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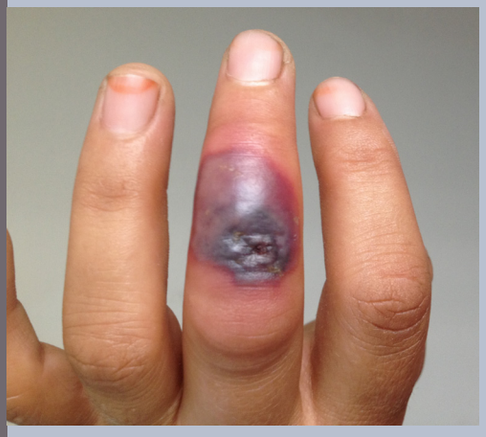

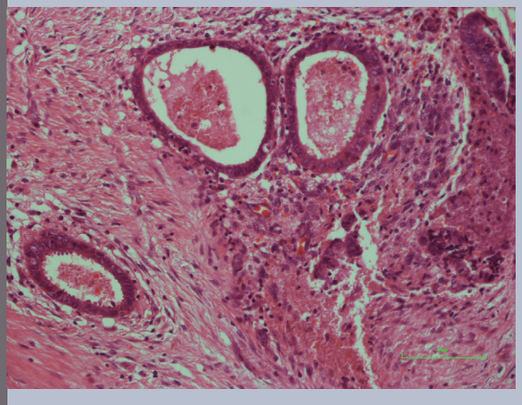

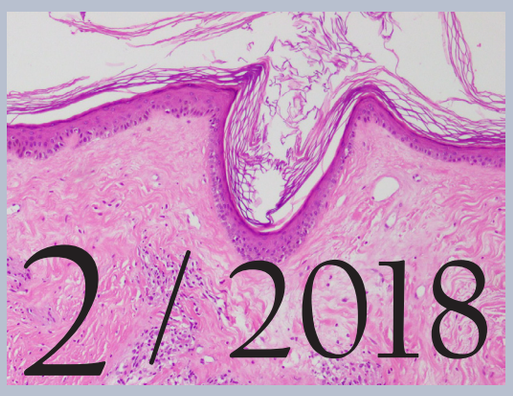

Editorial Pages

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Original Article

A novel nonsense ATP2C1 mutation causes Hailey-Hailey disease in a Tunisian family

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ABSTRACT

Background: Hailey-Hailey disease (HHD) is an autosomal dominant blistering skin disorder that manifests in the third to fourth decade of life. The *ATP2C1* has been identified as the pathogenic gene of this disease since 2000. **Materials and Methods:** We report here a three generations Tunisian pedigree, where almost all males are severely affected and present with complete penetrance of HHD, while only one female shows a mild disease's phenotype in her fourth decade. A molecular study using Whole exome sequencing and direct sequencing was performed to this family. **Results:** By whole exome sequencing and direct DNA sequencing, a novel nonsense mutation in *ATP2C1* (c.2698A>T; p.Lys900Ter) was identified in all patients, supporting that alterations in *ATP2C1* are causative of HHD. Unexpectedly, this mutation was found in one female who was initially not diagnosed for HHD. Our observations would be in line with incomplete penetrance and variable expressivity between male and female of this disease, or evidence for genetic modifiers. **Conclusion:** We report here a novel nonsense heterozygous mutation in *ATP2C1* gene in 5 patients with HHD. Interestingly, one woman carries the nonsense *ATP2C1* mutation but displays a mild phenotype of HHD. This could indicate a variation in pattern and expressivity between male and female developing HHD phenotype which should be considered when providing genetic counselling to family members carrying such mutations.

Keywords:*ATP2C1;* Hailey-Hailey disease; benign familial chronic pemphigus.

INTRODUCTION

Hailey-Hailey disease (HHD), or benign familial chronic pemphigus, which was described by the Hailey brothers in 1939, is characterized by recurrent vesicles and erosions, usually affecting the neck, axillae and groins [1]. HHD is an autosomal dominant blistering skin disorder that manifests in the 3rd to 4th decade of life with a prevalence of 1 in 50,000 [1,2]. Vesicular, itchy and painful lesions on an erythematous base characterize the rash and affect the flexural areas of the skin. Friction, sweating, heat, stress, UV exposure and cutaneous infections trigger the rash. The disease has a fluctuating course with remissions and exacerbations [3]. In rare instances, the rash may become generalized and the skin lesions may even develop into squamous cell carcinoma [4]. Penetrance in adults affected by HHD is complete but expressivity is variable [2]. Histopathological features show a widespread loss of cell-to-cell adhesion (acantholysis) in the suprabasal layer of the epidermis. Ultrastructural studies demonstrate perinuclear aggregation of keratin intermediate filaments, which have retracted from the desmosomal plaques in the acantholytic cells [5,6].

Two different research groups have linked HHD to mutations in the gene *ATP2C1*, which encodes the human secretory pathway Ca2+/Mn2+ ATPase

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(hSPCA1) [7,8]. SPCA1 is ubiquitously expressed in human tissues with the highest abundance in keratinocytes [9]. SPCA1 localizes to the Golgi-apparatus and controls its Ca^{2+} stores along with SERCA transporters. In primary keratinocytes, the role of SPCA1 is more significant than in other cell types [10,11], explaining why only the skin is affected in HHD patients. In this report we investigated a three generations Tunisian pedigree where males are severely affected with HHD. By whole exome sequencing followed by Sanger sequencing, we identified a novel nonsense mutation (c.2698A>T; p.Lys900Ter) in the *ATP2C1* gene that causes Hailey-Hailey phenotype in this family

MATERIALS AND METHODS

Subject and informed consent

We performed molecular studies in a Tunisian family with HHD where males are severely affected in three generations (Fig. 1a). After obtaining the informed written consent and IRB approval (NUS IRB 10-051), blood samples were taken from four affected males and two unaffected females. Genomic DNA (gDNA) was extracted using Qiagen® kit.

Whole exome sequencing

For mutation analysis, 1ug of gDNA of proband III:8 was used for exome capture with Ion TargetSeqTM Exome and Custom Enrichment Kit. The exome library was prepared on an Ion OneTouch System and sequenced on an Ion Proton instrument (Life Technologies, Carlsbad, CA, USA) using one ION PI chip. Sequence reads were aligned to the human reference genome [Human GRCh37 (hg19) build] using Torrent Mapping Alignment Program (TMAP) from the Torrent Suite (v4.2.1). The variants were called using the Torrent Variant Caller (TVC) plugin (v4.2.1) and were annotated using the "annotate single sample variants" workflow, including the associated gene, variant location, quality-score, coverage, predicted functional consequences, protein position and amino acid changes, SIFT [12], PolyPhen2 [13], and Grantham [14] prediction scores, phyloP [15] conservation scores and 5000 genomes Minor Allele Frequencies. Annotated variants were filtered for common SNPs using the ClinVar "common and no known medical impacts" database (ftp://ftp.ncbi.nlm.nih.gov/pub/clinvar/vcf_GRCh37/) and the Exome Aggregate Consortium (ftp://ftp.broadinstitute.org/pub/ExAC_release/release0.2/). Variants were next compared to an in-house database of 485 previously sequenced samples, and those that were present in more than 1% of the previously sequenced samples were removed.

Sanger sequencing

Sanger sequencing was performed using 2 different sets of primers (Table 1) to verify the segregation of the identified ATPC2 nonsense mutation. After amplification, products were purified and sequenced on DNA sequencing system (model 3730XL; ABI).

RESULTS AND DISCUSSION

For all the affected members from this family (Fig. 1a) the disease began within the third decade of life. All affected individuals exhibited the typical clinical features of HHD with late onset of the rash characterized by itchy and painful lesions on an erythematous base and affect the flexural areas of the skin (Figs 1b and c). All the patients mentioned that friction, heat or sweating exacerbated the disease and the symptoms were worse during summer or if they were under stress. It is noteworthy that HHD penetrance is complete in adults affected with a variable expressivity.

To look for the causative mutation, 1ug of gDNA of patient III:8 was used for exome capture with Ion TargetSeqTM Exome and Custom Enrichment Kit. Whole exome sequencing of this proband generated a total of 14.5 Gb with an average read length of 155 bp. An average coverage of 188X was achieved across the exome, with 96% of the targeted sequences covered at ≥20X. A total of 37,131 variants were identified across protein-coding exons, UTRs, splice sites and flanking introns. After applying a series of filters and following an autosomal dominant mode of inheritance, a final set of 28 heterozygous variants were selected (Fig. 2a), including a deleterious mutation in *ATP2C1*, known as the causative gene of HHD. By Sanger sequencing, we confirmed that the nonsense mutation c.2698A>T (p.Lys900Ter) was heterozygous in all affected male members and unexpectedly in the female III:10 who was initially not diagnosed for HHD (Fig. 2b). This result prompted us to re-examine this individual over time, and noted that she has started developing a milder HHD phenotype, less severe than her male siblings and father despite the fact that she is in

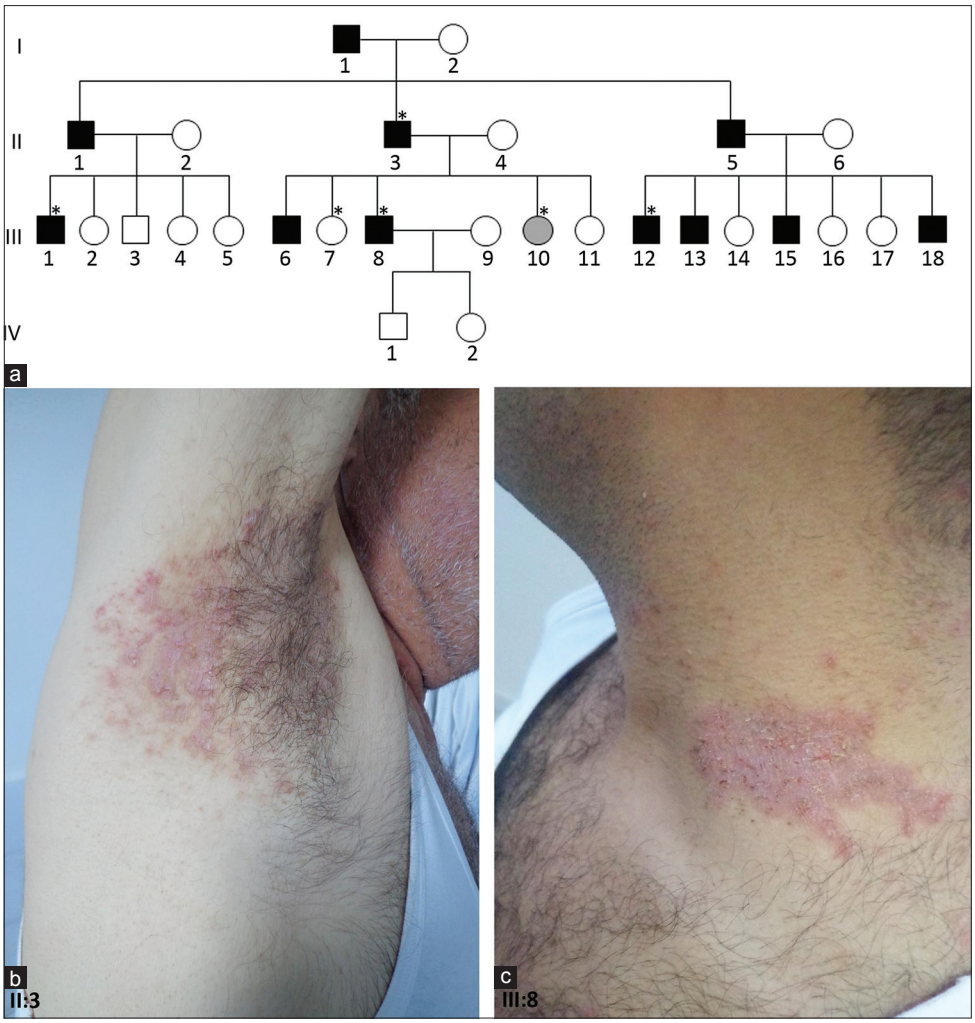

Figure 1: Clinical manifestations of autosomal dominant Hailey-Hailey disease. (a) Pedigree of a Tunisian family presents with an autosomal dominant HHD, where only males are severely affected on three generations (filled black symbols: affected individuals, square: male, circle: female, stars: studied individuals). (b) and (c) patients II:3 and III:8 with HHD features: rash that is characterized by painful lesions affecting the neck and the axillary.

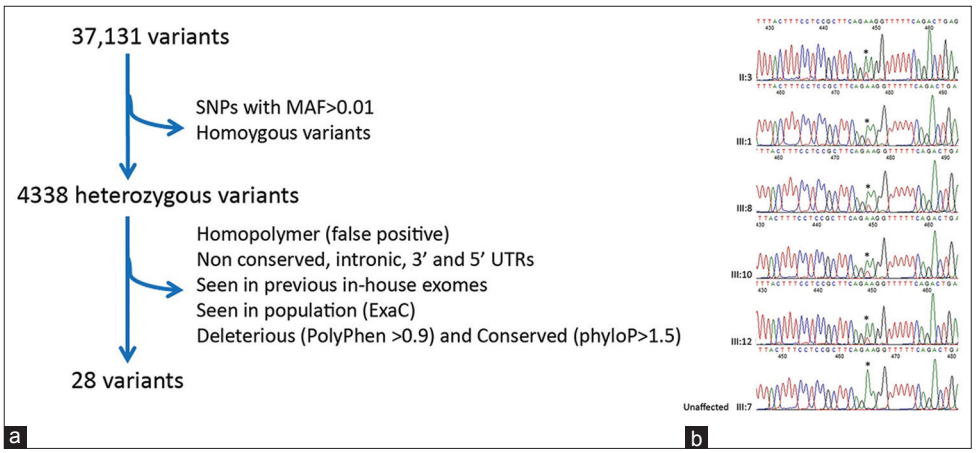

Figure 2: A nonsense mutation p.Lys900Ter in *ATP2C1* causing HHD. (a) Whole exome sequencing results workflow. (b) Sanger sequencing results confirmed the segregation of the c.2698A>T mutation with HHD phenotype in 4 affected males and one female.

her late forties. To date, all individuals who have been reported as carriers of *ATP2C1* mutations and present the complete clinical features of this disease. Even though HHD is transmitted as an autosomal

Table 1: Primers used for Sanger sequencing.

Primer	Sequence
Set1 Forward	5' CACACAATTAGGTCCATTCTCCA 3'
Set1 Reverse	5' TCATTCCTCACACCTACACAG 3'
Set2 Forward	5' AGCGAATTCTCACTAATTGACCA 3'
Set2 Reverse	5' TTCTGCTATTTGGTCAGACTGA 3'

dominant disease, variation in the pattern and the severity of symptoms within the same family have been reported [2]. The variability of age of onset and the different expressivity of the phenotype that we report here may provide evidence for genetic modifiers as well as for environmental factors such as sun, heat, stress and friction that affect the HHD development [2].

CONCLUSION

We report here a novel nonsense heterozygous mutation in *ATP2C1* gene in 5 HHD-patients supporting that alterations in the human secretory pathway Ca2+/ Mn2+ ATPase are causative for this disease. It is worth noting that in this family one woman carries the nonsense *ATP2C1* mutation but displays a mild phenotype of HHD. Our observations would thus be in line with variation in pattern and expressivity between male and female developing Hailey-Hailey disease phenotype or evidence for other modifiers. This aspect should be considered when providing genetic counselling to family members carrying such mutations.

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Conflict of Interest: None declared.

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Original Article

The prevalence of pediculosis capitis in Makkah city Saudi Arabia

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ABSTRACT

Background: In Saudi Arabia, there is limited data regarding the epidemiology of head lice. We aim to measure the prevalence of head lice particularly in Makkah city along with assessing the risk factors associated with it. **Material and Methods:** A descriptive cross-sectional survey was conducted from 1st of June to 1st July 2017, among all males and females with no age restriction living in the city of Makkah, Saudi Arabia. **Results:** Of the 438 participants (mean age, 25) the majority were female 78.5% (344), and 21.5%(94) were male. The prevalence of P. capitis infection was 64.2% (281/438) among them 85.8%, (178/281) confirmed transition of disease to another family member mainly to their sisters. The prevalence of pediculosis was significantly elevated among females 88.3 % (248) than males 11.7 % ($p<0.0001$). Also, it was higher among the nonemployees 82.6%(232/281) ($p<0.015$). The infestation was significantly associated with long hair $p<0.0001$. 71.2%, participants with a history of lice infestation confirmed sharing personal belongings with others ($p<0.0001$). **Conclusion:** The infected participants were predominantly females, with a sizeable proportion confirming transition of infestation to another family member. P. capitis was commonly associated with long hair and higher among nonemployee and those who share their personal belongings.

Key words: Head lice; Pediculosis capitis; Hair disorders; Makkah

INTRODUCTION

The close relationship between lice and human is explained by a long history back to approximately 100,000 years ago. With early human migrants from Africa where they found that P. humans show a genetic evidence of expansion from Africa. This was evidenced by the oldest human head louse nit that was found from a hair sample dated to 8000 B.C. in northeastern Brazil; another oldest hair sample was found 9000 years old, obtained from a corpse found in the Nahal Hemar cave [1].

There are two types of lice parasitizing humans: Pthirus and Pediculus. The latter one caused a significant public health problem, and it has two ecotypes: the body louse and the head louse, where head lice diverge into three clades (A, B and C), each clade has a unique geographic distribution throughout the world [2].

P.hummanus var. capitis is an infestation of human hair, scalp, and neck by the head louse. An obligatory blood-feeding ectoparasite, transmit by sharing personal belongings like head scarfs, brushes, pillows or even head to head contact which is the most common mode of transmission [2].

They cause blood loss, itchiness, redness, discomfort, social and psychological distress. Usually, the most common symptom of head lice is pruritus; Although head lice do not transmit any disease the possibility of secondary bacterial infections occurring at scratch sites with impetiginization [3].

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According to many studies, head lice are considered one of the common infection in school-age children all around the world [4]. Annually in the United States of America, Pediculosis capitis, affects six to twelve million individuals. In Saudi Arabia, some studies were reported but with a limitation in the data regarding the epidemiology of head lice.

In a recent study in Jeddah city, the prevalence of pediculosis was 11.26%, and distribution of infestation among males and females was broadly similar [5]. In Abha city, the most common transmissible skin diseases were P. capitis (9.6%) with an increase in the prevalence as the age decreases [6].

In our study, we aim to measure the prevalence of head lice in Makkah city among all ages. Additionally, the effect of risk factors associated with head lice.

MATERIALS AND METHODS

It is a Descriptive cross-sectional survey that was successively conducted from 1st of June to 1st July 2017, Among All males and females with no age restriction living in the city of Makkah, Saudi Arabia. We included all Makkah residents who participated in the survey and excluded all non-Makkah residents from the study.

After seeking the approval from the institutional review board (IRB) of King Abdulaziz Hospital, Makkah, Saudi Arabia. An electronic questioner was created using google docs and was distributed. The survey included an overall information about the previous occurrence of pediculosis capitis, demographics and risk factors such exposure to animal pets and sharing personal belongings.

Statistical analysis

After importing the data from the Microsoft Excel file, the data was analyzed using Statistical Package for the Social Science SPSS version 23. Frequency tables were made to measure the prevalence of each variable. The risk factors were analyzed by chi-squares to measure its significances in relation to the primary dependent variables and to sit the P.value to be =0.05

Ethics statement

This manuscript describes original work and is not under consideration by any other journal. All authors approved the manuscript and this submission, also all the authors made significant contribution to the study.

