell carcinomas (multiple types such as morpheaform, micro nodular, nodular) (Figs. 2a - 2c).

The skull x-ray (AP view) showed a linear vertical radiopaque line in the midsagital plane of the cranium suggestive of calcified falx cerebri (Fig. 3a). The chest x-ray (PA) showed bifida rib (Fig. 3b).

Based on clinical, radiographic and microscopic data the diagnosis of nevoid basal cell carcinoma syndrome was established. Panoramic radiograph revealed well defined unilocular radiolucencie in maxilla (Fig. 4).

Patient underwent multiple surgical excisions with primary closure and flap for his lesions and we recommended him for regular check-ups and protection from sun exposure.

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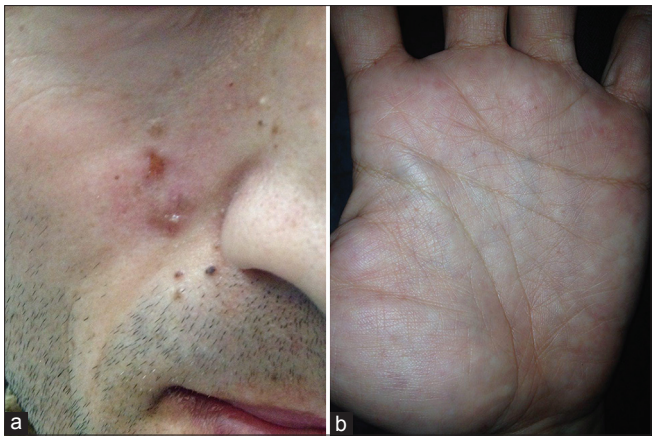

Figure 1: Multiple facial BCC (a), Palmar tiny pits (b).

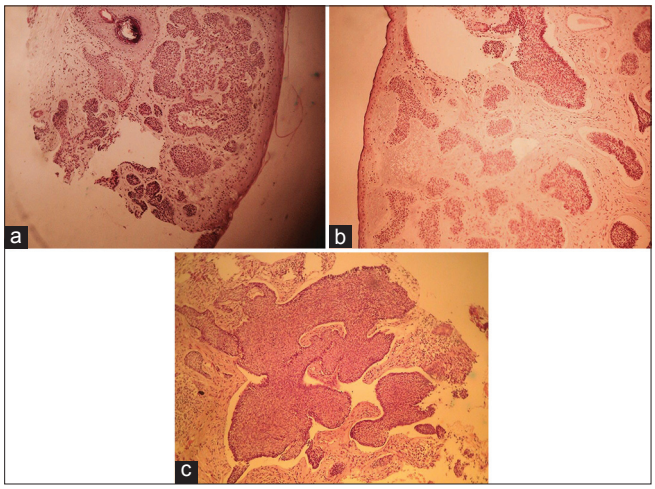

Figure 2: Biopsy of lesions microscopically showed basal cell carcinomas.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

DISCUSSION

Nevoid basal cell carcinoma syndrome (NBCCS) or Gorlin-Goltz syndrome (GS) was described for the first time in 1894 by Jarisch and White [3]. This syndrome represents a series of multi-organ abnormalities known to be the consequence of abnormalities in the PTCH gene located in long arm of chromosome 9q22.3-q31 [4]. This gene has important roles in neurologic and anterior-posterior axis development as well as hair follicle formation. This explains the broad range of developmental anomalies including craniofacial, skeletal defects and the propensity for meduloblastomas, odontogenic keratocysts and multiple basal cell carcinomas to arise [5].

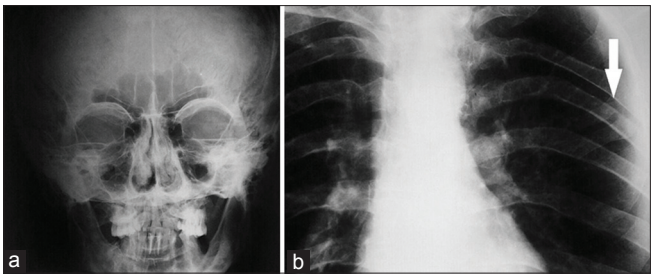

Figure 3: Calcified falx cerebri (a) bifida rib (b).

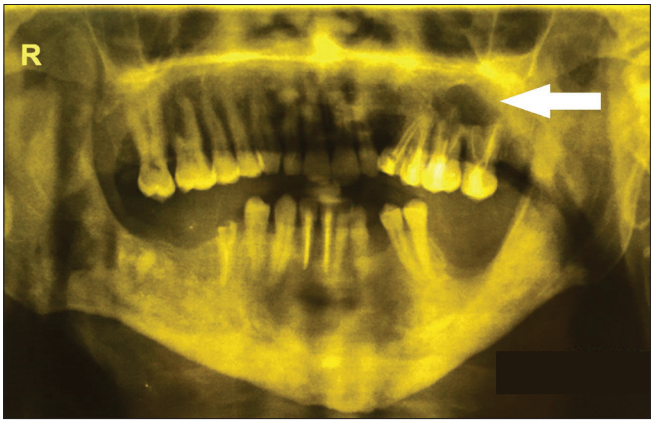

Figure 4: Unilocular radiolucencie in maxilla.

Evans et al proposed a diagnostic criteria that was further modified by Kimoni et al in 1997 (Table 1) [6,7]. The clinical diagnosis is made in the presence of two major criteria or one major associated with two minor.

Basal cell carcinomas favour the sun exposed areas of the face neck and trunk but may occur in sun protected sites too [5].

In general,the mean age of onset is about 25 years of age but it usually occur between puberty and 35th year of life [1]. And suspicion for Gorlin syndrome should be high if basal cell carcinoma are detected in pediatric age range as in our patient which is reported the onset of his lesions from fourteen years old.

Isolated cases are frequent, despite the hereditary characteristics of syndrome almost 60% of patients have no known affected family members and the absence of positive family history doesn't exclude the diagnosis [1].

Palmoplantar pits occur in 30-65% of the patients and are 2-3 mm in diameter and 1-3 mm in depth they are found on palms commonly than soles (77% versus 50%) [8]. Lamellar calcification of the falx cerebri occur in 70-85% of these patients [9].

Table 1: Diagnostic criteria that was modified by Kimoni et al. in 1997[6]

Major criteria
1. More than 2 BCCs or 1 BCC in a person younger than an age of 20
2. Odontogenic keratocysts of the jaws before age 15
3. Pitted depression on hand and feet (palmar plantar pits) (three or more)
4. Lamellar calcification of the flax cerebri under age 20
5. Fused bifid or markedly splayed ribs
6. First degree relative with NBCSS
7. PTCH gene mutation in normal tissue
Minor criteria
1. Macrocephaly determined after adjustment for height
2. Congenital malformation; Frontal bossing; Coarse face; Moderate to severe hypertelorism; Cleft lip/cleft palate
3. Skeletal abnormalities; Sprengel deformity; Marked pectus deformity; Marked syndactyly of digits
4. Radiological abnormalities; Bridging of sells turcica; Lucencies of hand and feet; Bifid and splayed ribs; Hemivertebra; Fusion of vertebral bodies
5. Ovarian fibroma
6. Medulloblastoma

BCC: Bassal Cell Carcinoma. NBCSS: Nevoid Basal Cell Carcinoma Syndrome. PTCH: Protein Patched Homologue 1

Odontogenic keratocyst (OKC) is the cystic lesion of odontogenic origin that illustrates the behaviour of a benign neoplasm with recurring propensity following surgical treatment. They are rarely symptomatic and can leads to the loss of bone and pathological fractures by losing the bone around the tooth [10,11].

Keratocysts may show a uni- or multilocular pattern and the cystic spaces may have a smooth or scalloped border [12]. Because of orthopantogram (OPG) findings we referred the patient to a dentist.

Through positive correlation of BCC with UV exposure the patients should be recommended for good UV protection with using sunglasses, sunscreen and avoiding from excess sun exposure.

Since the criteria of multiple BCC, multiple palmar pits, bifid rib in CXR, falx cerebri calcification, macrocephaly were present in our patient the final diagnosis of Gorlin syndrome was given.

The patient with Gorlin syndrome must be managed by several specialists such as dentist for odontogenic keratocysts, dermatologists for skin lesions and neurologists so familiarity with this syndrome is important for clinicians because of propensity of these patients to develop multiple neoplasms including basal cell carcinoma and meduloblastoma.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Case Report

Bell's palsy in a case of Darier's disease - a rare disease association or coincidental finding?

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ABSTRACT

Darier's disease (DD) is a rare acantholytic dyskeratotic autosomal dominant genodermatosis characterized by the presence of warty, brown papules and plaques affecting the seborrhoeic areas. Frequent bacterial, fungal and viral particularly herpes simplex virus (HSV) infections complicate DD. Bell's palsy is an acute onset, idiopathic facial paralysis resulting from a dysfunction anywhere along the peripheral part of the facial nerve. Reactivation of HSV is considered to be the main cause of Bell's palsy. This case represents, to the best of our knowledge, the first case of DD presenting with Bell's palsy. This case underlines the importance of recognizing HSV infection in DD.

Key words: Darier disease; Bell's palsy; Skin disease

INTRODUCTION

Darier's disease (DD) is an autosomal dominant condition characterized by a persistent eruption of hyperkeratotic papules, histological examination of which shows suprabasal acantholysis with dyskeratosis. Patients with DD have an increased susceptibility to herpes simplex infection. A number of clinical studies have described co-occurrence of various neurological and psychiatric symptoms with DD, but occurrence of Bell's palsy with DD has never been reported [1]. We report here a case of DD with Bell's palsy.

CASE REPORT

A 20 year old man presented with a 3-year history of itchy greasy yellow brown papules and plaques over face, neck, shoulders, upper back, axillae and groins (Fig. 1). On examination patient also had minute Palmar pits (Fig. 1) and V- shaped nicking of the free edge of the nails, characteristic of DD (Fig. 2). A skin biopsy was done from back to support clinical diagnosis, came consistent with diagnosis of DD (Fig. 3). Five days later, the patient presented with pain and weakness over right side of the face, was diagnosed as Bell's palsy (Fig. 4) and treated with oral steroids and valacyclovir. Serological test for both HSV-1 and HSV-2 were non-reactive. A week later, there was improvement in both facial palsy and skin lesions (Fig. 5).

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

DISCUSSION

Darier's disease was described independently by White and Darier in 1889. It is an autosomal disorder with variable penetrance, related to mutations in ATP2A2 gene at chromosome 12q24.1, which encodes the sarco-endoplasmic reticulum calcium AT4Pase type 2 (SERCA2). This defect results in impaired intercellular adhesions [2,3].

It has world-wide distribution, with the prevalence estimated to vary from one in 36,000 to one in 100,000 and an incidence of new cases of four per million per 10 years [2,4].

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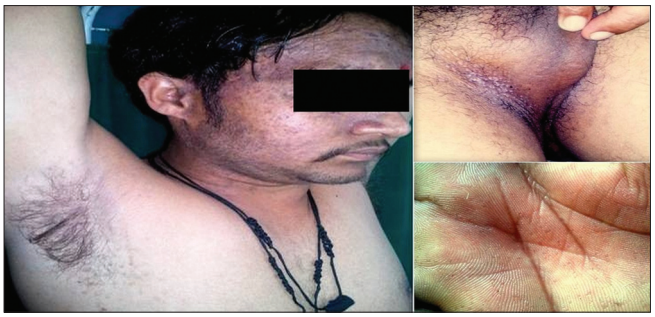

Figure 1: Greasy yellow brown papules and plaques over face, neck, shoulders, upper back, axillae and groins, with minute palmar pits.

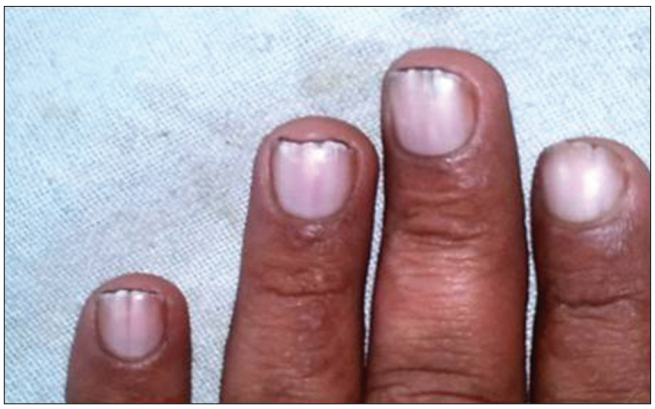

Figure 2: V- shaped nicking of the free edge of the nails.

Clinically, the distinctive lesions are firm, rather greasy, crusted papule that is skin-coloured or yellow–brown. Papules coalesce to produce irregular warty plaques or papillomatous masses, which, in the flexures, become hypertrophic, fissured and malodorous. Lesions are distributed in seborrhoeic pattern. The palms and soles may show punctate keratoses and minute pits. Nail changes include nail fragility, longitudinal ridging and splitting [2].

DD, either active or in remission, predisposes to some infectious complications, such as herpes simplex virus (HSV), varicella-zoster virus (VZV) and pox virus infections. It may present as atypical clinical presentation that frequently delay the recognition of viral infection and postpone adequate antiviral treatment. Increase in the susceptibility to various infections in Darier's disease has been linked to impaired cellular and/or humoral immunity [5].

Bell's palsy the most common disease of facial nerve, is an acute idiopathic peripheral facial paralysis. It is the cause of 60-75% of cases of unilateral facial paralysis with incidence of 20-30 cases per 100,000 per year [6].

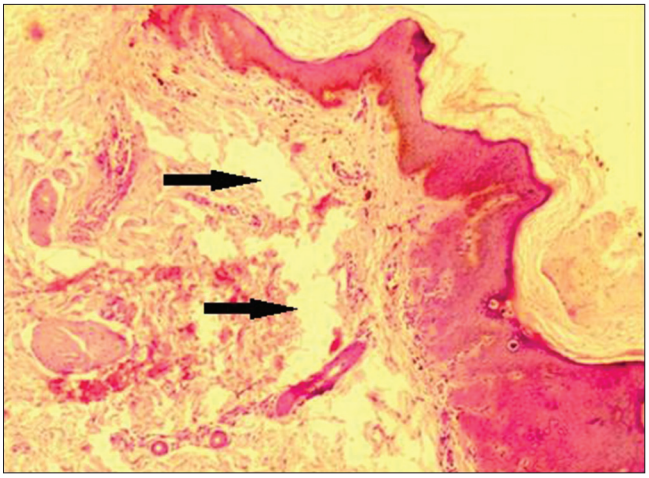

Figure 3: Skin biopsy showing suprabasal cleft with acantholytic dyskeratosis.

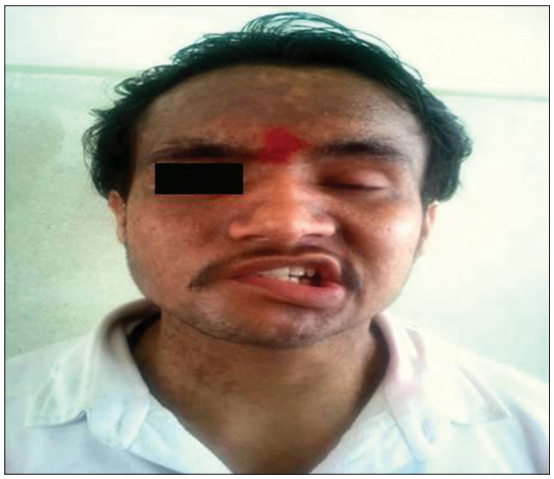

Figure 4: Weakness over right side of face (Bell's Palsy).

The diagnosis can be easily established in patients with unexplained unilateral isolated facial weakness. The onset is sudden and symptoms typically peak within a few days. Additional symptoms may include pain in or behind the ear, numbness or tingling in the affected side of the face usually without any neural deficit, hyperacusis and disturbed taste on the ipsilateral anterior part of the tongue [7].

Autoimmune process, viral infections, and even ischemia are the cause of initiation of inflammation. Different viruses from herpes virus family, herpes simplex virus -1 (HSV-1), HSV-2, human herpes virus -6 (HHV-6), and varicella zoster virus (VZV) have been considered to play role in bell's palsy. HSV has been considered particularly as the etiological agent in recent studies. Despite studies in favour of seropositivity of HSV in Bell's palsy, most studies could not find any definite association between antibody titres and Bell's palsy [8,9].

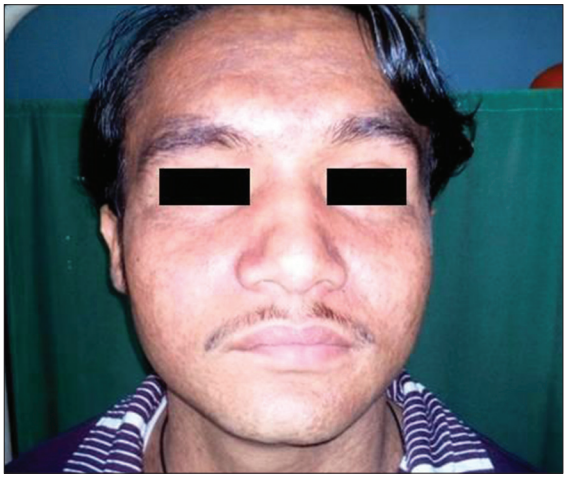

Figure 5: Improvement in both facial palsy and skin lesions after treatment.

The main aim of treatment in the acute phase of Bell's palsy is to speed up the recovery and to prevent corneal complications. Strategies to speed recovery include physical therapy, corticosteroids and antiviral agents. Inflammation and edema of the facial nerve are implicated in causing Bell's palsy. The rationale for the use of corticosteroids in acute phase of Bell's palsy is that they have a potent anti-inflammatory action which should minimize nerve damage and thereby improve the outcome. The rationale for the use of antiviral agents is the evidence that the inflammation of the facial nerve in Bell's palsy might be related to the HSV [10].

CONCLUSION

This case represents, to the best of our knowledge, the first case of DD presenting with Bell's palsy. The cause of Bell's palsy in this case, whether it is the reactivation of HSV due to disease per se or due to stress after skin biopsy remains eluded. This case underlines the importance of recognizing HSV infection in DD and also highlights the possible therapeutic effect of antiviral agents in DD.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Case Report

Rosacea-like tinea faciei

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ABSTRACT

Tinea faciei, is a facial superficial mycosis. The most frequent etiological agents are *Microsporum canis*, *Trichophyton rubrum* and *T. tonsurans*. It is often overlooked when considering the differential diagnoses of Rosacea. The most well known dermatology textbooks list acne, LE, perioral dermatitis, nasal sarcoidosis, carcinoid syndrom and other conditions but do not mention TF.

We describe 3 patients with lesions that clinically appeared to be Rosacea.

Key words: Tinea; dermatophytoma; Rosacea

INTRODUCTION

Tinea faciei (TF) is a dermatophyte infection of glabrous skin of the face sometimes displaying a wide and variable range of clinical features from erythema, patches, induration, vesicles, pustules, papular and circinate lesions. So, this infection is often deceptive and may clinically mimic other facial dermatoses. Discoid lupus erythematosus, lymphocytic infiltration, seborrheic dermatitis, granuloma annulare and contact dermatitis are the most frequent misdiagnoses [1].

We describe 3 patients with lesions that clinically appeared to be Rosacea.

CASE REPORT

Case 1

A 66-year-old woman complained of a 2 years history of facial eruptions – " - *an allergy* " - that had initiated by the nose (Fig. 1a). She was treated firstly with antibiotics for acne rosacea, but with no improvement, and then prednisone (20 mg daily) still for presumed acne rosacea was tried, also unsucessfully (Fig. 1a). Physical examination revealed extensive erythematous lesions on the face with some pustules. Clinically, the lesions appeared to be acne rosacea. The well defined delimited edges of the forehead lesion lead to the suspicion of tinea, and KOH examination was positive. The patient improved considerably in just two weeks of teatment. We used oral terbinafine and ciclopirox olamine cream.

Case 2

A 32-year-old woman came to the office with facial erythema to which had been prescribed cortisone creams and again no improvement was noted by the patient (Fig. 2a). Clinically, the lesions seemed to be irritant dermatitis, eczema or rosacea. This time,

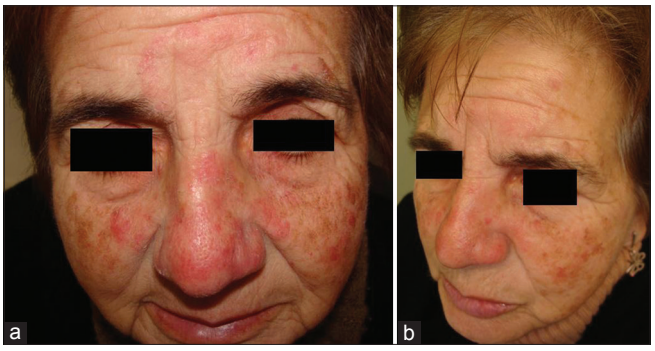

Figure 1: (a) June 2015-Papular and pustules lesions with flat patches of erythema on the nose and cheeks and annular circinate lesions with a raised margins on the forehead where KOH examination was positive. (b) One week later-The patient improved considerably in just two weeks of teatment; we used oral terbinafine and ciclopirox olamine cream.

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instead of well defined edges of *case 1-patient*, we could note a fine dusty scaling that suggested micotic scales and KOH examination was again positive. We used oral terbinafine and ciclopirox olamine cream. The patient returned 8 days after and already a good improvement was easily detected (Fig. 2b).

Case 3

A 68-year-old woman came to the office with facial erithema, telangiectasia and some papulo-granulomatous indurated lesions to which again had been prescribed betamethasone creams (Fig. 3). Clinically, the lesions seemed a bit of sarcoidosis or rosacea, but not tinea. Scales were scrapped and KOH examination was again hyphae positive. We used oral terbinafine and ciclopirox olamine cream. The patient still did not return but improvement is expected since this medication works well when direct mycologic test is positive.

DISCUSSION

Superficial fungal infections of the face seem not to be sufficiently or routinely investigated as differential diagnosis possibility and as a result, some facial dermatoses are often incorrectly diagnosed. As many as 70% of patients with tinea faciei are initially misdiagnosed as having other dermatoses. In a 20-year survey of tinea faciei, Nicola et al [2] examined 107 cases of tinea faciei. Typical forms were 57.1% whereas in 42.9% atypical forms were observed, mainly mimicking discoid lupus erythematosus (9 cases), polymorphous light eruption (8 cases) and Rosacea-like presentations [1,2]. In another study [1], in 100 cases of tinea faciei in adults, 52 mimicked a discoid lupus erythematosus, 15 lymphocytic infiltratation and four polymorphous light eruption

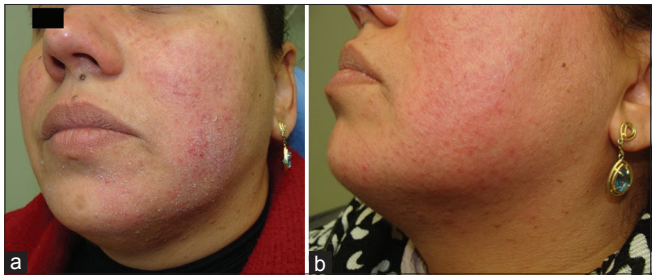

Figure 2: (a) Skin lesions with flat patches of erythema on the nose and cheeks. (b) Lesions in 8 day of treatment by oral terbinafine and ciclopirox olamine cream

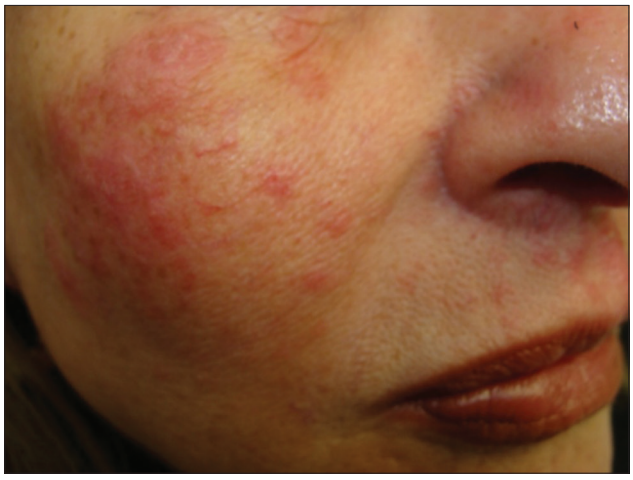

Figure 3: Erythema on the face with telangiectasia and some papulo-granulomatous indurated lesions.

Although tinea faciei is tinea itself, some authors claim it as a distinct entity. Facial anatomy, physiology characteristics, exposure to sunlight, repeated washing and the use of cosmetics often determine atypical clinical presentation that leads to incorrect diagnosis.

"Four-in-one" *anti: fungal-bacterial-histamine+ corticosteroid topical preparations* are widely used by nondermatologists in the treatment of superficial fungal infections, but may be associated with persistent/recurrent infections [3].

Therapy with terbinafine associated with topical anti-fungal therapy leads to healing within 3–6 weeks without relapse or side effects. An added benefit of using ciclopirox and terbinafine is their anti-inflammatory effect [4].

Our patients had pruritic erythematous facial eruptions and had been treated for other dermatosis but with no improvement. Direct microscopy revealed multiple hyphae in all 3 cases. The first patient is a poor elderly woman coming from a peripheral village where it is difficult to get lab tests, so for practical purposes and the patient immediate relief we chose the commencement of treatment even to the detriment of a culture examination in order not to delay the relief of the symptoms. The patient improved considerably in just 8 days. Complete resolution is expected after 6 weeks of therapy. The same option was made for the other 2 patients.

Tinea faciei accounts for 3–4% of cases of tinea corporis and is often initially misdiagnosed and treated as other dermatoses in as many as 70% of patients [5]. It may be easily diagnosed or excluded by KOH preparation. It is important that we are aware of the possibility of tinea faciei and that diagnostic procedures are put in

place to identify, or eliminate tinea facei from the final diagnosis.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Case Report

Intravascular fasciitis in foot – a rare entity in a rare site

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ABSTRACT

Intravascular fasciitis is a rare lesion involving blood vessels, showing proliferation of myofibroblasts, involving the superficial or deep fascia. We report a case of middle aged male patient presenting with callosity over the toe, with features of intravascular fasciitis, which is a rare site.

Key words: Intravascular; Fasciitis; Foot

INTRODUCTION

Intravascular fasciitis is a rare benign condition, involving arteries and/or veins, characterized by reactive myofibroblastic proliferation arising from the superficial or deep fascia [1]. Intravascular fasciitis was first described by Patchefsky and Enzinger in 1981 [2]. It is a rare variant, seen in less than 3% of nodular fasciitis [3]. It usually occurs in the small veins and arteries of the limbs, trunk, head and neck region. Only a few cases involving the foot have been reported [4].