RESULTS

A total of 438 participants were included in the analysis. The average age of participant was 25 years. 78.5% (344) of them were female, and 21.5% (94) were male. The overall prevalence of P. capitis infection was found to be positive in 64.2% (281/438). The prevalence of pediculosis was significantly elevated among females 88.3% (248) than males 11.7% ($p<0.0001$). Also, it was higher among the nonemployees 82.6%(232/281) in comparison to the employees 17.4% (49/281) ($p<0.015$).

On the contrary, no significant association between head lice infection and having a pet or animal at home ($p>0.414$), or any other demographic data (Table 1).

As for the frequency of hair washing three times a week it was higher among those who previously had head lice 51.6% (145/281) ($p=0.033$).

Regarding the type and length of hair, both oily and long hair were great in association with head lice accounting for 39% (112/281) and 59.9% (168/281) respectively. However, only hair length showed a significant association with head lice infestation $p<0.0001$ and not so much with the type of hair (Table 2).

From the 281 people with lice 241 confirmed transition of infection to another family member by 85.8%, (178/281) 63.3% were their sisters ($p<0.0001$). However, 97,9% (275 out of 281) denied transition of

Table I: Demographic data

variables	Previous head lice		P value
	Yes (%)	No (%)	
Sex			
Male	33 (11.7)	61 (38.9)	0.000**
Female	248 (88.3)	96 (61.1)	
Are you employed?			
Yes	49 (17.4)	42 (26.8)	0.015*
No	232 (82.6)	115 (73.2)	
How many family members?			
2	5 (1.8)	4 (2.5)	0.687
3	13 (4.6)	10 (6.4)	
4	30 (10.7)	20 (12.7)	
>4	233 (82.9)	123 (78.3)	
Where do you live?			
Owned	184 (65.5)	109 (69.4)	0.231
Rented	97 (34.5)	48 (30.6)	
Presence of animals/pets			
Yes	36 (12.8)	22 (14.0)	0.414
No	245 (87.2)	135 (86.0)	

infection to another site in their body rather than their head (p<0.0001).

Also, 71.2%, participants with a history of lice infestation confirmed sharing personal belongings with others (p<0.0001) while around 40% share their bed with others (p=0.021).

Scalp itchiness and redness were the major significant symptoms for head lice infestation (p<0.0001 and p=0.011) (Table 3).

Table 2: Hair

variables	Previous head lice		P value
	Yes (%)	No (%)	
How many times do you wash your hair weekly?			
Three times	145 (51.6)	60 (38.2)	0.033*
Daily	86 (30.6)	67 (42.6)	
Less than three times	50 (17.8)	30 (19.1)	
Hair type			
Curly	31 (11.0)	20 (12.7)	0.551
Dry	110 (39.1)	51 (32.5)	
Dily	112 (39.9)	67 (42.7)	
Straight	28 (10.0)	19 (12.1)	
Hair length			
Long	168 (59.8)	64 (40.8)	0.000**
Short	112 (39.9)	93 (59.2)	

Table 3: Infectious data

Variables	Previous head lice		P value
	Yes (%)	No (%)	
Itchy scalp			
Yes	126 (44.8)	44 (28.0)	0.000**
No	155 (55.2)	113 (72.0)	
Redness in the scalp			
Yes	39 (13.9)	10 (6.4)	0.011*
No	242 (86.1)	147 (93.6)	
If yes, has another family member been infected?			
Yes	241 (85.8)	1(.6)	0.000**
No	39 (13.9)	0 (0.0)	
If yes, what is kinship?			
Brother	39 (13.9)	3 (1.9)	0.000**
Father	3 (1.1)	0 (0.0)	
Mother	15 (5.3)0	0 (0.0)	
Sister	178 (63.3)	12 (7.6)	
Did infection move to another place rather than your head?			
Yes	6 (2.1)	0 (0.0)	0.000**
No	275 (97.9)	0 (0.0)	
Sharing personal belonging with other			
Yes	200 (71.2)	84 (53.5)	0.000**
No	81 (28.8)	73 (46.5)	
Sharing beds at home			
Yes	115 (40.9)	45 (28.7)	0.021*
No	165 (58.7)	112 (71.3)	

DISCUSSION

There are Important factors affecting the rate of infestation by head lice, such as socioeconomic factors, personal hygiene, and gender. Some of them have been approved previously by researchers to contribute to the transition of lice from head-to-head which we included in our research. The rate of infestation in unemployed people were higher at different ages by 82.6% comparing to the employees. Which reveals that people of low income can somehow be more prone to infestation, this finding has been approved as well in Korea where they found that even the occupation of parents can affect the prevalence of head lice [7]. We wanted to elaborate further on the income side by asking about the ownership of their property whether it was rented or owned. However, the majority responded by owned which caused a low sensitivity in the desired result. On the other hand, the rate of infestation increased among bigger families which known to lower the income, 233 previously infected people said to live amongst a large family of more than four household members.

The rate of infection is primarily affected by personal hygiene. In our case, the type of hair and length which require more effort and care had a significant impact on both the prevalence and rate of infestation. In our study, the rate of infestation encompassed 59.8% in those with long hair and oily hair by 39.9%. In the analysis of data, the girls over twelve years of age in Sindh province of Pakistan indicated that infrequent bathing in summer was associated with higher rates of infestation in Sindh province which shows the importance of hygiene when it comes to lice [3].

The head infestation was found to be higher among females by 88.3% than males. Also, the transition of infection from an infected person to another was higher by 63.3% among their sisters, which means that P. capitis infection has a higher rate among females. This might be explained by the nature of females and their close contact with each other, unlike males they share their headscarf accessories [8]. Again this is also was seen in our results, sharing personal belongings had a higher rate of infestation by 71.2%. In many studies like the cross-sectional study that was conducted in the urban area of Athens in Greece. The overall infestation rate (5.30%) was mainly attributed to females, which manifested a higher rate (4.84%) than male children [9].

CONCLUSION

In summary, our study found out that among people who are affected by pediculosis capitis in Makkah region were mostly female. Other risk factors were the frequency of hair washing, type, and length of hair, a high number of family members, sharing personal belongings, beds, and transition of infestation between family members, in addition to employment and socioeconomic status. The results that gathered here, using a questioner reflected a lower sensitivity of this technique in relation to direct scalp examination, microscopic examination for nets, and taking hair samples from barbershops. We expect for future research to put that in consideration, also to establish the prevalence of pediculosis capitis in Saudi Arabia as well as launching educational campaigns in schools, hospitals, and public areas to increase the awareness hopefully to prevent and control head lice.

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Our Dermatology Online

Original Article

Severe cutaneous drug reactions in Guinean children: a monocentric retrospective study of 35 cases

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ABSTRACT

Background: Data on Severe cutaneous drug reactions (CADRs) are not common among in sub-Saharan Africa children. The purpose of this study was to document the clinical, etiological and evolutionary aspects of Severe CDRs in children hospitalized at the dermatology department of university hospitals of Conakry. **Material and Methods:** Retrospective study, conducted from 1 January 2000 to 31 December 2014. Were included all children aged 0-17 years hospitalized for severe CARDs. The data collected were Socio-demographic, clinical, para-clinical and evolution variables. The data was entered and analyzed using the Excel 8.0 software. **Results:** During a study period, 4437 patients of all ages was hospitalized in dermatology department. 35 patients were included with an average age of 11.3 years and a sex ratio of 1.5.The main clinical patterns were: Stevens Johnson syndrome 37.14% (13/35) Lyell syndrome 25.71 % and generalized bullous fixed eruption 22.85%. The drug was identified as 32 patients (91.42%): Sulfadoxine–Pyriméthamine 40.62%, cotrimoxazole 21.85%, nevirapin 12.5%, ampicillin 6.25%, traditional Pharmacopoeia 6.25% and griseofulin 3.12%. It was taken following self-medication in 14 patients, including a parental initiative in 9 patients. 7 patients had a history of drug allergy and 4 were HIV positive. We recorded 5 deaths. **Conclusion:** Our study confirms the rarity of severe CADRs in children. The importance of the sulfadoxine-pyrimethamine in the occurrence of severe CADRs in children is the particularity of our series.

Key words : Severe cutaneous drug reaction; Children; Toxic epidermal necrolysis; Sulfadoxine pyrimethamine; Guinea

INTRODUCTION

Cutaneous adverse drug reactions (CADRs) are mucocutaneous complications secondary to enteral, intravenous, subcutaneous or intramuscular administration of drugs [1].

They are the cause of hospitalization in dermatology services for about 1.5% of patients in France [2], 4.11% in Tunisia [3], 27% in Togo [4], 10.40% patients in Guinea [5] and have a high degree of semiological variability.

Two per cent of these can be severe, with severe sequelae or life threatening [6].

These severe forms include the bullous cutaneous adverse drug reactions of Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN or Lyell's syndrome), drug reaction with eosinophilia and systemic symptoms (DRESS), and acute exanthematous pustulosis (AGEP) [7].

They can occur in all individuals without distinction of age; however, they are less frequent in children with

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an incidence of 0.9% in the Thiesen et al series in England [8].

Data on severe CADRs are not common among in sub-Saharan Africa children.

To our knowledge, no data on this subject are available in specialized hospitals in Guinea. This study was intended to document the clinical, etiological and evolutionary aspects of CADRs in children in hospitals specializing in dermatology.

MATERIAL AND METHODS

This monocentric, retrospective study was conducted during the period of January 2000 to December 31, 2014 in the Dermatology and Venereology Department of the teaching hospital of Conakry (Guinea). It is the only hospital dermatology service for a country of 11663627 million habitants. Were included all children aged 0-17 years hospitalized for severe cutaneous drug reaction during the study period.

We have considered as severe CADRs all the CADRs that lead to hospitalization or prolongation of hospitalization, life threatening, death, inability or invalidity, or other serious medical condition [9].

Were retained the Stevens-Johnson syndrome (SJS) and Lyell syndrome, DRESS (drug reaction with eosinophilia and systemic symptoms), acute generalized acute exanthematous pustulosis (AGEP) and generalized bullous fixed pigmented erythema.

The diagnosis of SJS and Lyell syndrome was selected based on the percentage of body surface peel-off: SJS <10%, overlap syndrome 10-30%, and Lyell syndrome > 30% [7]. Cases of erythema multiforme and incomplete files were not included.

The data, collected from the subjects were age, sex, antecedents of allergy, name and number of the suspect drug (s), the circumstances of taking the drug (s), the time between taking the drug and the occurrence of dermatological lesions, the clinical type of cutaneous drug reaction, the evolution of the CADRs. When possible, additional tests (HIV serum, blood count, blood ionogram, liver function) were performed.

In order to establish a possible causal link, the French method was used. It is based on semio- logical and chronological arguments [10]. Due to insufficient technical platform, any allergological exploration has been carried out.

The present study was approved by the ethics committee of the medical faculty of Gamal Abdel Nasser University.

The data was entered and analyzed using the Excel 8.0 software.

RESULTS

After applying the exclusion criteria, a total of 35 patients aged 17 and under who had severe CADRs were enrolled the study. During the same period, a total of 4437 of patients of all ages was hospitalized, ie a hospital prevalence of severe CARDs in children of 0.78%. Of the 35 patients, 18 were male and 17 were female. The sex ratio was 1.5. The mean age was 11.3 years and extremes of 4,6 years and 17 years.

Nineteen (54.1%) patients were adolescents with an age range of 13-17 years, 9 patients (26.1%) were great children aged between 7 and 12 years and 7 patients (19.8%) were of young children with an age of less than 7 years. The most frequently detected children severe cutaneous drug reactions were Stevens Johnson syndrome (37.14%), Lyell syndrome (25.71%) (Fig. 1) and generalized bullous fixed eruption (22.85%) (Table 1).

The interview revealed the circumstances of drug intake in all patients. This was the result of a medical prescription in 21 patients (60%) and self-medication in 14 patients (40%), including a parental initiative in 9 patients. In 7 out of 35 patients, there was a history of drug allergy.

Sulfadoxine-Pyrimethamine and cotrimoxazole were the most frequently involved drugs with respective

Table 1: Type and frequency of severe cutaneous drug reactions in children

Type of severe CDRs (N=35)	Effective	Frequency (%)
Toxic epidermal necrolysis:		
- Steven-Johnson Syndrome	13	37.14
- Lyell Syndrome	9	25.71
- Overlap SJS-Lyell	2	5.71
Bullous fixed pigmented erythema	8	22. 85
Acute generalized exanthematous Pustulosis	1	2.85
Dress	1	2.85
CDRs not specified	1	2.85
Total	35	100

proportions of 40.62% (13/32), cotrimoxazole 21.85% (7/32). The clinical types of severe CADRs identified and the main drugs involved are detailed in Table 2.

The lesions appeared between one and three weeks in 19 patients, in less than one week in 8 patients. The time to onset was not specified in 8 patients. HIV serology was performed in 21 patients (60%). It was positive for HIV 1 in 4 patients, ie 19%.

We recorded 5 deaths (14.2%), all of which occurred in patients with Lyell's syndrome (55.5%), of which 3 were infected with HIV. Two cases of symblepharon sequelae following Lyell syndrome were noted.

DISCUSSION

This study describes the clinical, etiological and evolutionary profile of severe cutaneous drug reactions in children based on the information contained in the medical records of patients hospitalized in the Department of Dermatology at the University Hospital of Conakry from 01 January 2000 to 31 December 2014. Although the service of dermatology is the only service of this type in Guinea, our data are not exhaustive.

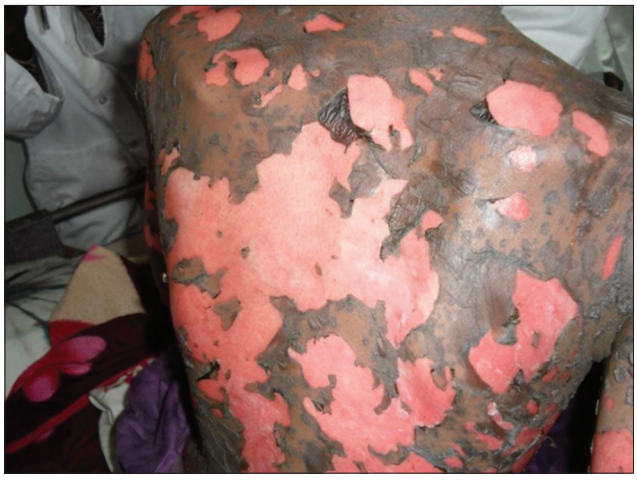

Figure 1: Lyell syndrome with dermal exposure in a 16-year-old patient.

It is possible that some children suffering from severe CADRs may outside hospital structures due to accessibility problems, both economic and geographical. To these are added cultural contingencies which make the patients first practice self-medication and thus rarely resort to a first-line medical consultation consulting first the traditional practitioners. However, with 35 cases of severe CDRS in children collected in 14 years, ie a hospital prevalence of 0.78% and a mortality of 14.2%, our results are similar to those of the literature on the severity of severe childhood CADRs and mortality severe forms of Stevens Johnson syndrome and Lyell Syndrome. These two CADRs were respectively 1st and 2nd severe CADRs in hospitalization in children. These two entities are the spectrum of one disease, differing only by their extent of skin detachment: SJS <10%, overlap syndrome 10 to 30%, and Lyell syndrome > 30%[7]. DRESS and AGEP were less represented with 1 case each. The weakness of the diagnostic facilities and the low frequentation of the hospital structures can explain this low representation.

For *Roujeau J C*, the prognosis of SSJ and Lyell syndrome in children is much better compared to the adult with a mortality of the order of 5% for all cases where the epidermal detachments exceed 10% of the body surface [11].

Our results suggest that in both children and adults in Africa, the mortality of severe CADRs remains particularly high, linked in part to insufficient technical platform and HIV infection, as already noted by other authors [5,12].

Table 2: Distribution of severe cutaneous drug reactions in children according to the drug in cause

Type of severe CDRs / Drug in cause	Toxic epidermal necrolysis			Bullous fixed pigmented erythema	AGEP	DRESS	CDRs not specified
	Steven-Johnson syndrome	Lyell syndrome	Overlap SJS-Lyell				
Sulfadoxine –Pyriméthamine n=13	7	3	1	2			
Salts of Quinine n=1		1					
Cotrimoxazole n=7	2	2		3			
Ampicillin n=2	1				1		
Aspirin n=2				2			
Nevirapin n=4	3	1					
Greseofulin n=1							1
Traditional Pharmacopoeia n= 2			1			1	
Medication found n=3		2		1			
Total	13	9	2	8	1	1	1

The comparison of the frequencies of the different types of severe CADRs encountered during our study in other countries is difficult because studies differ method of recruiting patients. Our study focused exclusively on the hospital. The forms of CADRs called benign as the fixed drug eruption or maculo-papular exanthema require hospitalization only when signs of gravity cause it. The forms of fixed drug erythema hospitalized in our series was generalized. Such a clinical forms will indicate a reintroduction of the inducing drug.

The proportion of the various drugs responsible for CADRs depends on the epidemiology of infantile diseases in the environment, the practice of physician and the use of these drugs in the general population, all of which are related to time [13].

The responsibility of sulfonamides anti-infectious such as cotrimoxazole in the occurrence of severe CADRs, as found in the present study, is a classic fact already reported by other authors [14,15].

However, in our study, as in that of *Sarah et al*. [16] in Côte d'Ivoire, the combination of sulfadoxine-pyrimethamine was the first drug to induce severe cutaneous drug reactions in children in Conakry.

Unlike the French and Tunisian studies where this first place is occupied respectively by vaccines and penicillin. The application of the accountability criteria has made it possible to standardize etiological research and to compare our results with those of other authors (Table 3). Antiretrovirals, in this case nevirapine, are the third most important drug responsible for severe CADRs in our study. This third place is occupied by psycho stimulants in France and anticonvulsants in India and Malaysia.

The important role of the sulfadoxine-pyrimethamine combination in the occurrence of children CADRs is the particularity of our series. In malaria-endemic areas such as Guinea, sulfadoxine-pyrimethamine is recommended for the prevention of malaria in combination with amodiaquine during the high malaria season to prevent disease, with the objective of maintaining concentrations treatment of antimalarial drug in the blood during the period when the risk of contracting malaria is higher [22].

The use of this molecule in monotherapy and often in self-medication as observed in 40% of our patients is an inappropriate practice. Indeed, for *Laroche et al*, self-medication and the illicit sale of medicines increases the inappropriate consumption of medicines favoring the occurrence of potentially preventable side effects [23].

Two percent of the patients in our series had a history of drug allergy. One might think that a prescription made under the conditions of a medical consultation taking into account the history would have avoided the occurrence of the cutaneous drug eruption 2% of the patients. In addition to the risk of occurrence of severe toxidermia at the occasional fatal outcome, the use of the sulfadoxine-pyrimethamine combination as monotherapy favors the emergence of resistance mutations of Plasmodium falciparum. This resistance makes the fight against malaria even more difficult in endemic areas [24].

Efforts in the fight against the illicit sale of medicines, coupled with public awareness and education campaigns on the harmful effects of self-medication, may contribute to reducing the prevalence of these severe cutaneous drug reactions in children.