CASE REPORT

A 56 year old male was referred to the surgical outpatient with a callosity over the left great toe and a skin patch on the left thigh. The clinical diagnosis was papilloma and corn respectively. Laboratory investigations revealed high cholesterol and triglyceride levels, with all other parameters being within normal limits. The patient underwent excision under local anesthesia and specimen was sent for histopathological examination. The gross specimen consisted of a skin covered tissue bit, which weighed about 2 gm, measured 3x0.5x1 cm. The cut section showed gray white, grey brown and haemorrhagic areas. Microscopy showed hyperkeratotic stratified squamous epithelium overlying dermal papillae along with few blood vessels with slit–like lumen, (Fig. 1) surrounded and compressed by an ill-circumscribed lobulated lesion composed of a mixture of fibroblasts and myofibroblasts in a myxoid background (Fig. 2), suggestive of intravascular fasciitis. Masson Trichrome stain showed the fibrous nature (Fig. 3) of the lesion. The associated papilloma of right thigh was histologically diagnosed as fibroepithelial polyp of Molle.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

DISCUSSION

Intravascular fasciitis is a rare tumour with only thirty three cases being currently reported in literature [1]. Intravascular fasciitis is most commonly seen in adolescent and young adult patients. Females and males are equally affected [3]. The predisposing factors may include thrombosis, trauma and pregnancy-related hormonal changes [1]. The most common anatomic locations of the lesion are the upper extremities, head and neck, occurring rarely in the foot [3]. The size of the lesions vary from 0.6 to 5 cms. The patient usually presents as a painless subcutaneous mass which is

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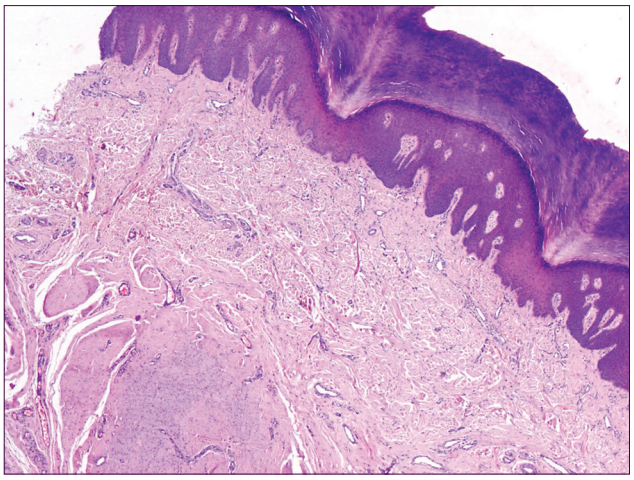

Figure 1: Hyperkeratotic epidermis overlying lobulated lesion in deep dermis, Haematoxylin and Eosin, x50.

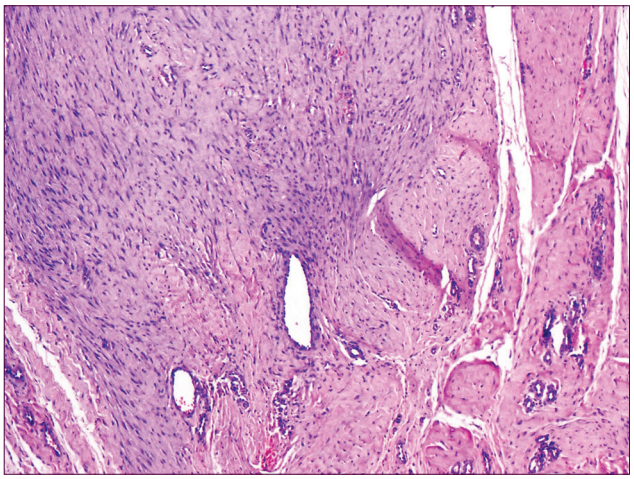

Figure 2: Mixture of fibroblasts and myofibroblasts in a myxoid background, Haematoxylin and Eosin, x200.

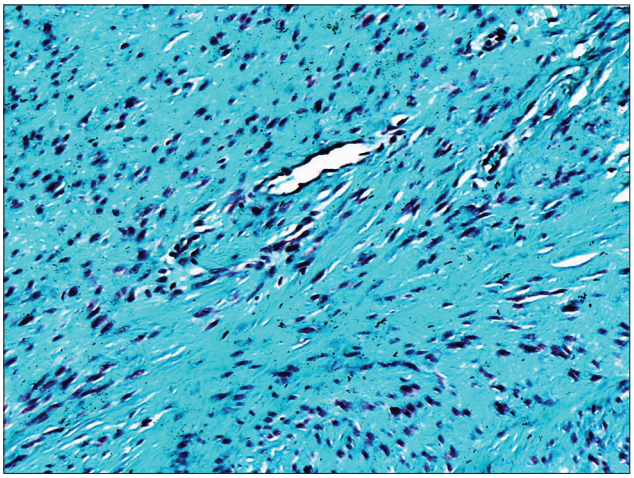

Figure 3: Masson trichrome stain showing fibrous nature of lesion, x 200.

gradually increasing in size for a duration varying from 2 weeks to 8 years [3,5].

Microscopically, intravascular fasciitis is characterized by plump spindle cells arranged in intersecting fascicles, a storiform pattern, or in a haphazard manner. The lesion may be present inside the lumen or associated with the walls of arteries or veins of all sizes. The separation of mass from the vessel wall may cause clefts [3,5]. Patchefsky and Enzinger reported that among their cases, the soft tissue component often was more prominent than the vascular component [2]. Though the infiltrative growth pattern resembles a sarcoma, it can be differentiated by the absence of significant cytologic pleomorphism and abnormal mitotic figures and the overlying epidermis is usually intact [1]. The background stroma varies from a dense hyalinized to edematous, myxoid appearance. Often associated are scattered multinucleated giant cells, lymphocytes and red blood cells [2].

The proliferative cells in intravascular fasciitis are of myofibroblastic phenotype, being positive to both vimentin and α-smooth muscle actin expression and negative for keratin, S100 protein, desmin, CD31, CD34, and c-kit. The multinuclear giant cells were CD68 positive, confirming the histiocytic origin. CD31 and CD34 can demonstrate the highly vascular stromal background and the vascular location of the lesion. Special stains for elastic fibers can be used to demonstrate the walls of associated vessels [2].

The pathogenesis of intravascular fasciitis is currently under study. According to Patchefsy and Enzinger [5], the lesion was due to the proliferation of myofibroblasts from the walls of systemic arteries and veins and they attributed the frequent extension into perivascular connective tissue to the "field effect" of myofibroblastic transformation [2]. The myofibroblastic origin of the spindle cells is confirmed by immunohistochemistry. Factors initiating myofibroblast proliferation may be preceding trauma, thrombosis, and high levels of estrogen and has to be further studied [1].

In the present case, the diagnosis was made by histology and use of special stains like Masson Trichrome. Differential diagnosis include sarcoma, benign fibrous histiocytoma, organizing thrombus, pyogenic granuloma, peripheral nerve tumours, spindle cell carcinoma, spindle cell melanoma, myxoid liposarcoma, fibrosarcoma and leiomyosarcoma [4]. This entity has been described as "pseudosarcoma" due to the misinterpretation of intravascular growth as vascular invasion by a sarcoma in addition to increased

mitotic activity, involvement of muscle and infiltrative borders of soft tissue [2].

The clinical behavior of this lesion is usually benign but a few cases have been reported to recur after excision. Since the soft tissue component comprises a much greater proportion of the tumor than the vascular component, diagnosis may be difficult in small biopsy specimens [1]. If blood vessel walls cannot be seen easily, the multinodular or serpentine growth pattern may be useful along with special stains for elastic fibers and smooth muscle [2].

CONCLUSION

Intravascular fasciitis is a rare variant of nodular fasciitis involving with blood vessels. The recognition of intravascular growth is required for diagnosis on small biopsy specimens and may be facilitated by special stains for elastic fibers or immunohistochemical stains for vascular markers. This case is reported with the intention of creating awareness of this lesion to avoid misdiagnosis and aggressive treatment.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Source of Support: Nil, **Conflict of Interest:** None declared.

Our Dermatology Online

Case Report

Porokeratosis of the scrotum

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ABSTRACT

Porokeratosis (PK) is disorder of keratinization characterized by annular lesions surrounded by raised sharply marginatedkeratotic borders with a characteristic histopathological finding named cornoid lamella. PK of genitalia is very rare condition and mostly reported among Asian population. The aim of present report is to document a new patient with localized scrotal PK with his father suffering from the same disease in the scrotum.

Key words: Porokeratosis; Cornoid lamella; Genitalia; Neurodermatitis

INTRODUCTION

PK is disorder of keratinization characterized by annular lesions surrounded by raised sharply marginatedkeratotic borders with a characteristic histopathological finding named cornoid lamella. It consists of a heterogeneous group of disorders inherited in a autosomal dominant fashion. PK has a wide variety of manifestations including classical plaque-type porokeratosis of mibelli, disseminated superficial actinic porokeratosis, linearporokeratosis, porokeratosispalmaris, plantaris, etdisseminata and punctate porokeratosis [1].

In Iraq, a special variety of pk has been reported affecting the face only called solar facial porokeratosis [2]. Localized pk of the genitalia is a rare in occurrence with 24 cases reported in the literature [3]. The aim of present report is to document anew patient with his father suffering from localized scrotal PK.

CASE REPORT

55-year-old male patient presented to Department of Dermatology, Baghdad Teaching Hospital in 20-8-2012 suffering from severely itchy genital lesions. Since 1982 the patient described a rash on the scrotum which was gradually enlarging in size and increasing in number. Also he mentioned the occurrence of exactly the similar disease of his father (dead) in genital area and this in favor of autosomal dominant inheritance. All the lesions are persistent with no history of spontaneous resolutions of any them. On examination, numerous papules, nodules and plaques distributed over the scrotum (>40 lesions) with none of them on the shaft of penis (Fig. 1). Those lesions were indurated skin colored while others were dark with typical annular configurations as the borders seemed more active while the center had the tendency towards atrophy. Some of the lesions showed thickened surface with pigmentation

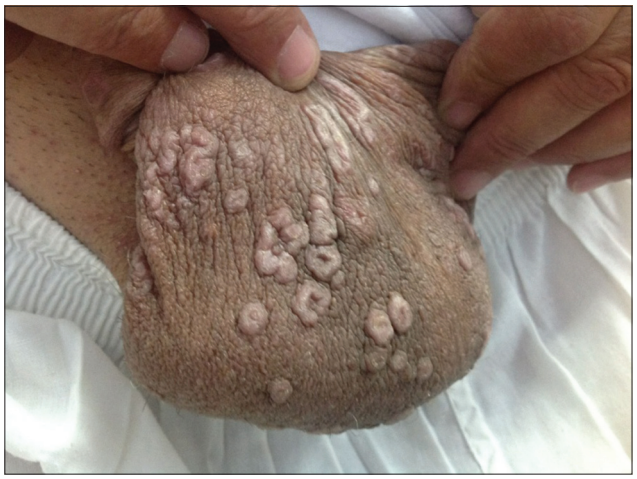

Figure 1: Numerous papules, nodules and plaques distributed over the scrotum.

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DOI: 10.7241/ourd.20161.22

without the typical annular ring of the most of the lesions and looked like neurodermatitis (Fig. 2).

The histopathology of the disease as follow: theepidermis wasacanthotic with basket weave hyperkeratosis. There was invagination of epidermis by column of keratin reaching the basal layer of epidermis. At the site of invagination there was absence of granular layer. This column consisted of parakeratotic cells, forming a typical feature of cornoidlamella. Many individual dyskeratoticcells were seen under the base of lamella reaching almost the basal layer of epidermis. While the dermis consisted of many dilated blood vessels with severe inflammatory reaction at the base of cornoid lamella, consisting of many lymphoid cells. In addition pieces of manydartos muscles were observed in the dermis (Fig. 3).

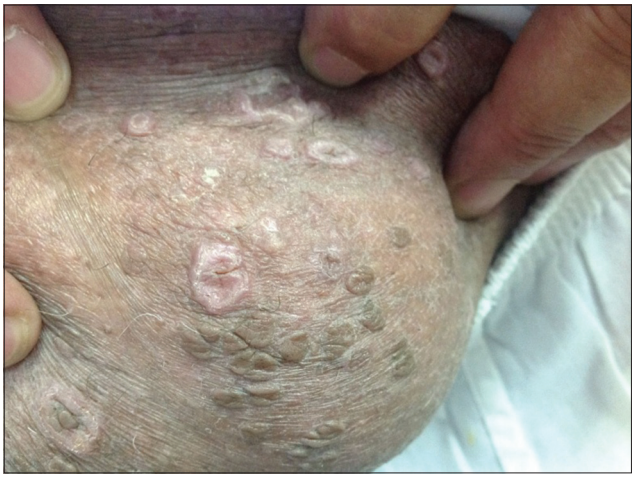

Figure 2: Some of the lesions showed thickened surface with pigmentation without the typical annularring of the most of the lesions and looked like neurodermatitis.

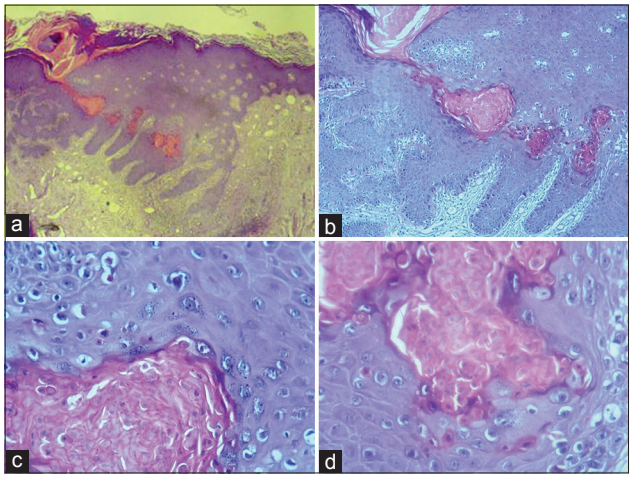

Figure 3: Histopathology showing epidermal acanthosis with basket weave hyperkeratosis. Invagination of epidermis by column of keratin reaching the basal layer of epidermis consisted of parakeratotic cells, forming a typicalfeature of cornoid lamella. Many individualdyskeratoic cells were seen under the base of lamella reaching almost the basal layer of epidermis. The dermis consisted of many dilated blood vessels with severeinflammatory reaction at the base of cornoid lamella, consistingof many lymphoid cells. [Hematoxylin and eosin stain; original magnification (a) x4, (b) x10, (c) x40, (d) x40.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

DISCUSSION

Porokeratosis of genitalia is very rare condition and mostly reported among Asian population [3-5]. The present report is the first case study that is being reported in Arab region. Family history of the present case was positive as his father had the similar condition and this might support the autosomal dominant inheritance like other types of pk. Itching is a prominent feature of present case and this was similarly reported [4]. There are many dermatosis involvingthe genital area like psoriasis, lichen planus and dermatitis and these are usually associated with itching. Lichen simplex is commonly superimposed on the top of these skin diseases [6]. Accordingly the present case was diagnosed and treated for 30years by most of dermatologists as case of lichen simplex chronicus. In most of reported genital pk, the lesions were scanty (1-3) while in our patient, numerous lesions were seen that widely distributed over the scrotum. Malignant transformation of pk had been reported [7], but fourtounitly no malignant changes were observed in present case.

There are no effective therapies of pk but many treatment had been suggested like topical steroid, cryotherapy, electrocautry, Co_2 laser and others. We recommend Co_2 laser removal as one of most effective modality [8,9], in order to relieve the patient complaint and to prevent the possibility of malignant changes.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Source of Support: Nil, **Conflict of Interest:** None declared.

Our Dermatology Online

Case Report

Phakomatosis pigmentovascularis with lower limb vascular abnormalities in a young Kashmiri male child-Report of a first child from Kashmir Valley (India) and review of literature

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ABSTRACT

Phakomatosis is a developmental abnormality simultaneously involving eyes, central nervous system, and skin. Phakomatosis pigmentovascularis (PPV) is a rare cutaneous disorder which is characterised by a combination of capillary malformations and pigmented anomalies. It arises sporadically. PPV was first described by Ota et al., in 1947. There is no sex predilection, but Japanese have been found to be affected more. There are four main types of PPV. Recently a fifth type with cutis marmorata and aberrant Mongolian blue spot has also been added to the classification. Here we report a case of PPV with Struge – Weber syndrome and Klippel Trenaunay syndrome in a young Kashmiri male child, which has been rarely reported in the literature.

Key words: Phakomatosis pigmentovascularis(PPV); Klippel Trenaunay syndrome(KTS); Struge – Weber syndrome(SWS)

INTRODUCTION

Phakomatosis, a neurocutaneous syndrome is a developmental abnormality which is characterised by the involvement of skin, eyes and central nervous system. Phakomatosis pigmentovascularis is a rare cutaneous disorder which was first described by Ota et al. in 1947 [1]. There is no sex predilection. Four types with two subtypes have been described, where subtype 'a' refers to cases presenting with only cutaneous manifestations and subtype 'b' is characterised by systemic association like in, Struge – Weber syndrome and Klippel Trenaunay syndrome [2]. Recently a fifth type with cutis marmorata and aberrant Mongolian blue spot has also been added to the classification.

CASE REPORT

A 12 – year old male child product of a non-consanguinous marriage presented with history of non-blanchable erythematous macules, which was noted to be portwine stain (naevus flammeus) on examination involving face, forearms, and right lower limb since birth (Fig. 1 and 2). There was history of increase in the size of these macules with age. The facial portwine stain was more marked and abundant on the left side.There was associated history of abnormal gait. However there was no history of headache, seizures, abnormal body movements or blurring of vision.The patient was noted to have hypertrophy of right lower limb with disparity in the length of legs. A visible vessel was seen in right groin crossing the midline (Fig. 3). Ophthalmological

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examination revealed raised intraocular pressure in left eye. Optical coherence tomography revealed severe thinning of Retinal nerve fibre layer (RNFL) in left eye, loss of neuro-retinal rim (NRR), gross asymmetry in cup-disc ratio and RNFL thickness (Fig. 4 and 5).

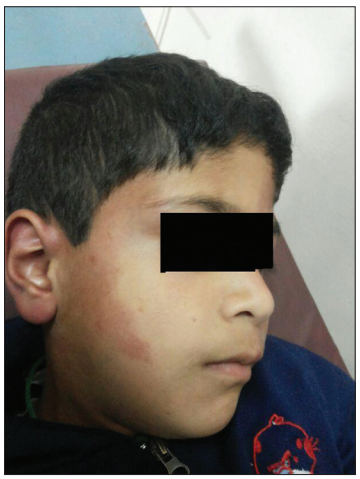

Figure 1: Portwine stain involving face.

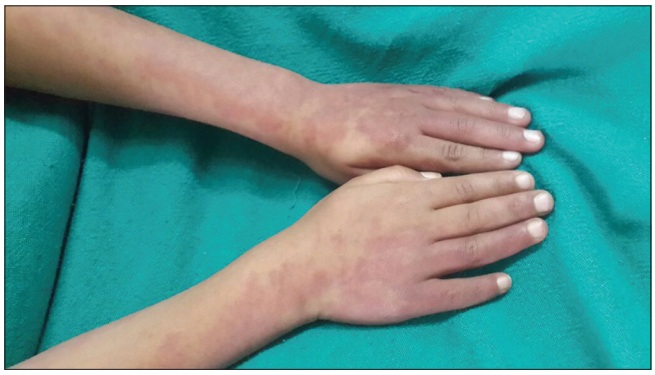

Figure 2: Portwine stain involving upper limbs.

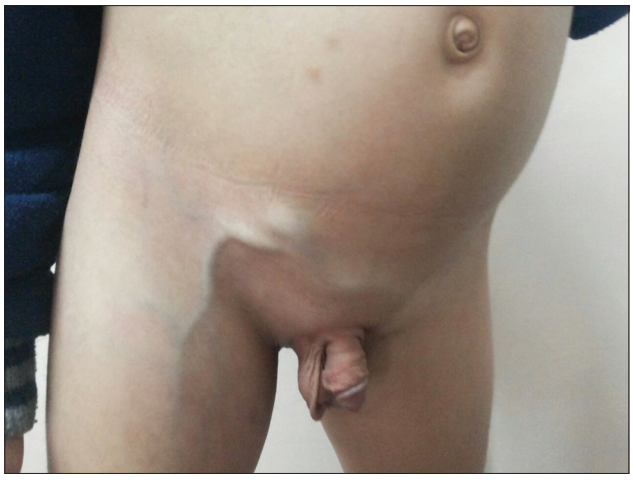

Figure 3: Picture showing a visible vessel was seen in right groin crossing the midline.

X ray of lower limbs showed limb lengthing in right limb, however, no bony hypertrophy was seen (Fig. 6). Colour Doppler and CT angiography of limbs revealed absence of right iliac vein with right femoral vein draining through collateral vein crossing midline into

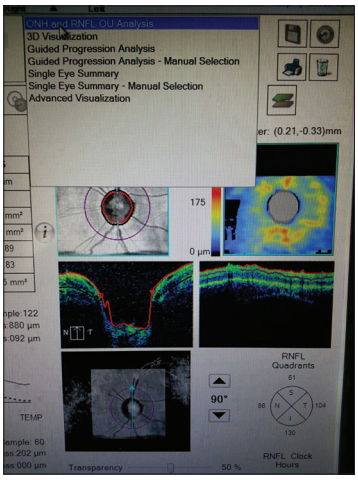

Figure 4: OCT showing severe thinning of RNFL.

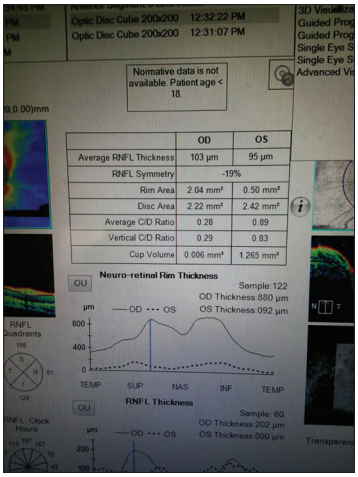

Figure 5: OCT showing ocular changes.

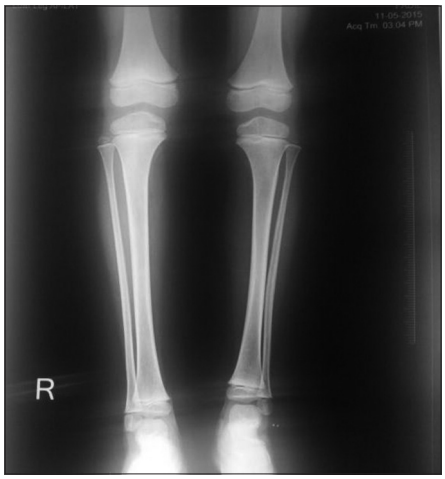

Figure 6: X-ray showing limb length disparity(R>L).

left external iliac vein. High origin of superficial femoral artery and deep femoral artery was also noted (Fig. 7). Non contrast CT of brain showed foci of dense gyral calcification in left temporoparietal region (Fig. 8).

DISCUSSION

The term phakomatosis refers to a developmental abnormality simultaneously involving skin, eyes and central nervous system. Phakomatosis pigmentovascularis is a rare syndrome characterised by the combination of a capillary malformation with pigmented naevi of various types. It is very uncommon and arises sporadically. Both, males and females are affected. There is no sex predilection, but Japanese have been reported to be affected more often [1,2]. Twin spotting phenomenon of Happle and Steijen has been proposed to explain this phenomenon (PPV). In this hypothesis,somatic mutations on nearby genes leads to mosaic spots in close proximity to one another [3]. PPV has been classified into four types namely; type I: Nevus flammeus and epidermal nevus, type II: Nevus flammeus, Mongolian spots, ± nevus anemicus, type III: Nevus flammeus, nevus spilus, ± nevus anemicus, and type IV: Nevus flammeus, Mongolian spots, nevus spilus, ± nevus anemicus. Recently a fifth type with cutis marmorata and aberrant Mongolian blue spot has also been added to the classification. Each type is subdivided into two subtypes; subtype 'a', cutaneous involvement only; and subtype 'b', both cutaneous and systemic involvement. Type II is reported to account for approximately 80% of all cases. PPV type I, III, and IV have very rarely been reported in literature.The capillary malformation may occur anywhere on the body and can be segmental.The aberrant Mongolian spots occur on atypical sites and tend to persist, unlike conventional Mongolian blue spots. It is estimated that 50% of patients with PPV present with systemic involvement. The cases that have been most studied are those associated with PPV type IIb. In the case of PPVs, neurologic anomalies develop in the first months of life. The most common ocular abnormality which has been found to be associated with PPV is ocular melanosis, which consists of unilateral or bilateral blue-gray pigmentation in the sclera (Naevus of Ota). Other ophthalmologic abnormalities are much less common. However, a large number of abnormalities have been published in relation to PPVs, but with the exception of Sturge-Weber syndrome, Klippel-Trenaunay syndrome, and ocular melanosis, the abnormalities are varied [4,5].

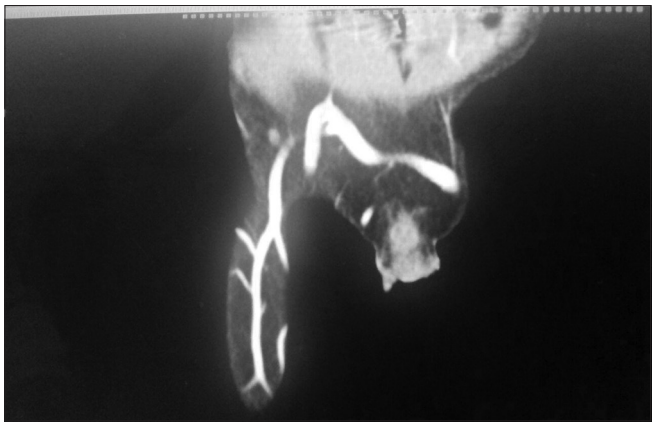

Figure 7: CT angiography absence of right iliac vein with right femoral vein draining through collateral vein crossing midline into left external iliac vein.

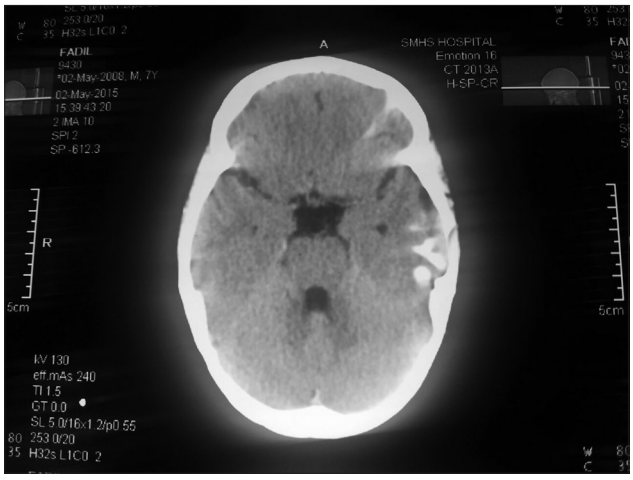

Figure 8: Non contrast CT of brain showing foci of dense gyral calcification in left temporoparietal region.

It is recommended that all the affected cases should be investigated depending on the sites involved, like for glaucoma if the capillary malformation affects the eyelids, or for Klippel-Trenaunay syndrome if there is limb involvement. A patient's quality of life may be improved by treating nevus flammeus using a pulsed dye laser and by treating pigmentary nevus using Q-switched lasers. Some authors suggest that the pigmentary nevus should be treated first [6].

Various systemic syndromes have been reported to be associated with PPV in the literature, and these include; Sturge-Weber syndrome, Nevus of Ota, and Klippel-Trenaunay syndrome [7].

Sturge –Weber syndrome (SWS) is defined as a facial portwine stain with ipsilateral vascular malformation of leptomeninges and eyes. Eye involvement is however not necessary for the diagnosis of SWS. Brain

changes show calcification with atrophy and increased vascularity of unilateral leptomeninges.

Naevus of Ota also known as naevus fuscoceruleus ophthalmomaxillaris is a dermal melanocytosis and is characterised by slate-grey or bluish hyperpigmentation involving the periorbital area, sclera and conjunctiva. It may be congenital or may develop around puberty.

Klippel Trenaunay syndrome is characteried by the association of cutaneous capillary malformation of limbs(naevus flammeus) and soft tissue swelling of limb with or without bone hypertrophy. Original description of this syndrome included vericosities of the limb [8,9].

Since its first description in 1947, 222 cases of PPV have been published, most of which are sporadic and from Japan, Mexico, or Argentina. The most common type is type IIb (45%), followed by IIa (30%). The rest are much less frequent and no isolated cases of PPV Ib, IVb, or Vb have been published. To date only two case series of PPV have been published. The largest series was published by Cordisco et al [10], who presented 25 patients in Argentina.In this series, type IIb was the most common type. Vidaurri-de la cruz et al [11] presented a series of 24 patients in Mexico in which 75% of the cases were type IIb, 25% were IIa, and the rest of the subtypes were not found.

Literature shows that till date only few well established cases of phakomatosis pigmentovascularis (PPV) have been reported, but PPV presenting with Sturge-Weber syndrome and lower limb vascular abnormalities is extremely rare. Sumita S et al [12] reported a case of PPV with Sturge–Weber syndrome and Klippel-Trenaunay Syndrome in a 13 year old female who presented with port wine stain over face, limbs and trunk, with a visible vessel along the lateral aspect of left lower leg together with associated hypertrophy of the involved limb.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Review Article

Medical leech therapy (Hirudotherapy)

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ABSTRACT

Leeches have been used in medicine long time before BC. In recent years medical leech therapy has gained increasing interest in reconstructive surgery and pain management and other medical fields. The possible indications and success rates of this treatment are discussed. There is a special interest in salvage of flaps and grafts by the use of medical leeches. Retrospective analysis indicates a success rate of >80%. Randomized controlled trials have been performed in osteoarthritis. Case reports and smaller series are available for the treatment of chronic wounds, post-phlebitic syndrome and inflammatory skin diseases. The most common adverse effects are prolonged bleeding and infection by saprophytic intestinal bacteria of leeches. Medical leech therapy is a useful adjunct to other measures wound management.

Key words: Acute wounds; Chronic wounds; Medical leech therapy; Reconstructive surgery; Hirudis medicinalis

Key Messages:
- medical leeches offer several advantages in venous congestive syndromes seen after reconstructive surgery
- medical leeches secrete substances which can reduce pain and itch
- infection control is necessary for safe medical leech therapy

HISTORY

Therapy with leeches is one of the oldest minor invasive procedures in medicine that was already mentioned 1,500 BC in Egypt. Sanskrit writings described leech therapy from 1,300 BC on. Hippocrates introduced leech therapy in Greece but the method was also known to ancient Mesopotamians, Egyptians and Aztecs, and Mayans. Medical leech therapy is part of the concept of the Greko-Arab Unani System of Medicine [1].

The procedure has seen a first renaissance in the 18th and early 19th century when it became extremely popular again. This has led to an eradication of naturally occurring leeches in Ireland where leech export was an important trade [2].

Medical leech therapy reemerged in the 70ies of the last century as an adjuvant to surgery. In 2004 the FDA approved medical leeches as medical devices in plastic and reconstructive surgery [3].

LEECHES

Leeches are hermaphroditic, bloodsucking annelid worms. Medicinal leeches belong to the order *Arhynchobdellida*, family *Hirudinidae*. Only 15 of the more than 600 of the known species are classified as medical leeches, such as *Hirudo medicinalis*, *H. verbana* and *H. orientalis* [4].

H. medicinalis has 33 to 34 body segments, is brown or black, and has six long reddish stripes on the back. The cylindrical body is slightly flattened and can measure up to 20 cm. Although they have 5 pairs of eyes they use the olfactory system to find their hosts. Adult animals have two suckers at the end of their body, a

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large posterior sucker and a smaller disc-shaped on the head that contains the mouths with tree jaws (Fig. 1).

Jaws consist of up to 100 teeth and salivary glands that release more than 100 known substances. Hirudin is the most powerful natural thrombin inhibitor. Hirudin works in synergy with two Factor Xa inhibitors, i.e. antistasin and ghilanten also found in the saliva of this worm. Calin is a platelet adhesion and activation inhibitor. Hyaluronidase supports the spread of active saliva compounds in the tissue. Destabilase dissolves fibrin. Bdellin, eglins and hirustatin are anti-inflammatory substances with protease inhibitory activity. There is a number of neurotransmitters like dopamine or serotonin in the saliva that reduce pain perception in the host. Acetylcholine works as a vasodilator [4].

Leeches can survive a year from a single blood meal, were they ingest about 10 times of their own body weight [4].

HOW TO USE MEDICAL LEECHES

Only medical leeches from serious suppliers should be used. Wild animals increase the risk of severe infections [5].

The site where leeches are to be placed should be clean and free of ointments, pasts etc. Cleaning can be done by sterile Ringer solution, physiological sodium chloride solution or sterile water.

Leeches are fast and elegant swimmers in water ("sweet water dolphins"). And they can move rapidly on the patient's skin as well. To apply them correctly, a 5 ml syringe were the nozzle was removed by scissor, can be used. The leech is placed into the prepared syringe and the syringe is directly applied on the skin surface to be treated with its open end. When the leech is feeding, the syringe is removed [6].

Some authors use anchoring sutures on medical leeches to ensure that the leeches stay in place. In a small trial no adverse effect on leech survival was noted by such a method [7]. The question remains whether this will negatively affect the feeding behavior.

The number of leeches applied depends on the size of the area that has to be treated. The Iowa Head and Neck Protocol recommends to applied leeches every 2 hours. The spots where the leeches are placed should be changed. The duration of leech treatment is until recovery of the compromised flap, mostly about one week [8]. For other indications different schedules may be more appropriate.

A single feeding lasts up to 60 min. After that the leech stops sucking and can gently be removed. Actively feeding leeches should not be pulled off. Forced detachment by a forceps or chemicals can lead to regurgitation of the leech stomach contents into the wound [4,6].

Wounds after leech biting lose some blood and may be oozing for about 24 hours [6] (Fig. 2).

Wounds should be disinfected and covered by a sterile dressing. Repeated application of heparin or sodium chloride has been recommended to stimulated hemorrhage from these wounds [6].

Although large randomized controlled trails are completely missing in the field of medical leech therapy

Figure 1: Disc-shaped sucker on the head of a medical leech.

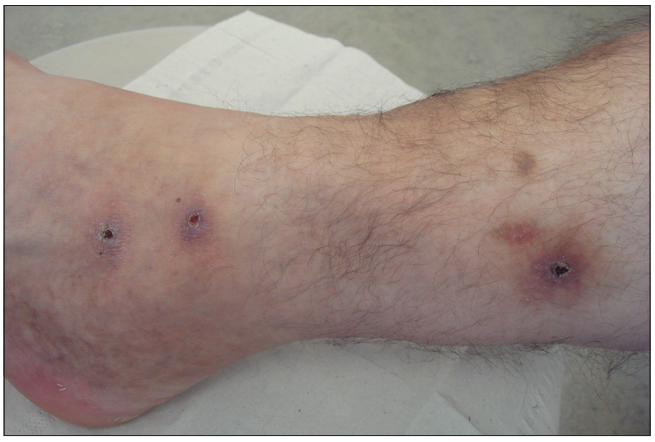

Figure 2: Three bite marks two days after medical leech therapy of post-phlebitic syndrome.

we will present indications and results. Please be aware of these limitations.

FLAPS

Medical leeches are extremely helpful in salvage of venous outflow compromised pedicle and free flaps. Leeches are used in the critical phase postoperatively in the dusky areas of a flap (Fig. 3). Animal studies suggested that leech therapy replaces congestive venous blood by fresh arterial blood improving survival of tissue [9].

Initially leeches may be necessary more often than just once daily for. Treatment is continued a couple of days until the flap recovers and venous congestion is overcome [10-17]. However, success rate is not 100% in particular in patients with lower hemoglobin levels and a need of more erythrocyte transfusions [13]. Leeches will not attach to dead flaps [14]. The success rate – complete and partial salvage – is 81.9 % (Table 1).

Leeches have also been used successfully to compensate venous congestion in replants of fingers, toes or nose [18]. Leech therapy increases perfusion resulting in hyperemia in dynamic and blood phases of Tc-99 m HDP bone scintigraphy [19]. Hirudin in leech saliva increases the level of messenger RNA for the vascular endothelial growth factor (VEGF) and VEGF expression in flap vessels [20].

HEMATOMA

Leeches can reduce postsurgical and posttraumatic hematomas (Fig. 4). This may help to salvage flaps and grafts [21] - or testicles in case of scrotal hematoma [22]. Evidence level is low (V) since no trials have been published.

Table 1: Salvage of flaps by medical leech therapy

Reference	No of patients	Salvage of flaps	Remarks
Jones et al. (2015)	1	1	Nasal flap
Pannucci et al. (2014)	4	1/4 (partial salvage)	Free flap breast reconstruction
Kim et al. (2013)	2	2/2	Paramedian forehead flaps
Nguyen et al. (2012)	27	18/27 (9 partial salvage)	Local, regional and free flaps
Whitacker et al. (2012)	277	216/277 (total and partial salvage)	Meta-analysis
Gröbe et al. (2012)	148	94 partial salvage 54 complete salvage	Local or pedicle flaps or grafts
Soengkar et al. (2012)	1	1/1	Free flap
Oh et al. (2012)	1	1/1	Free-style propeller flap
Our data	4	3/4	Local flaps
Total	465	381	

PAIN MANAGEMENT

Leeches are used in pain syndromes of various origins. The pain relief is rapid and sometimes long-lasting [23]. There are reports on successfully leech therapy in severe cancer pain [24]. Studies in osteoarthritis argue for symptomatic improvement by leech therapy by analgesic and anti-inflammatory effects [25].

An open trial in 32 patients with osteoarthritis leech therapy improved pain, stiffness and movement of joints [26]. In a randomized trial with 52 patients with either leech therapy or transcutaneous electrical nerve stimulation (TENS) their osteoarthritis of the knee responded significantly better to leeches. That was in particular relevant in pain reduction and improvement

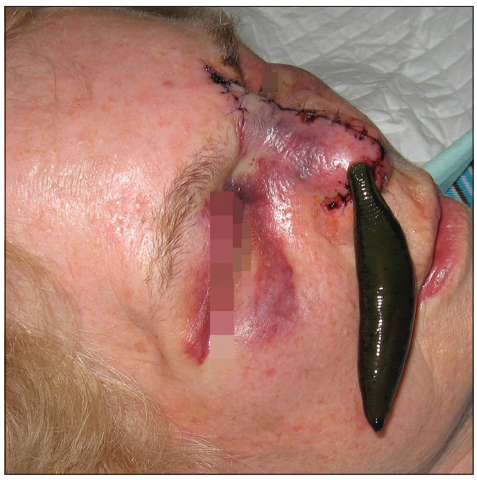

Figure 3: Medical leech for nasal flap salvage. The treatment resulted in complete flap survival.

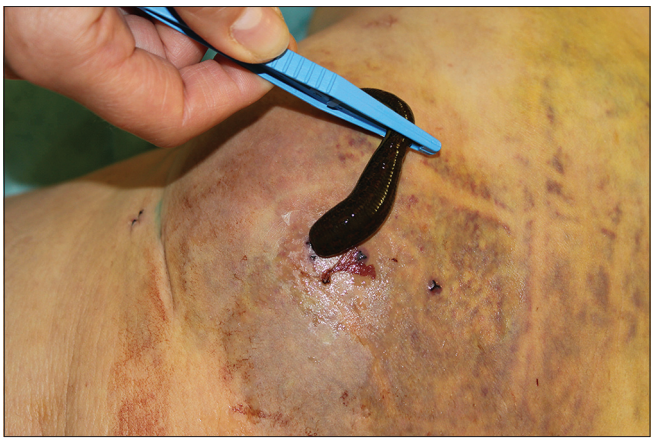

Figure 4: Application of a medical leech on a postsurgical hematoma.

of the Lequesne's index [27]. The Lequesne's index scores the severity of knee osteoarthritis by three dimensions: (I) pain or discomfort, (II) maximum distance walked, and (III) activities of daily living.

In a meta-analysis of four trials where 237 patients with osteoarthritis were included there was strong overall evidence for immediate and short-term pain reduction. The authors also found evidence for immediate improvement in patients' physical function, and both immediate and long-term improvement in joint stiffness. Leech therapy was not associated with any serious adverse events [28].

VARICOSE VEINS, LEG AND FOOT ULCERS

Application of medical leeches for the treatment of varicose leg ulcers decreased edema, limb girth, and improved ulcer healing [29]. In a randomized controlled study 50 patients with varicose veins were treated either by compression therapy or by medical leech therapy. After two months the group with medical leech therapy (n = 30) achieved a significant reduction in pain, edema and hyperpigmentation [30]. Leeches diminish the time of healing in post-phlebitic syndrome [31]. Since anticoaguagulant therapy is a contraindication for medical leeches (see below) this may limit the use of medial leech therapy in particular in venous leg ulcer patients. On the other hand, there is evidence level I for compression therapy for venous leg ulcers and leeches are not a substitute for medical compression.

There are case reports on the successful use of medical leech therapy for diabetic foot ulcers to salvage the leg [32]. Here, the monitoring of the treatment must be especially careful to avoid leech-borne infection.

SKIN DISEASES

In an open label study 27 patients with atopic eczema participated. Medical leech therapy was performed once a week for at least four times. The effects were measures by Eczema Area and Severity Index (EASI) score, SCORing of Atopic Dermatitis (SCORAD) Index, and Dermatology Life Quality Index (DLQI). The number leeches gives dependent on the size of the lesions. No concomitant treatment was applied. Reduction of EASI was 54.5%, of SCORAD 55%, and DLQI improved by 62.4% [33].

Leech application on five times reduced the hyperpigmentation of a nevus of Ota in a 23-year-old female patient from India [34].

ADVERSE EFFECTS

Prolonged bleeding of up to ten hours can be observed from time to time after medical leech therapy [21,35]. Usually, bleeding can be stopped by pressure but sometimes a primary suture might be necessary [36]. Repeated application of large numbers of leeches can cause anemia that needs transfusion. Control of hemoglobin during medical leech therapy is recommended [35].

Medical leeches may cause primarily local infections at a rate of 2 to 25 %. The infections are due to microbiota of the leech. A number of species have been characterized form the midgut of *Hirudis* spp. *Morganella morganii*, *Rikenella*, and *Aeromonas veronii* are dominant members but bacteria like *Magnetospirillium* spp. and *Roseospira marina* have also been detected [37].

In a US multicenter trial antimicrobial prophylaxis was documented in 91.5% of the included patients. Surgical site infection was observed in 11.9% of patients – all of them received antibiotic prophylaxis. In four infections *Aeromonas* spp. was isolated. *Aeromonas* spp. are Gam-negative bacteria that live symbiotically in the leech intestine. Those isolates were resistant to the antibiotic agents used in prophylaxis. It was found that trimethoprim-sulfamethoxazole and ciprofloxacin were equally effective appear in preventing leech-associated infections [38]. However, ciprofloxacine-resistant *Aeromonas* infections have been reported [39,40].

In a retrospective study from Belgium wound infections were registered 27.5%. Levofloxacin has been recommended for prophylaxis of *Aeromonas* infection [41]. Other authors suggested dipping the leech in 0.02% chlorhexidine solution to reduce the infectious risk [35].

The development of inflammatory epidermal cysts has been observed after leech therapy with unidentified species, probably due to infection [42].

A very rare adverse effect of medical leech therapy is the development of diffuse pseudolymphoma [43, 44].

Allergic reactions have been described in rare cases. In selected patients hirudin was identified as possible

allergen, in others the cause remained obscure [45, 46]. Local irritant contact dermatitis is another possible adverse reaction [47].

Another aspect of safety has been stressed by Siddall et al. (2007) who investigated commercially available leeches by molecular methods. They identified *H. verbana* in a number of cases instead of ordered *H. medicinalis* [48].

CONTRAINDICATIONS TO MEDICAL LEECH THERAPY

There are some disorders that are absolute contraindications to medical leech therapy such as blood clotting disorder (e.g. hemophilia), severe anemia, arterial insufficiency, hematological malignancies, hypotension, septic disorders, known allergic reaction to active ingredients of the leech saliva (hirudin, hyaluronidase, destabilase, etc.), and patient refusal to leech therapy. Pregnancy and lactation are contraindication due to the risk of infection and bleeding. Patients on anticoagulants and immunosuppressive therapy should not be treated with leeches. Anticoagulants will increase the risk of major bleeding. Immunosuppression increases the risk of infections. Since erythrocyte transfusions may be necessary during medical leech therapy, patients who refuse transfusions should not be treated by leeches [6].

CONCLUSIONS

Medical leech therapy is an effective treatment modality in plastic and reconstructive surgery for flap salvage, in the treatment of hematomas, post-phlebitis syndrome, and possibly for chronic wounds. There is very limited experience for other diseases. The most important adverse effect is the risk of leech-borne infection with *Aeromonas* spp. Prophylactic antibiosis can decrease this risk factor. The risk of anemia warrants hemoglobin control.

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Our Dermatology Online

Review Article

Melanocytic lesions and dermoscopy in childhood: diagnosis, therapy and foloving

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ABSTRACT

Early detection of malignant skin tumors, particularly malignant melanoma in childhood, which is most malignant of all, is of utmost importance for the prevention, treatment and outcome of the disease. If an observed lesion is characterized as melanocytic, the second step can be performed in the so called "*two-step dermoscopy*" algorithm, which includes the differentiation between a benign pigmented lesion (nevus) and a malignant lesion (melanoma). Gigantic nevi, as a subgroup of large congenital nevi, may pose a huge psycho-social burden for children and their parents, primarily because of potential complications. Melanoma can develop in healthy children but also in those with identified genetic disorder or immune diseases. Dermoscopy, at the same time, has reduced the number of unnecessary biopsies and excisions of benign melanocytic lesions in childhood.

Key words: Nevi; Melanoma; Dermoscopy; Childhood; Therapy; Foloving

Knowledge of pigmented lesions dates back to ancient times beginning with Hippocrates and Celsius, who called such outgrowths *naevi nigricans*. In the 16th century they were described by Russius Lauretius and then in 17th century by Highmore as tumors resembling coal. During the 20th century those tumors had different names: melanotic tumor, melanotic cancer, anthracite cancer etc. One of recent publications is the thesis by Bonnta in Lion (1911) titled "Melanosis and melanotic tumors". In 1967 an international body within the World Health Organization was established for the evaluation of diagnostic methods and treatment of melanoma, and under its auspices comparative clinical research has been conducted worldwide [1].

Moles (nevi) are confined, circumscribed inherited skin anomalies, occurring as a consequence of embryonic development [2]. Nevus cells appear in the shape of a nest on epidermal border and epidermis, and unlike melanocytes, they have no dendritic endings, tonofilaments and desmosomes. On the other hand, melanocytes and nevocytes produce the only endogenous autochthon skin pigment – melanin. The process of melanogenesis is monitored by genes, hormones and UV radiation [3].

Early detection of malignant skin tumors, particularly malignant melanoma, which is most malignant of all, is of utmost importance for the prevention, treatment and outcome of the disease. With the production of the first portable dermatoscope in 1958 by Leon Goldman a new era in the diagnostics of pigmented skin lesion and early detection of melanoma has begun [4].

Dermatoscopy (also known as dermoscopy, epiluminescence microscopy, surface microscopy, video dermoscopy) is a diagnostic method in dermatovenerology performed by covering of a skin lesion with mineral oil, alcohol or water and examination of the lesion with handheld dermatoscope, stereo microscope, camera or digital imaging system. It enables *in vivo* visualization of skin structures that cannot be seen with the naked eye such as epidermis, dermoepidermal borders and papillary dermis [5,6].

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In the past decades different epiluminescence microscopy (ELM) parameters have been set to differentiate melanocytic (moles, melanoma) from non-melanocytic lesions (basalioma, spinalioma, dermatofibroma, hemangioma and seborrheic keratosis), which is the so called first step in dermatoscopy.

If an observed lesion is characterized as melanocytic, the second step can be performed in the so called *"two-step dermoscopy"* algorithm, which includes the differentiation between a benign pigmented lesion (nevus) and a malignant lesion (melanoma). With that regard several different melanocytic algorithms have been established [7].

- Pattern analysis
- ABCD rule
- Menzies method
- 7-point checklist
- 3-point checklist
- ABC-point list

Classical dermatoscopic algorithm in the diagnostics of pigmented skin lesions is the so called *pattern analysis* set by Pehambergera et al. in 1987 and further modified and supplemented by Argenzian et al. in 2000 [8,9]. It is established on the analysis of numerous dermatoscopic structures, and based on the analysis of different colors and patterns, along with the so called *clue*, can lead to a specific diagnosis. The structures are often described metaphorically (e.g. *blue-whitish veil*) and they are difficult to translate into our language, for example, structureless blue zone vs. blue veil [10].

Melanocytic neoplasia in childhood can be divided into three classes: congenital moles (small, medium size, big and gigantic), acquired moles and melanoma. Congenital melanocytic nevi are found in 1-6% of newborns and are defined as benign neoplasia present already *in utero*. They include moles found during the birth but also tardive nevi, which become visible shortly after the birth. According to their size they are divided to small nevi, up to 1.5 cm in diameter, medium-size 1.5-19.9 cm and large ones of 20 cm and over [11].

Gigantic nevi, as a subgroup of large congenital nevi, are found in 1:500.000 newborns and they pose a huge psycho-social burden for children and their parents, primarily because of potential complications: melanoma, rhabdomyosarcoma, and manifestations of neurocutaneous melanocytosis. Nevus spilus and segmental speckled lentiginous nevi are also classified as congenital melanocytic lesions found in 0.2% of newborns. They need to be distinguished from agminated nevi, which lack brownish background. In early childhood sebaceous nevi composed mainly of fully formed sebaceous glands located in dermis are commonly found on the scalp. In around 30% of cases they can develop into basal cell carcinoma. From the time of birth or during early childhood a verrucous mole with hyperkeratotic surface, linear and/or bizarre shape located mainly unilaterally may occur. Its prognosis is good and malignant alteration is not known [12].

Specific dermoscopic structures, which can be found in congenital nevi include hypertrichosis, perifollicular hyper/hypopigmentation, multi-component pattern and structures that are found in seborrheic keratosis, so called *milia-like cysts* [13].

Melanoma incidence in children below the age of 14 is generally low worldwide, however, its increase has been evident over the past decades. Around a half of melanoma in children occur *de novo*, and a half from pre-existing melanocytic lesions [14].

Melanoma can develop in healthy children but also in those with identified genetic disorder or immune diseases. A special risk comes from the gigantic congenital nevus of over 50 cm in diameter, and a relatively high risk of malignant alteration exists for a small congenital nevus of 1.5-2 cm in diameter [15].

In terms of clinical but also dermoscopic examinations, pitfalls occur in vascular lesions (capillary and cavernous hemangioma) in childhood, particularly if they are thrombosed. In case of an injury of such lesions, they can be of livid and black color, they can bleed and be affected by a secondary infection and definitely resemble melanoma or pyogenic granuloma [16]. Differential diagnosis may also include angiokeratoma, which is a benign vascular lesion with dilated capillaries located below hyperkeratotic and acanthotic epidermis. Multiple angiokeratoma appear mainly before puberty and they are mostly symmetrically distributed on the skin of the scrotum, gluteus and periumbilically [17].

Furthermore, melanocytic nevi, which occur several months or years after the birth (so called acquired melanocytic nevi), such as the blue nevus (*naevus coeruleus vs. blue naevus*), Spitz and Reed *naevus*,

and Clark's nevus are a clinical, dermoscopic but also histopathological challenge for establishing a proper diagnosis.

Atypical vs. dysplastic nevi are defined as acquired melanocytic nevi with diameter over 6 millimeters, irregular borders and different colors, therefore, in terms of dermoscopy several types of such moles can be differed. Special attention should be paid to moles with an eccentric hyperpigmented patch or focally pronounced or prominent reticular pigment network [18].

Nevocellular nevus of *"Halo"* type (Sutton's *naevus*), which is clinically presented as depigmented halo around the junction mole, is relatively common in childhood. The mole in the center of the halo may completely disappear leaving behind a leucodermic patch resembling vitiligo. Children with a higher number of such lesions need to be observed and dermoscopic monitoring should be recommended [19].

A real challenge in clinical diagnosis is posed by hypopigmented and depigmented melanocytic lesions, because in everyday clinical practice, melanoma is rarely suspected in children, especially amelatonic or hypomelanotic type of tumors. Juvenile melanoma is a special form of a complex nevocellular nevus in children, which is most often located on the face. Clinically, it is recognized as a dome-shaped, red and brownish in color, with smooth surface and hard to elastic consistence. Although malignant alteration is very are, surgical treatment is recommended, i.e., full removal of the lesion and histopathological verification [20].

Neurocutaneous melanosis is a very rare disease in children manifested with rather large hyperpigmented areas located mainly in the bathing trunk site. It is described within the neurocutaneous syndrome, which involves nevocellular nevi on the brain, spinal cord and meninges. Having in mind symptoms originating from the central nervous system (e.g. convulsions) and the development of hydrocephalus at a later stage, the prognosis is very bad and there is no successful treatment. Malignant alteration is possible [21].

A special risk is found in children with a higher number of moles (more than a hundred) and atypical nevus syndrome, especially in case of a hereditary AMS type (a*typical mole syndrome*). Anatomic site of the moles and child's phototype are of special importance for several reasons. Namely, regions exposed to light as well as sites of permanent or occasional mechanic irritation are given special importance, particularly in terms of recommendations for prophylactic excision biopsy. On the other hand, dermoscopically, such lesions have special algorithms for recognition because they belong to so called *specific-site lesions*, which particularly involve lesions of the face, hairy part of the head, palms, soles and nails as well as all *milky-line lesions* affecting *mammae* areola, umbilicus, vulva and penis [22].

Dermoscopy, as a non-invasive and pain-free diagnostic procedure, has given a new morphological dimension to nevi, their dermoscopic monitoring and as well as early detection of atypical melanocytic lesions and melanoma in childhood. At the same time, it has reduced the number of unnecessary biopsies and excisions of benign melanocytic lesions in childhood.

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Source of Support: Nil, **Conflict of Interest:** None declared.

Our Dermatology Online

Clinical Image

A case of onychomadesis following hand, foot, and mouth disease

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We report a 3-year-old girl with onychomadesis on big toes, 2nd left finger, and 1st right finger since 4 weeks (Fig. 1). Five weeks before that, she suffered from fever, maculopapular and vesicular rash involving his hands, feet and mouth that was diagnosed as Hand-foot-mouth disease (HFMD). Onychoscopy with polarized light and magnification x20 well demonstrated the separation of nail plate from the proximal nail bed with subsequent shedding of the nails (Fig. 2).

HDMD is an acute infection caused most often by Coxsackie A virus type 6 and Enterovirus 71. It is more common among children than elderly. The disease is characterized by maculopapular and vesicular lesions on the hands, feet, and mouth. Onychomadesis is a shedding of the nail beginning at its proximal end, caused by temporary arrest of the function of the nail matrix. Since the first description in 2000, there are several reports of nail changes following HFMD. Recently, Shin et al. observed a group of 13 children with median age of 33 months. They found an average interval from HFMD to the nail changes 5.9 weeks (range, 3 to 12 weeks), average number of involved digits 7.4 (range, 2 to 14) and the most common involvement of left great toe (85%). The nail changes varied from transverse ridging of the nail plate (Beau's lines) up to complete nail shedding (onychomadesis). The mechanism of nail involvement remains unclear but it is considered that the inhibition of nail matrix proliferation is due to direct inflammation spreading from skin eruptions around nail. This was supported by the observation of Shikuma et al. that onychomadesis was developed only on the fingers and toes having cutaneous eruptions around nails. Onychomadesis occurring after HFMD is temporary with spontaneous normal regrowth [1-4].