Table 3: Main drugs involved in descending order in different series of cutaneous adverse drug reactions (CADRs) in Children

Rank	1st Rank	2nd Rank	3rd Rank
our serie (Guinée)	Antimalarial (Sulfadoxine –Pyriméthamine)	Sulfonamides (Cotrimoxazole)	Antirétroviraux (Névirapine)
Adegbidi et al Benin [13]	Sulfonamides (Cotrimoxazole)	-Penicillin -Vaccine -Acetaminophen	non-steroidal anti-inflammatory drug (Aspirin)
Khaled A et al Tunisie [17]	Penicillin	Non-steroidal anti-inflammatory drug (Aspirin)	Antiepileptics (Phenobarbital)
Dilek et al Turkey [18]	Penicillin and its derivatives	Acetaminophen	Vitamins and minerals
Damien et al Saint-E´tienne [19]	Vaccin BCG (bacille de Calmette et Guerin)	Anti-neoplastic and immunomodulating agents(Asparaginase)	Psychostimulant (Méthylphénidateà
Rosli R et al Malaysia [20]	β-Lactam Antibacterials, Penicillins	Analgesics and Antipyretics	Antiepileptics
Qayoom et al Inde [21]	Antimicrobials(ofloxacin)	Non-steroidal anti-inflammatory drugs(Piroxicam)	Antiepileptics (Phenytoin)

In three patients, the inducer drug was not found, which increases the risk of accidental reintroduction of the drug.

CONCLUSION

Severe cutaneous drug reactions are relatively rare in specialized hospitals in Conakry. As in adults, severe and HIV-associated forms have a poor prognosis. The etiological profile of the 35 cases of severe CADRs observed in the children of our serie is generally comparable to those already reported in the literature. However, the importance of the sulfadoxine-pyrimethamine combination in the occurrence of severe CADRs in children is the particularity of our series.

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Our Dermatology Online

Original Article

Melanoma and medical education: student's perceptions of skin cancer screening in three medical schools

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ABSTRACT

Background: This study evaluated how the level of medical education affected confidence of counseling on skin health, performing skin examinations, and the likelihood of using those skills in future screenings. **Material and Methods:** An online survey was distributed to students at three midwest medical schools that assessed sun protection behaviors, and the student's perception of education and comfort on the complete skin examination and counseling on skin health. **Results:** 51.6% (n=243) indicated they were in the last two years of medical school and were considered advanced students, compared to less advanced students in the first two years. Advanced students were significantly more comfortable in performing a complete skin examination and counseling patients on skin health when compared to less advanced students ($p < 0.001$). However, they were also statistically less likely to indicate they would provide skin screening on future patients when compared to less advanced students ($p < 0.001$). Only 29.5% (n = 139) and 60.7% (n = 286) of students indicated they had been educated on performing a complete skin examination and counseling on skin health, respectively. **Conclusions:** Although advanced students were more likely to report education and comfortability in skin cancer screening they were not as likely to indicate they would practice these skills in the future compared to less advanced students. Also, only a small number of students perceived themselves to be educated in these aspects.

Key words: Skin health; Melanoma; Complete skin examination; Medical education; Primary care

INTRODUCTION

Although melanoma is the third most common form of skin cancer behind basal cell carcinoma and squamous cell carcinoma, it is the deadliest [1-4]. Over 63,000 people are diagnosed annually and nearly 9,000 people die from this affliction yearly [4,5]. In 2016, the estimations of melanoma incidences and mortalities were 76,360 and 10,130, respectively [1]. It is one of the most common cancers among young adults and is currently the fifth most common cancer in males and sixth in females [4,6,7]. Overall, melanoma is most prevalent among caucasians, older adults, and males, although, in individuals below 50 years of age, it is more frequent in females [2,4,8]. Although the five-year survival rate has been improving over the past two decades, at about 90%, this rate varies among different stages of disease. For example, over 90% of patients with local disease survive the five years compared to less than 20% of patients surviving with metastatic disease [4,7,9]. These variations demonstrate the significance of early

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detection and prevention and how these aspects can reduce the progression of melanoma.

The outcome of melanoma, a potentially fatal disease, is dependent on early diagnosis. When detected by a physician, that physician is almost always a primary care physician (PCP) rather than a specialist [3,10,11]. It has been found that most patients with newly diagnosed melanoma have visited their PCP at least once the year prior to their diagnosis [6,10,12,13]. Even so, only 20% of these patients report receiving a skin cancer examination, indicating that this diagnosis could have been detected sooner [10,12]. Also, the majority of Americans do not go to the dermatologist regularly, so without finding high-risk lesions themselves or via PCP, these patients would not be able to have the early detection that is crucial with a melanoma diagnosis [6,9]. Of melanomas found by physicians, over 75% are detected by PCPs [13]. Therefore, PCPs are important targets for skin cancer prevention practices and education. They are important for early detection as well because melanoma lesions diagnosed at later stages are associated with a greater risk of death. However, evidence is lacking regarding the best way to conduct early detection in order to reduce melanoma mortality and is a controversial subject currently [4].

At this point, there is conflicting information between medical organizations regarding prevention recommendations and skin cancer screening and little research has been done to delve into the obstacles against the CSE. Whereas some organizations recommend screening of all adults, others suggest screening high-risk adults only, and others do not promote screening at all. There is also not much research in the area of early detection or which modality works best. These inconsistencies and the minimal amount of research in this area prompted the authors to further investigate. We studied medical school education as this is where future medical professionals would typically get their first exposure to dermatological education. The survey examined aspects on whether the level of medical education influenced the student's perception of their desire to tan, their own sun protective practices, comfort-level and amount of education on counseling in skin health risk factors and performing the complete skin examination, as well as the likelihood to use these skills on future patients. The purpose of this study is to analyze and discuss how the comfort level of a physician can influence the performance and use of sun safety screening and education. This emphasizes the need for PCPs to widen their scope of practice into the dermatological field, especially with early detection of skin cancer.

MATERIAL AND METHODS

The survey had to be developed and the questions determined before it was emailed to approximately 1,200 medical students at the University of Kansas School of Medicine, University of Missouri—Kansas City School of Medicine, and Kansas City University. The survey had two primary sections and included a total of 22 questions administered to all medical students at the three medical schools via email. The survey consisted of 22 questions divided into demographics, personal attributes/behaviors, and curriculum sections. The demographics section assessed the student's gender, age, skin type, personal history of skin cancer, and year of medical education. The personal attributes/behaviors section assessed student's own sun protection behaviors, and view of complete skin examination (CSE) and counseling patients about skin cancer risk factors. When discussing counseling throughout this paper, please note that it is referring to counseling patients on risk factors and prevention of skin cancer. Curriculum assessed the student's perception of education, comfort, and level of importance on the CSE and counseling on skin cancer risk factors. This survey was administered to all medical students at three medical schools via email.

The University of Missouri-Kansas City hosted REDCap electronic data capture tool – a secure, web-based application designed to support data capture for research studies, providing an interface for validated data entry, audit trails for tracking data manipulation, and automated export procedures and procedures. Also used was a Stata statistical software package (SE version 9.2; Stata Corp., College Station, Texas) for all analyses. These modalities were used to compare medical students in medical school years 3 or 4, considered advanced students to students that indicated medical school years 1 or 2, considered less advanced students. The University of Kansas Institutional Review Board (IRB), University of Missouri-Kansas City IRB, and Kansas City University IRB approved this study and an IRB approval exemption was obtained from each school.

The chi-square test generated descriptive statistics and correlations performed evaluated direction and magnitude of any significant associations found

between survey items. We conducted sequential and systematic stratified univariate and multivariate logistic and multinominal regression analyses, as appropriate, to assess for significance, confounding and effect modification on selected items utilized as outcome variables. The final regression models used all variables found to be statistically significant or to be associated with confounding/interaction. Regression modeling determined odds ratios (OR) and 95% confidence intervals (CI) and all tests for significance were two-tailed with an a priori level of 0.05 (p</= 0.05).

RESULTS

A total of 1,200 students were surveyed with a response rate of 39.25% (n=471). The survey responses were reviewed for completeness and blank surveys (n=2) were not included in the final study population or descriptive statistics. Of the responders, 51.6% (n=243) of students indicated they were advanced students, compared to 48.2% (n=227) that indicated they were less advanced students. Female (54.8% (n=258)) Male (44.6% (n=210)) The indications of age groups were as follows: 17-21 years 6.4% (n=30), 22-26 years 68.6% (n=323), 27-31 years 19.5% (n=92), 32-36 years 3% (n=14), 37-41 years 0.8% (n=4), and >/= 42 years 1.1% (n=5). Skin types based on the Fitzpatrick scale are as follows: Type 1 (white, very fair) 6.4% (n=30), Type 2 (white, fair) 23.4% (n=110), Type 3 (beige, very common) 46.3% (n=218), Type 4 (beige, with brown tint) 18.5% (n=87), Type 5 (dark brown, very rarely burn) 4.9% (n=23), and Type 6 (black, never burn) 0.6% (n=3).

92.6% (n=436) have utilized sun protection in the past year compared to 6.8% (n=32) who did not. 56.5% (n=266) spent time in sun with intent to tan in past year compared to 43.1% (n=203) who did not. 10.4% (n=49) utilized a tanning booth in past year compared to 88.7% (n=418) who did not. 19.5% (n=92) utilized artificial tanning products compared to 79.8% (n=376) who did not. 36.3% (n=171) believed that their medical education changed their desire to tan compared to 39.5% (n=186) who did not, and 23.8% (n=112) who indicated that this was not applicable due to no desire to tan. 5.1% (n=24) indicated personal history of skin cancer whether suspected or confirmed compared to 94.9% (n=447) who did not. 25.5% (n=120) indicated they had a first-degree relative history of skin cancer compared to 74.3% (n=350) who did not.

Advanced students were statistically more likely to report more sun protective practices ($p = 0.008$, $r = 0.169$) and to indicate that medical education has changed their desire for tanned skin ($p < 0.001$, $r = 0.181$) when compared to less advanced students. Advanced medical students were also significantly more comfortable in performing a CSE and counseling patients on skin health, or skin cancer screening, when compared to less advanced students ($p < 0.001$ for both variables) ($r = 0.229$ for CSE) ($r = 0.218$ for counseling). Although, advanced students were statistically less likely to indicate that they were very likely or somewhat likely to perform skin cancer screening on future patients when compared to less advanced students ($p < 0.001$, $r = 0.289$ for CSE) ($p < 0.001$, $r = 0.252$ for counseling).

However, regarding performing a CSE and counseling patients about risk factors of skin cancer, only 29.5% (n = 139) and 60.7% (n = 286) of medical students, respectively, indicated that at their level of education had been educated on these topics. Only 33.1% (n=156) of respondents indicated that to this point in their medical training they feel comfortable in performing a CSE and 73.7% (n=347) indicated they felt comfortable in counseling patients. The majority of respondents, 80.9% (n=381) indicated that they believed it was very important or somewhat important for patients to receive an annual CSE. This is compared to 14.6% (n=69) that felt it was neither important nor unimportant, 4.5% (n=21) that indicated somewhat unimportant and 0.0% (n=0) to indicate very unimportant for patients to receive an annual CSE. The majority of respondents, 67.7% (n=319), indicated very likely or somewhat likely to perform an annual CSE on their future patients, compared to 16.6% (n=78) indicated neither likely nor unlikely, 15.7% (n=74) indicated somewhat unlikely or very unlikely to perform annual CSE on their future patients. The majority of respondents, 98.5% (n=646) indicated that they believed it was very important or somewhat important for patients to be counseled on risk factors for and prevention of skin cancer. While, 0.8% (n=4) indicated neither important nor unimportant and 0.0% (n=0) indicated somewhat unimportant or very unimportant for patients to receive counseling on risk factors for and prevention of skin cancer. The majority of respondents, 88.9% (n=419) indicated they were very likely or somewhat likely to counsel their future patients on the risk factors, 6.2% (n=29) indicated neither likely nor unlikely and 4.8% (n=23) indicated

somewhat unlikely or very unlikely to counsel their further patients.

Respondents who indicated that they had been educated on performance of CSE were significantly more likely to indicate that they felt the CSE was the responsibility of a patient's PCP ($p=0.029$, $r=0.122$). These respondents that indicated they were educated on performance of CSE were also significantly more likely to indicate they felt it was very important or somewhat important for patients to receive an annual CSE ($p=0.040$, $r=-0.115$). They were also significantly more likely to indicate that it was very important or somewhat important to counsel patients ($p=0.040$, $r=-0.110$). Also, these respondents had a statistically significantly higher number of total sun protection practices ($p=0.003$, $r=0.182$) and that they were educated on how to counsel ($p<0.001$, $r=0.370$). It was significantly more likely for those that had indicated being educated on counseling patients to also indicate they were likely to perform a CSE on their future patients ($p=0.001$, $r=0.190$).

DISCUSSION

Melanoma is the deadliest form of skin cancer and skin cancer is among the fifth and sixth leading causes of death in men and women, respectively [4,6,7]. Although not proven through randomized clinical trials at this point, early detection has been associated with a better mortality and increased survival rate [8,14,15]. There are inconsistencies with the guidelines on early detection and how to approach skin health screening in general, especially primary practice [5,14,15]. Therefore, more research is needed in this area to determine the best way to conduct early detection and screening [5].

The survey showed that although advanced medical students were statistically more likely to indicate that they felt educated on and comfortable with skin cancer screening, they were not as likely to indicate that they would practice these in the future when compared to less advanced medical students. Although, only a small number of advanced medical students perceived themselves to be educated at their point in training on both of these aspects of skin health. This could suggest a number of things, namely that students do not perceive their experiences accurately. It could also suggest that the students personally would not plan to carry out these practices in the future due to a specialty that are planning on pursuing since there was no option on the survey for future specialty. Or this could suggest that students do not see a need for dermatology in primary care. Advanced students were more likely to indicate higher number of sun protection practices and that their medical school education had changed their desire for tanned skin. This could suggest that education on risk factors of skin cancer and general dermatological education has an effect on sun protection practices.

There were some limitations in our study such as, we did not ask students their future specialty. This would confound results if students were planning on a future specialty that would not encompass skin health counseling or CSE. We also did not ask students their opinions on ways to improve the dermatological education within their respective schools. Although this was not a necessity, it would have been useful and relevant data. There were also only three schools surveyed and all confined to the Midwest and a metropolitan area. Further research is needed to be able to generalize our data to a national scale.

Early detection for skin cancer is a controversial topic with no clear guidelines and more research is needed in this area to determine best modality for early detection [4,5]. Therefore, this area was of interest. We looked into medical schools to investigate the beginning of dermatological education and grasp the perceptions of future physicians. We recommend further research in the education of medical students on the importance of early detection and on the role of the primary care physician in dermatology. Another recommendation is investigation into the best deliverance methods of dermatology education in medical schools. Some suggestions from the authors include incorporating use of live patients, grand round approach and minimal rotational experience in dermatology in addition to the basic science lectures. Ulman, Binder and Borges proposed similar modifications to curriculum after administration of a dermatology quiz to medical students [11]. Further investigations in the curriculum would help ensure that new physicians would be able to identify new cases of skin cancer.

CONCLUSION

Students' own perceptions of their education regarding skin cancer screening indicated that they were lacking in certain areas. Although, the majority of students

reported that they were less likely to utilize skin cancer screening in their future practice. This possibly suggests the need for medical school curriculums to increase training and practicing of the clinical skills needed in order to recognize early stages of skin cancer as well as increased ability to counsel patients on the risk factors of skin cancer. This could include curriculum reform in medical schools regarding dermatology education. It could also mean educating students regarding the importance of dermatology in primary practice. However, it is difficult to know whether students reported specific answers due to future desired specialties. Even so, future investigation is needed in this area to determine if this is needed and the mode of education that would most benefit students.

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Our Dermatology Online

Original Article

The number of dysplastic and common nevi in patients with malignant melanoma

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ABSTRACT

Background: There is no data about melanocytic lesions in Turkish patients with melanoma. We aimed to investigate number of melanocytic nevi in melanoma patients compared with control subjects. **Methods:** We performed a retrospective study in 78 melanoma patients and 78 control subjects. Clinical data were recorded for all participants. **Results:** As compared with control subjects, patients had more common nevi than control subjects on the legs. The patients and control subjects had similar number of overall and dysplastic nevi. **Conclusions:** Furthermore, the number of nevi is not a risk factor for our patients, and patients with few or no moles may develop MM, all patients need attention.

Key words: Dysplastic; Nevi; Melanoma

INTRODUCTION

The dysplastic nevus was first described in the 1970s in families prone to develop melanoma. It was believed that these lesions had a higher risk of transformation to melanoma than common nevi [1,2]. In 1992, Consensus Conference at the National Institutes of Health (NIH) described the criteria for dysplastic nevus [3]. Some of the authors suggest that nevus known as dysplastic nevus are actually congenital nevus. It is established that patients with melanoma have more Dysplastic nevi and common nevi than patients without melanoma [4-7]. We aimed to determine the number of nevi in melanoma patients.

MATERIALS AND METHODS

We performed a retrospective study on patients with malignant melanoma, who were recruited from those consecutively attending the outpatient clinics of department of dermatology at Bezmialem Vakif University, during January 2005 through December 2016. Seventy eight melanoma patients and age-gender matched 78 control subjects, were enrolled in the study. The study was approved by the local ethics committee. Informed consent was obtained from all participants, and the study was conducted according to the Declaration of Helsinki Principles. Patients and controls fulfilled the following criteria: (1) no positive family history of melanoma; (2) not referred for another skin cancer; (3) no previous history of phototherapy or any immunosupression therapy and; (4) no previous history of malignancy (5) with a Fitzpatrick skin type 4 or 5.

For all participants, melanocytic nevi which have atypical dermoscopic and clinical features at first visit (from 5-15 mm in diameter, flatness, indistinct borders, and color irregularity) followed up for 5 years and more without melanoma development, and common nevi (size >2 mm, sharp border, homogeneous light to dark brown color) were counted on all body sites (including palms, soles, scalp, and genital areas) and recorded using categories (0; 0-10; 11-50; 51-100; >100).

Dermoscopic images of all the lesions were acquired at a 30 X magnification and were stored in a digital imaging system (Fotofinder). All digital images were examined by experted dermatologist in a blinded fashion and were evaluated for global and local features.

All melanomas included in the analysis were; 3 (1.9%) patients with lentigo maligna melanoma, 7 (4.5%)

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with acral lentiginous melanoma, 40 (25.6%) with superficial spreading melanoma, 13 (8.3%) with nodular melanoma, 3 (1.9%) patients with spitzoid melanoma, 8 (5.1%) patients with in situ melanoma and 4 (2.6%) with unknown origin.

Statistical Analysis

Continuous variables were compared using Student t test and Mann-Whitney-U test. For categorical variables, different groups were compared using chi-square tests. All statistical tests were 2-tailed and considered significant for P values of 0.05 or less. The analysis was carried out using SPSS.

RESULTS

Characteristics of the Study Population

A total of 156 participants, 78 melanoma patients [31 male and 47 female; median age: 55 years (range 16-89 years)] and 78 control subjects [31 male and 47 female; median age: 50 years (range 16-74 years)] were included in the study (Table 1).

We found a wide variation in the total number of nevi both in melanoma and control groups (from 0 to 226). Melanoma patients had more nevi than control subjects, both in the overall nevus count and in the number of atypical nevi, but both of them were not statistically significant (P:0.260 and P:0.382, respectively) (Table 1).