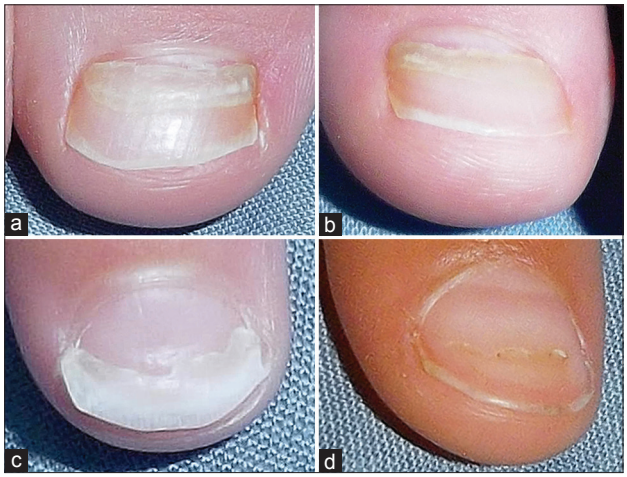

Figure 1: Clinical images of onychomadesis of the right big toe (a), left big toe (b), 2nd left finger (c), and 1st right finger (d).

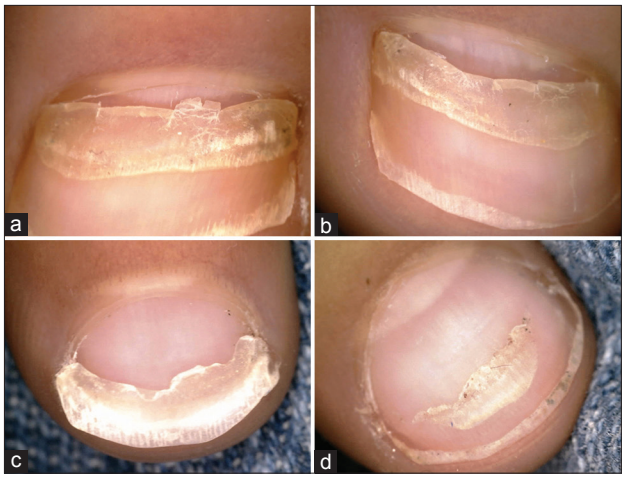

Figure 2: Onychoscopy images of onychomadesis of the right big toe (a), left big toe (b), 2nd left finger (c), and 1st right finger (d).

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Clinical Image

Plaque with pearly raised borders on the forearm

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A 29-year-old male presented with an annular plaque on his arm for 7 months. Physical examination revealed 6 mm annular plaque with raised borders and a shiny stellate-purple center (Fig. 1). Dermoscopic evaluation revealed a ring of white spiked structurless areas at the periphery; an intense structurless steel purple area was observed at the center of the lesion (Fig. 2). Hystopathological examination showed a band-like infiltrate of lymphocytes, pigment incontinence, saw tooth rete ridges, hypergranulosis and hypekeratosis (Figs. 3 and 4).

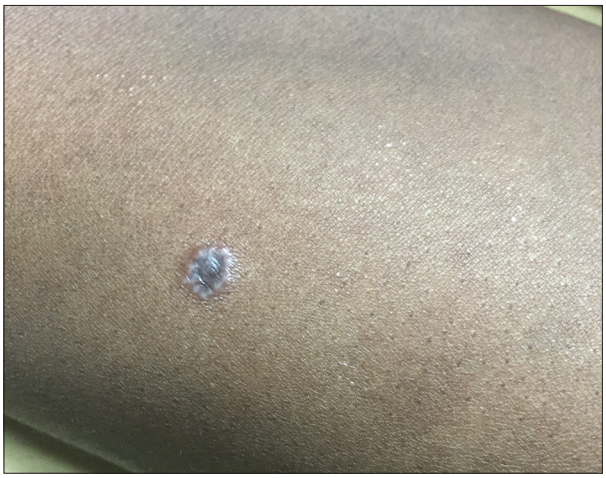

Figure 1: Annular plaque with raised borders and a shiny stellate-purple center.

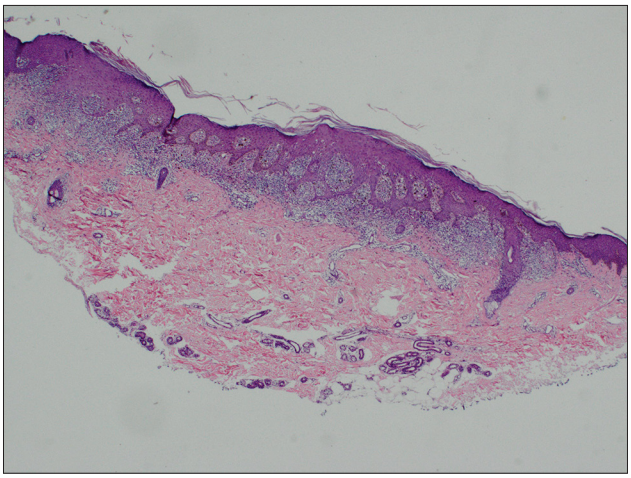

Figure 3: Band-like infiltrate of lymphocytes, pigment incontinence, saw tooth rete ridges hypergranulosis.

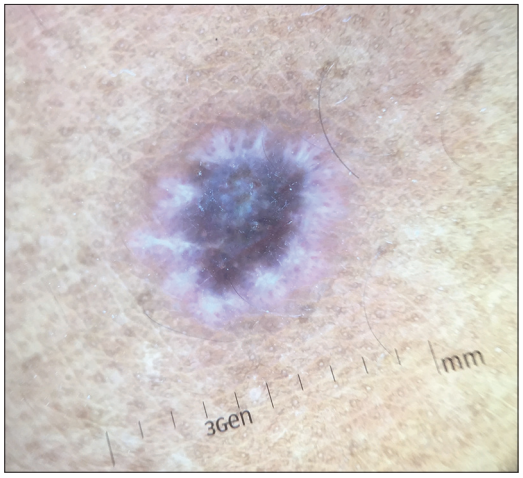

Figure 2: Ring of white spiked structurless area at the periphery; with an intense structurless steel purple area at the center.

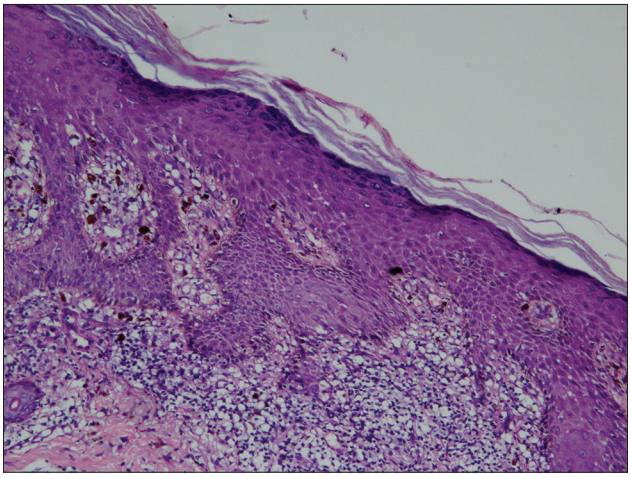

Figure 4: Band-like infiltrate of lymphocytes, pigment incontinence, saw tooth rete ridges hypekeratosis.

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Lichen planus annularis (LPA) is a rare variant of lichen planus; it might comprise less than 7% of LP cases [1,2]. Most frequently described locations include the genital and intertriginous areas, however, lesions on the trunk, extremities, eyelids and neck have been reported as well [3].

The pathogenesis of LPA has not been clearly elucidated nevertheless published data has revealed that the lichenoid tissue reaction might be triggered by a sequential activation of Langerhans cells, ultimately triggering the release of activated T cells which in turn migrate to the dermis and release several cytokines such as interleukins 1 and 2, and interferon gamma [4].

Interesting dermoscopic findings were the spiked white structurless areas in a circular disposition corresponding to Wickham striae; histopathologically it may correlate to the overall hyperplastic epidermis. Pigmentation at the center of the lesion is better appreciated dermoscopically showing a structurless steel purple area, corresponding histopathologically to areas of pigment incontinence in the papillary and reticular dermis. The black color of melanin is perceived as steel purple due to a tyndall effect. Long wavelengths are not reflected to the open air whereas shorter wavelengths are reflected and perceived by the eye as violet or blue [5].

In conclusion, interpretation of dermoscopic findings may help to infere histopathological changes to render an accurate diagnosis.

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Letter to the Editor

Pediculosis Capitis. Report of 2 cases

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Sir,

Pediculosis capitis is a common parasitic infestation of the scalp. pediculus humanus is the etiologic agent involved. We hereby present two classic cases of pediculosis capitis, for its numerous amount of parasites, in different stages of the disease. Both patients were treated with Ivermectin 200 mcg per Kg, a single dose.

CASE 1

Female patent 80 years who came to emergency due to urinary tract infection, during her clinical examination we found patient abandoned in poor hygienic conditions without any family or personal history importance.

During her clinical examination a patient in bad condition and neglected was found with the presence of numerous parasites on scalp and hair, observing many hairs with the presence of nits (Fig. 1 a-c) and microscopic view of the multiple nits (Fig. 2). Rest of the physical examination was normal

CASE 2

Female patient 45 years old hospitalized due to ovarian cancer, during her clinical examination head lice was seen, dermatological examination showed the presence of nits, lice walking on her hair (Fig 3), Dermatoscopic and microscopic view of parasite walking (Fig. 4) Rest of the physical examination was normal.

Both patient received ivermectin 200 mcg/kg one single dose and cured their pediculosis capitis.

DISCUSSION

Pediculosis capitis is a head lice infestation and it is an endemic parasitosis affecting many countries in

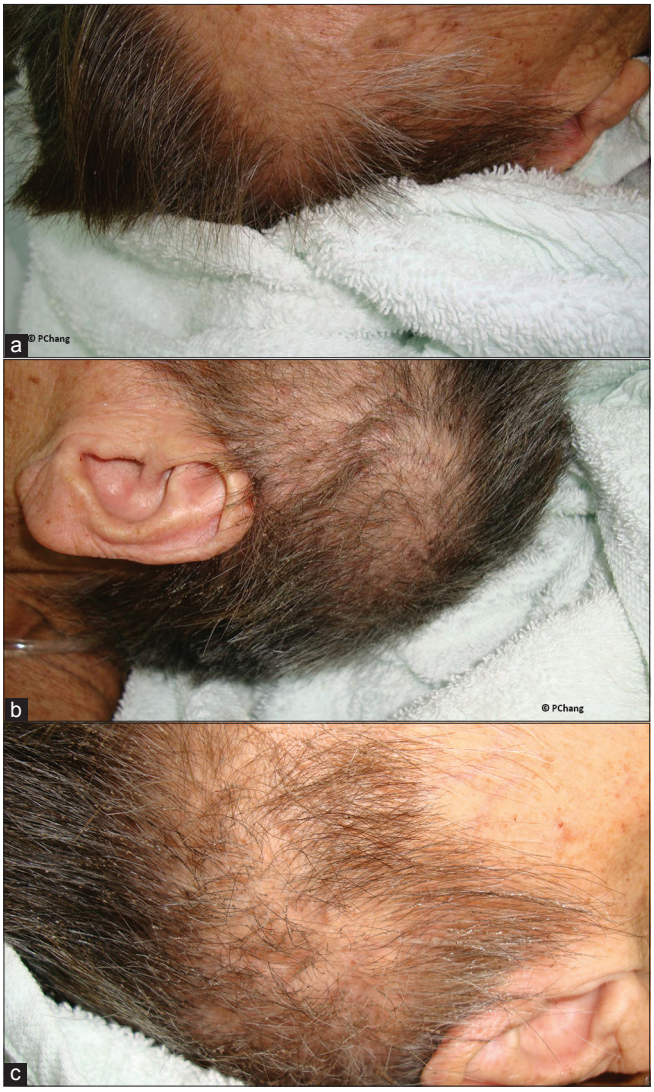

Figure 1: (a-c) Multiple nits on hair.

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DOI: 10.7241/ourd.20161.28

Figure 2: 4 Microscopic view of the nits.

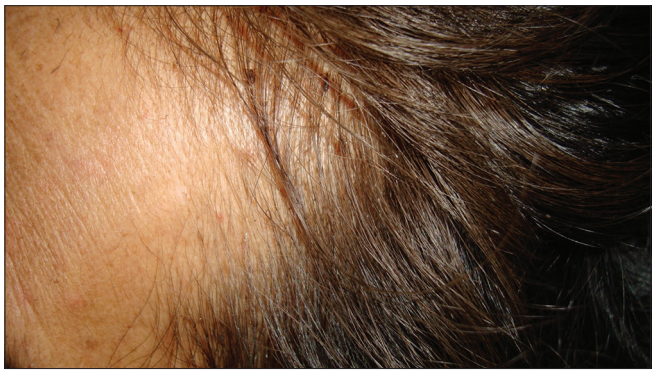

Figure 3: Parasites walking on the hair of the patient

Figure 4: Dermatoscopic and microscopic view of the louse

the world. It is one of the most common infections in childhood, the incidence has been estimated to be 800 and 2400 new cases per 10 000 children every year [1].

The etiologic agent is the pediculus or lice, which come in three varieties, being Pediculus humanis capitis the one who causes head infection. The other two cause body and pubic infections. Pediculus human capitis can only propagate on human scalp [2,3] and if they are away from the host head for more than 2 days, they die.

Transmission of head lice requires intimate head to head contact, and it also occurs through fomites (combs, hats, clothing, etc) but it has less relevance [2,4]. It occurs that way because these wingless ectoparasites cannot jump, but they move from hair to hair quickly when the hair is dry [4].

Pruritus of the scalp is the most common symptom of head lice infestation, and it increases depending on the number of lice present and the duration of the scalp infestation [2,4]. Usually patients carry less than 20 mature head lice, which survive for 30 days [4] other signs may be reddish and itching papules frequently surrounded by erythema. These primary signs can be delayed for 4 - 6 weeks, which means that the infestation cannot be diagnosed on the early phase. In the case of reinfection, primary signs may appear within 24 – 48 hours due to the immune mediated reaction against the components of the lice saliva [2].

If the patient is not treated and continues scratching, secondary lesions as excoriations may appear, and they reduces the natural barrier function of the epidermis and this leads to a superinfection typically by *Staphylococcus aureus* and *streptococci*. The chronic infestation and persistent scratching may lead to chronic impeginization of the scalp or regional lymphadenopathy (less frequent) [2,4]. Many head lice infections cause no symptoms, and probably less than half cause itching, so this symptoms are unreliable as indicators of lice presence [3].

There are two methods to confirm the presence of pediculus human capitis on human scalp: visual inspection and diagnostic combing. In clinical practice the visual diagnosis is the most common, which consists in the direct observation of juvenile or adult head lice on the scalp. Detection combing can be performed on dry or wet hair. Combs must have parallel sided teeth and a distance of <0.3mm between teeth so that the even juvenile lice are caught [2,5]. The most important matter is to diagnose active head lice infestation in order to prescribe appropriate treatment, and the wrong diagnosis may lead to a potentially antilouse or pediculicide formulations over applied [5].

The treatment consists of manual removal and topical or systemic treatment. The first one consist in the extraction of the lice with a comb of short space between each teeth, there are also electric combs that can be used [6].

A pediculicide (lindane 1%, pyretrhin, permetrhin 1%, etc) should be applied, and all household members with a ctive infestation should be treated. A reapplication of the treatment is recommended 7-10 days after the initial procedure. In children under 2 years old, manual removal of lice, eggs and nits is recommended [4].

There are several different kinds of topical treatment available: chemical, physical and natural. Permetrhin is the most commonly used and the least toxic for humans. It should be applied during 10 to 30 minutes and the procedure must be repeated seven days later. Ivermectin may be used as oral treatment for pediculosis, and it must be administered as a single dose of 200mcg/kg.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Letter to the Editor

A black nodule on the temple

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Corresponding author: Dr. Hiroo Hata, M.D., Ph.D., E-mail: hata07jp@yahoo.co.jp

Sir,

A 56-year-old female was referred to our hospital complaining of a nodule on the left temple. In the history-taking interview, she reported that the lesion had appeared 3 years before referral to our hospital as a tiny pinkish papule and had gradually enlarged. Physical examination found the nodule to be dark purple to black, well circumscribed, hemispherically raised and 5 x 5 mm in diameter (Fig. 1a). Dermoscopic examination revealed reddish-blue lacunae (Fig. 1b). We suspected hemangioma, venous malformation, apocrine hidrocystoma and basal cell carcinoma. We surgically removed the lesion for the correct diagnosis. In the histopathological findings, there were large vascular channels filled with abundant erythrocytes from the upper dermis to the subcutaneous tissues (Fig. 2a). There was no evidence of epidermal change. The vascular channels were lined by a single layer of endothelial cells or a thin wall of fibrous tissue (Fig. 2b).

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

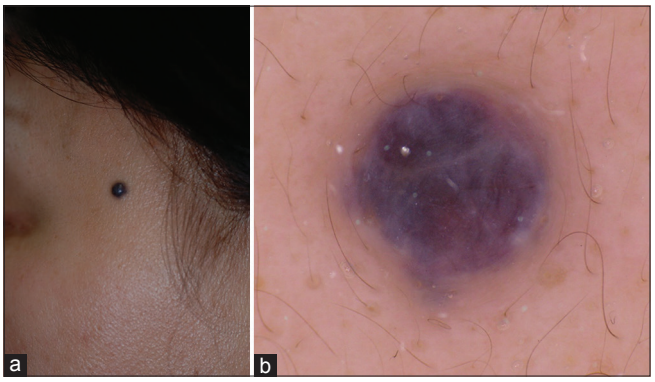

Figure 1: (a) There is a solitary soft, dark, well-circumscribed nodule and an area of 5x5 mm on the left temple. (b) In dermoscopy, the lesion presents red-blue lacunae.

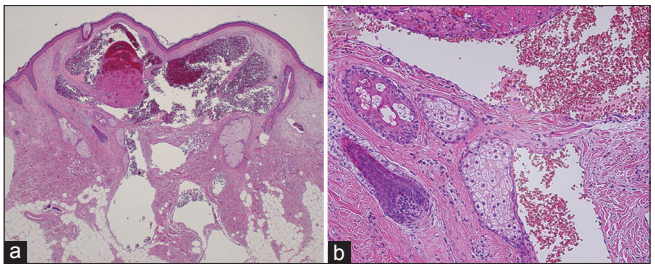

Figure 2: (a) Histopathorogy, low magnification. (hematoxylin and eosin stain; x20). (b) Histopathorogy, high magnification. (hematoxylin and eosin stain; x200).

DIAGNOSIS

Venous lake

DISCUSSION

Venous lakes were first described by Bean and Walsh in 1956 [1].

They noted that venous lakes clinically present as soft, dark gray papules of a few millimeters in diameter with domed surface, often on the face, head and neck area, especially on the lower lips of elderly people. Histopathological study shows one to several dilated spaces with erythrocytes, and the lesions have very thin, fibrous walls. Usually, severe sun damage is evident in adjacent dermis. The pathogenesis of venous lake is still unclear. It is speculated that they venous lakes occur as a result of severe sun damage to collagen fibers that prevents venous dilation [2].

Venous lake clinically resembles angiokeratoma, apocrine hidrocystoma, basal cell carcinoma and malignant melanoma. It is comparatively easy to rule

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out basal cell carcinoma and malignant melanoma with dermoscopy; however, it is difficult to dermoscopically distinguish venous lake from other non-melanocytic tumors.

Venous lake often histopathologically mimics many kinds of vascular tumors. Angiokeratoma histopathologically demonstrates numerous dilated, thin-walled capillaries, mainly in the papillary dermis, whereas the epidermis presents acanthosis with elongated rete ridges and hyperkeratosis [3]. Epidermal changes and the location of the lesions are different from those in venous lake. Apocrine hidrocystoma is clinically similar to venous lake and histopathologically shows dilated ducts in the dermis; however, the walls of apocrine hidrocystomas are lined by rows of columnar cells and show decapitation secretion indicative of apocrine glands [4]. Apocrine hidrocystoma clinically resembles venous lake, although the pathological features are different.

Above all, we confirmed the diagnosis as venous lake. Venous lake usually appears on the lower lips, and if this lesion had been located on the lower lip, the diagnosis would have been easy. However, in this case, the black nodule was located on the left temple, so the diagnosis was more difficult.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Source of Support: Nil, **Conflict of Interest:** None declared.

Our Dermatology Online

Letter to the Editor

Treatment option of advanced of vulvar carcinoma with cisplatin, 5-FU, and TS-1

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Sir,

The incidence of vulvar squamous cell carcinoma (SCC) has been increasing, [1,2] and remarkable progress has been made in the surgical management of resectable vulvar SCC to the date [3-5]. These modifications in surgical management have maintained oncologic outcomes while significantly reducing morbidity. However, there has been no improvement in survival for those diagnosed with advanced or recurrent disease to the date. Some chemotherapy regimens have been used for treating advanced cutaneous vulvar SCC today; there is unfortunately no current standard treatment. Herein we describe a case of vulvar SCC with lung metastasis that had treated and kept stable disease (SD) with chemotherapy by cisplatin and 5-fluorouracil (high dose FP), and TS-1 as the additional chemotherapy after surgical treatment.

A 67-year-old Japanese woman referred to our hospital complaining of a reddish nodule on her vulva with four months history. Physical examination revealed a reddish nodule measuring approximately 20 x 20 x 8 mm on the right labia minora (Fig. 1a). A biopsy specimen from the nodule showed typical findings of SCC (Figs. 1b and c). Computed tomography (CT) scan showed no evidence of metastasis to the other organs. The tumor was completely excised by using the butterfly excision and both inguinal region lymphadenectomy. One year after the primary operation, CT scan showed multiple pulmonary nodules and they were diagnosed as multiple pulmonary metastases (Figs 2a and c). The treatment with cis-daimminedichloroplatin (cisplatin) and 5-fluorouracil (5-FU) (high dose FP; cisplatin 80 mg/m^2, 5-FU 800mg/m^2) was started.

After 6 cycles of administration, the lung metastases did not show any change in size or number, and the condition of lung lesions were evaluated for SD. 2 cycles of treatment with TS-1 as the additional chemotherapy were done after 6 cycles of high dose FP. She had been examined by CT scan every 3 months, and her disease was kept for SD more than one year after the treatment (Figs 2b and d). There were no serious side effects throughout the chemotherapy.

Vulvar SCC is extremely rare, which is account for only about 2% of malignant neoplasm of the female genital tract and 3.3% of all SCCs. As the background vulvar dystrophy and lichen sclerosis are known to be as pre-existing phenomena in some patients [6]. Owing to the low incidence, randomized trials have not been carried out and current therapeutic strategies have been based on retrospective studies. The role of surgery has been done at the initial stages. At the early stage, single external beam radiotherapy or even brachytherapy has achieved excellent results in improving 5-year survival rate. However, 5-year survival rate in advanced cases are still poor, being 52.2% for patients with stage 2, 42.5% for patients with stage 3, and 20.5% for patients with stage 4A disease [7]. Only a few previous studies of concurrent chemoradiation therapy for primary SCC of the vagina have been reported, but the conclusions from these studies were limited by their small numbers [8]. Only one prospective study evaluating the use of single chemotherapy in the adjuvant setting was published by Bellati [9]. Consisting of single chemotherapy, radiation, or both of them, has been recommended.

In our presenting case, the patient had treated with high dose FP and TS-1 as the additional chemotherapy after surgical treatment without radiation therapy, and

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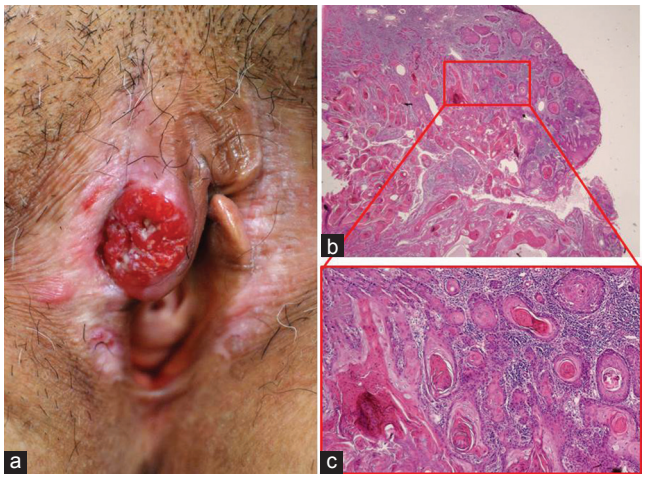

Figure 1: Clinical and histopathological findings. (a) A reddish nodule is observed on the right side of labia minor, (b,c) Histopathology findings showed atypical squamoproliferative lesion composed of atypical, keratinizing tumor islands and infiltrating strands with moderate- to high-grade nuclear atypical and miotic figures, consistent with squamous cell carcinoma. (Hematoxylin and eosin staining, original magnification x40 (b), x100 (c)).

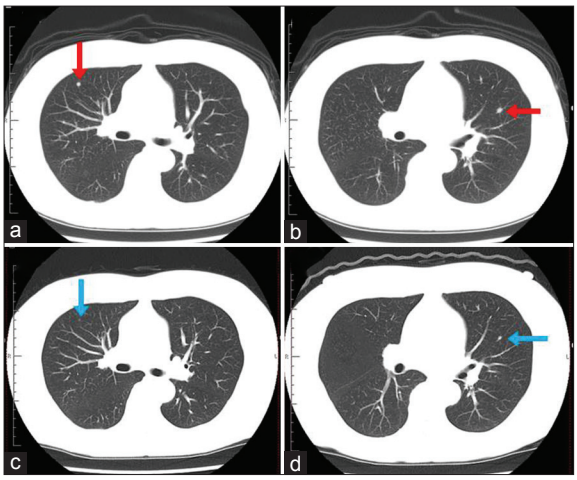

Figure 2: Computed tomography scan imaging. (a) (b) There are multiple pulmonary nodules in her lung one year after the primary operation, (c) (d) Multiple pulmonary nodules are kept for SD more than one year after finishing chemotherapy.

her condition has been kept for SD for three years up to the present.

In conclusion, the regimen of combination with high dose FP and TS-1 might be new treatment option for advanced vulvar SCC.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Letter to the Editor

A case of purpura annularis telangiectodes of Majocchi

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Sir,

Pigmented purpuric dermatoses (PPD) are a group of disorders which have histopathologically similar but morphologically different appearance. The five main clinical subtypes of PPD has been described. The perivascular lymphocytic infiltration of superficial dermal vessels with extravasations of red blood cells are common histopathological characteristics for all types of PPD. The etiopathogenesis of disease remains obscure yet [1,2]. We report a case of purpura annularis telangiectodes of Majocchi (PATM), a very rare type of PPD, with possible drug etiology of diclofenac sodium.