In detail, 1 of 78 (1.28 %) patients versus 1 of 78 (1.28%) control subjects had no nevi, 26 of 78 (33.3 %) patients versus 13 of 78 (16.7%) control subjects had 1 to 10 nevi, 30 of 78 (38.5%) patients versus 47 of 78 (60.3%) control subjects had 11 to 50 nevi, 12 of 78 (15.4%) patients versus 14 of 78 (17.9%) control subjects had 51 to 100 nevi, and 10 of 78 (12.8%) patients versus 4 of 78 (5.1%) control subjects had more than 100 nevi.

In melanoma patients, common nevi were more prevalent on the legs than in control patients (13 vs. 7). The common nevi count didn't differ for other sites between melanoma patients and controls. For Dysplastic nevi, they were most often located on trunk for both melanoma patients and controls, but the nevi count on trunk, legs didn't differ statistically between the groups (P:0.420) (P:0.114).

Logistic regression analysis, showed no specific and statistical significant feature for melanoma (Table 2).

Table 1: Clinical features of patients and controls

	Patients	Controls	P value
Age	55.50±12.91	49.30±15.21	0.062
Common nevi	40.96±37.81	36.66±28.56	0.260
Head and neck	9.62±8.37	7.25±5.29	0.115
Upper extremities	8.25±8.24	10.27±9.60	0.074
Trunk	9.73±9.31	11.94±9.28	0.084
Lower extremities	13.36±11.89	7.2±4.39	0.035
Clark nevi	3.37±4.77	1.85±1.68	0.382
Head and neck	0.64±1.72	0.36±0.62	0.075
Upper extremities	0.45±1.54	0.11±0.71	0.093
Trunk	2.08±1.10	1.01±0.13	0.420
Lower extremities	0.19±0.41	0.37±0.22	0.114

Table 2: Risk factors for Melanoma

	P value	Odds ratio	95% confidence interval
Age	0.53	0.962	0.938-0.987
Gender	0.806	0.917	0.461-1.826
Overall nevi count	0.376	1.006	0.993-1.018
Clark nevi count	0.054	0.799	0.643-0.994

DISCUSSION

Dysplastic nevus (DN) was defined as a clinical and dermatopathological described lesion in melanoma-prone families. B-K mole syndrome was defined as in 15 patients with melanoma, as having <10 to >100 nevi, with various sizes (from 5-15 mm in diameter), irregular border, and multiple colors in Dysplastic et al's report [1]. On the other side, in their report two individuals developed melanoma without having atypical nevus.

The presence of a large number of nevi (>50) with several, atypical clinical features was accepted as a criteria of Familial Atypical Multiple Mole Melanoma (FAMMM) syndrome or Dysplastic Nevus Syndrome (DNS) in 1992 National Institutes of Health Consensus [3]. Having more than 100 nevi with a diameter of 2 mm or larger; more than 50 nevi if the patient is younger than 20 years of age were scored as one point according to the British group dysplastic nevus syndrome score [8].

The Dutch Working Group, described the atypical nevus as being >5 mm in size, or having a vague border, asymmetric shape, irregular pigmentation, and red hue [9].

Harada, Ackerman and Kittler were the first to indicate the importance of congenital melanocytic nevus (CMN), and proposed that these DN are actually CMN, in 2007 [10,11]. It is accepted now, 'dysplastic

nevus,' cannot be defined by clinical or dermoscopic examination and therefore it is a histological term. Therefore, we preferred to use the term 'Dysplastic nevus' instead of 'Dysplastic Nevus' in this study.

While these hypotheses are still debated, we aimed to investigate the association between nevus count (both dysplastic and overall) and melanoma development. There is sufficient clinical evidence indicating that melanoma most commonly develops de novo, infrequently melanoma arises from preexisting nevi, which may be either common nevus or Dysplastic nevus. According to the literature, it appears that 20% to 30 % of melanomas appear to arise from nevi [12].

Based on current data, there is evidence that a high nevus count correlates with a higher risk of melanoma [6]. In our study of Turkish population, we didn't find an elevated melanoma risk in patients with high nevus counts. Our results are in contrast with the results of several case-control studies showing that high total-body nevus counts are the major risk factor for the development of MM [6,7]. This result may be due to genetic factors, Fitzpatrick skin type of Turkish population, and increased sun exposure resulting from the geographic region of our country. The populations in other studies mostly had Fitzpatrick skin type 1, 2 or 3, whereas in our country, Fitzpatrick skin type 4 and 5 are common in general population. The habit of sunscreen usage are lower than the individuals from Europe, the USA and Australia.

Two genetic variants, at 9p21 and 22q13, have recently been identified by genome-wide association studies (GWAS) to be associated with melanocytic nevi development [13]. A recent GWAS identified a novel susceptibility locus known as nidogen 1 (NID1) on 1q42 for nevus count and melanoma risk [14]. Results of the study indicated that increased expression of nidogen in one variant NID1 SNP (rs10754833 T allele) was significantly associated with decreased melanoma risk (OR, 0.86). Since these studies were carried out in Europe and USA, these genetic mutations were not identified in Asian patients. We propose that the similarity of nevus count between patients and controls originate from genetic alterations of different geographic regions.

In conclusion, our data show that (1) high counts of common nevus on the legs in melanoma patients; (2) no difference for number of common nevus; (3) no difference for number of Dysplastic nevi; (4) similar risk of melanoma for individuals with few or more moles. All the patients require a careful screening for melanoma.

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Our Dermatology Online

Original Article

Home remedies for Pediculus humanus capitis infection among schoolchildren

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ABSTRACT

Background: Head lice (HL) is still signifying problem, its incidence is greater in crowded places like schools and day cares. **Material and Methods:** A total of 1791 school children, from five schools (three elementary and two intermediate) in Kirkuk province, Iraq, were eligible for screening of head lice. The heads were examined carefully by naked eye or with assistance of magnifying lens. After detecting the infected heads, natural and chemical products were used for treating the infection. **Results:** 255 samples were found to be positive for head lice, with rate of 14.2 %. Females were significantly more infected than males. The most age group which was infected in both genders was 7-8 and 9-10 years old. parents occupation or the educational status of the infected children, were not associated with the incidence of the infection. The effect of the natural plant oils were intensified when used with head cap. The most effective plant oil was olive and anise followed by thyme and sesame they were effective at 48 hours after usage. Very little effect had resulted with garlic oil. Petrol was effective in killing the lice with or without head cap. The anti-lice shampoos were effective after repeating treatment for four to five days. The ordinary hair shampoos were effective at killing the lice only when used with head cap. Olive oil and sesame are effective for head lice treating, but the commercial normal hair shampoos are better for completely removing the infection, if used with hair cap. **Conclusion:** The recommendation is that, the ordinary hair shampoos can be used instated of natural oils or anti-lice shampoos that contain insecticides and may have side effects.

Key words: Head lice; Home remedies; School children

INTRODUCTION

Head lice belong to Anoplura (sucking lice), it's an obligate ectoparasite. Human being is the only known host for the head, body and crab lice, however, there is evidence that crab lice can infect gorilla [1]. Head lice cannot fly or jump. Therefore, it transmit from one to another by direct contact, or by using hair combs, brushes, hats and bedding of infected person [2]. HL sucks human blood for nourishment causing head itching and discomfort. The infection may also cause anemia particularly in individuals with low hemoglobin level. A greater importance is that, their saliva may led to allergy, psychological effect and absence from school [1,3]. Factors as crowd and poverty may enhance the infection [4]. Despite the huge number of chemical components used for treating the infection, HL is still prevalent worldwide [5]. In Iraq and Arab countries the parasite rate is fluctuating. In Kirkuk city, in 16 primary schools, of 1130 pupils (828 girls, 302 boys) examined for HL. A rate of 20% was positive [3]. From eight elementary schools in Baghdad, 540 boys and girls, the total rate of head lice was 13.5% [6]. The head lice prevalence among 5150 Jeddah city Saudian girls were11.26% [7]. The overall prevalence of pediculosis was 23.32%, among 747 schoolchildren from12 primary school of Ladkrabang district, Bangkok, Thailand [8].

Preschool and elementary school aged children were most common infested with head lice in the United States [9]. The prevalence was 43% in a slum and 28% in a fishing village in Brazil [4]. Many chemical products were investigated for their effects on HL, in United States the most studied pediculicide was Permethrin, the least toxic to human [10]. In 2009, 5% benzyl alcohol was inspected as a treatment for HL

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in children older than 6 months [11]. Three head lice treatment options: malathion, pyrethrins and piperonyl butoxide was conducted and studied in children who had severe head lice infestations in Queensland primary schools, [12]. But no or very little effects were recorded in all mentioned studies. Anise, ylang-ylang, coconut oils and isopropyl alcohol was found to be at least as effective as the permethrin [13]. Because of the urgency for new therapies, due to resistance phenomenon appeared in HL against chemical product, the absence of eradicative treatment and HLincreases in human population, the aim of the present study was to detect the rate of head lice in Kirkuk province and practice some natural and chemical products HL.

MATERIALS AND METHODS

Methods

Population Study: From September 2015 to April 2016, a total 1791 school children were screened for head lice presence. The children were from five schools (three elementary and two intermediate)in Kirkuk province, Iraq. Data including gender, age, head or body itching, parents occupation, economic and educational status of all infected children were recorded.

Head screening and sample collection: The head of all children were examined carefully by naked eye or with assistance of magnifying lens. After detection of insect stages (nit, nymph, adult), they were transferred into disposable cup with tide lid. The specimens were bring to the laboratory for microscopic examining.

Treatment groups: 190 person of heavy or moderate infected were agreed to be chosen for treatment experiments. they were divided in to two main groups (120 infected person for natural products and for 70 person for chemicals). From each of the two groups a number of 10 infected person were used for each product (two person for each time used).

Treatment experiments: Different natural and chemical components were used in lice treating. The natural products, composed of plant oil (olive, thyme, anise, sesame, garlic and apple vinegar), these was obtained from local markets. Chemicals were petrol, anti-lice shampoos (Sali, lice therapy and lycid). In addition to ordinary or traditional hair shampoos. All products except anti-lice shampoos were tested with or without using a plastic head cap. The cap when used, was completely covered the head along with the ears and well tide retaining the air form reaching the head. The natural components were left on the infected head to different times (2, 8, 12, 24, 48 hours) while the chemical components was used for 1-5 days (anti-lice shampoos were used according to instructions on their bottles). A towel was put around the plastic cap, to prevent the leak out. After applying a massive amount of plant natural oils or shampoos on the infected head. They were mixed thoroughly with all parts of the hair. After each time or day of treating, the heads were examined periodically for the presence or absence of the lice stages. Afterword (two to eight weeks) the treated heads were reviewed for the recurrence of the infection. Along with hair treatment the families of treated groups were asked to expose the clothing and bedding to heat or to sun for two to five days.

2-4-Statistical Analysis: T test and χ^2 (chi-square) test in style of independent and in style of homogeneous were used manually. The significant level used was $P< 0.01$ or 0.05.

RESULTS

This study had designed to find out the prevalence of head lice in children of Kirkuk city. All stages of the HL were seen and magnified microscopically, Figs. 1-3 denotes these stages.

A significant differences of head lice infection was noted between female and male, Table 1. The rate of infection was higher in female (23.6%)comparing with that in male (3.2%). The overall frequency rate of HL among children of Kirkuk province was 14.2 %.

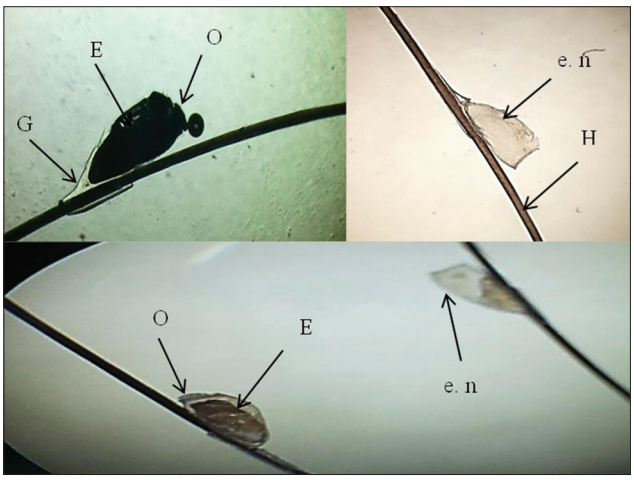

Figure 1: The embryo containing or empty nits of head Lice. (E= embryo, O= opercula, G= glue used for egg attaching with hair, e. n= empty nit, H=hair shaft).

Figure 2: The nymph stage of head lice.

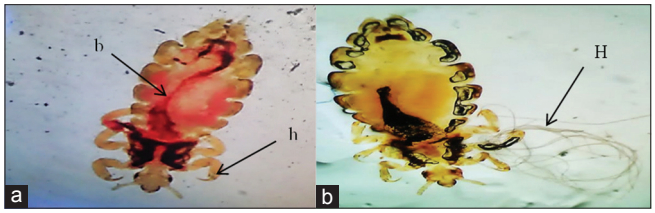

Figure 3: The adult stages of head lice (a) a blood sucked adult, (b) none blood sucked adult) b= blood, H=hairs, h= hanging claw on the leg.

The result in Table 2 indicates, that there were significant differences in HL infection between age groups. The most age group which was infected in both females and males were 7-8 and 9-10 years old. With a rate of 41, 32.8 % for females and a rate of 13,7 % for males in each age group respectively.

The recurrence of HL infection is illustrated in Table 3. The result was significant for those had continuous infection comparing with children infected for first or second time. In female the maximum infection rate (65%)was for continuous infection, while in male was for those infected for first time with rate of 73.1%.

HL infection had well related with head or body itching, Table 4. A rate of 44.9% of infected persons had suffered from head itching and a rate of 15.3% had suffered from both head and body itching, whereas 40% of them had no head or body itching.

As indicated in Table 5, the rate of heavy infected persons were significantly higher (64.3%) than moderately (20.8%) or lightly (14.9%) infected.

The parents occupation or educational status of the infected children, were significantly not associated with the incidence of the infection. Both employed or un employed parents or parents at different levels of educational status had infested child with nearly equal rates.

All families with different levels of economic status had infested child. But the greatest percentage was for intermediate level with rate of 66.5%, followed by bad level with rate of 20.4%, the lowest rate was among families of good level with rate of 14.35.

The effect of the natural plant oils were intensified when used with head cap. All plant oils were not effective at

Table 1: Head lice frequency among school children

Gender	Total no. examined	+ve (%)	–ve (%)
Female	971	229 (23.6)	742 (76.4)
Male	820	26 (3.2)	794 (96.8)
Total	1791	255 (14.2)	1536 (85.8)
T value	Evaluated T value=29.5, T value of P<0.05=6.31 (significant**)		

Table 2: Head lice frequency according to pupils age

Age group in years	Gender	
	Female	Male
	+ve (%)	+ve (%)
7-8	94 (41.0)	13 (50)
9-10	75 (32.8)	7 (26.9)
11-12	42 (18.3)	6 (23.0)
13-14	18 (7.9)	0 (0)
Total	229	26
T value	Evaluated T value=2.55, T value of P<0.05=2.35 (significant)	

Table 3: Head lice frequency according to head or body itching

Gender	No. had head itch's (%)	No. had head and body itch's (%)	No. had no head or body itch's (%)
Female	94 (41.0)	37 (16.1)	98 (42.8)
Male	20 (76.9)	2 (7.7)	4 (15.4)
Total	114 (44.9)	39 (15.3)	102 (40)
χ^2 value	Evaluated χ^2 value=12.21, χ^2 value of P<0.01=9.21 (significant)		

Table 4: Some natural oils effect on head lice viability

Natural oils	Time in hours				
	2	8	12	24	48
Olive					
W	A	A	A	A	A
Wc	A	A	A+D	A+D	D
Thyme					
W	A	A	A	A	A
Wc	A	A	A	A+D	D
Anise					
W	A	A	A	A	A
Wc	A	A	A+D	D	D
Sesame					
W	A	A	A	A	A
Wc	A	A	A+D	A+D	D
Garlic					
W	A	A	A	A	A
Wc	A	A	A	A+D	A+D
Apple vinegar					
W	A	A	A	A	A
Wc	A	A	A	A+D	D

W=without cap, Wc=with cap, A=a life head lice, D=dead head lice, A+D=a life more than dead, a+D=very few alive.

Table 5: Some chemical components effect on head lice

Chemical components	Days of washing or treating and combing the hair				
	1	2	3	4	5
Petrol					
W	A	A+D	A+D	D	D
Wc, 2-3h	A+D	D	D	D	D
Anti- louse shampoos					
Wm					
Sali	A+D	A+D	A+D	A+D	D
Lice therapy	A+D	A+D	A+D	D	D
Lycid	A+D	A+D	A+D	A+D	D
Ordinary hair shampoo (2-3 h. each time)					
W	A	A	A	A	A
Wc	A+D	D	D	D	D

W=without cap, Wc=with cap, Wm=with comb, A=a life head lice, D=dead head lice, A+D=a life more than dead, a+D=very few a life, h=hour

the maximum time used (48 hour), when used without cap. The most effective was olive and anise, followed by thyme and sesame they were effective at 48 hours after usage. No or very little effect had resulted with garlic oil treatment. Also low effect was recorded with apple vinegar. (Nearly all of natural products treated persons had recurrence infection after two to eight weeks post treatment, except olive and anise).

Petrol was effective at killing the lice, with or without head cap especially after repeating treatment. The anti-lose shampoos were effective after repeating treatment for four to five days. The ordinary hair shampoos had no effect in killing the lice when used solely, however their effect were very greatly intensified when used with head cap. Their effect were appeared after 2-3 hours of usage. Repeating this process for two to three days had completely removed the lice. Fig. 4 shows the dead removed lice after hair shampoos usage as appeared with naked eye. (All of chemical treated persons had no recurrence infection after two weeks to one month post treatment). The insect adult had the ability to still alive on beddings and clothes for two days in very sunny Summer day, but they were die after one day when put in a small glass bottle with strong tide cover.

DISCUSSION

Pediculosis is remain a public health problem in our country as in some others. In our province the incidence of the infection among children under the study was 14.2%. Females were significantly more (23.6%)affected than males (3.2%). The majority

Figure 4: The dead removed lice stages as appeared with naked eye.r.

of the studies had showed identical results. A study in Kirkuk, indicated a prevalence rate of 20%. And girls were infested more (27.29%)than boys (0%) [3]. An overall rate of infestation was 13.5% in a Baghdad, and the incidence was significantly higher in girls (17.33%)than in boys (8.75%)[6]. In similar study in Jeddah, Saudi, among 5150 school girls examined, HL infection rate was 11.26% [7]. Also HL infestation rate was 23.32%, the infestation rate was higher in girls (47.12%)than in boys (0%). [8]. A report of Centers for Disease Control (CDC) indicated that 6 to 12 million people each year will experience head lice infection in the United States. And that girls have an increased risk rate of infection than boys [14]. Despite the huge development in human life styles and high educational levels of populations, HL is still prevalent. This may be in one hand due that, most of parents are employers or workers and spend a lot of times outdoors. On the other hand, they may be careless and not periodically checking their children heads. In addition to that the direct contact among children in crowded areas as schools and day cars may amplify the chance of transmission. However was the prevalence of HL, females always have head lice infection more than males [3,6,8]. This may possibly because, the long hair in females offers good humidity and breeding environment for the insect. A confirmatory datum from a study indicated that, length of the hair is of a very high significant effect on the infection rate. The study found highest rate (22.2%)in long haired, compared to 10.7% in medium and 0% in short haired girls [6]. In current study the most age group which was significantly more infected in both females and males was 7-8 and 9-10 years old, with a rate of 41, 32.8% for female and a rate of 13,7% for males in each age groups respectively. A study found that the age group 3-10 years were most infested with head lice [14]. A lowest infestation rate (12.62%) was found in12 years old while the highest (30.13%)

was in 8 years old [8]. High incidence (6.01-7.61%)of head lice was delimited in age 6-9 years [3]. A highest rate (18.7%)of infection was recorded in age group of >8-10 years [6]. This age limitation is probably because of that, the child in this age start to have a type of dependency for bathing and taking care of their cleaning. In such age they may not be able to clean their heads or bodies in a proper way. And this is why the infection rates is lower in smaller or bigger ages in all communities [3,6,14,15]. Furthermore, in this age the contact between children will be more, because of playing groups and attending daycares or schools. In the present study, continuous infection significantly was more (65%)among girls than in boys. The long of the hair had significantly related with the infestation rate of infection. The highest (22.2%)was for long haired compared to 10.7% for medium and 0% for short haired [6]. This result may be due that, boys in school age in our community will not allowed to have long hair cut. This will aid in noticing the infection easily, and treating it quickly by shaving the hair as done in this area. Also in the current investigation, HL infection had well related with head or body itching. A rate of 44.9% of infected persons had suffered from head itching and a rate of 15.3% had suffered from both head and body itching, while 40 % of them had no head or body itching. Similarly, other studies indicated that head itching occur in some persons while some others never experience it [9,16,17]. The itching sensation in sensitized person is most likely due to, the saliva secreted form the insect before feeding process or due to the insect soil or other substances. These substances can led to head or localized irritation and erythema. Moreover, however head lice cannot transmit diseases to human but it can cause scalp pyoderma resulted from mechanically transmitted Streptococci and Staphylococci found on the skin, scratching may also cause impetigo or other skin infection, which can resulted in local adenopathy [15,18]. Delayed itching sensation to 4-6 weeks post infection may miss early identification and delay treatment process [19,20]. In contrast to the results of most of the studies, that showed significant effect of occupation and educational status [6,7]. In current study, the occupation and educational status of the parents were significantly not associated with the frequency of the infection. All families with different levels of economic status had infested child, but the greatest percentage was for intermediate level with rate of 66.5%, followed by bad level with rate of 20.4%. The lowest rate was among families of good level with rate of 14.35%. Most of the studies were agreed with this result [3,6,7]. This may because poorer families or families with low outcome may not have good life style. The effect of the natural plant oils in this study were intensified when used with head cap, especially the olive and the anise oils. Anti-lice shampoos were effected without head cap. The anise, ylang-ylang, coconut oils, and isopropyl alcohol were effective at least as permethrin when used in Frankowski et al. (2010)study [9]Several other products have been studied such as andiroba oil, quassia vinegar, melaleuca oil [tea tree oil], lavender oil)[2]. Some home remedies were used in a study, like vinegar, isopropyl alcohol, olive oil, mayonnaise, melted butter and petroleum jelly, tub margarine and thick hair gel. None of these substances were effective in eradicating head lice comparing to insecticides [6,7,15]. In Israel some other natural products (coconut oil, anise oil and ylang ylang oil)were as efficacious as the pediculicide. But a vast number of investigators were focused on using of chemical products (insecticide)for head lice treatment [2,8,9,15,16]. The effect of the chemicals is because of their toxic effect on the lice that may affect human health too. The petrol is very good and effective but its toxic and flammable. The natural oils action may be because of their ability to exclude air openings in the insect, which led to anoxia and death. But they need repainting washing with a liberal amount of ordinary shampoos to remove them from the hair. In addition to that their toxic effect are not approved yet. Treating infected head with the ordinary hair shampoos with a plastic hair cap is the most recommended, its safe, available and not toxic or flammable.

CONCLUSION

The petrol is very good and effective but its toxic and flammable. The natural oils action may be because of their ability to exclude air openings in the insect, which led to anoxia and death. But they need repainting washing with a liberal amount of ordinary shampoos to remove them from the hair. In addition to that their toxic effect are not approved yet. Treating infected head with the ordinary hair shampoos with a plastic hair cap is the most recommended, its safe, available and not toxic or flammable.

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Our Dermatology Online

Brief Report

Cryosurgery in a dermatology setup: a hospital based study

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ABSTRACT

Background: Cryosurgery is the well-aimed and controlled destruction of diseased tissue by application of cold. It has been shown to be effective and efficient in various skin diseases. We studied the indications and outcome of this modality of treatment in our set up. **Materials and Method:** All patients who were subjected to cryosurgery for different clinical indications were included in the study. **Results:** There were total of 133 patients. Commonest age group presentation (between 16 to 25 years) constituted about 42%. 52% of patients were students. There were total of 91% who were from Kathmandu remaining 9% were from outside of Kathmandu Valley. Cryosurgery was indicated commonly in warts for 82% patients followed by 12% for keloid & hypertrophic scar, benign conditions in 4% and in premalignant conditions for 2% patients. Number to treatment (or sessions) for indicated disease with resolution of disease was single in 81% patients followed by between 2 to 5 treatment session in 14% and more than 5 treatments was necessary in 5%. Side effects like pain was seen in 64% patients, erythema in 8% and blistering 4%. 24% patients did not have any side effect. **Conclusion:** Cryotherapy is considered as one of the effective modality of treatment for various skin conditions with various limitations. Further prospective clinical trial with large population group should be carried out.

Key words: Cryosurgery; Liquid nitrogen; Keloid; Warts

INTRODUCTION

Cryosurgery is a procedure in which there is controlled destruction of diseased tissue by application of cold. It provides an effective and efficient treatment option for various skin diseases like infections, benign skin growth, premalignant condition and some malignant skin tumors and provides high cure rates and good cosmetic results with a few contra-indications and low incidence of complication [1]. The biological changes that occur during and after cryosurgery is caused by tissue injury which is induced by cell freezing and by the vascular stasis followed by cryo-reaction, the immunological phase. There are no study regarding use of cryosurgery in dermatology done in the literature of Nepal. So with this study we want to know the clinical indications and outcome of the procedure.

MATERIALS AND METHOD

All patients who visited Dermatology outpatient department of Nepal Medical College and Teaching hospital and were subjected to cryosurgery for different clinical indications were included in the study. Details of patients who had undergone cryotherapy, their demographic profile including, name, Hospital reference number, age, sex, address, occupation, marital status, indication for treatment, number of treatment, side effects were obtained from the data maintained in the dermatology department outpatient record from fiscal year June 2016-June 2017. Patients having other systemic illness, Pregnancy or lactating were excluded from the study. Statistical study using SPSS version 16 with descriptive analysis was carried out.

RESULT

There were total of 133 patients who underwent cryosurgery during the one year study period. 42% patients were between 16 to 25 years, followed by 27% patients between 26 to 35 years, 25% less than 15 years and 6% more than 36 years. Cryosurgery was indicated mostly in students (52%) followed by 24%

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who were housewife, in 22% who were employed and in 2% unemployed. There were total of 91% who were from Kathmandu, out of which 68% from south of Kathmandu, 13% from north of Kathmandu, 6% from west of Kathmandu and 4% east of Kathmandu, the remaining 9% were from outside of Kathmandu Valley.

Cryosurgery was indicated commonly in warts 82% out of which plantar warts were in 36%, verruca vulgaris 34%, palmar 14%, genital 13% and periungual 3% patients. Second common indication was in keloid, hypertrophic scar and post burn scar, which together constituted 12% followed by in benign conditions like acne cyst, corn, De Morgan spots, Seborrheic keratoses, linear porokeratosis in 4% and in premalignant conditions like Bowes disease in 2%. The duration of the disease was less than one month in 46%, between 6 month to 1 year in 33% and more than one year in 21% patients. Number of sessions required to treat the indicated disease with resolution of disease was single in 81% patients followed by between 2 to 5 treatment sessions in 14% and more than 5 sessions was necessary in 5%. Side effects like pain was seen in 64% of the patients, erythema in 8% and blistering in 4 % patients. There was no side effects seen in 24%.

DISCUSSION

Cryosurgery is a well-aimed and controlled destruction of diseased tissue by application of cold. It has been shown to be effective and efficient in various skin diseases, provides high cure rates and good cosmetic results with a few contra-indications and low incidence of complications. The biological changes that occur during cryosurgery are tissue injury in which intracellular ice formation damages mitochondria and endoplasmic reticulum, leads to an irreversible cell destruction known as homogenous nucleation, followed by heterogenous nucleation, vascular stasis and tissue anoxemia resulting in ischemic necrosis [2].

There are still only few studies on the immunological cutaneous response to cryosurgery. Langerhans cell activity was found to be increased in mouse skin after treatment. In keloid cryosurgery leads to tenascin expression while IFN-g expression was depleted [3,4]. Cryosurgery is considered the treatment of choice in hypertrophic scars, keloids, infantile hemangioma, isolated actinic keratoses and granuloma annulare and an alternative therapy for various conditions like warts, superficial basal cell carcinoma and Kaposi sarcoma. There is increasing trend for use of cryosurgery in cutaneous malignancy, premalignant conditions and benign neoplastic diseases [5].

In a study by Menedez et al. total cure rate was seen in 87.7% with various cutaneous indications. In cutaneous warts cryotherapy was found to be useful in 50%-70%. In our study we found effective in 82%. Stefanaki et al found 67.3% improvement in children treated with cryotherapy [6-7]. A comparative study with immunotherapy and cryotherapy, Kozeimeh et al found immunotherapy more effective then cryotherapy [8]. Punch reduction prior to cryotherapy in patients with viral warts showed decrease the number and cost of treatment by reducing the size of warts and inducing local inflammation to accelerate resolution [9].

In a questionnaire based study cryotherapy was the first choice of treatment for 73% responders in palmar warts, 49% in plantar warts and 72% in warts on other location [10]. In our study we found cryotherapy being used for plantar warts (36%), verruca vulgaris (34%), palmar (14%), genital (13%) and periungual (3%).

Meymandi et al in a comparative study found that cryotherapy as well as intense pulse light laser were equally effective in treatment of hypertrophic scar and keloid [11]. Cryotherapy in combination with intralesional steroid has been the most popular classic treatment for both keloid and hypertrophic scars. It also helpful to heal small scars like acne scars. Moreover, the common side effect of cryotherapy is permanent hypopigmentation. Although it is used in multiple sessions every month, the least number of sessions the better chance for post-operative healing. Further, the success rates after 2 sessions have ranged from 30 to 75% either by using spray or contact cryosurgery with liquid nitrogen, and it is lower in keloid than in hypertrophic scars [12-13]. In a clinical trial, 79.5% of patients responded very well with a volume reduction of the initial mass of greater than 80% after a median of 3 treatments (range 1-9). In our study it was indicated in 12% patients. A good result was obtained in 14.5% of lesions, while results were unsatisfactory in 6% of cases. The main adverse effects reported were atrophic depressed scars and residual hypo pigmentation 75% of cases. No recurrences arose during the follow-up period (12-72 months) as per Russian et al. [14]. In our study we found side effects like pain in 64% of the patients, erythema in 8% and blistering in 4 % patients. There were no side effects in 24%.

Cryotherapy also has been tried and found to be useful in parasitic infection like leishmaniasis. In a randomized clinical trial, Farajzadeh found that cryotherapy in combination with oral terbinafine was less effective then cryotherapy with systemic Meglumine Antimoniate. In a meta-analysis, per lesion efficacy of 67.3% and 67.7% were reported for cryotherapy and pentavalent antimonials respectively [15]. In a case series for multiple recurrent Actinic keratosis sequential treatment of diclofenac 3% gel with cryotherapy was found to be an effective treatment [16].

In a multricentric intralesional randomized trial cryotherapy was found to have higher efficacy than topical methyl aminolaevulinate- photodynamic therapy by Kaufmann et al. [17]. A case series of loco-regional cutaneous metastasis of melanoma combination therapy of cryosurgery and imiquimod was found to be successful [18]. Cryotherapy as a useful procedure used for other dermatological indications in case series and case reports mentioned in the literature are Xanthelasma palpebrarum, dermatofibrosarcoma protuberans, Refractory cutaneous Rosa Dorfman disease, cutaneous sporotrichosis, and also in oral lesions in children [19-20].

In our study the other indications were benign conditions like acne cyst, corn, De Morgan spots, Seborrheic keratoses, linear porokeratosis was seen in 4% and in premalignant conditions like Bowes disease in 2% patients.

There are various limitations in our study, being retrospective we couldn't assess the complete clearance of the disease, follow-up of the patients and recurrence rates.

CONCLUSION

Cryotherapy is one of the effective modality of treatment for various skin disease ranging from warts, hypertrophic scars, benign skin conditions to premalignant conditions. Further prospective clinical trial with large population group with long term follow-up can help us identify cure rates with number of treatment session required for remission and recurrence rates.

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Our Dermatology Online

Brief Report

The way to cure a complicate syndrome caused by an insect (parasite) using a cerumen extracted from another insect (no parasite, that lives in some countries where the first disease is endemic)

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ABSTRACT

Scope of our research is to demonstrate how a neglected tropical desease (that in several countries is endemic) and that is very dangerous and perilous and is caused by an insect (a parasite called Tungia penetrans) may be treated using a wax another insect (no parasite, but that lives in one of these countries where the disease is endemic) produces. The importance of the fact resides only in the tempestivity of discovering (by the usage of a simplest lens) yhe moment when the insect penetrates into skin (generally toes or sole of kids under 10 years old). After one week, the stages of incubation of the Yungia does not permit a resolution. This wax could be proposed as a preventive agent with regards to the parasite itself.

Key words: Tungia penetrans; Tungiasis; KMnO4; Dimethicone; Chinese wax

INTRODUCTION

Tungiasis (sand flea disease) is a neglected tropical disease, prevalent in resource-poor communities in South America and sub-Saharan Africa. It is caused by an inflammatory response against the pinch caused by the penetration in mild wounds or underneath nails (especially of feet, in co-presence of onychomycosis too) of the female sand fleas (Tunga penetrans). Although associated with debilitating acute and chronic morbidity, there is no proven effective drug treatment. By consequence patients attempt to remove embedded sand fleas with non-sterile sharp instruments, such as safety pins, a procedure that represents a health threat by itself [1].

Common remedies are to be considered:

Application of topical anti-parasitic medications such as ivermectin, metrifonate, and thiabendazole
Locally freezing the lesion using liquid nitrogen (cryotherapy).

Even if criotherapy may be used only till the second Fortaleza's stage (see below), it is indisputable that sand flea cannot tolerate freezing and for this, criotherapy should be preferred to other medicaments.

The chief aims of this research is to show that chriotherapy must be effectuated only when it is the just moment to do it (we will explaiin that this period might not exceed 14 days).

It has been referred that tungiasis is present worldwide in 88 countries with varying degrees of incidence [2].

A governmental action in Kenya forecasts the application of a diluted solution of KMnO4 onto the feet of pupils at rural primary schools.

This disease is of special public health concern in highly endemic areas such as Nigeria, Trinidad and Tobago, and Brazil, where its prevalence, especially in poor communities,even if the disease has not to

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be considered endemic, has been known to approach 50% [3].

A topical application of a mixture of two dimeticones of low viscosity (NYDA) onto one foot of 47 school children in Kenya [1] has been experimented and a comparison was made with the application of a 0.05% solution of KMnO4 onto the other foot of the same 47 school children. The efficacy of the treatment was assessed during a follow up period of seven days using viability signs of the embedded parasites, and must be driven just when the children noticed that they have been pinched and adverted slight pain and erythema around the spot of the penetration of the parasite and not after two weeks when alterations in the natural development of lesion morphology and the degree of local inflammation outcome more evidently. Seven days after treatment, in the dimeticone group 78% (95% CI 67–86%) of the parasites had lost all signs of viability as compared to 39% (95% CI 28–52%) in the KMnO4 group ($p<0.001$). In the dimeticone group 90% (95% CI 80–95%) of the penetrated sand fleas showed an abnormal development already after 5 days, compared to 53% (95% CI 40–66%; $p<0.001$) in the KMnO4 group. Seven days after treatment, signs of local skin inflammation had significantly decreased in the dimeticone group ($p<0.001$). The AA asserted that the topical application of dimeticones of low viscosity (NYDA) was an effective means to kill embedded sand fleas, in view (they herald) of the efficacy and safety of the topical treatment with dimeticone, the mechanical extraction of embedded sand fleas using hazardous instruments is no longer warranted.

The symptoms of this disease include:
- Severe pruritus
- Pain
- Inflammation and swelling
- Lesions and ulcerations, with black dots in the center.

Left untreated, secondary infections, such as bacteremia, tetanus, and gangrene, can occur.

In all cases, tungiasis by itself only caused morbidity, though secondary infection may lead to mortality.

A person assaulted by a single flea may present immediately when, though the erythema is barely perceptible, a boring pain and the curious sensation of pleasant itching occurs. This inflammatory reaction is the initial immunological response to the infestation and is sudden.

It must be stressed that meanwhile Tunga males are still mobile after a blood meal like all other fleas, the female flea burrows head-first into the host's skin, leaving the caudal tip of its abdomen visible through an orifice in a skin lesion. This orifice allows the flea to breathe, defecate, mate and expel eggs while feeding from blood vessels. It lives in the cutaneous and subcutaneous dermal layer.

It is suggestive to remember that the male flea dies after copulation.

Fortaleza identified 6 stages or steps of the infection evoked by the penetration of Tunga p., and we can herald that Tungiasis may be cured only before the passage from II to III Fortaleza's step.

The first step of Tungiasis is the Tunga penetration even if too often it is not noticeable by the host, who can suppose the slight pain is due to the pinch of whichever else (mosquitoes or other fleas).

The very first stage of the first step lasts two days and a treatment with dimethicone or any other insulating grease or wax must be begun at this first step, in order to massacre definitively the flea.

Anyway caudal tips and abdomen can be observed thanks a lens and if the person who has been pinched by the Tunga notices this "penetration" subitaneously, can immediately spread over the skin, where the flea is penetrated, paraffinic waxes, avoiding the complete life cycle of the flea.

The complete lifecycle of Tunga p. is presented as follows.

Heavily infested patients may not notice a stage 2 infection due to the other fleas' causing irritation as well. Feces may be seen, but this is more common at the 3rd stage,(at the second week of incubation) when it is too late for healing. Around the third day after penetration, erythema and skin tenderness are felt, accompanied by pruritus (severe itching) and a black furuncular nodule surrounded by a white halo of stretched skin caused by the expansion of the flea.For this matter it is better to begin to apply parafinic waxes right the first or the second day after the aggression of Tunga p., in order to suffocate the parasite, since fecal coils may protrude from the center of the nodule where the flea's anus is facing upward. They should be washed off quickly as the feces may remain in the skin

unless removed.If neglected, during this 3a substage, pain can be severe, especially at night or, if the nodule is on the foot, while walking. Eggs will also begin to be released and a watery secretion can be observed. The radical metamorphosis during the 3rd to 6th day after penetration, or neosomy, precedes the formation of a small caldera-like rim rampart as a result of the increased thickness of the flea's chitinexoskeleton. During the caldera formation, the nodule shrinks a bit and it looks as if it is beginning to dry out; this takes 2 weeks and comprises stage 3.