A 52-years-old man presented with asymptomatic skin eruption that had been present for two months. The patient had a history of oral diclofenac sodium intake ten days before the lesions appeared. He also had hepatitis B infection recognized at childhood and he had been followed by gastroenterology department with regular interval. The physical examination revealed several, symmetrical, nonpalpable, nonblanchable, erythematous, annular purpuric patches with a peripheral rim of dark red telangiectatic puncta distributed symmetrically on the upper legs and inguinal areas. The annular patches showed central clearing with minimal hyperpigmentation and atrophy (Fig. 1).

The laboratory investigations were within normal limits except hepatitis B antigen positivity. The histopathological examination revealed a moderately bandlike infiltration accompanying a prominent perivascular lymphocytic infiltration in superficial dermis. There was no evidence of vasculitis (Figs. 2A and 2B). Also, hemosiderin deposits was demonsrated within papillary dermis and macrophages

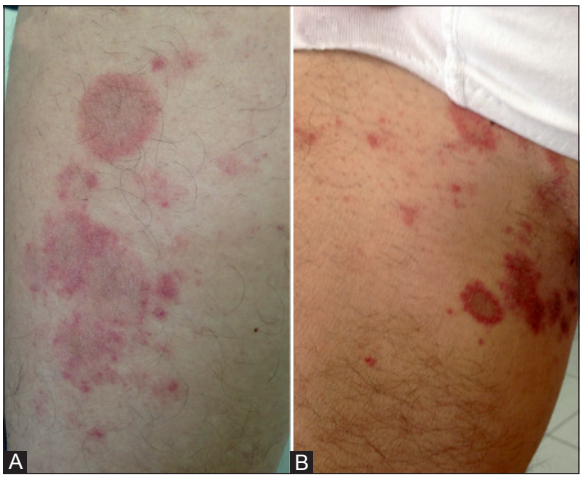

Figure 1: (A) Symmetrical, nonblanchable, erythematous, annular purpuric patches with central clearing and minimal hyperpigmentation and atrophy on the left and (B) inner side of upper leg.

by using prussian blue staining (Figs. 2C and 2D). These findings suggested a diagnosis of pigmented purpuric dermatosis. Together with clinical and histopathological appearance, diagnosis of purpura annularis telangiectodes of Majocchi was made. The drug was discontinued and topical corticosteroid was started as a treatment. Partial healing was obtained after 2 weeks of therapy.

PATM is an uncommon clinical variant of pigmented purpuric dermatosis [2]. A variety of diseases had been suggested to be associated with PPD, including Hepatitis B and C infection [3]. We consider that the skin eruption of our patient might not occur as a result of Hepatitis B virus, because he was positive for hepatitis B antigen since childhood but the lesions appeared first time two month before, after taking diclofenac sodium tablets.

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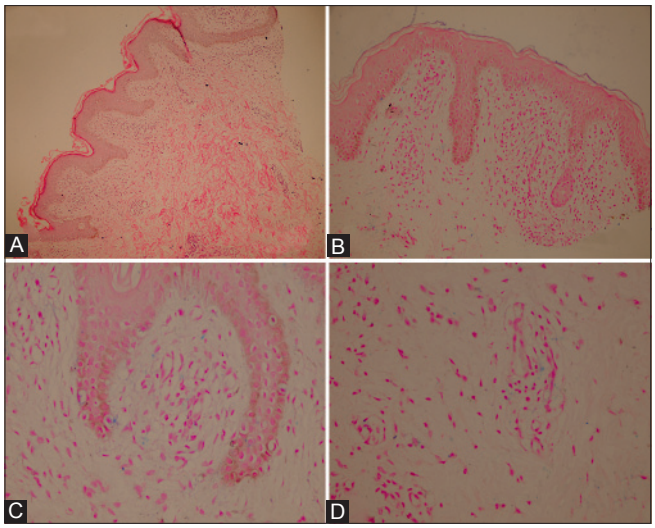

Figure 2: (A) A moderately bandlike and prominent perivascular lymphocytic infiltration in superficial dermis. (H&E, x100), (B) The closer view of infiltration, the pseudovasculitis appearence of lymphocytic infiltration encircling vessels without fibrin deposition (H&E, x200), (C,D) The presence of blue coloured hemosiderin within the papillary dermis freely or in macrophage system (Prussian blue, x40).

A wide variety of medications including NSAIDs, diuretics, sedatives, stimulants antibiotics, cardiovascular drugs, vitamins, antihyperglycemics, isotretinoin have been implicated as drugs responsible for the development of PPD [4,5]. It was suggested that as a result of antigen-antibody complex deposition in which drugs act as a hapten, an immune system-mediated vascular damage occur and this results in capillary leakage and erythrocyte extravasation [5]. The skin eruption of our patient appeared at 10th day of oral diclofenac sodium treatment. So, we thought it might be the possible cause of skin eruption in our patient. Also, as in our case, the drug induced cases are more likely to resolve after discontinuation of drug in contrast to chronic or relapsing nature of idiopathic ones [5]. Due to possible risk of provocation of a systemic reaction [4] we couldn't confirm the diagnoses by oral challenge test or patch test.

We intended to present this case because of the rarity of the PATM and it should be kept in mind that diclofenac sodium could play role in etiopathogenesis.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Our Dermatology Online

Letter to the Editor

Graham little picardi lassueur syndrome

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Sir,

A 27-years-man presented with progressive and wide spread hair loss over scalp, axillae and pubic area for the last 3-4 years. A multitude of therapies did not benefit him and the initial hair loss over occipital area had progressed to involve the whole scalp and other body sites. His medical history was unremarkable and no other family member had similar problem. Cutaneous examination (Figs. 1 - 4) showed smooth and whitish parchment-like scalp skin, areas of variable brownish pigmentation, atrophy, scarring, and minimal scaling, and was devoid of hair. The cicatricial alopecia involved the whole scalp with few hair strands and tufts of remnant hairs particularly at scalp margins. The skin interspersed between intact hairs too had similar texture. Hair pull test was positive. The eyebrows were sparse at lateral half. Numerous erythematous-brownish follicular papules were noted over scalp margins, beard area, neck, and whole trunk. There was complete non-cicatricial alopecia in both the axillae and the pubic region showed partial non-cicatricial alopecia. Nails and mucosae were normal. Systemic examination and routine laboratory parameters including complete blood counts, serum biochemistry, ANA and thyroid function tests were normal. Histology showed features of lichen planopilaris (Figs. 5 and 6). Treatment with systemic prednisolone 30mg/day was initiated after counselling for long-term follow up in view of protracted clinical course and prognosis.

Graham Little Picardi Lassueur Syndrome is a very rare presentation of lichen planopilaris. Clinically, the triad of progressive cicatricial alopecia of the scalp, non-cicatricial alopecia involving axillae and groin, and follicular keratotic papules on the glabrous

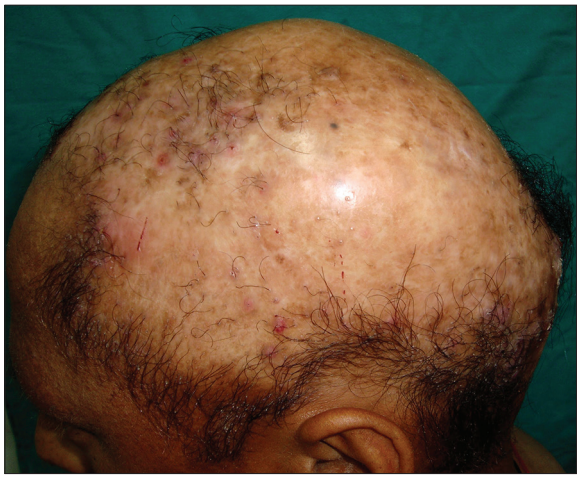

Figure 1: Late lichen planoplaris showing diffuse hair loss, whitish atrophic scarring, few follicular plugging, and residual single hairs and tufts of hair especially at scalp margins.

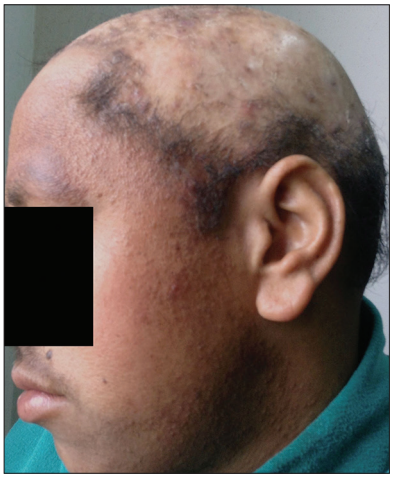

Figure 2: Red-brownish follicular papules over fronto-temporal area, cheek and side of neck. Note: alopecia involving lateral eyebrows.

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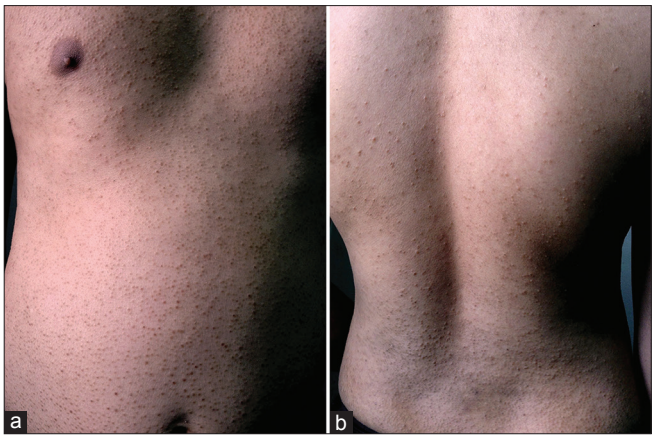

Figure 3: (a and 3b) Disseminated red-brownish follicular papules over front and back of trunk.

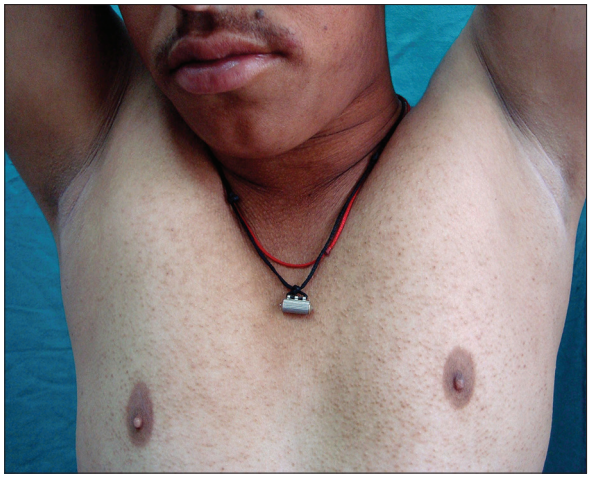

Figure 4: Non-scarring hair loss of axillae. Similar changes were noted over pubic skin.

skin is characteristic. These three clinical features usually appear concurrently but scalp alopecia may precede the follicular keratosis in most instances. Pruritus, when present, is often severe. Follicular inflammation destroys hair follicles permanently with hardly any possibility of hair re-growth causing substantial scarring alopecia and significant cosmetic embarrassment leading to anxiety, psychological distress and psychosocial morbidity necessitating treatment that is more aggressive. Its exact etiology remains obscure and there is no underlying systemic disorder except for one report of its association with androgen insensitivity syndrome [1]. Most patients are females between 30 and 70 years while young males are affected very rarely [2]. There is no racial predilection but familial cases have occurred [3]. Its association with the hepatitis B vaccination too has been speculated [4]. It is considered immune mediated on the analogy of its other more common variant, cutaneous or mucosal lichen planus that occurs

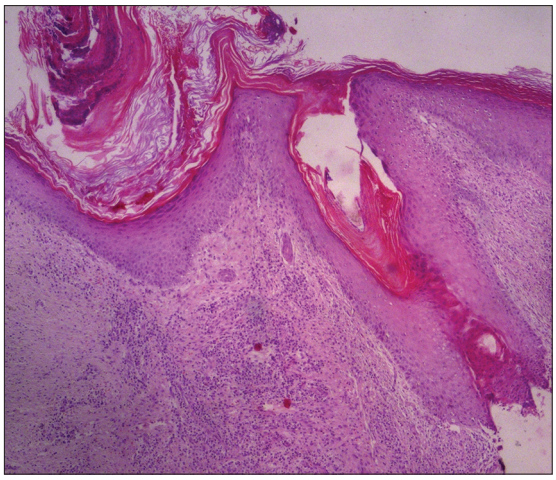

Figure 5: Histology shows infundibular hyperplasia, follicular plugging, wedge-shaped hypergranulosis, mild pigment incontinence, and dense perifollicular lichenoid infiltrate extending around its base (hugging type) and comprising mainly lymphocytes (H&E, x10).

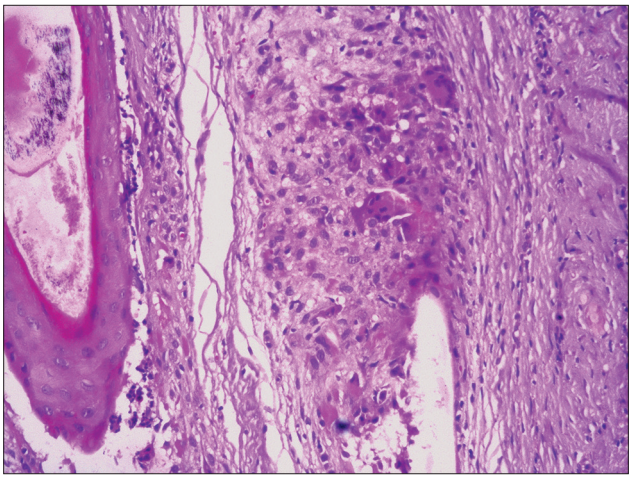

Figure 6: The lichenoid infiltrate is permeating lower follicullar epithelium and vacuolar changes of the outer root sheath are seen. (H&E, x40).

concurrently in about 50% cases [5]. The histologic features of infundibular hyperplasia, follicular plugging, wedge-shaped hypergranulosis, mild pigment incontinence, and dense perifollicular lichenoid infiltrate extending around its base (hugging type) are characteristic. Perifollicular lymphocytic infiltrate at the level of the infundibulum and the isthmus along with vacuolar changes of the outer root sheath are of early lichen planopilaris. More developed lesions show perifollicular fibrosis and epithelial atrophy at the level of the infundibulum and isthmus giving rise to a characteristic hourglass configuration. Alopecia with vertically oriented elastic fibres that replace the destroyed hair follicles is characteristic of advanced stage of the disease. The disease has a chronic unrelenting clinical course and needs differentiation

from folliculitis spinulosa decalvans, keratosis pilaris atrophicans, pityriasis rubra pilaris, pseudopelade of Brocq and discoid lupus erythematosus. The treatment is usually symptomatic and targeted to arrest progression of disease and alopecia. Topical, intralesional or systemic corticosteroids, oral retinoids, PUVA therapy, and antimalarials have been used with a limited success. Few reports on efficacy of cyclosporin (5 mg/kg/day) are also available [6]. However, the claimed efficacy of thalidomide in lichen planopilaris remains unsubstantiated [7,8].

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Letter to the Editor

Would you consider pilomatricoma as a differential diagnosis?

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Sir,

A 26-year-old man presented with a pedunculated, ulcerated, reddish and partially yellowish tumor of 6.5 cm in length by 6.0 cm in width by 3.0 cm in height on his left upper arm (Fig. 1a). The tumor had gradually developed during the half-year before the patient came to us on referral. There was no past history. We performed surgical removal of the entire tumor with a 1.0-cm margin, and the lesion was found to be a histopathologically poorly demarcated tumor without a capsule. There were darkly stained basophilic cells and so-called shadow cells with missing nuclei. There was no obvious nuclear atypia, and calcium deposits were found in the lesions. Based on these findings, we made the final diagnosis of pilomatricoma.

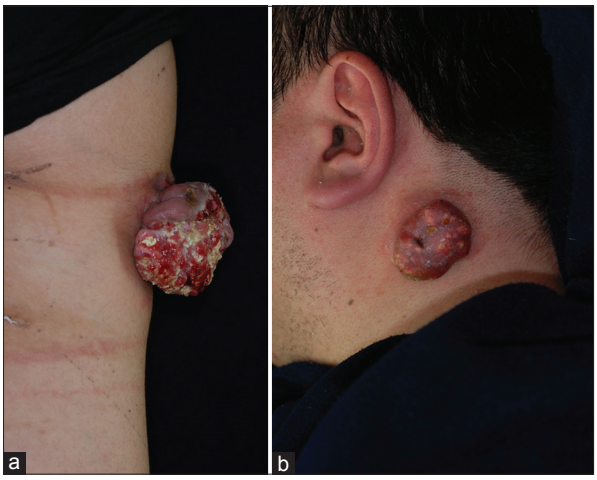

Figure 1: (a) A pedunculated, ulcerated, reddish and partially yellowish tumor of 6.0 cm in length by 5.0 cm in width by 4.0 cm in height is seen on the left arm. (b) A pedunculated, non-ulcerated, reddish tumor of 5.0 cm in length by 2.5 cm in width by 0.8 cm in height is seen on the left upper neck.

In a second case, a 42-year-old man presented with a pedunculated, non-ulcerated, reddish tumor of 5.0 cm in length by 2.5 cm in width by 0.8 cm in height on the left neck (Fig. 1b). We were able to see small yellowish dots on the surface. The tumor had gradually developed during the 3 months before the patient came to us on referral. The entire tumor was excisionally removed, and the surgical specimen showed the typical features of pilomatricoma.

Pilomatricoma is a common tumor derived from hair matrix cells; it is most often diagnosed in young children but may also affect adults. The clinical presentation is characterized by a subcutaneous nodule, usually up to 1 cm in diameter, with or without high mobility and calcification within the lesion that makes it feel hard and bony. For these reason it is easily diagnosed [1]. However, once the tumor is larger than 5 cm as the Case 1 and 2, they are called 'giant pilomatricoma', and it may become very difficult to make an accurate clinical diagnosis [2,3]. To date, several cases of giant pilomatricoma have been reported [4,5].

In Case 1, we initially suspected sebaceous carcinoma, squamous cell carcinoma or dermatofibrosarcoma protuberans, because of the unusual clinical manifestations. However, we considered pilomatricoma as an initial diagnosis when we encountered the Case 2, so we were able to avoid over-treatment on Case 2.

In conclusion, we should keep giant pilomatricoma in mind as a differential diagnosis when we see pedunculated, firm reddish tumor of >5.0 cm in size, with or without ulceration. This knowledge can avoid misdiagnosis and unnecessary examination and treatment.

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Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Letter to the Editor

Cutaneous creeping eruption in a child

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Sir,

A 6 year-old boy from coastal area presented with history of intensely pruritic skin lesions over the right forearm for last 3 days. There were no other symptoms. The boy used to play on the beach barefoot daily. Clinical examination revealed an erythematous, serpentine lesion on the dorsal aspect of the right forearm (Fig. 1). Systemic examination was unremarkable. Hemogram was normal except for eosinophilia (absolute eosinophil count-1400/mm^3). A diagnosis of cutaneous larva migrans was made and he was treated with single dose of albendazole (400 mg) and ivermectin (6 mg). Lesion had healed during his subsequent followup after 1 week.

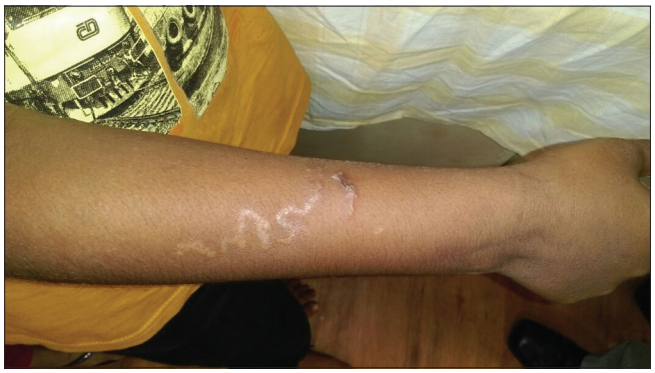

Figure 1: Raised curvilinear serpentine lesion of cutaneous larva migrans.

Cutaneous larva migrans (CLM) also called creeping eruption, plumber's itch, is characterized by classical serpentine skin lesions in a tropical setting [1]. Bare foot walkers, farmers, children playing in beaches, sandy and moist arears are at high risk. CLM is mainly caused by infection with larvae of animal hookworms like *Ankylostoma caninum* and *A. braziliens*. Other offenders include *A. ceylonicum*, *Bubostomum phlebotomum* etc [2,3]. Larval penetration of skin and migration cause itchy erythematous, raised vesicular and serpentine cutaneous lesion. The disease is usually self-limiting and lasts for 4-6 weeks until the larva dies and humans are accidental and 'dead-end' host [2,4]. Severe infestations manifest as Loeffler's syndrome of pulmonary eosinophilia and rarely as eosinophilic enteritis [5]. Biopsy is of no value as the larvae advance ahead of the clinical tract. Optical coherence tomography is a non-invasive modality for diagnosis [6]. We treated with a single dose of ivermectin and albendazole [7]. Other treatment regimens include oral albendazole (400 mg) daily for 3 days and topical application of 10% thiabendazole [8]. CLM can be prevented by avoiding skin contact with soil contaminated with animal feces and adequately covering the feet when visiting sandy and moist areas.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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Letter to the Editor

Perinatal varicella

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Sir,

Neonatal varicella infection has been rarely reported since vaccine introduction. Neonatal varicella occurs in the first 28 days of life and is due to infection developed by mother during last 5-7 days of delivery and 5- 7 days post delivery or in case of similar infection acquired by other siblings [1,2].

Neonatal varicela should be differentiated from "fetal varicella syndrome, congenital varicela with high morbidity and mortality.

Rapid and accurate diagnosis is of great importance on therapy and prognosis.

Two cases of neonatal varicela treated with systemic acyclovir are presented.

Case 1: A 28-day old female patient was seen in Dermatology Hospital for vesicular rash distributed on the limbs and trunk, few vesicles were scattered mostly on the superior part of the body (Fig. 1).

No systemic symptoms, usual lab analysis within normal limits for the age. Tzanck smear showed multinucleate giant cells, history of recent chicken pox in older brother was admitted by the mother, and serology confirmed the diagnosis of neonatal varicella.

Good response and complete recovery was achieved under systemic administration of acyclovir 20mg/kg iv 8 hourly for 10 days.

Case 2: A 17-day old male premature born baby was referred to Dermatology Unit for vesicular lesions

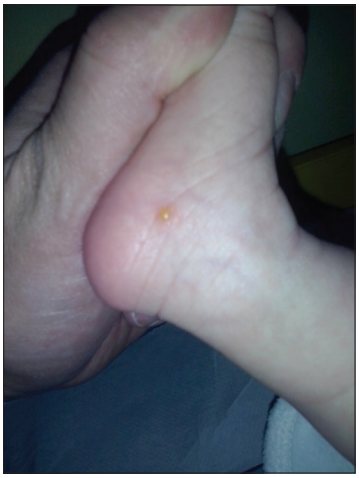

Figure 1: Vesicle on the left foot.

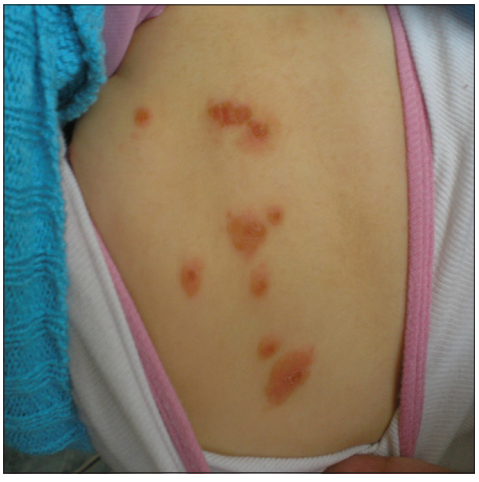

Figure 2: Crops of vesicles on the trunk.

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spread on the limbs, trunk and scalp accompanied by high fever (Fig. 2). His mother was treated for varicella one week before admittance to the hospital, meaning approximately 10 days after delivery.

Based on clinical aspect of the lesions, close contact with mother infected with varicella 10 days postpartum, the presence of typical multinucleate giant cells on Tzanck smear a diagnosis of neonatal varicella was admitted and a treatment with acyclovir iv was started. Serology was negative. For the following 14 days the baby boy has received acyclovir 20 mg/kg 8 hourly with good response and no complications. A close follow-up was recommended.

DISCUSSION

If maternal infection occurs during 8–20 weeks of pregnancy fetus has a risk of developing varicella embryopathy known as "fetal varicella syndrome" characterized by ophtalmological, muscle and skeletal anomalies, mental retardation, microcephaly, urological defects [3,4].

Congenital varicella occurs if the mother got the infection at 13-20 weeks gestation and presents in new borns with skin scars, growth retardation, limb defects, chorioretinitis, neurological involvement [1]. The prognosis is poor, mortality high, around 30% [2].

When the maternal infection occurs after 20 weeks of gestation some reports highlighted an increased risk of herpes zoster during childhood [5].

Neonatal varicella occurs in the first 28 days of life and is due to infection developed by mother during last 5- 7 days of delivery and 5- 7 days post delivery or in case of similar infection acquired by other siblings [3,4].

Two types of neonatal varicela exist: if the mother has been infected less than 5 days before birth or 2 days after delivery, varicella-associated antibodies are not transmitted to the newborn and the infection is severe; if the mother has got the infection more than 5 days prior to birth, antibodies have been transmitted to the child causing a less severe form of the disease [2].

It is of outmost importance to consider the moment of varicela virus infection that will guide the therapeutical approach.

Maternal treatment includes oral acyclovir within 24 h of onset [6].

If maternal infection occurs 5-7 days before delivery or 2 days after giving birth a prophylactic approach to the child is mandatory using varicella zoster immunoglobulin or intravenous acyclovir, if immunoglobulin therapy is not available or the diagnosis was made 96 hours after exposure [6].

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Letter to the Editor

The fascination of mineral pigments in organic and natural eye shadows and compact cakes: are they risky or innocuous?

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INTRODUCTION

A recent study conducted at the Ahmadu Bello University of Zaria, Nigeria [1] refers that different samples of made in China eye shadows randomly selected from products available in the shops at several towns of Nigeria, were analysed to determine the levels of heavy metals (Pb, Cd, Ni, Cu, Zn, Cr, Co, and Mn).

Moreover an avalanche of experiments conducted all over the world have been showing since immemorial times that a plentitude of decorative make up products for eyes that had been examined contained huge amounts of hazardous heavy metals [2].

Here follows Table 1 where the results of these analysis are referred [2].

Purpose of my research, indeed, is to determine the levels of certain minerals that are usually included (because legally admitted) in the formulas of eye shadows, kohls and encaustics (crayons) which are retrievable in human blood after application onto eyelids, eyelashes and eyebrows of the aforesaid decorative products.

I intend to discover the pool of heavy minerals and other chemical constituents that are reputed dangerous for Human health in volunteers' blood, even though my attention is not focused on the accidental presence of heavy metals, but actually on the same metals that are admitted in the Strasbourg's International Nomenclature Cosmetics Index, and therefore are designed as inorganic pigments.

Table I: Elemental Results for 30 Eye Shadows from 19 Manufacturers: Mean results (mg/kg)

Sample	As	Cd	Cr	Co	Pb	Hg	Ni
ά	0.36	0.07	18	5.5	5.0	NF	16
έ	0.62	NF	8.5	7.9	4.4	NF	16
ή	0.65	0.14	15	8.1	3.9	TR	20
ί	0.38	TR	3.8	1.9	4.7	0.07	2.9
ΰ	0.50	NF	7.7	4.0	3.4	NF	9.4
α	0.19	NF	2.0	0.35	0.05	NF	0.91
β	0.78	NF	7.7	1.6	6.5	0.008	3.1
γ	0.36	NF	12	3.7	0.66	0.04	8.2
δ	0.63	0.36	8.3	1.7	6.7	0.0075	4.8
ε	0.24	NF	4.5	0.88	4.7	TR	2.5
ζ	0.25	TR	11	5.3	0.77	0.03	42
η	TR	NF	3.9	1.3	3.1	NF	4.8
θ	0.45	TR	21	2.8	5.7	0.03	9.6
ι	0.62	NF	18	5.5	4.1	0.0085	15
κ	0.62	NF	9.6	2.1	3.6	0.006	7.5
λ	0.30	NF	1100	2.7	4.2	NF	13
μ	0.41	NF	7.4	2.4	3.4	0.01	8.8
ν	1.0	NF	11	1.6	5.9	NF	10
ξ	0.36	TR	12	7.8	27	TR	32
ο	0.33	NF	16	0.94	3.9	0.02	12
π	0.69	TR	420	4.9	8.6	0.0085	10
ρ	1.7	TR	350	64	2.1	TR	1600
ς	0.47	TR	22000	1.1	2.4	NF	17
σ	0.26	0.11	6.6	6.8	2.8	0.02	14
τ	0.07	NF	4.7	0.31	2.2	NF	5.0
υ	0.79	TR	3500	2.9	7.5	0.015	16
φ	0.29	NF	40	13	4.2	TR	18
χ	0.48	TR	5.1	2.4	5.7	TR	9.4
ψ	1.2	TR	32	0.64	14000	40	9.0
ω	NF	NF	1.1	0.11	0.11	NF	1.3

All the inorganic pigments useful to decorate eyes belong to the category of colours (designed as Colour Index, whereof the acronym is C.I.) that is comprised between the progressive number 77000 and 77999, throughout the entire world, unless only a minor part of those is allowed by EC and is referred below, quite completely (Box 1):

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77000 White, Aluminium powder	77335 Blue Mixture of cobalt zincate and zincoxide
77002 White, Aluminium hydroxide sulphate	77343 Blue Cobaltous/chromium/alumino oxide
77004 White, Kaolin	77346 Blue Cobaltous aluminate (Dresden blau or bleu de Thénard)
77007 Blue, Lazurite (Ultramarine)	77357 Yellow Potassium cobalt nitrite
77015 Red, Aluminium silicate plus Ferric oxide	77368 Blue Cobaltous stannate
77120 White, Barium sulphate	77377 Blue Cobalt titanate (Cobaltous/nickel/zinc/titanium alumino oxide
77163 White, Bismuth oxychloride	77400 Brown, Copper dust
77199 Yellow Calcined Cadmium Sulphide	77480 Brown, Burnish gold, AURUM
77205 Yellow Cadmium zinc sulphide	77489 Orange, Orange Iron oxides (ferrous)
77220 White, calcium carbonate, Rügener kreide	77491 Red, Red Iron oxides (ferric)
77231 White, calcium sulphate dyhidrate	77492 Yellow, Yellow Iron oxide (ferrous)
77266 Black, Carbon black, Nerofumo, (benzopyrenes, benzo[ghi] perylene, coronene, fluoranthene and pyrene can be adsorbed onto carbon black particle surface, and are recognized as human carcinogens.Carbon black materials are categorized as acetylene black, channel black, furnace black, lampblack or thermal black according to their manufacturing process.Opinion of SCCS,12 December 2013)	77499 Black, Black Iron oxide (ferric)
77267 Black, Bone charcoal	77510 Blue, Ferric ferrocyanide
77268 Black, Carbo vegetabilis, cendres végétales	77520 Blue Ammonia ferrocyanide (Japan)
77288 Green Chromium sesquioxide	77713 White, magnesium carbonate,
77289 Green Hydrated chromium sesquioxide	77742 Violet, Mangan (iii) ammonium-Diphosphate
77310 Brown Antimony/chromium/titanium oxide	77745 Red, Trimanganese bis (orthophosphate)
	77820 White, Silver dust
	77891 White, Titanium dioxide
	77947 White, Zinc oxide

We want to stress that pigments, with relative C.I. number, written in bold italics are those that are to be reputed toxic and risky, even though I have recorded even the pigment C.I. 77520, that is extremely carcinogenic, that is fully admitted in Japan and in many other countries and consequently eye decorative products containing it are globally retrievable in the markets and boutiques of all the world.

MATERIALS AND METHODS

In order to determine how the matrix of excipients and bulking agents can be reputed responsible of the release of the same heavy metallic ions, I have chosen three different formulas of eye shadows, (designed as pressed powders or compacts or cakes) which contain diverse inert powders, starch, a natural binder and the mineral pigments.

The first of those is considered a fully "classic and chemical fard aux paupières" and the other two are designed as "organic and natural metallic compact powdered cakes".

It is suggestive to emphasize that starch and the natural binder, that is the caprylic/capric glycerides, appear always in all the three formulations, that the pigments are always the same but only two ingredients, corresponding to the inert powders, change in the formulas and exactly these are supposed to interfere with the same release, since they can behave as linkers and sequestrers of the metallic ions.

Formula num ONE:
Talc
Zinc stearate
Zea mays starch
Caprylic/capric glycerides

Formula num TWO:
Shellac
Calcium Magnesium silicate
Zea mays starch
Caprylic/capric glycerides.

Formula num THREE:
Karaya gum
Magnesium carbonate
Zea mays starch
Caprylic/capric glycerides

The following is the list of the pigments I have employed to give four compact eye shadows blue with golden sparkling and fluorescent reflections:

a. Cadmium zinc sulphide
b. Chromium sesquioxide
c. Cobaltous aluminate (Dresden blau or bleu de Thénard)
d. Cobaltous stannate
e. Ammonia ferrocyanide

The total amount of the powdered pigments represents the 7.00% in each formula;the remnant is constituted by inert powders, bulking agents and the binder.

We have a class of galenic preparations laboratory and the class numbers 21 pupils, 18 of whom are female and the other 3 are male.

Even the boys do not distaste to wear eye shadow when go out to dance on Saturday night and so it has been easy to let them prepare the three formulas and pray them to decorate their own eyelids by the aids of three simple sponge-tipped plastic rods (in order not to inquinate the different products) and to undergo two mineralogram blood tests, the former before and the latter after the application of the cosmetic products onto their eyelids.

It is well known that skin penetration of whichever chemical substance is performed through the conjunctival sac and through the stratum corneum of the eyelid itself, that is the thinnest epidermal layer in all the human body.

Since these toxic substances are often ingested or inhaled accidentally a list of tolerable limits of their presence in blood exists and is to be considered official; and therefore is plotted in Table II:

The three types of coloured cakes (pressed powders) were prepared during the class of laboratory preparations and were shared in 63 small tureens, and each of every pupil had three tureens available, to spread onto his own eyelids in three different occasions, during the Carnival feasts and precisely on Thursday, on Sunday and next Tuesday night (Jeudi Gras, Samedi Gras and Mardi Gras).

The successive morning of every night of amusement, at 7.00 a.m., before classes, the pupils were invited to have their blood examined to detect the plot of heavy minerals and tiocyanates.

Here follow Tables III and IV where all the scores regarding the classic and chemical "compact fard aux paupières" and regarding the organic and natural compacts cakes are reported:

Table 2: Standard Levels and tolerated limits of Heavy metals in human serum according to Test 34506 :Heavy Metals Screen with Demographics, Blood Clinical Information Mayo Foundation for Medical Education and Research

Type of chemical substance	Pool of chemical substance tolerated in non exposed individuals
Cadmium	4 µg/lt
Chromium	0.05-0.5 µg/ml (serum)
Cobalt	1.8 µg/lt
Tin	2 ng/ml
Ferrocyanide (as thiocyanate, biotransformed in the liver)	15-70 µmol/lt

Table 3: Values of risky substances levels retrieved in blood, after application of the chemical eyeshadow (Formula num ONE)

Pupils	Cadmium µg/lt	Chromium µg/ml	Cobalt µg/lt	Tin ng/ml	Tiocyanates µmol/lt
1F	4.5	0.7	2.0	2.2	77
2F	4.2	0.9	2.1	2.2	81
3F	3.9	0.2	2.2	2.3	88
4F	5.0	0.6	2.4	2.1	92
5F	4.6	0.1	1.9	2.8	65
6F	4.4	0.09	2.3	2.3	89
7F	3.8	0.4	2.2	2.7	93
8F	4.7	0.6	2.1	2.5	67
9F	4.3	0.8	2.2	2.0	99
10F	4.1	0.6	2.4	2.1	91
11F	4.0	0.8	2.2	2.0	78
12F	5.1	0.9	2.0	2.8	83
13F	4.2	0.08	2.1	2.9	77
14F	3.8	0.6	2.1	2.3	82
15F	4.3	0.7	2.3	2.1	75
16F	4.1	0.5	2.7	2.5	69
17F	5.0	0.47	2.4	2.3	73
18F	4.9	0.08	2.1	1.8	88
19M	3.3	0.03	1.8	1.4	101
20M	3.7	0.04	1.7	1.5	105
21M	3.1	0.1	1.8	1.9	121

Table 4: Average values of risky substances levels retrieved in blood, after application of the organic natural eyeshadows (Formula num ONE)

Pupils	Cadmium µg/lt	Chromium µg/ml	Cobalt µg/lt	Tin ng/ml	Tiocyanates µmol/lt
1F	3.8	0.14	1.1	0.2	18
2F	3.3	0.13	1.4	0.3	21
3F	2.9	0.14	1.6	0.1	22
4F	3.7	0.16	1.7	0.4	34
5F	3.9	0.08	1.8	0.6	11
6F	4.0	0.09	1.3	0.2	23
7F	3.3	0.18	1.6	1.0	24
8F	3.6	0.09	0.9	0.8	17
9F	3.7	0.13	0.9	0.5	16
10F	4.0	0.12	1.1	1.1	22
11F	3.2	0.17	1.6	1.4	36
12F	2.9	0.09	1.7	0.8	23
13F	3.1	0.12	0.9	0.9	17
14F	3-0	0.13	1.1	0.7	14
15F	2.8	0.07	1.3	1.2	15
16F	3.3	0.05	1.4	1.6	11
17F	3.1	0.1	1.0	1.4	14
18F	3.7	0.03	1.5	1.2	15
19M	4.1	0.02	1.8	1.9	56
20M	4.2	0.01	1.7	2.1	65
21M	3.9	0.04	1.8	1.9	77

DISCUSSIONS

It is evident that formula num ONE that contains Talc and Zinc stearate, as prime bulking agent and excipient, releases substantially the metallic ions and the ferrocyanides, cause of their intrinsic chemical structure, being inert powders, that is that are not prone to sequestrate the ions, as no free electric charges are available on their molecules.

Therefore high levels of Cadmium, Chromium, Cobalt, tin and ferrocyanide (as tiocyanates) are present in the blood of volunteers that applied this chemical formula of pressed powder.

On the other hand, the natural cakes can boast about the presence of gum resins and bioplymers as shellac and/or karaya gum.

Shellac, a resin secreted by the female lac bug, on trees in the forests of India and Thailand, according to manifold A.A. [3-7]. contains polyhydroxy acids and it has been suggested that these might occur as lactides or as other inter-esters‘ in producing the resin., giving its peculiar liquid-viscous texture, with the capability of linking bivalent and polyvalent metallic ions.

Karaya gum, instead, is a vegetable gum produced as an exudate by trees of the genus *Sterculia*. Chemically, gum karaya is an acid polysaccharide composed of galactose, rhamnose and galacturonic acid, so that the carboxylate ions are capable to capture and sequester the risky ions and ferrocyanides.

It is suggestive to observe that as far as tiocyanates levels (derived from the biotransformation of ferrocyanides in liver and detectable in human plasma) are higher in man than in female, and several are the A.A. which have attempted to clarify this obscure phenomenon [8,9].

Finally, male can more easily intoxicated by Cobalt and Tin, and this matter of fact is utterly unclear.

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Our Dermatology Online

Historical Article

Nomenclature in medicine; a perspective

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There are several types of nomenclature used in medicine [1-3]. These include eponyms and acronyms. This journal already published two supplements for eponyms in dermatology.

There are, also, several origins for the medical terms. This may include places (e.g. Lyme disease) or food (e.g. Salmon patch) [4].

In this manuscript, We shall present a perspective on medical terminology.

TERMS ORIGINATED FROM ANIMALS

There are several names in medicine which originated from animals names. The genes in humans have been named after some of the animals like mice, drosophila, and hedgehog [5].

An example for a term related to animal is ''Elephant man syndrome''.

Sir Frederick Treves first showed Joseph Merrick, (Fig. 1), the famous Elephant Man, to the Pathological Society of London in 1884.A man with gigantic growth, thought to have neurofibromatosis or Proteus syndrome [5,6].

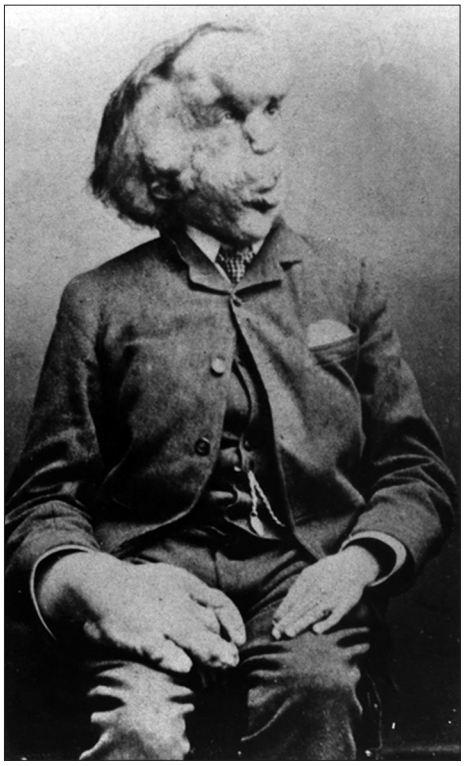

Figure 1: Joseph Carey Merrick (1862-1890).

NAMES ORIGINATED FROM FAMOUS CHARACTERS OR STORIES

Researchers who named the medical things are also influenced by public characters or stories. So it is not strange to find some medical terms based on non-medical things.

For example; Kabuki syndrome, which is a congenital disorder with multiple anomalies and intellectual disability. It is named Kabuki Syndrome because of the facial resemblance to Kabuki actor's mask [7], (Fig. 2). Kabuki is a Japanese traditional theatrical form.

Another example for the above subheading is Rapunzel syndrome, which is a gastric trichobezoar with a tail extending up to the jejunum, ileum, or ileocecal junction [8]. It was first described by Vaughan et al. in 1968 It is named after the eponymous heroine of a German fairy tale written by the Grimm Brothers in 1812 about a 12-year-old princess imprisoned by a witch in a tall tower with neither stairs nor doors for many years; the princess lowered her long hair to the ground

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from her window, allowing a young prince to climb up and rescue her [8] (Fig. 3).

NAMES ORIGINATED FROM A PATIENT'S NAME OR THINGS RELATED TO THE PATIENTS

Rarely a disease may be named after something related to the paients. For example; Ambras syndrome (AS), which is a form of hereditary, generalized hypertrichosis. The Ambras name was given because the family portraits of Gonzales (The first recorded case of AS) were discovered in Ambras castle, (located in Austria) amongst an art collection started by the archduke Ferdinand II (1529-1595) [9].

Petrus Gonzales (Fig. 4) was born in the Canary Islands in 1556. Out of curiosity, Petrus was brought to France where he was presented as a gift to the nobles. He subsequently produced offspring with similar AS features. Currently, the same paintings of Gonzalez which were in Ambras castle, hang in the Kunsthistorisches museum in Vienna [9].

Figure 2: Two Kabuki actors.

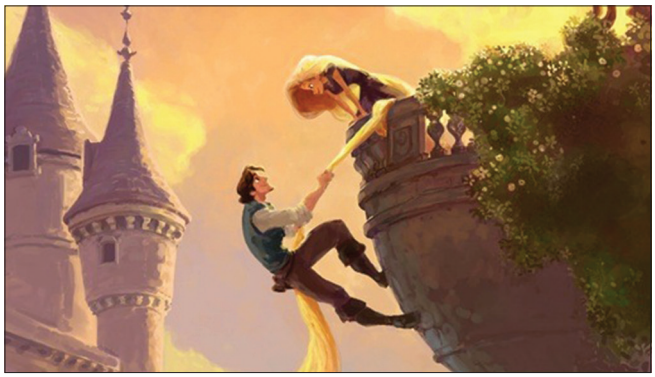

Figure 3: Rapunzel story. From: Walt Disney Studios Motion Pictures and New Media Strategies.

Figure 4: Petrus Gonsalvus.

Figure 5: Henry Louis Gehrig (1903-1941).

Table 1: Selected medical terms in which the name of the disease refers to the patient's name

The term	Remarks
Auberger's blood group[10]	This is a type of human blood group in which the Aua antigen is expressed. It is found in 82% of Caucasians. It may be related to the Lutheran antigen system The blood group was named after patient Auberger, who was a 59 year old French woman with oesophageal varices
Christmas disease [11]	Named after Stephen Christmas, the first patient described with this disease in 1952 .This term is a synonym for Hemophilia B, which is a blood clotting disorder
Cowden disease [12]	This is a synonym for multiple hamartoma syndrome, which is an autosomal dominant genodermatosis. It was first described in 1963, and named after a young women ''Rachel Cowden'', and her family in which it was first reported
Galli-Galli disease [13,14]	It is a rare genodermatosis in the spectrum of reticulate hyperpigmentation.It is regarded as an acantholytic variant of Dowling-Degos disease. It was originally reported by Bardach, Gebhart, and Luger in 1982. The term Galli-Galli is an eponym, derived from the family name of the two brothers being originally described with this genodermatosis
Hartnup disorder [15]	This is an autosomal recessive abnormality of renal and gastrointestinal neutral amino acid transport .Clinical features include photosensitive pellagra-like skin rash, cerebellar ataxia and other neurological symptoms. It is named after an English family described in 1956
Lou Gehrig's disease [16]	It is another name for amyotrophic lateral sclerosis, which is a late-onset neurodegenerative disease. Named for Lou Gehrig, [Figure 5]; a famous baseball player. He was diagnosed in 1939 and passed way, at the age of 37 in 1941 due to the disease
Mortimer's disease [17]	This term might be applied to a case of polymorphic cutaneous and systemic sarcoidosis. In 1898, Jonathan Hutchinson (1828-1913), an English physician, coined the term 'Mortimer's Malady'. He coined the term after the name of his patient Mrs. Mortimer who presented with multiple, raised, dusky-red, non-ulcerative and persistent patches, which Hutchinson considered different from tuberculous affliction. Though other eponyms like 'Boeck's sarcoid' have also been in vogue, now the term 'sarcoidosis' is the most accepted for this condition

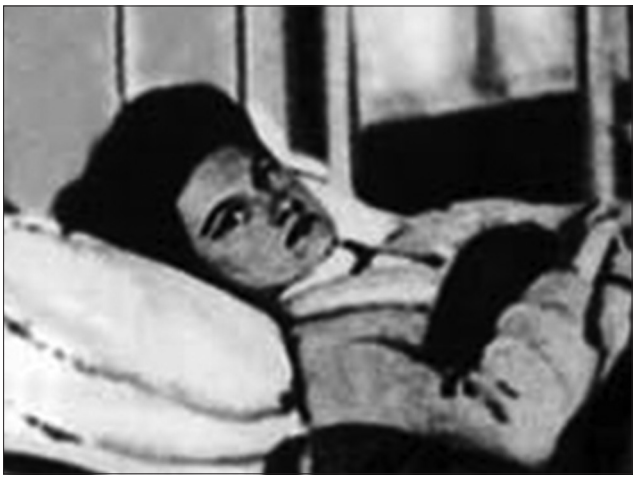

Figure 6: Mary Mallon (1869-1938).

Patients have been immortalized by having their names or initials incorporated; for example, B-K mole syndrome, where the letters B and K refer to the two patients in whom the condition was first described. The same is true of anti-Sm, anti-La, and anti-Ro antibodies, each of which is derived from letters of a patient's name. In table1 [10-17], I listed selected medical terms in which the name of disease refers to the patient's name.

Out of curiosity a reverse might happens, and a person might carry the name of the disease.This is what happened to Mary Mallon (1869-1938), (Fig. 6), who was the first person in the United States identified as an asymptomatic carrier of the pathogen associated with typhoid fever. Mary Mallon was then best known as Typhoid Mary [18].

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ADOLFO ARTHUR NOUEL

ENFERMEDADES DE LA MUCOSA ORAL

Atlas

Atlas de Enfermedades de la Mucosa Oral

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ÍNDICE 512

Prólogo

Es un gran honor escribir el Prólogo del libro "Atlas de Enfermedades de la Mucc
lista en Cirugía Bucal y Ortopedia Maxilar y Médico Estomatólogo, Dr. Adolfo Ar
vendrá a enriquecer el no muy amplio cúmulo de información bibliográfica y grá
campo de la medicina. Lo acepto, siendo lo más importante, más por razones de
por conocimientos específicos del tema, cuyo análisis formal lo dejo a la consider
También lo hago con una visión de compromiso social, ya que el ejercicio de la pr
solamente una actividad individual, sino que más bien requiere vivenciar los distint
en la vida cotidiana, los cuales tienen influencia en el desarrollo de la ciencia y tec
Es una obra completa y actualizada con temas en constante evolución, con múltiple
que sus afecciones clínicas producen alteraciones morfológicas tisulares con signc
En mi calidad de Director General del Instituto Dermatológico y Cirugía de Piel '
Diaz" (IDCP), he crecido en la búsqueda desinteresada del conocimiento, del rigc
la investigación, de la reflexión en torno a los valores y de la dimensión ética de l
ahí, nuestra deuda de gratitud con el profesor meritísimo de la Facultad de Cien
Universidad Autónoma de Santo Domingo, Dr Arthur Nouel; ya que la dermatol
medades de la mucosa oral y de la mucosa genital, convirtiéndose en un referen
público general, pero muy especialmente para los médicos en formación de nuestra
en cursos de postgrado en dermatología de la República Dominicana.
Con más de 48 años de carrera docente, profesor invitado a numerosos Congresos
cionales, su dedicación institucional al Patronato de Lucha Contra la Lepra y el Ins
Dominicano y Cirugía de Piel "Dr. Huberto Bogaert Díaz", su trabajo hospitalario y
la calidad suficiente para la autoría de esta obra.
Esta primera edición viene a llenar un vacío en el haber científico Latinoamer
eminentemente didáctico, con una presentación vanguardista, de lectura sintética
diversa, traduce en sí el trabajo diario del autor, fruto de sus manos hábiles y expe

En 11 capítulos y 520 páginas, el profesor Arthur Nouel nos entrega en su libro nítidas e impecables ilustraciones fotográficas representativas de la patología de la mucosa oral, de manera concisa y ordenada, apoyada en un contenido temático con más de 250 citas bibliográficas. Con una excelente puerta de entrada, el capítulo primero muestra los elementos esenciales de las manifestaciones semióticas de las áreas anatómicas, la inspección, la palpación y la olfacción que, complementadas con las pruebas diagnósticas sugeridas, colocan al lector en el camino correcto en el abordaje diagnóstico del enfermo.

Las lesiones elementales representativas de las características clínicas o signos clínicos y morfológicos de las enfermedades de la mucosa oral, se tipifican de manera magistral en el segundo capítulo. Su división y clasificación se expresan en un contenido llano y visual exquisitos. Fruto de su rica experiencia, a partir del capítulo tercero, el autor se fundamenta en la morfología de las lesiones orales para agrupar en cada uno de ellos una verdadera expresión del conjunto de afecciones inflamatorias, infecciosas, traumáticas, displásicas, artefactas, tumorales benignas y malignas que convierten este texto en una referencia obligada en la lectura de las enfermedades de la mucosa oral.

Promoviendo un enfoque de direccionamiento estratégico; al finalizar el capítulo 11, el Dr. Arthur establece diagnósticos certeros, lo que reduce la probabilidad de error en la práctica médica, al mostrarnos tablas de Diagnóstico Diferencial, donde se podrá inspeccionar la distinción entre dos o más Enfermedades de la Mucosa Oral de síntomas similares mediante la comparación sistemática de dichos síntomas.

Finalmente, parafraseando a Francis Bacon, escritor y filósofo; aprovecho para desearle al autor éxitos y unirme a las voces de felicitación que imagino por cientos, y también animarle a que persevere en su labor docente y médico en ejercicio, dado que ya a estas alturas de la vida debe saber que "la lectura hace al hombre completo; la conversación lo hace ágil y el escribir lo hace preciso".

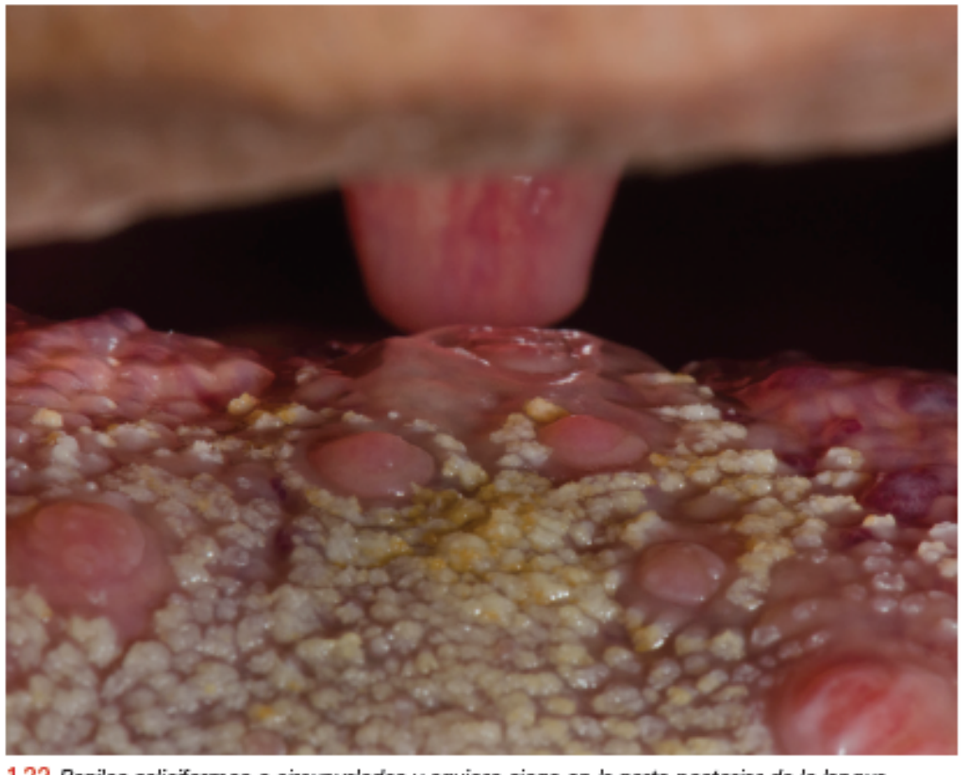
Con la puesta en circulación de este libro "Atlas de Enfermedades de la Mucosa Oral", en víspera de la conmemoración del 50 aniversario del IDCP, bajo la autoría de un miembro ilustre de la familia del Patronato de Lucha contra la Lepra/IDCP, tenemos motivos para celebrar con júbilo la publicación de este libro, conscientes de que es una nueva contribución fortaleciendo el sistema de salud como elemento básico para el desarrollo de las poblaciones afectadas.