At the third week after penetration and stage 4, the eggs' release will have stopped and the lesion will become smaller and more wrinkled. As the flea is near death, fecal and water secretion will stop altogether. Pain, tenderness, and skin inflammation will still be present. Around the 25th day after penetration, the lesion looks like a black crust and the flea's carcass is removed by host repair mechanisms and the skin begins to heal. With the flea gone, inflammation may still persist for a long time.

Anyway caudal tips and abdomen can be observed thanks a lens and if the person who has been pinched by the Tunga notices this "penetration" subitaneously, and thus can immediately spread onto the spot of the skin, where the flea is penetrated, paraffinic waxes, avoiding the flea drives her complete life cycle and disease grows perilous and perhas letal.

After the first applications (three pro day) the treatment must last almost 7 days.

It is possible to behold the lack of faeces or eggs owing to the lens after the 4th day of applications.

The flea is completely exterminated.

We have attempted to employ the same wax even as preventive agent, especially in some parts of the child's body that can be assaulted by Tungia p.

For instance, we prayed the kid to spread the wax onto a little wound onto hand and between one finger and another and convinced him to play with sand in the foreshore close to a dirty river.

MATERIALS AND METHODS

We have recruited a Brazilian child (7 y.old) who once upon a time was pinched in his right foot by a Tungia penetrans and for the fact he did not inform his parents of the disadventure, underwent to a drastic cure by metronidazole and amoxicillin (with all the carreau of risky side effects that buid up a perennial morbidity), after 2 months from the assault of the sand flea and the complete Calvary due to the syndrome of intections.

This volunteer uses to play with sand (as all childrens do) along the a river on the sand.

It is wellknown that Tungia p. likes lukewarm temperatures (till 30°C) and humid environmental conditions.

We have scrutinised the paper Thielecke and Nordin (1) published in 2014 and attempted to discover if it is possible to treat Tungiasis throuhout all the lifecycle of the insect by occlusive paraffines, like dimethicone, for the sake of the diachysis of all paraffines that resemble human sebum (the AA refer that the employ of the mix of viscous dimethicone is advisable during the passage from II and II Fortaleza's stage) and we can assert that it is utterly true and a complete tretament throughout all the 6 Fortaleza's stages is not possible at all, nevertheless we have revealed that if the treatment is made using an occlusive wax (chinese wax) that is a fatty, solid substance, produced by bees from Far East, and employed by them in the construction of their comb, results can be amazing. This cerumen is first excreted, from a row of pouches along their sides, in the form of scales, which, being masticated and mixed with saliva, become whitened and tenacious. Its natural color is pale or dull yellow.

Since the melting point of chinese wax is quite elevated we have prepared an emulsion with macadamia nut oil, in order to render it spreadable more easily.

It is suggestive to recall that chinese wax is produced by an insect, the coccus sinensis, and so this typology of healing could reflect the Dantesque retaliation.

The malaise evoked by an insect (a parasite) can be cured with a secrete obtained from another insect.

The applications began the very first day the child noticed a slight pain and itch in his left hallux (he has no more the right big toe) and were repeated three times pro day.

We used a lens the very first day noticing that the abdomen of the Tungia was mobile and flapping alla round the point of penetration of the parasite.

RESULTS

Effectively for the first two days the entire caudal tip and abdomen were well visible under the lens.

After the third day, we observed only a little reddish spot, where the flea was penetrated and the child asserted he felt a little itch but not pain.

At fourth day, even pruritus disappeared and after the 7th day the skin was clear as no insect had never penetrate the cutis.

The childat the end of our study is safe and shows any symptom of infection nor itching or pain.

DISCUSSIONS AND CONCLUSIONS

For the fact that Tungia p. hits children under 10 years and they do not possess the ability to notice that something odd or wrong is happening in their organism, and even that too often they had played in certain places or zones prohibited by their parents, and in these places there is a real chance to encounter the sand fleas, this situation complicates the cursus of eventual operations of diagnose and a prompt and early intervent.

Praffinic waxes and occlusive jellies and especially chinese wax find a notable interest if applied at the very beginning of the Tunga penetration until the beginning of the third Fortaleza's step.

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Our Dermatology Online

Case Report

The first reported case of a variant of Mal de Maleda of the Gamborg-Nielsen type in an Egyptian origin patient

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ABSTRACT

Mal de Meleda is a rare genodermatosis with an autosomal recessive inheritance. Mutations in the SLURP1 gene are the cause of this disease. Clinically, it is characterized by progressive palmoplantar hyperkeratosis exhibiting a transgradiens pattern extending to the dorsal aspects of the hands and feet in a glove and stocking pattern. It is also associated with hyperhidrosis, nail changes, subungual hyperkeratosis and perioral erythema. Here we report the first case of Gamborg-Nielsen variant of Mal de Meleda disorder in a patient of an Egyptian origin.

Key words: Mal de Meleda; Keratoderma; Genodermatosis; SLURP1 gene

INTRODUCTION

Mal de Meleda is a type of palmoplantar keratoderma (PPK) which has an autosomal recessive (AR) pattern of inheritance. Clinically, the patient present with bilateral diffuse PPK in a stocking and glove pattern with sharp margins and a yellow tone complicated by hyperhidrosis and microbial infection leading to malodorous and painful lesions [1]. Nail anomalies are the most common associated feature in the form of subungual hyperkeratosis, Beau's lines, onycholysis, hyperconvexity, and koilonychias [1,2]. Other associated features such as psoriasiform lesions on the knees and elbows, perioral erythema, angular cheilitis, arched palate and rarely malignant melanoma within the areas of hyperkeratosis may be seen [3-7]. Mal de Meleda can be diagnosed based on clinical features, family history and genetic analysis. Here we report the first case of Gamborg-Nielsen variant of Mal de Meleda in an Egyptian origin patient in the English medical literature.

CASE REPORT

A 13-year-old Egyptian boy presented to the dermatology clinic of King Fahd Hospital of the University with erythematous patches affecting the palms and soles. The condition started at the age of 3 months when he developed palmoplantar erythematous erosive lesions with fluctuating course. There was no history of blisters or fissures. That time he was seen by several dermatologists and treated with oral antihistamines and topical steroids. According to the parents the condition worsened and became palmoplantar macerated hyperkeratotic lesions. Past medical history was irrelevant. There was no history of atopy. The mother and father are first degree relatives with no similar condition reported in the family. The

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patient had normal growth milestones and doing well in school. On examination there was bilateral symmetrical palmoplantar waxy hyperkeratotic lesions with irregular borders extending to the dorsum of both hands and feet with erythematous background (Figs. 1 and 2). There was hyperhidrosis that was not malodorous. There were no nail changes or contractures or autoamputation of the digits. The face showed scaly slightly erythematous patches on the cheeks and nose. The lips were slightly dry without perioral erythema. His gingiva, teeth and tongue were normal. Hair was normal. Besides, there was erythematous hyperkeratotic patches on the extensor surfaces of the arms, legs, thighs and back but the knees and elbows were free of lesions. There were no hearing loss or eye abnormalities. Systemic examination showed normal findings. Laboratory blood tests including complete blood count, liver function test, renal function test, thyroid function tests, serum iron, total iron binding capacity, urinalysis and stool analysis were within normal range. But total IgE level was high (5000 IU/ml). ECG and echocardiogram were normal. Biopsy from the trangredient border of keratoderma revealed mild to moderate acanthosis, orthokeratosis, and prominent hypergranulosis without epidermolytic changes. Besides, there was mild perivascular lymphocytic infiltrate in the upper dermis, Interestingly, the presence of less severe hyperkeratosis with the transgredient pattern to the dorsum of the hands and feet, lack of nail involvement and lack of distant keratosis we think that our case is a variant of Mal de Meleda called hereditary PPK of the Gamborg-Nielsen type. Genetic analysis was our next step to detect the affected gene but due its unavailability we decided to postpone and follow the patient every 6 months as a case of Mal de Meleda. Currently, the patient is being treated with strong emollients.

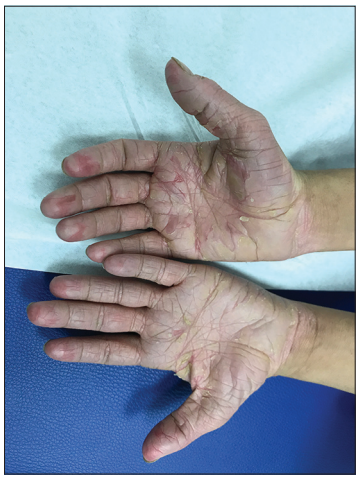

Figure 1: Bilateral thickened palmar keratoderma with mild peripheral erythema on the palmar surface.

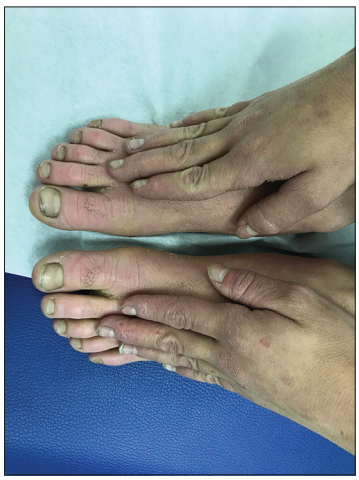

Figure 2: The transgrediens progression of hyperkeratoses to the dorsal surfaces of the hands and feet.

DISCUSSION

Mal de Meleda is a type of palmoplantar keratoderma (PPK) which has an autosomal recessive (AR) pattern of inheritance with an estimated prevalence of 1 case per 100,000 population [8]. The disease has been reported in many countries including Croatia, Algeria, Chile, China, Germany, India, Indonesia, Italy, Japan, Korea, Laos, Libya, The Netherlands, Pakistan, Saudi Arabia, Scotland, Sweden, Tunisia, Turkey, and the United Arab Emirates [3,9].

Clinically, the patient presents with bilateral diffuse PPK in a stocking and glove pattern with sharp margins and a yellow tone. Usually, symptoms begin soon after birth as palmoplantar erythema and progress to the characteristic thickened, exfoliative hyperkeratosis that has a transgrediens nature [2]. This can be complicated by hyperhidrosis and microbial infection leading to malodorous and painful lesions [1]. Nail anomalies are the most common associated feature in the form of subungual hyperkeratosis, Beau's lines, onycholysis, hyperconvexity, and koilonychias [1,2]. Digital disorders including fifth-finger dysplasia, tapering of the digits, contractures, knuckle pads and pseudoainhum may be present [2,4,10]. Other associated features such as psoriasiform lesions on the knees and elbows, perioral erythema, angular cheilitis, arched palate and rarely malignant melanoma within the areas of hyperkeratosis may be seen [3-7]. Histologically, there is hyperkeratosis, parakeratosis, orthokeratosis,

hypergranulosis and acanthosis without evidence of epidermolysis [11-14]. Mal de Meleda can be diagnosed based on clinical features, family history and genetic analysis. Fischer et al, identified the gene that encodes the secreted LY6/urokinasetype plasminogen activator receptor (uPAR)-related protein-1 (SLURP-1) to be the cause [15]. Originally, there were three different gene mutations of SLURP-1 a homozygous single nucleotide deletion, a homozygous point mutation and a homozygous splice site mutation, However, 14 other mutations have been identified in 19 other countries later [10,16,17]. Pathophysiologically, when the SLURP-1 protein is non-functional, hyperkeratosis results due to improperly regulated keratinocyte apoptosis and the macrophage and keratinocyte release of tumor necrosis factor (TNF)-a is inhibited producing generalized inflammation [18,19]. There are presently over 25 identified hereditary PPK diseases that can be separated into five basic categories: (1) diffuse PPK; (2) diffuse mutilating PPK; (3) focal PPK; (4) ectodermal dysplasia with PPK and (5) syndromic PPK [20]. Specifically, in hereditary PPK of the Gamborg-Nielsen type which was reported in the Swedish population and is referred to as a variant of Mal de Meleda with milder hyperkeratosis, lack of nail involvement, and no distant keratoses. But the transgredient pattern of hyperkeratosis to the dorsum of the hands and feet and knuckle pad formation are variably present [21]. Clinically, our case is similar to the Gamborg-Nielsen type. Though we could not explain the high IgE in the absence of atopy, it could be a new associated finding. Other related conditions include: Naxos syndrome and Carvajal syndrome which are both syndromic PPK diseases with congenital cardiomyopathy, Papillion-Lefevre syndrome (PLS) is another AR PPK typically with periodontitis and early tooth loss, Nagashima-type PPK is a non-progressive hyperkeratosis usually terminates at puberty, Greither's disease present later in childhood and Olmsted syndrome presents in early childhood with a mutilating-type PPK with a tendency to involve the perioral area [22,23]. Treatment include topical corticosteroid, lactic acid, retinoid acid, emollients, keratolytics with topical urea-based ointments and oral 13-cis retinoid acid [9]. Recently, Gruber et al, showed effective treatment with oral acitretin 20 mg/day plus topical antimicrobial and keratolytic therapy [14]. Surgical treatment of hyperkeratosis can be an option with excision and subsequent placement of a full thickness skin graft [5]. Genetic counselling is warranted in these families. The absence of symptoms in biological parents is most likely suggestive of either an AR inheritance pattern or a spontaneous mutation.

Our patient is the first reported case of Gamborg-Nielsen variant of Mal de Meleda in an Egyptian origin patient in the English medical literature. Genetic analysis is being undertaken to confirm the clinical diagnosis.

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Our Dermatology Online

Case Report

Werner's syndrome: A case report and review of literature

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ABSTRACT

Werner's Syndrome also known as Pangeria is an autosomal recessive disorder characterized by premature aging, increased risk of malignancies and atherosclerosis. The Global incidence rate is less than 1 in 100,000 live births. The incidence is higher in Japan and Sardinia affecting 1 in 20,000 – 40,000 live births and 1 in 50,000 live births respectively. Individuals with this disorder typically grow and develop normally until they reach puberty. Affected individual usually do not have a growth spurt, resulting in short stature. The characteristic aged appearance typically begins to develop when they are in their twenties and includes graying and loss of hair, a hoarse voice, and thin, hardened skin. They may also have a facial appearance described as "bird-like facies." Werner's Syndrome has been described as "Caricature of ageing" by Epstein et al. 1996. An OMIM number # has been assigned to Werner's Syndrome with phenotype MIM number 277700 (OMIM#277700).

Key words: Pangeria; Premature ageing syndrome; Bird like facies; OMIM277700

INTRODUCTION

Werner's Syndrome (WS) is an autosomal recessive disorder affecting the connective tissue of the whole body. It is also known as Progeria adultorum and Pangeria. Werner's Syndrome is considered one of the genomic instability syndromes. It is characterized by short stature, senile appearance, cataracts, joint contractures, early menopause, and premature arteriosclerosis, Scleroderma like features, premature canities, baldness, ulceration and increased risk of malignancy.

The highest incidence of WS reported is in Japanese patients (1000 of 1300 cases reported worldwide). We report a 33 year old male patient presenting to our outpatient department with history of keratosis over both the feet.

CASE REPORT

A 33 year old unmarried male presented with history of painful lesions over pressure bearing areas of both the feet since 5 years. Patient gives history of hoarseness of voice since 6 months. He had undergone surgery for cataract 10 years back. Patient had shown to a surgeon where he had advised a Doppler study for both the limbs one month back. The Doppler study showed early arteriosclerotic changes of both lower limbs. Venous system was normal.

Patient was born of consanguineous marriage. He has one female sibling affected with similar disease. On physical examination the skin over both the limbs were shiny, with mottled pigmentation couldn't pinched off (Fig. 1). Multiple hyperkeratotic lesions were seen over pressure bearing areas of both the limbs (Fig. 2). Contractures were seen over toes of both the feet but more on left feet. The trunk was normal. There was premature greying of eyelashes (Fig. 3) with receding scalp hair (Fig. 4). Biopsy was taken from the skin over left leg.

Histopathology showed sparse superficial perivascular and periappendageal lymphohistiocytes with occasional plasma cells. There is marked thickening of collagen

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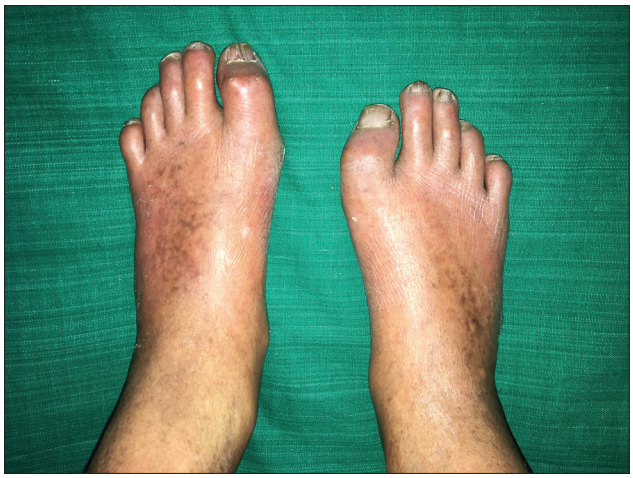

Figure 1: Scleroatrophy of skin on dorsum of both the feet.

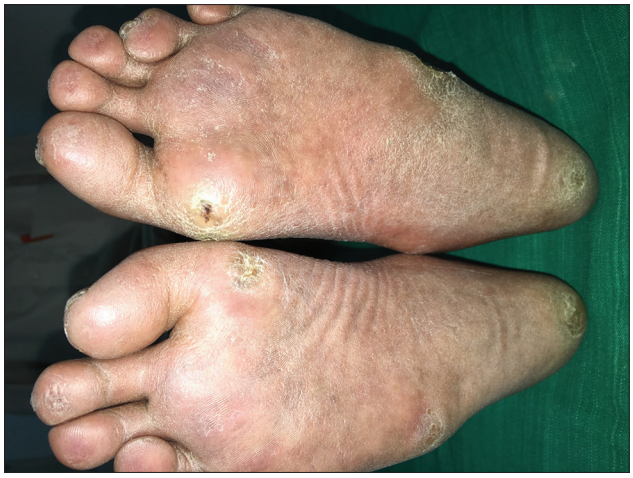

Figure 2: Keratoses at pressure points on both the feet.

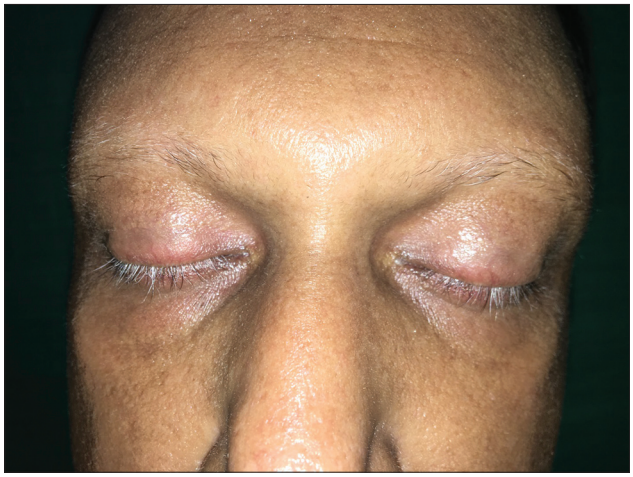

Figure 3: Premature greaying of eyelashes.