Rafael A. Isa Isa

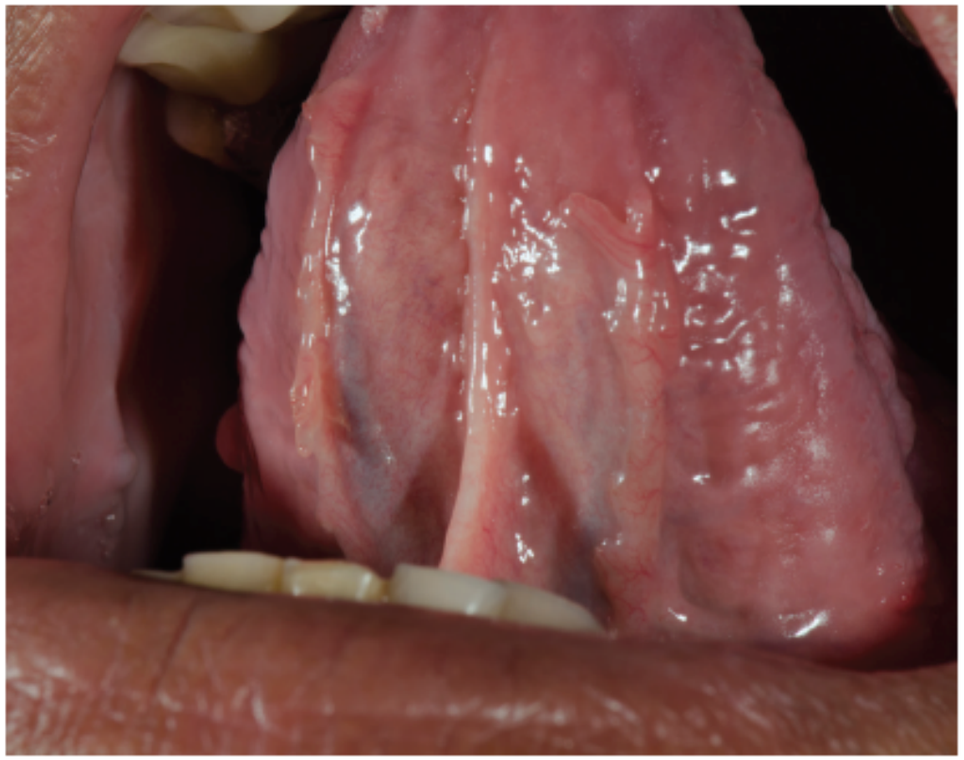
1 Examen del paciente

El examen del paciente se inicia desde que éste ingresa a la consulta. Observar como camina, se sienta y expresa su queja principal. La historia de la enfermedad es de fundamental importancia para llegar al diagnóstico presuntivo. Los antecedentes personales y familiares deben incluirse en el interrogatorio. Examinar cuidadosamente la cara, aspecto y proporciones de todas sus partes, asimetrías verticales u horizontales, la piel que la recubre, la cantidad y aspecto del pelo en las regiones habituales, los pliegues naturales, movimientos de los ojos, la conjuntiva y si la nariz presenta desviaciones o si por ella sale alguna secreción espontáneamente.

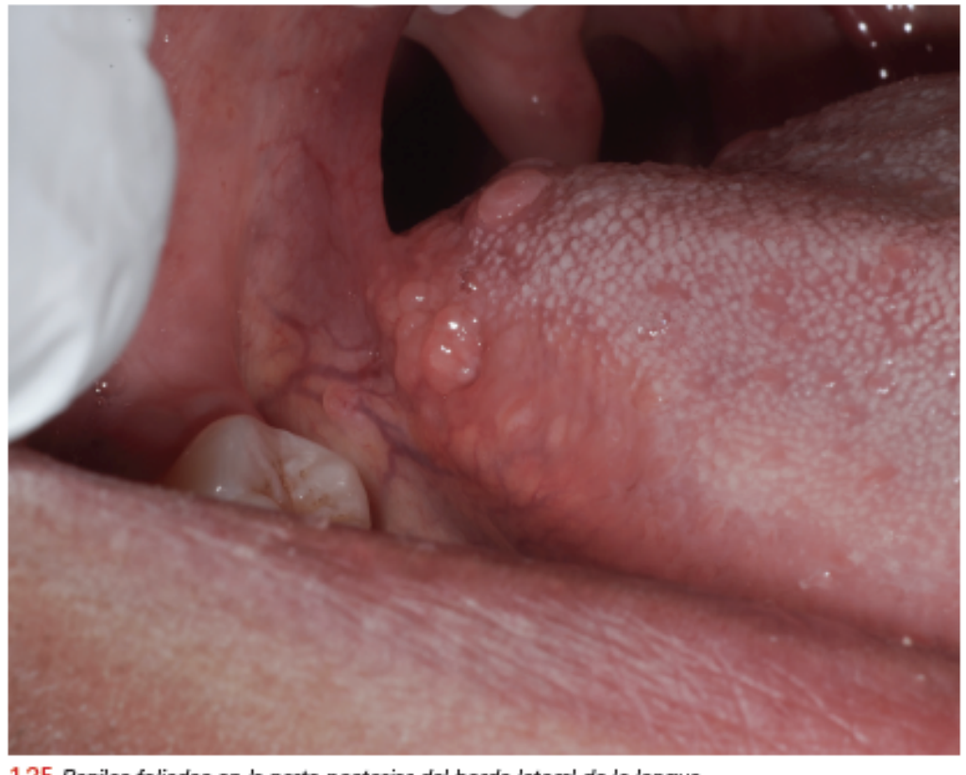
- Áreas anatómicas a examinar
- Exploración diagnóstica
- Pruebas diagnósticas



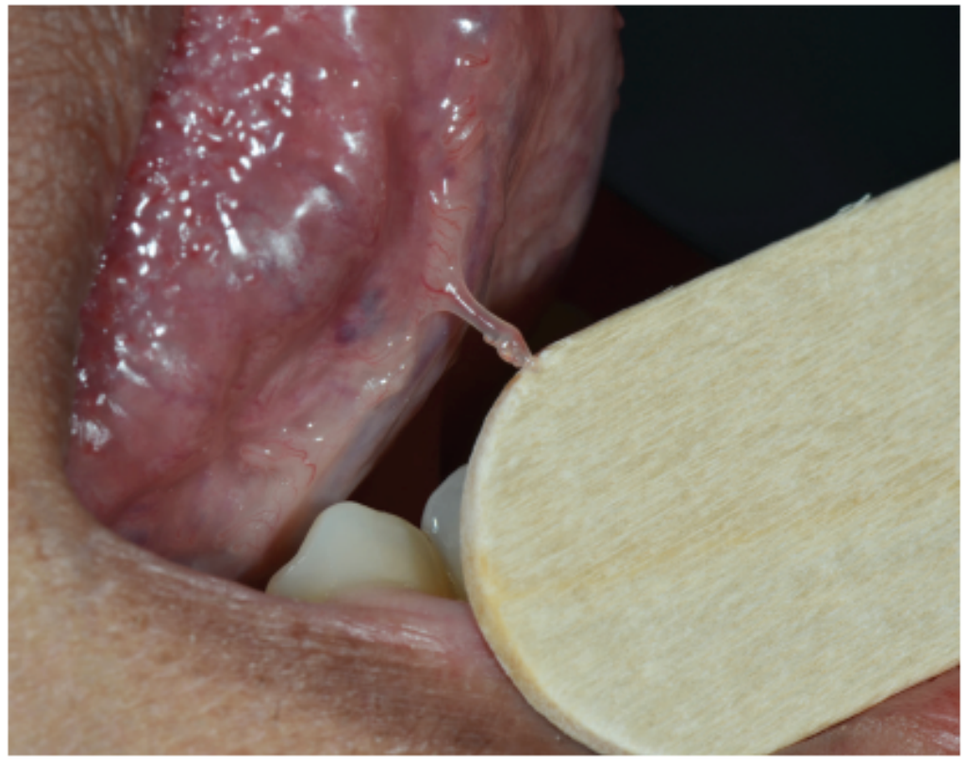
1.23 *Papilas caliciformes o circunvaladas y agujero ciego en la parte posterior de la lengua.*



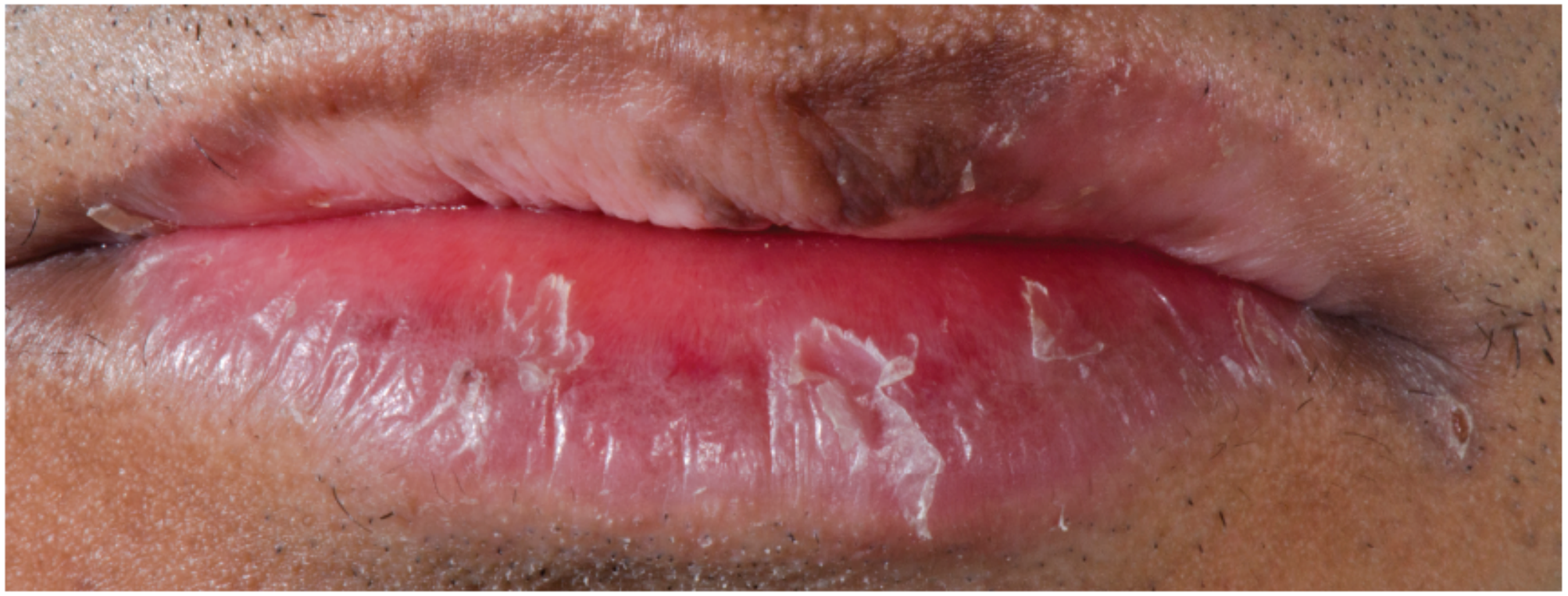
1.24 *Frenillo lingual, venas raninas y pliegues fimbriados en la cara ventral de la lengua.*



1.25 *Papilas foliadas en la parte posterior del borde lateral de la lengua.*



1.26 *A gran aumento pliegue fimbriado transluciendo su contenido salivar.*



2.28 *Finas escamas en labio inferior que se desprenden fácilmente en paciente con queilitis descamativa.*

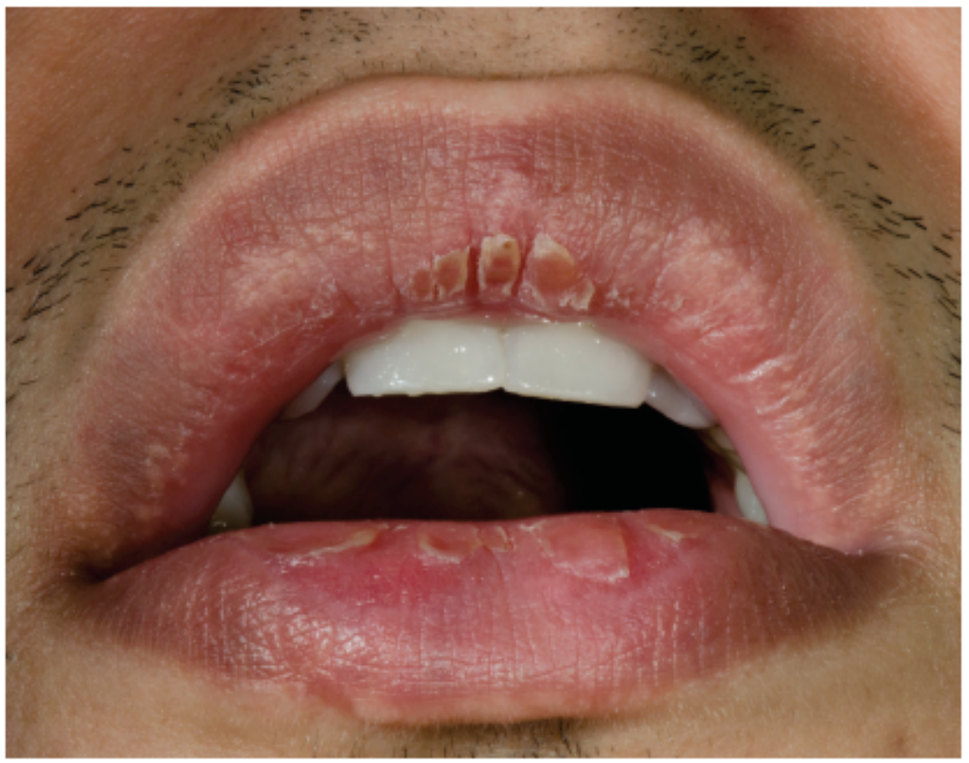
Escama y queratosis

La escama es la lesión elemental que consiste en el desprendimiento de las capas superficiales del epitelio en forma visible, mientras que las queratosis son acumulaciones de escamas que no se desprenden.

Dentro de los procesos escamosos se encuentran: la queilitis exfoliativa, el lupus eritematoso crónico, la psoriasis y la pitiriasis rosada de Gilbert. Algunos procesos que producen escamas pueden también incluirse dentro de las lesiones queratósicas.

La queratosis semiológicamente es una mancha blanca engrosada, frecuente de ver en la leucoplasia grado II, en el liquen queratósico, la queratosis friccional y la poroqueratosis de Mibelli.

Los síndromes malformativos congénitos pueden dar lesiones queratósicas.



2.29 *Escamas bien adheridas y gruesas en paciente con queilitis descamativa y fisurada.*

Estomatitis aftosa recurrente

La estomatitis aftosa recurrente (EAR) no tiene una etiología específica pero puede estar asociada a desórdenes sistémicos, deficiencias nutricionales y de inmunoglobulina A, enfermedades gastrointestinales, inmunodepresiones incluyendo el síndrome de inmunodeficiencia humana VIH-SIDA, enfermedad de Reiter, síndromes como el de Sweet, MAGIC (úlceras en boca, genitales con inflamación del cartílago) y PFAPA (fiebre, adenopatía, faringitis, y aftas) o formar parte de la aftosis de Touraine, la aftosis bipolar de Neumann, Ulcus Vulvae Acutum, enfermedad de Behçet y enfermedad de Crohn.

Aftas menores

Las aftas menores o de Mikulicz, ocurren en el 80% de los casos de estomatitis aftosa recurrente y se localizan en la mucosa de labios, carrillos, piso de la boca, cara ventral y borde lateral de la lengua, paladar blando y pilares amigdalinos, con excepción de la encía y el paladar duro.

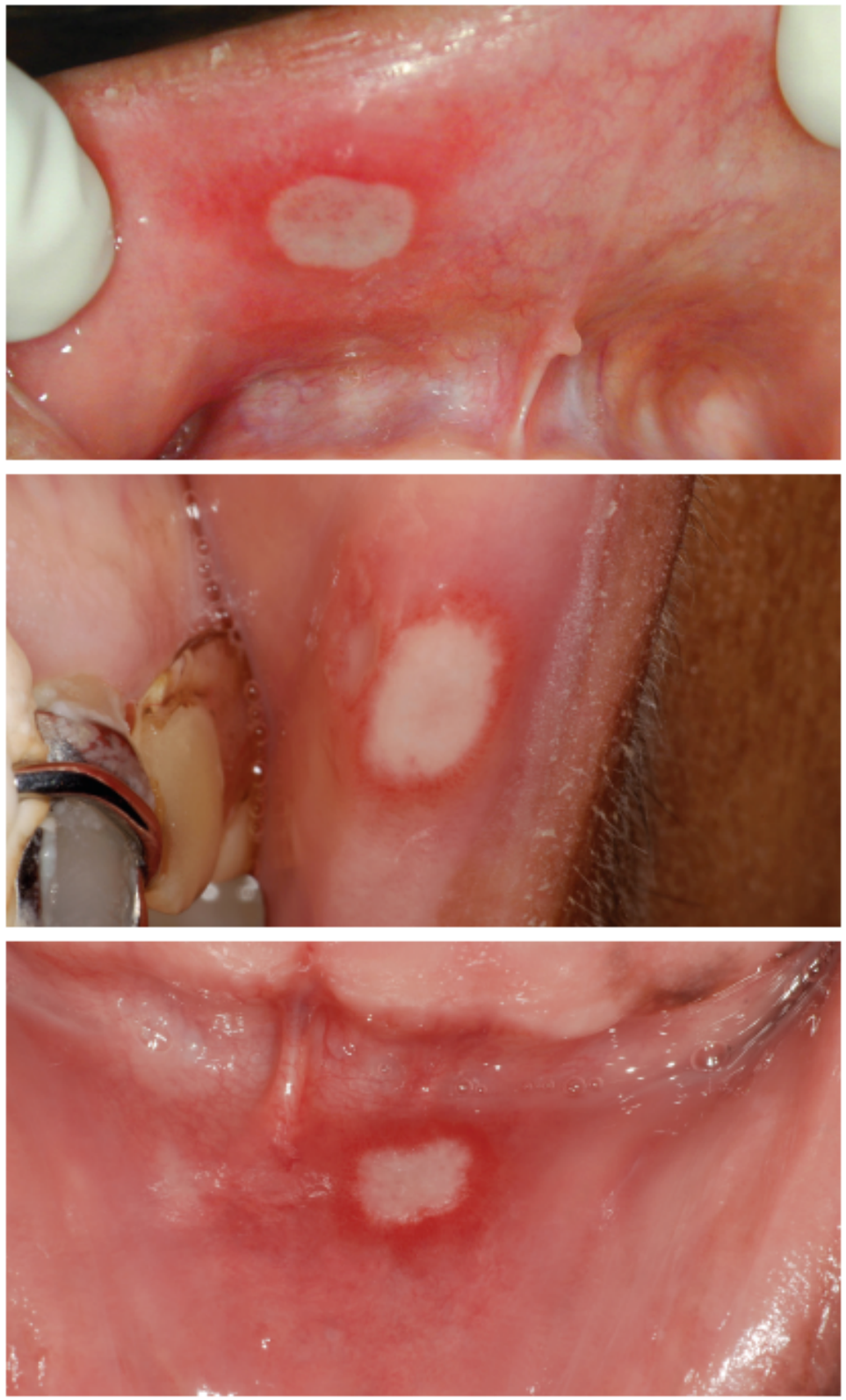
Las lesiones generalmente están precedidas por los síntomas de ardor, picazón o escozor, con la aparición de una mácula eritematosa.

Se presentan como erosiones o ulceraciones superficiales, redondeadas u ovales de 3 a 10 mm de diámetro, con el fondo amarillento de tejido necrótico rodeadas por un halo rojizo inflamatorio.

Su número es variable, son muy dolorosas y curan espontáneamente en 8 o 10 días sin dejar cicatriz, para luego reaparecer un tiempo después.

Su etiología es desconocida, se la relaciona con el estrés, la menstruación, trastornos digestivos, alergias alimentarias o microbianas, mecanismo autoinmune y otros.

El factor de necrosis tumoral (FNT) representa una de las causas principales en la formación de las aftas.



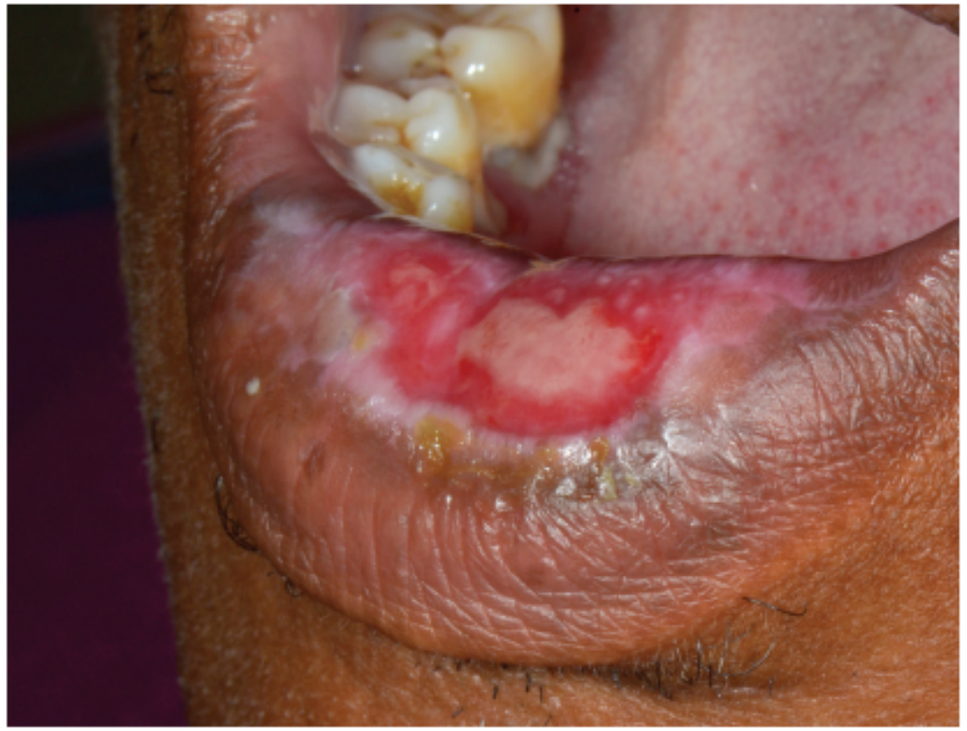
6.38 *Aftas menores características, en diferentes localizaciones de la mucosa oral.*

Queilitis actínica

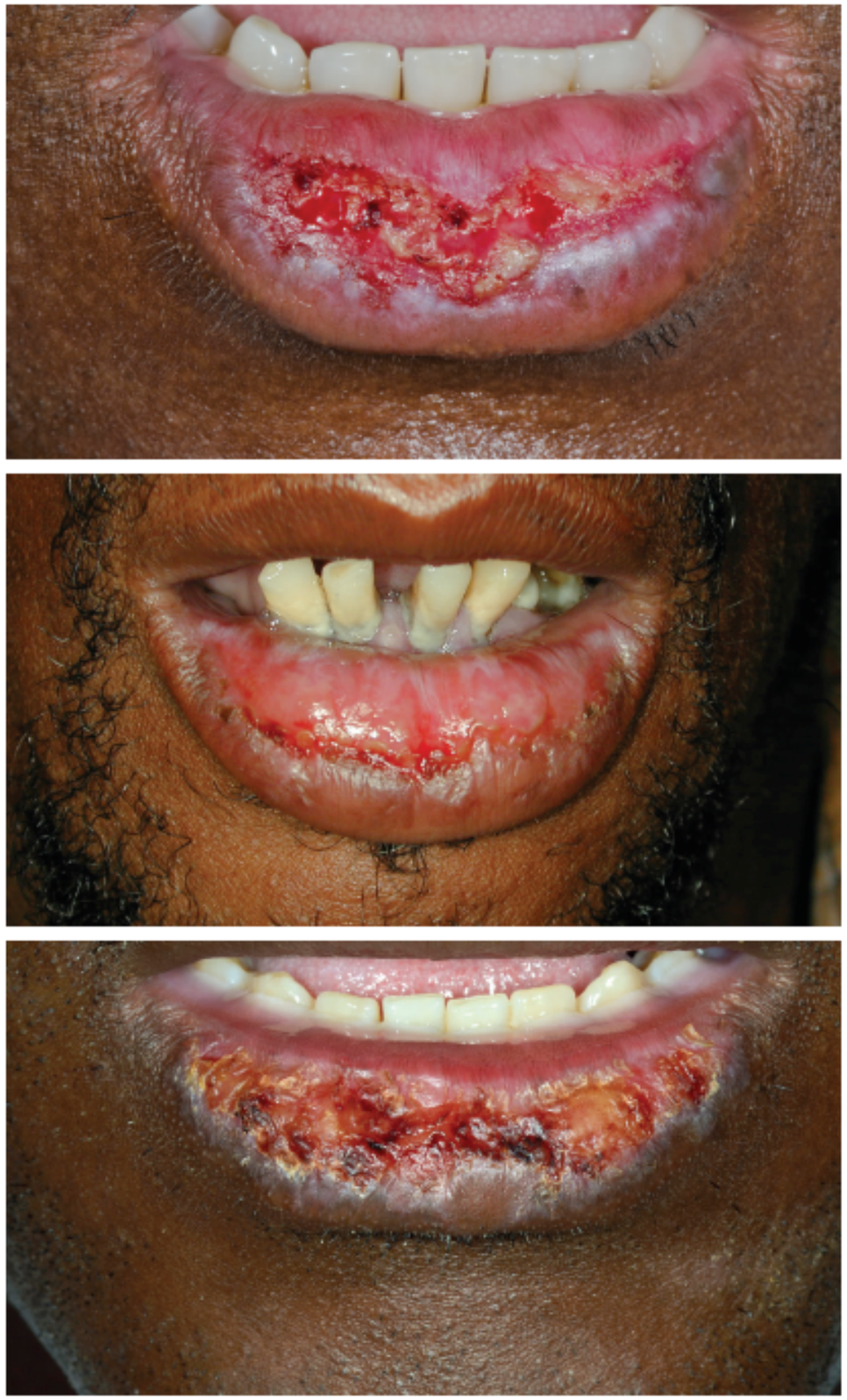
La queilitis actínica es la enfermedad inflamatoria del labio, causada por la exposición al sol. Afecta principalmente el bermellón del labio inferior y es la más común de las lesiones potencialmente cancerizables del labio. El mayor número de cáncer de labio inferior está asociado a una queilitis actínica previa. El factor más importante en su transformación depende de la severidad de la displasia epitelial.

La queilitis actínica se desarrolla lentamente en un extenso período de tiempo con episodios agudos y crónicos en individuos que trabajan o practican deportes expuestos al sol. El uso del cigarrillo potencializa los efectos de las radiaciones actínicas.