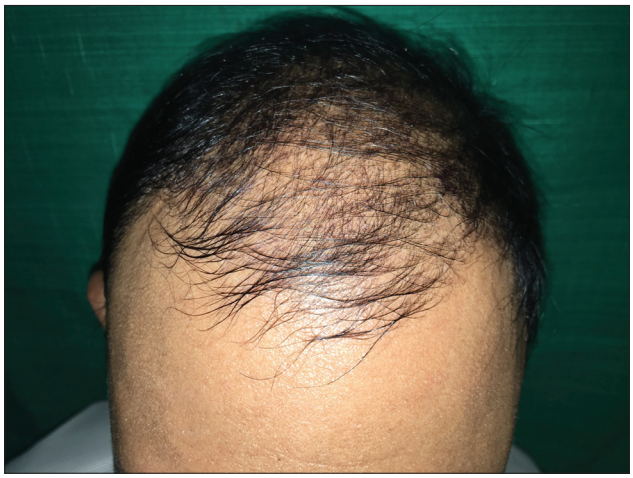

Figure 4: Recession of Scalp hair.

bundles in upper reticular and papillary dermis. The thickened bundles are closely packed so as to give hyalinised appearance. The upper dermis shows telangiectasia.

DISCUSSION

Werner's syndrome was first described by Otto Werner at 1904 [1]. He reported four cases of brothers and sisters, where he observed juvenile cataract, Pachyderm like alteration of the extremities, small stature, premature ageing of the face, juvenile grey hair and genital hypoplasia. In 1934, Oppenheimer and Kugel [2] described additional endocrine abnormalities such as osteoporosis and type 2 diabetes mellitus. The diagnostic criteria proposed for Werner syndrome by International registry for Werner syndrome 2000- 2005 given in Table 1.

Werner's syndrome is a genetic disorder transmitted in an autosomal recessive pattern that affects males and females in equal numbers. The disorder frequency has been estimated at 1 to 20 per one million individuals in the Unites States. In the Japanese population it is 1 per 20,000 to 1 per 40000.

Werner Syndrome is caused by null mutations of WRN gene located on the short arm of chromosome 8 (8p12- 11.2) [3]. More than 80 different mutation of WRN gene have been identified. WRN gene codes for a member of Rec Q family of DNA helicases [7]. The disease is associated with abnormal metabolism of connective tissue. There is excessive synthesis of collagen I and III which is dependent on increased mRNA levels. Fibroblasts exhibit genomic. Instability, increased sensitivity to DNA damaging agent, lengthened S phase, accelerated replicative senescence.

Chromosomal instability including dysfunction of telomere maintainance is more prominent in WS cell than in normal cell. The accumulation of DNA double

Table1: Diagnostic criteria for werner syndrome

Cardinal signs and symptoms (onset over 10 years old)
1. Cataracts (bilateral)
2. Characteristic dermatological pathology (tight skin, atrophic skin, pigmentary alterations, ulceration, hyperkeratosis, regional subcutaneous atrophy and characteristic facies (bird facies)
3. Short stature
4. Parental consanguinity (3rd cousin or greater) or affected family
5. Premature greying and or thinning of scalp hair
6. Positive 24 hour urinary hyaluronic acid test when available
Further signs and symptoms
1. Diabetes mellitus
2. Hypogonadism (secondary sexual underdevelopment, diminished fertility, testicular or ovarian atrophy)
3. Osteoporosis
4. Osteosclerosis of distal phalanges of fingers and / or toes (x-ray diagnosis)
5. Soft tissue calcification
6. Evidence of premature arteriosclerosis (e.g. history of myocardial infarction)
7. Mesenchymal neoplasms, rare neoplasms or multiple neoplasms
8. Voice changes (high pitched, squeaky or hoarse voice)
9. Flat feet
Definite: All the cardinal signs and two others
Probable: The first three cardinal signs and any two others
Possible: Either cataracts or dermatological alterations any four others.
Exclusion: Onset of signs and symptoms before adolescence (except stature, because current data on pre-adolescent growth patterns are inadequate)

strands breaks (DNBs) [3] at G1 phase including those at telomere foci is accelerated in WS cells even at a low senescence level. These results indicate that WS cells are prone to accumulate DSBs spontaneously due to defect of WRN gene which leads to increased chromosomal instability that could activate check points resulting in accelerated senescence. The accelerating ageing in WS is due to increased levels of inflammatory cytokines produced by senescent Cells. The sclerodermatous skin and blood vessel wall calcification contributes to the development of non-healing ulcers and keratosis of the limbs. Individuals with WS develop normally till puberty. The onset can be in the mid- term but can be seen even after the age of 30 years. Patient with WS show increased risk for the development of malignant lesions and malignancy is a frequent complication of WS. The frequency of malignant lesion reported ranges from 5.6% to 25%. For general population epithelial cancer has an incidence of 10 times that of mesenchymal sarcoma, but among patients with Werner Syndrome incidence is approximately equal [5]. An increased risk of thyroid cancer, malignant melanoma, osteosarcoma and soft tissue sarcoma is also noted. Defective homologous recombination is believed to be the primary reason for chromosome abnormalities and genomic instability thus causing greatly increased risk of cancer. The severity of WS is due to its several complications dominated by atherosclerosis. Patients may develop different forms of atherosclerosis specially that affecting the coronary arteries leading to myocardial infarction that is the first cause of death in WS [4]. Increase in the blood hyaluronic acid levels is responsible for sclerodermatous changes and cardiovascular abnormalities [6].

The clinical manifestation starts with greying at the temples as early as 14 to 18 years of age. The skin manifestations are usually noticed at 18 and 30 years of age. The sclerodermatous skin changes gives rise to thin spindle shaped limbs contrast with the normal trunk. The facial appearance shows beaking of nose with bird like facies, shallow orbits giving rise to proptosis. The joints become fixed; with scelordactyly and acral gangrene can be present. Keratosis over pressure points on the feet and ankle separate to leave indolent ulcers. The voice may be high pitched and hoarse because of thickening of vocal cords. Most patient are of short stature and hypogonadal, some may achieve normal stature and successful pregnancies. Diabetes is seen in atleast 30% of patients. Cataracts are seen at the age of 20 and 38 years and mostly of posterior and subcapsular cataracts [8]. The radiological changes show calcification of arteries, ligaments, tendons and subcutaneous tissue with osteoporosis of the legs.

The differential diagnosis include other premature aging syndromes like Progeria, Acrogeria, Rothmund-Thomson Syndrome, Cockayne syndrome, hypohidrotic ectodermal dysplasia and Huriez Syndrome. Progeria is a rare condition with onset as early as 2 years of age. Affected individual shows short stature, large bald head with prominent veins, prominent eyes with

bird like facies, coxa valga. Acrogeria is characterized by cutaneous atrophy and loss of subcutaneous fat particularly over distal extremities. Micrognathia, hollow cheeked and owl- eyed appearance. The affected individual has normal stature, normal life expectancy and scalp hair. Cockayne Syndrome is characterized by photosensitivity, Mickey Mouse facies, and normal scalp hair with disproportionately large extremities. In Rothmund Thomson syndrome the features are atrophy, telangeiectasia and mottled pigmentation most intense on sun exposes areas scalp hair is absent. Bilateral cataracts have developed between fourth and seventh year of life. Hypohidrotic ectodermal dysplasia is another differential where conical teeth, with reduced and absent sweating is seen. Huriez syndrome is an autosomal dominant transgrediant keratoderma characterized by keratoderma with scleroatrophy and nail changes.

Our patient had history of cataract surgery at the age of 20 years. The skin over the lower extremities was thin shiny with keratosis on pressure points. There was hoarseness of voice with proptosis of eyes and receding scalp hair. Doppler showed atherosclerotic changes of arteries of lower limb. These findings led us to make the diagnosis of premature aging syndrome. The histopathology showed thickening of collagen bundles with hyalinization just like sclerodermatous changes. Excluding all other premature aging syndrome considering his clinical features a diagnosis of Werner syndrome was made. Treatment given for the patient was mainly symptomatic. The patient has been referred to cardiologist and endocrinologist for further management.

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Our Dermatology Online

Case Report

The spectacular presentation of orf disease

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ABSTRACT

Orf is a viral zoonotic skin disorder which is also called ecthyma contagiosum. The disease primarily affects sheep and goats, however it can be transmitted to humans through close contact with infected animals. Orf presents with characteristic skin lesions and the diagnosis of orf is usually made based on its clinical features. Hereby, we present a 21-year-old female patient with orf disease with a characteristic clinical lesion on her hand.

Key words: Contagious; Ecthyma; Orf; Skin; Viral

INTRODUCTION

Orf is an infectious disease which spreads by direct contact with sheep and goats [1]. The etiopathogenic agent is Orf virus. It belongs to the Parapoxvirus genus of the Poxviridae family. Orf virus is usually transmitted through the breaks and abrasions on the skin. The disease is generally localised to the hands. Systemic involvement has not been previously reported [2]. Farmers, butchers, sheep shearers and veterinarians are prone to disease [3]. Orf has a characteristic clinical pattern including inflamed macules, papules, vesicles, nodules, verrucous lesions and scar tissue. The incubation period can vary from 3 to 5 days. The disease first appears as erythematous macules which evolve to papules with a target appearance in 7 to 14 days. Afterwards, the lesions become nodular and vesicular which ulcerate in 2-3 weeks. The lesions are usually asymptomatic, however secondary infections can lead to pain and discomfort. Orf disease usually heals by itself completely in 4-8 weeks [4].

CASE REPORT

A 21-year-old Caucasian female patient was admitted for further clinical evaluation of the lesion on her hand. The lesion first appeared as an erythematous macule and it increased in size gradually within the last 2 weeks. The lesion was asymptomatic. The patient whose father was a farmer was living in a rural area. The patient stated that she helped her family to raise sheep. The medical history was unremarkable. Dermatological examination revealed a grey-black colored plaque with erythematous-purple colored sharp borders and ulcerated-crusted area at the center on the third finger of her left hand (Fig. 1). The lesion had concentric zones and a targetoid appearance. We made the diagnosis of orf based on its clinical features and physical examination. The patient was started on topical mupirocin ointment to prevent secondary bacterial infections. The orf lesion healed completely after 6 weeks.

DISCUSSION

Orf is a self-limited zoonotic infection which can be transmitted to people by direct/indirect contact with infected animals and animal products [5]. Orf usually presents as a solitary lesion on the hand. However, multiple lesions have also been described in the medical literature [6]. The diagnosis of orf is usually made based on its clinical features and the exposure history of the patient [7]. In suspicious cases, a skin biopsy and polymerase chain reaction may be performed [8]. No specific therapy is necessary for orf. However,

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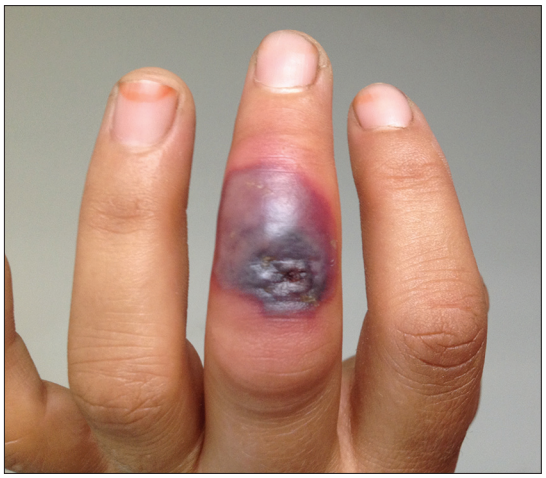

Figure 1: A targetoid plaque with erythematous-purple colored borders and grey-black colored, crusted center on the extansor surface of the third finger of the left hand.

the disease can easily be misdiagnosed as a bacterial infection or a neoplasm [5]. In immunocompromised patients, persistent lesions may require excision. It has been suggested that 1-week topical imiquimod therapy accelerated the healing of the lesions [9]. Geerinck et al. described a renal transplant patient with a growing orf lesion who was succesfully treated with topical cidofovir [10]. Orf has a benign course, however the lesions may reoccur in immunosuppressed patients [6]. Moreover, Duchateau et al. described autoinoculation with Orf virus in a patient who took azathioprine because of Crohn's disease [8]. Systemic symptoms do not occur, however, complications including fever, lymphangitis and lymphadenopathy have been reported. Furthermore, erythema multiforme and autoimmune bullous disease may be associated with orf [4].

CONCLUSION

Hereby, we present a case of orf disease with typical clinical course and appearance. Despite its rarity, orf disease should be included in the differential diagnosis of erythematous nodules, targetoid or crusted skin lesions of the hands.

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Our Dermatology Online

Case Report

A case of breast carcinoma presenting as carcinoma en cuirasse

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ABSTRACT

Carcinoma en cuirasse is a rare and unique form of cutaneous metastases, which surrounds the trunk in a firm and leathery fibrotic fashion, like an armor. We report a case of carcinoma en cuirasse in a 53-years-old female patient who was admitted to Dermatology outpatient clinic with pruritic, red, firm papules and plaques on the left breast and arm, extending to upper abdomen and medial part of right breast. At time of admission the patient did not have a history of an internal malignancy. Histopathological diagnosis of the incisional biopsy material from the plaque lesion was consistent with invasive ductal carcinoma. Imaging studies showed axillary, paraaortic, aortocaval lymph node involvement, pleural effusion, lung and liver metastases. The patient was referred to Medical Oncology Department and was treated with cyclophosphamide and adriamycine chemotherapy. This case of carcinoma en cuirasse preceeding the diagnosis of metastatic breast carcinoma and leading to diagnosis of malignancy is rare and important. Prognosis depends on the type of primary tumor and it's biological behavior, predictably, poor prognosis is expected in this case of metastatic invasive ductal carcinoma.

Key words: Cutaneous metastases; Carcinoma en cuirasse; Breast carcinoma

INTRODUCTION

Cutaneous metastases of internal malignities, constitute 2% of all skin tumors and is important in detecting undiagnosed malignancies and relapses of inadequately treated malignancies [1]. Carcinoma en cuirasse is a rare and unique form of cutaneous metastases, in which the cutaneous metastases cause lymphatic blockage and eventual thickening of chest wall skin, dermal and subcutaneous tissue fibrosis, resulting in armor-like wrapping of the trunk [2]. It generally occurs months to years after the diagnosis of primary cancer. However, rarely it may be the primary symptom of cancer [3].

CASE REPORT

A 53 –years-old female patient was admitted to Dermatology outpatient clinic with an erythematous rash on trunk and left breast. Patient's history revealed that she had suffered from hardening and shrinking of the left breast for 2 years and it was accompanied by a pruritic rash on trunk and edema in the left arm for the last 3 months. She didn't have any concomitant systemic diseases or use of any drugs.

Dermatological examination revealed distortion of the left areola, contraction and shrinking of the left breast, edema of the left arm and forearm. Dark red and purple colored, infiltrated, semi-firm, non-fading plaque and papules causing peau d'orange appearance was noticed on an area of approximately 50x35 cm on the left breast extending to the medial aspect of right breast, neck and abdomen (Figs. 1 and 2).

Routine blood tests, tumor markers, breast ultrasonography and mammography were performed. Overall report was BI-RADS-5, recommending tissue sampling from both breasts.

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A punch biopsy with the prediagnosis of carcinoma en cuirasse was performed from the lesions on the trunk. It was reported as carcinoma metastasis, tumor cells immunostained with pan CK. Approximately 20% stained with Estrogen Receptor (ER), 10% stained with Progesterone Receptor (PR). CerbB2 score was zero. The samples showed no staining with Mammoglobine and GCDFP15. These findings were consistent with breast cancer metastasis (Figs. 3-7). Incisional biopsy material from both breasts were consistent with invasive ductal carcinoma.

Bilateral enlarged axillary lymph nodes, pleural effusion and adjacent collapsed lung areas were detected in thorax and abdominal computed tomography. Multiple nodules were present in bilateral lungs. Multiple mass lesions showing contrast retention compatible with metastases were visualised in liver. Paraaortic and aortocaval lymph nodes were present.

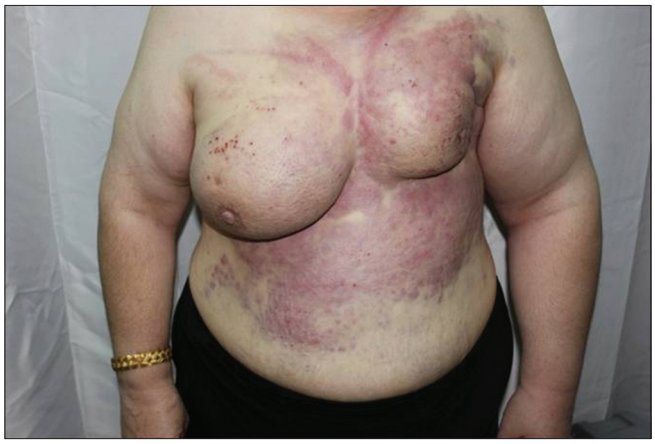

Figure 1: Infiltrated, plaque and papules causing peau d'orange appearance on the left breast extending to the medial aspect of right breast, neck and abdomen.

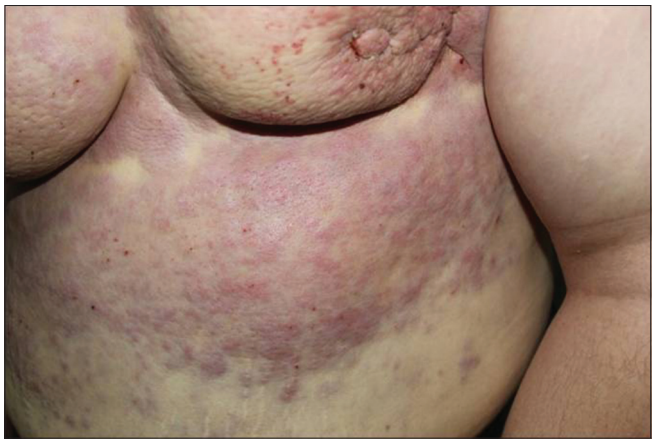

Figure 2: Close up image revealing infiltrated erythematous plaque and papules.

Patient was referred to Medical Oncology Department and chemotherapy with the diagosis of metastatic invasive ductal carcinoma was initiated.

DISCUSSION

Cutaneous metastases of internal malignities are relatively rare encountered conditions and of diagnostic importance for undiagnosed malignancies [3]. In female patients, most frequently breast cancer (69%), colon cancer (6%), melanoma (5%) and cervix cancers (2%) were accused of this condition [4].

Cutaneous breast cancer metastases can be seen in different morphological patterns such as papulonodular lesions, erysipeloid or sclerodermoid infiltrations [5]. Carcinoma en cuirasse is a special fibrotic form of cutaneous metastases, spreading rapidly and wrapping the trunk like a leather armor [6]. Lymphatic drainage

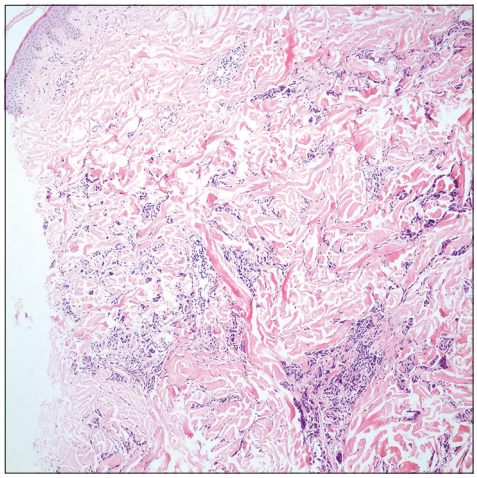

Figure 3: Infiltration of tumoral cells in dermis (HE x10).