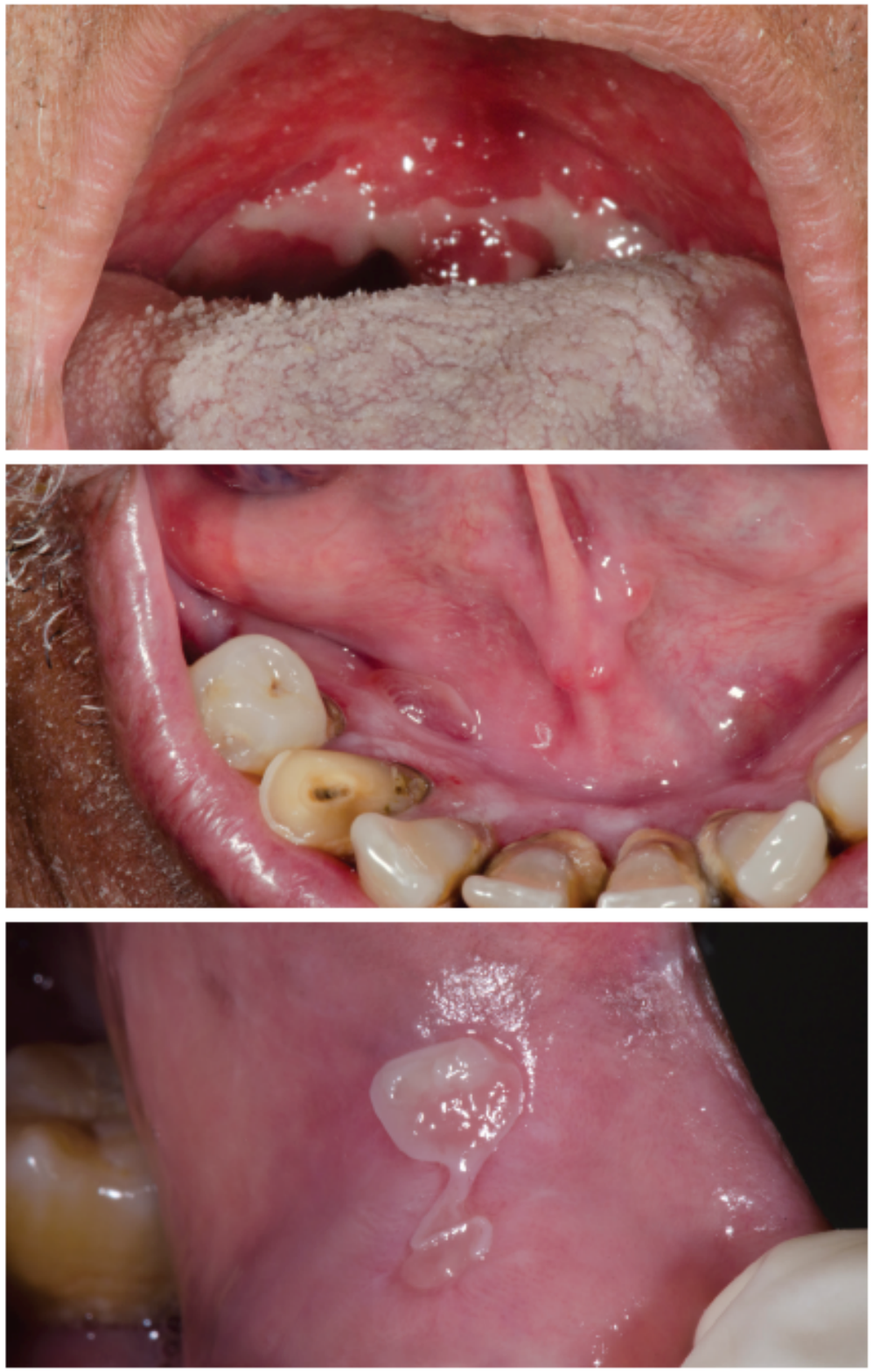
La histopatología es característica: displasia, elastosis solar, inflamación, vasodilatación, hiperplasia y atrofia epitelial. Productos locales o sistémicos (fenotiazinas, sulfamidas y antibióticos) pueden comportarse como sensibilizantes al sol.

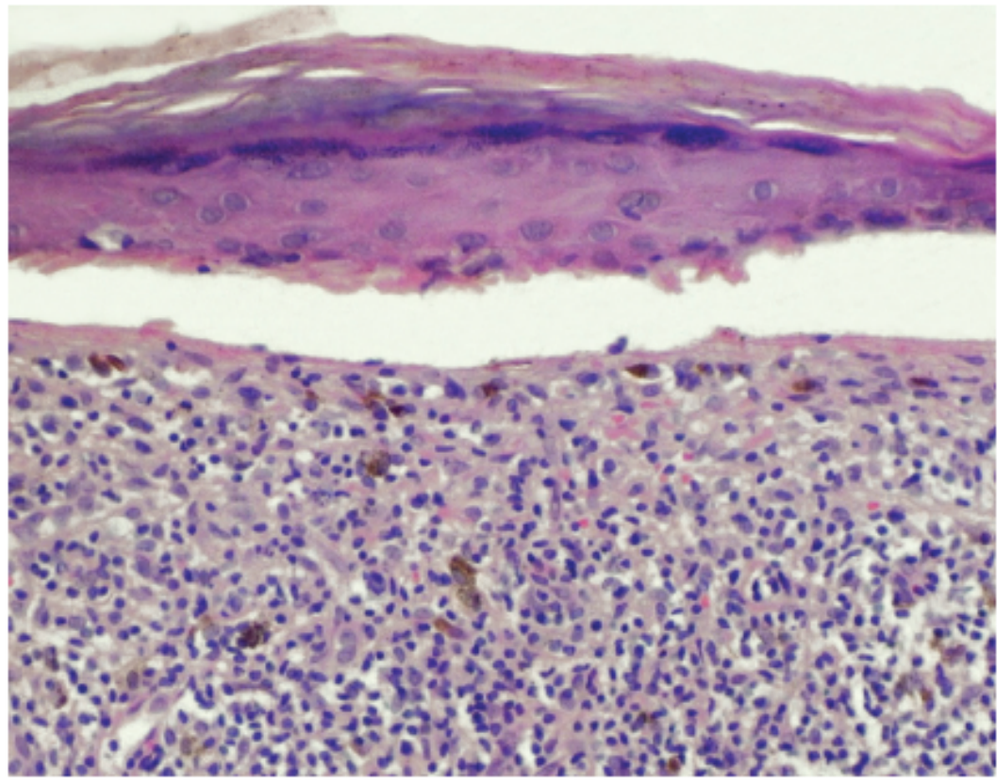


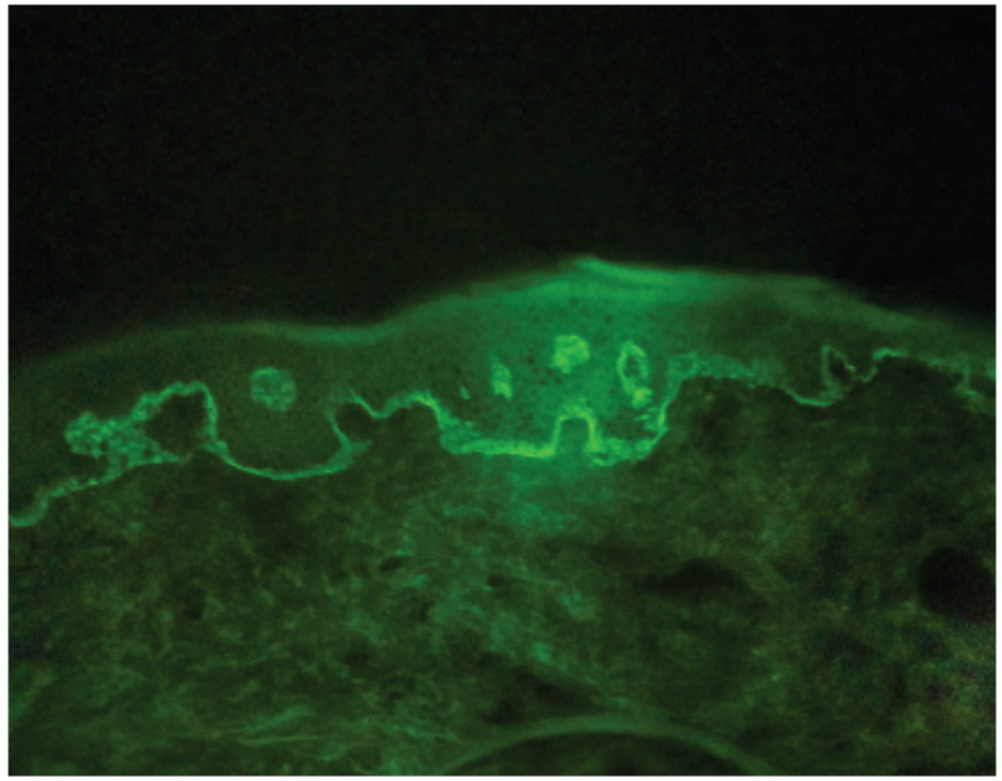
6.69 *Queilitis actínica aguda caracterizada por atrofia, erosión y ulceración de la semimucosa.*



6.70 *Diferentes aspectos clínicos de queilitis actínica aguda en semimucosa labial.*







7.31 *Ampollas en paladar blando, úvula, encía y carrillo del paciente 7.30 en consultas sucesivas. La histopatología muestra ampolla subepidérmica cubierta por epidermis con hipergranulosis triangular. En el dermis superficial medio: infiltrado liquenoide de linfocitos, melanófagos y algunos eosinófilos, en la inmunofluorescencia directa, depósitos de IgG (+++) en la zona de la membrana basal.*

Síndrome de Goltz

El síndrome Goltz es un inusual desorden genético dominante ligado al cromosoma X por mutaciones en el gen PORCN, que afecta piel, huesos, dientes y ojos.

Las lesiones de piel consisten en áreas de atrofia asociadas a telangiectasias, hipo e hiperpigmentaciones que siguen la trayectoria de las líneas de Blaschko's.

Crecimientos de tejidos parecidos a la "frambuesa" se forman en cualquier parte del cuerpo, siendo más común en labios y alrededor de los genitales y el ano.

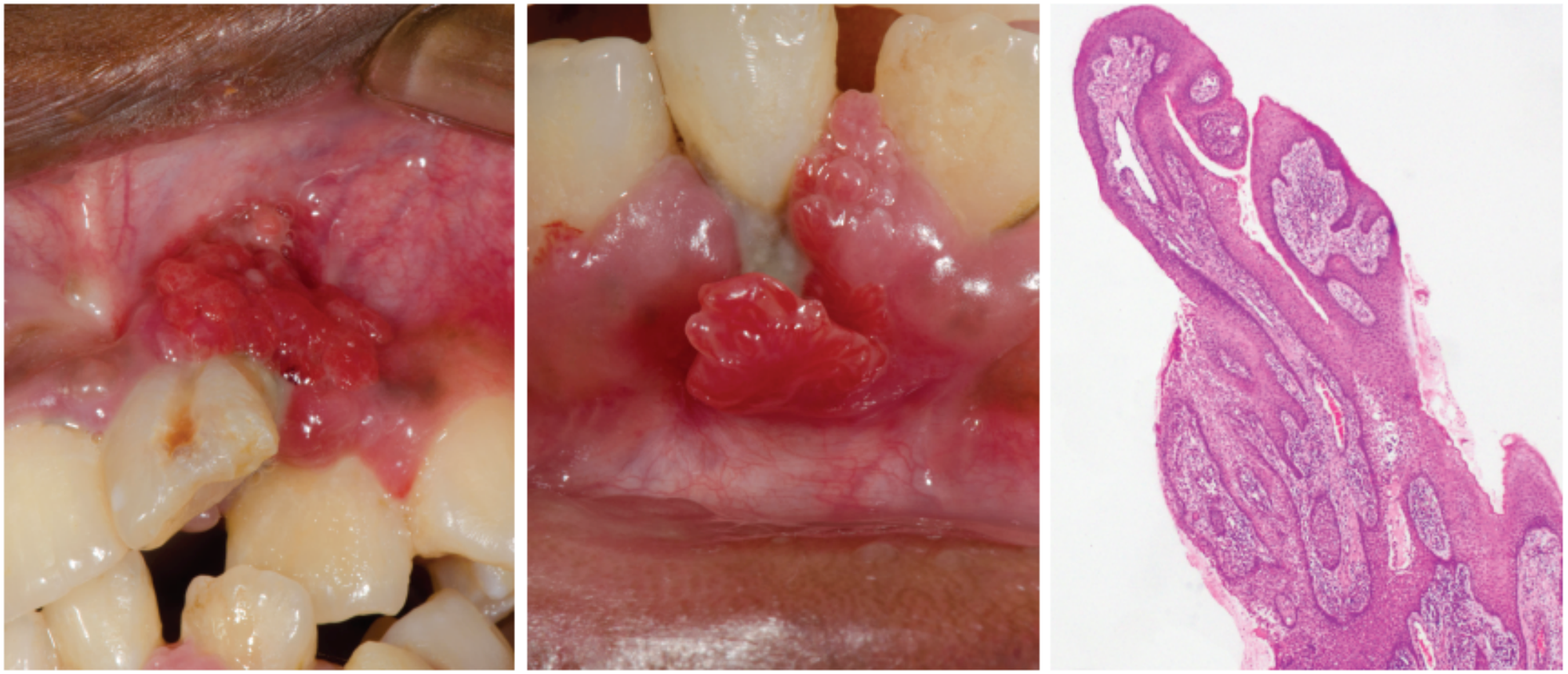
El pelo es escaso, las uñas distróficas con fisuras longitudinales e hipoplásicas.

Las alteraciones dentarias son: microdoncia, geminación, hipoplasia del esmalte y agenesias. En los huesos se traducen en manos y/o pies en "pinzas de langosta" y osteopatía estriada en las extremidades inferiores.

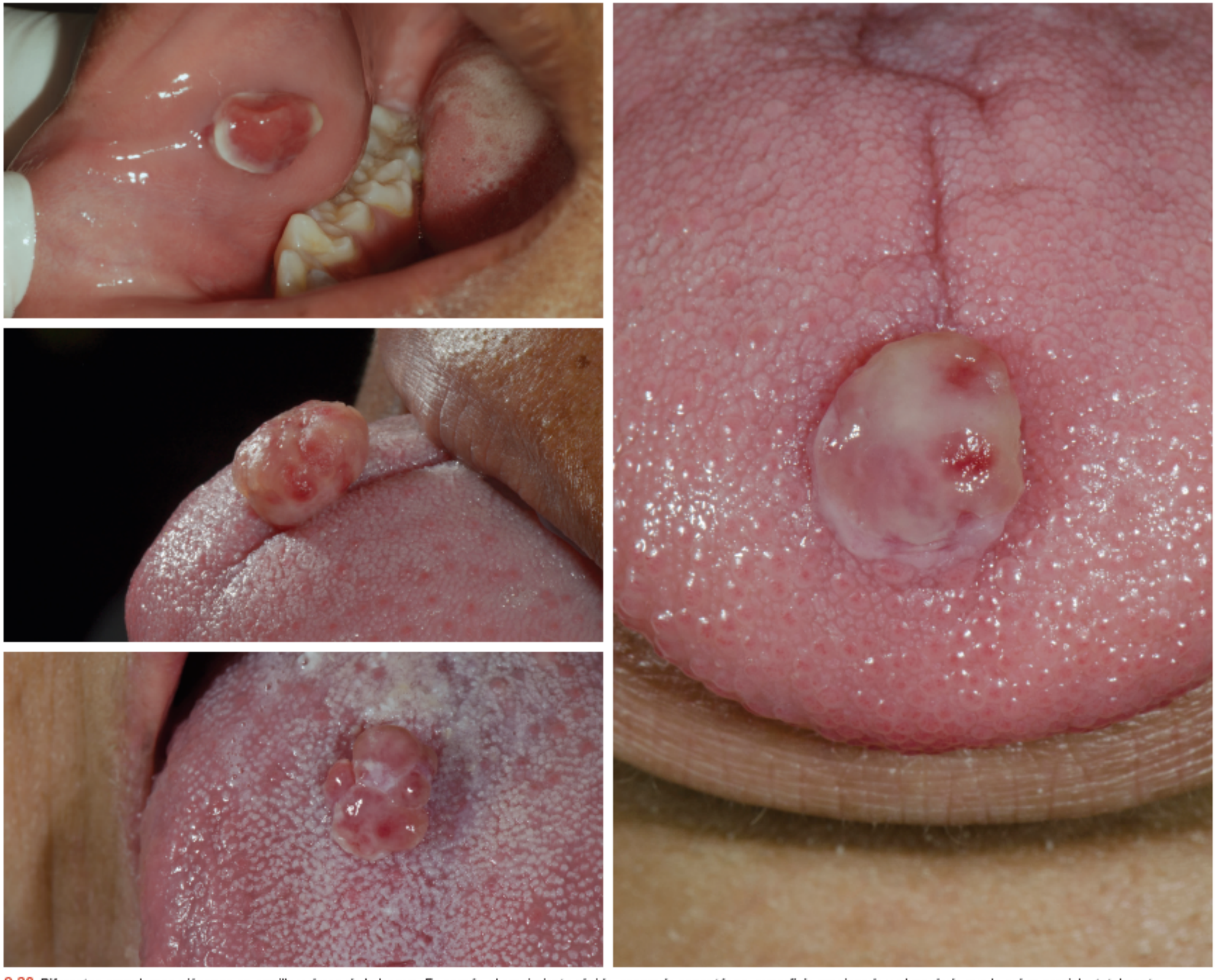
La cara es dismórfica, alargada, con alteraciones en los ojos, a menudo unilaterales que incluyen microftalmia, anoftalmia, aniridia y coloboma.

Las orejas son largas y deformes y el ala de la nariz es deprimida, labio y paladar hendido, sordera, aplasia cutis.

Otras alteraciones pueden aparecer en el aparato genitourinario y gastrointestinal.



8.76 *Lesiones papilomatosas con superficie lisa, roja y lobulada similares a la "frambuesa," en la encía vestibular de los incisivos del lado izquierdo, de niño con síndrome de Goltz. Lesiones de hipoplasia del esmalte se observan en el incisivo central superior izquierdo y el lateral inferior. Histopatología de la lesión que muestra la hiperplasia pseudoepiteliomatosa.*

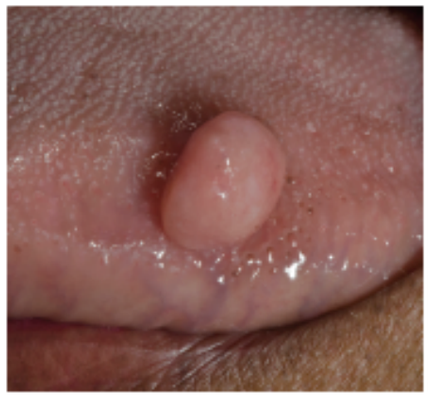


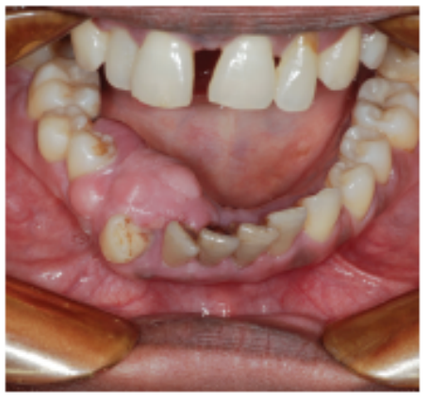
9.20 *Diferentes granulomas piógenos en carrillo y dorso de la lengua. Es común el crecimiento rápido, sangrado espontáneo, superficie erosionada y el exudado que lo cubre parcial o totalmente.*

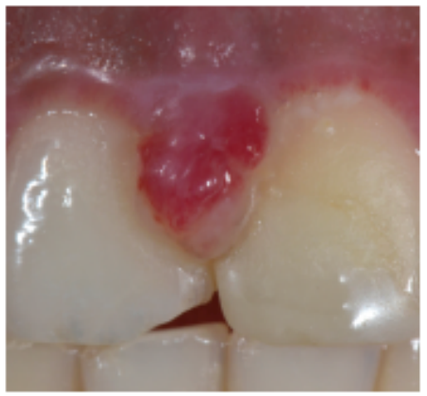
Apéndice

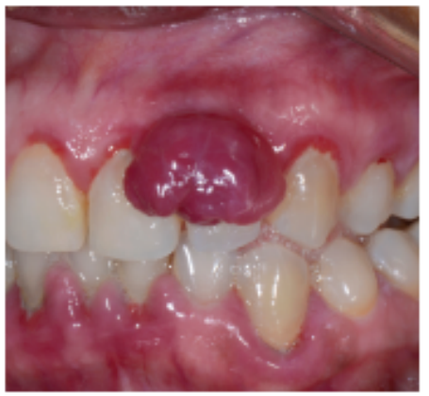
Tablas de Diagnóstico Diferencial

CRECIMIENTOS DE TEJIDOS BLANDOS









	Fibroma por irritación / Pág. 332	Fibroma periférico osificante / Pág. 336	Granuloma piógeno / Pág. 340	Granuloma periférico de células gigantes / Pág. 345
Etiología	Traumática irritativa.	Algunos autores lo relacionan con granulomas piógenos que se han fibrosado y calcificado.	Irritativa local que desencadena un crecimiento inusitado de tejido de granulación.	Relacionada a traumatismos y a extracciones dentarias.
Localización	Carrillos, labios, lengua y encías.	Encía.	Encía, labios, lengua y carrillos.	Encía, a partir de la papila interdentaria.
Edad	Adolescentes y adultos.	Adultos principalmente.	Cualquier edad. Más frecuente en niños, adolescentes y adultos jóvenes.	Adolescentes y adultos.
Sexo	Ambos sexos.	Más común en la mujer.	Más común en la mujer.	Ambos sexos.
Aspecto clínico	Crecimiento único de tamaño variable, forma esférica o hemisférica, base pediculada o sésil. La mucosa que lo recubre es lisa y brillosa, del color de la mucosa o más pálida, por el traumatismo puede cambiar a blanco por la queratinización del epitelio o ulcerarse.	Agrandamiento de tejido firme, rojo rosado, duro a la palpación, superficie lisa de base sésil y siempre localizado en la encía.	Lesión muy sangrante, de diferente tamaño, usualmente pediculada, superficie lisa y algunas veces lobulada, recubierta por una membrana amarillenta y en algunas partes con costras hemáticas especialmente los que están en la semimucosa de los labios.	Crecimiento de tejido con base ancha o pediculada, superficie lisa o lobulada, rojo azulada, fácilmente sangrante con historia de crecimiento en varias semanas o meses y algunos casos desarrollados luego de una extracción dentaria.
Síntomas	Asintomático.	Asintomático.	Muy sangrante en su inicio.	Fácilmente sangrante.
Evolución	Crecimiento lento.	Crecimiento lento.	Crecimiento rápido para luego detenerse y con el tiempo fibrosarse.	Semanas o meses.
Histopatología	Lesión con denso colágeno recubierta de epitelio escamoso estratificado normal.	Proliferación de tejido fibroso con algunos o numerosos focos de calcificación, recubierto por epitelio pavimentoso estratificado normal.	Compuesta en su mayor parte por pequeños vasos sanguíneos, tejido conectivo, células plasmáticas, linfocitos y gran infiltración de neutrófilos. El epitelio que lo cubre parcialmente es estratificado escamoso. Una membrana fibrinopurulenta cubre las áreas ulceradas.	Tejido conectivo recubierto de epitelio pavimentoso estratificado, con denso infiltrado de células plasmáticas, linfocitario y células gigantes multinucleadas con cierta cantidad de hemosiderina En algunas áreas puede faltar el epitelio por ulceraciones en la superficie.

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ENFERMEDADES DE LA MUCOSA ORAL
Atlas

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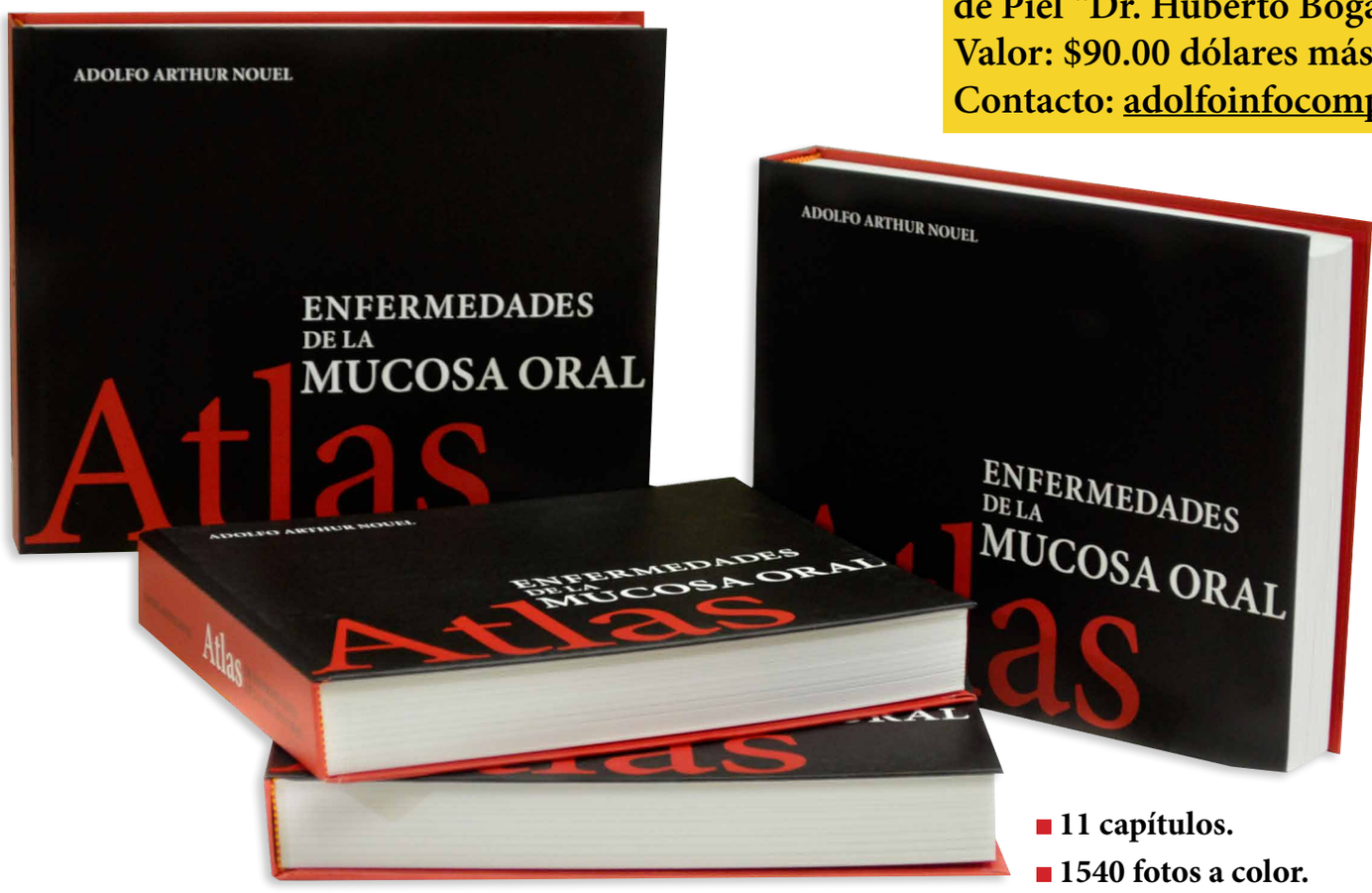

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- Lesiones rojas.
- Lesiones pigmentadas.
- Lesiones vesiculares, erosivas y ulceradas.
- Lesiones ampollares.
- Lesiones vegetantes y verrugosas.
- Crecimientos de tejidos blandos.
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O u r D e r m a t o l o g y O n l i n e

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1.2016 (07.January.2016)