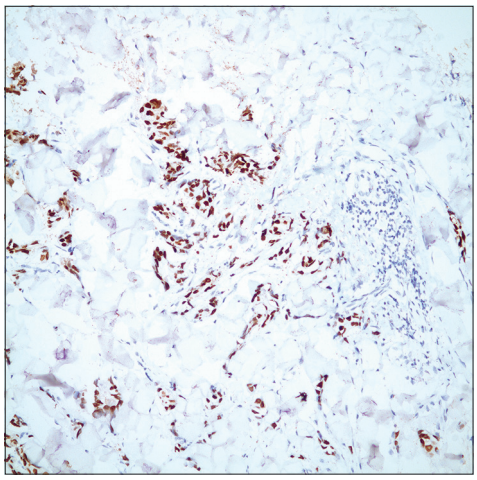

Figure 4: Tumour cells showing cytoplasmic staining with pan cytokeratin antibody (IHK x20).

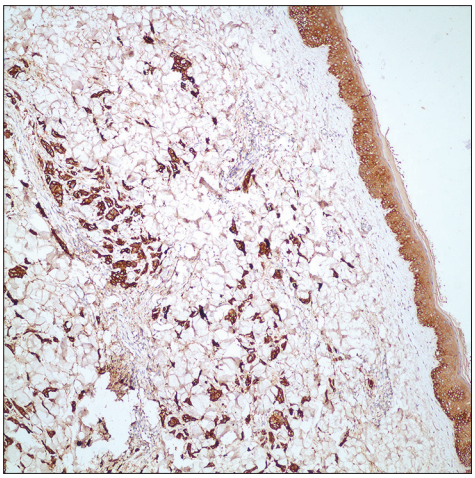

Figure 5: Tumour cells showing nuclear staining with estrogen receptor (IHK x20).

is disturbed, causing thickening of chest wall skin and edema, eventually resulting in fibrosis of the dermis and subcutaneous tissue [2]. This situation causes lymphedema, pitting of the skin and peau d'orange appearance [4,5]. Duration from cancer diagnosis to cutaneous metastasis may differ, however, generally it's seen in 3 years following the diagnosis [5]. Rarely, in some patients cutaneous metastases may be seen as the first and/or only symptom of the malignancies [3].

CONCLUSION

This patient is worth presenting due to carcinoma en cuirasse along with breast distortion and areola retraction being the preceding symptom of breast cancer. In this case, recognizing the cutaneous metastasis was important for initiation of the treatment and preventing further metastasis.

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

Our Dermatology Online

Case Report

Acute HTLV-1 leukemia/ lymphoma in a 33 year old grenadian migrant: A case report

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ABSTRACT

We present a case of an acute HTLV-1 leukemia in a 33-year-old Grenadian immigrant. Her diffuse skin nodules, and her coming from an area endemic for HTLV-1, namely the Caribbean made the clinical diagnosis of HTLV-1 leukemia/ lymphoma more feasible. Her disease was rapidly progressive, she having survived for one month, and fits into the acute classification of ATLL as proposed by Shimyoyama M. Immigrants from areas endemic for HTLV-1 who presents with skin rashes should be investigated for HTLV-1 neoplasms.

Key words: HTLV-1 leukemia/lymphoma; Skin manifestation; Endemic areas; Migrants

INTRODUCTION

HTLV-1 is a retrovirus which is associated with HAM/TSP [1-3] and which causes leukemia/ lymphoma [4].

The neoplastic process presents in the early stages with a skin lesion as the primary site [5,6] and progresses to the advance stages with multiple organs involvement of the reticuloendothelial system such as the liver, spleen, and, bone marrow. There are indeed other presentations as in the case in discussion that presented as an acute leukemia with diffuse skin involvement.

CASE REPORT

A 33-year-old Grenadian national was on a six months visit to Trinidad when she developed a generalized pruritic skin rash and abdominal pain following consumption of shrimps. She visited the General Hospital for the above complain three days following the above symptoms.

On examination she was found to be icteric and with pale mucous membrane. Her pulse was 80/min, blood pressure 90/60 mm Hg, respiratory rate 15/min and temperature 36.5°C.

She was not in discomfort. She had generalized nodules distributed on the upper and lower limbs, chest and face (Figs. 1 and 2). They were non-erythematous, and non-tender. No cervical, axillary or inguinal lymph nodes were palpable.

She was mildly tender in the left hypochondrium, with a markedly palpable spleen.

Her genitourinary and central nervous systems examination were essentially normal. She was started on analgesic and antipruritic oral medications and was admitted to the medical ward for further investigation.

Laboratory investigations revealed an Hb of 7.5 g/dl (11.7-15.5), hematocrit of 27.1%. (9 34.5-46.30), MCHC 27.6 g/dl (342.5-35.2), MCV 83 fl (80-40-95.5) MCH 22.9 pg. (27.2-33.5), WBC 131.6 10^3/ UL (4.10-11.20), with 83.6% Lymphocytes. (18.8-50.8), 13.3% (39.9-73.9) Neutrophils, 7.9% (0.3-1.8) Basophils, 0.1% (0.9-6.0) Eosinophils, 2.9% (0.3-0.9) Monocytes, platelets 263 x 10^3/UL. (159-388.0). There were polylobated lymphocytes, " flower cell "in the peripheral blood smear, but no hairy cells or blasts. She was hypercalcaemic, calcium 13.1 mg/dl, Na+ 127 mmol/L., K+ 4.8 mmol/L.,

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Figure 1: Skin nodules in the lower extremities.

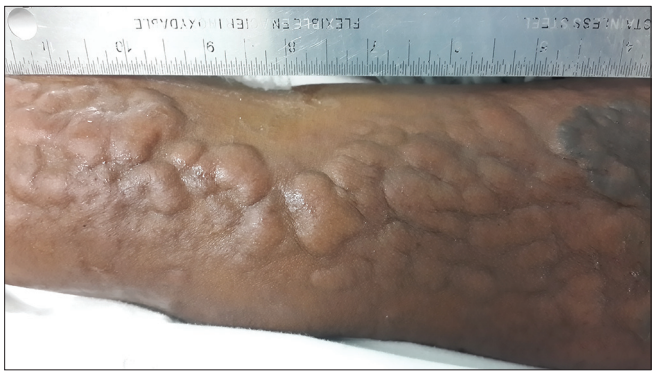

Figure 2: Higher magnification of skin nodules.

Chloride 90 mmol/L., AST 91 IU/L., ALT 16 IU/L., LDH 2901 U/L) 135-225), GGT 68 U/L., Alkaline phosphatase 123 U/L., Total protein 6.0 g/dl, Albumin 3.3d/dl, Globulin 2.7 gm/dl (3.5-5.5), Total Bilirubin 4.6 mg/dl, direct bilirubin 1.2 mg/dl, indirect bilirubin 3.4 mg/dl.

A bone marrow aspirate and trephine biopsy showed an infiltrate of malignant T-lymphocytes. There were no blast cells.

A skin biopsy of the nodule revealed a moderate infiltrate of small to medium sized lymphocytes with pleomorphic nuclei and sparse minute eosinophilic nucleoli arranged in a nodular fashion involving the dermis and subcutaneous tissue (Fig. 3). Admixed with this lymphocytic infiltrate were histiocytic cells (Fig. 4). Epidermotropism was absent.

The lymphocytes expressed T-cell markers with CD3, CD5, and CD4 positivity and were negative for CD7 and CD34.

Her HTLV-1 was positive. She was started on hydroxyurea and referred to the oncologist for further management.

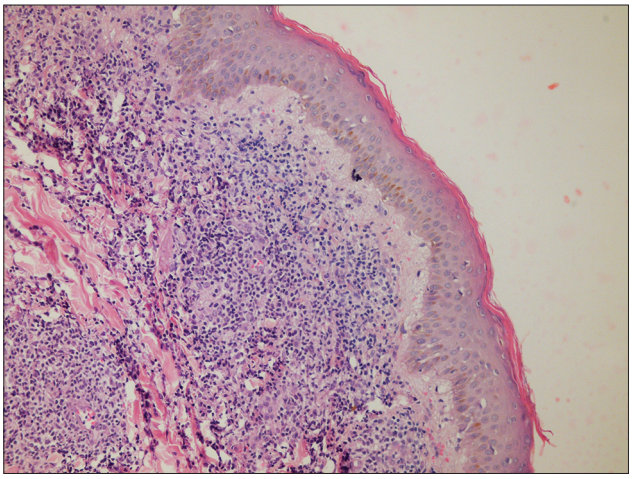

Figure 3: The sections show skin with hyperkeratosis. Within the dermis is a nodular infiltrate of atypical lymphocytes which is separated from the epidermis by a grenz. zone.

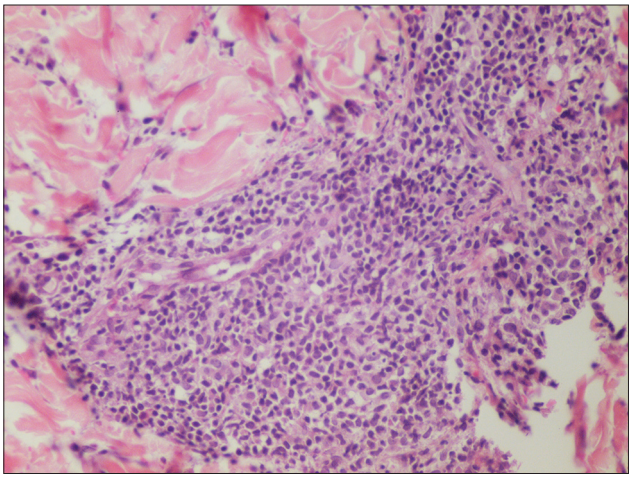

Figure 4: At higher magnification the atypical infiltrate was that of a T-cell lymphoma which is composed of small and medium size lymphocytes with pleomorphic nuclei and sparse eosinophilic nucleoli. Mitoses were infrequent. Admixed with these lymphoma cells were histiocytic cells.

The patient died one month after diagnosis while undergoing chemotherapy.

DISCUSSION

Human T-cell Lymphotrophic virus-1 is endemic in the Caribbean, Japan [7,8] and is seen now in most countries amongst immigrants from endemic countries [9,10].

The virus causes HAM/TSP and is the causative agent of HTLV-1 leukemia/lymphoma [1].

The acute, chronic, smoldering and lymphoma are the four different clinical manifestations of the neoplasm that have been identified [2].

We describe here a Grenadian national who presented in Trinidad W.I with splenomegaly and an acute HTLV-1 leukemia, having a white blood cell count of 131 x 10^3/UL with 83.6% atypical lymphocytes within the peripheral blood and a diffuse nodular skin lesion which histologically contained malignant HTLV-1 infiltrate. Hairy cell leukemia and chronic myeloid leukemia both have splenomegaly of the magnitude seen in this patient. However these two leukemia were eliminated from the diagnosis on account of the HTLV-1 positivity, the immuonhistology of the malignant cells, the absence of hairy cells and myeloblast in the peripheral blood and bone marrow aspirate. The skin nodules were composed of malignant infiltrate of HTLV-1 cells, which had no correlation with the consumption of shrimps. In addition to the lymphocytosis, she also presented with hypercalcaemia and a markedly elevated LDH, which are poor prognostic markers. This patient only survived one month after diagnosis.

HTLV-1 skin manifestation are varied and includes infective dermatitis, maculo-papular hyper pigmented or hypo pigmented rashes, nodules or ill-defined rashes, which mimics Mycosis fungoides, Psoriasis, Eczema and a list of common dermatoses, [11] and may be an early presentation of this neoplastic disease entity [5,6].

Clinicians should have a high index of suspicion of HTLV-1 related disease in patients from endemic regions who presents with a skin rash.

CONCLUSION

HTLV-1 is endemic in the Caribbean and Japan. Immigrants presenting with a skin rash from endemic regions should be investigated for HTLV-1 Leukemia/Lymphoma and it's other related diseases [12].

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Our Dermatology Online

Case Report

Case report on xeroderma pigmentosum with squamous cell carcinoma in a ten year old child

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ABSTRACT

Xeroderma pigmentosum (XP)is a rare inherited skin disorder characterized by a heightened sensitivity to the DNA damaging effects of ultraviolet radiation (UV). The main source of UV is the sun. The symptoms of XP can be seen in any sun-exposed area of the body. The effects are greatest on the skin, the eyelids and the surface of the eyes but the tip of the tongue may also be damaged. In addition, approximately 25% of XP patients also develop abnormalities of the nervous system manifesting as progressive neuro-degeneration with hearing loss. People with XP have a 10,000-fold increased risk for developing skin cancer including basal cell carcinoma, squamous cell carcinoma and melanoma. They also have a 2000-fold increased risk for cancer of the eye and surrounding ocular tissues. These symptoms appear early in life, typically before age 10 years.This case is being presented to highlight the rarity of a case of xeroderma pigmentosum with squamous cell carcinoma in a ten year old child.

Key words: Xeroderma pigmentosum; Squamous cell carcinoma in a child; Carcinoma

INTRODUCTION

Xeroderma pigmentosum (XP) was first described in 1874 by Hebra and Kaposi. In 1882, Kaposi coined the term xeroderma pigmentosum for the condition, referring to the characteristic dry, pigmented skin seen in these patients [1,2]. XP is a genodermatosis characterized by photosensitivity, cutaneous pigmentary changes, premature skin aging and the development of cutaneous and internal malignancies at an early age. These patients exhibit enhanced sensitivity to ionizing radiation [1,3,4].

CASE REPORT

A ten year old child born of a second degree consanguinity Presented to the Dermatology OPD at MVJMC & RH with Complaints of. Difficulty in opening eyes to broad day light since six months of age. Multiple dark and pale coloured discolouration present all over the body predominantly over sun exposed area. Swelling arising from the right eye since four months, associated with redness and watery discharge from both the eyes.

Patient was apparently normal at birth later cried excessively on bring out to the sun since six months of age. Patient would also refuse to open her eyes as a child to broad day light since six months of age. Patient also complaints of multiple dark and pale coloured discolouratioin associated with scales being present diffusely. Predominantly over the sun exposed area first starting over the face. Complaints of a swelling arising from the right lower eyelid since four months, which was insidious in onset initially of the size of a pea, which gradually progressed over a duration of three months to attain the present size of approximately 2x3 cm.

No H/o of application of native medicine. No H/o trauma to the eye. No H/o of known allergy to food, drug.

Family History

The Patient is a second child born to second degree consanguineous marriage.

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DOI: 10.7241/ourd.20182.14

Birth History

FTVD.

Developmental History

Attained developmental milestones.

Immunization History

Received as per schedule.

General Physical Examination

A moderately built and moderately nourished child who is consious and co-operative,Well oriented to time place and person.

Vitals

Normal.

On Examination

Extensive erosions with haemorrhagic crusts over the face, diffuse freckling, atrophic hyper and hypopigmented macules, ciliary and conjunctival congestion associated with watery discharge from bilateral eyes (Figs. 1 – 3).

A ulcero proliferative growth of size 3x4cm arising from the lateral aspect of right lower eyelid, the swelling is immobile, tender firm to hard in consistency. Eyelashes partially destroyed over b/l eyes. Systemic examination within normal limits.

With this we arrived a provisional diagnosis of xeroderma pigmentosus with squamous cell carcinoma.

Management

Initally patient was started on systemic and topical antibiotics analgesics and sunprotection. For further management patient was referred to ophthalmologist and plastic surgeon. Ophthalmologist prescribed tear drops, homatropine eye drops, antibiotic eye drops. Plastic surgeon did a wedge excision of the growth under local anaesthesia and specimen was sent for biopsy, which revealed well differentiated squamous cell carcinoma (Fig. 4).

Histopathology Report

Growth showed well differentiated squamous cell carcinoma – right eye lid and cheek (Figs. 5 and 6).

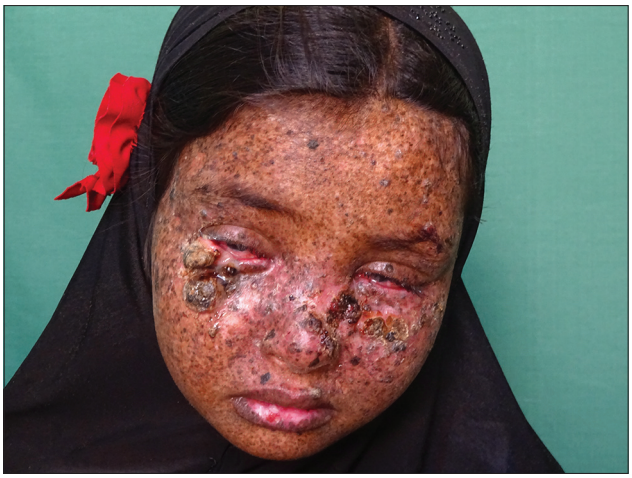

Figure 1: A single ulceroproliferative lesion arising from the lateral aspect of right lower eyelid.

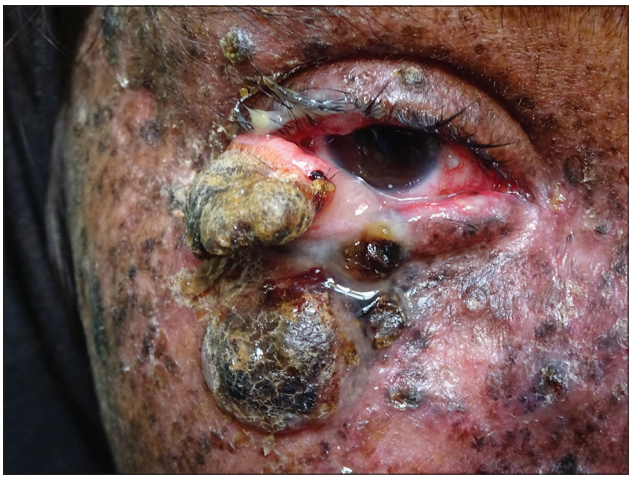

Figure 2: Haemorrhagic crusts over the face, diffuse freckling, atrophic hyper and hypopigmented macules and ciliary and conjunctival conjestion.

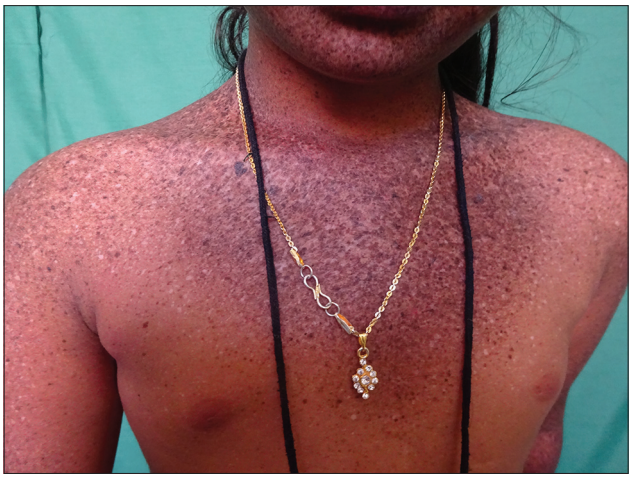

Figure 3: Atrophic hyper and hypopigmented macules.

Superolateral and superomedial margins are involved by tumour, with staging $pT_2N_xM_x$.

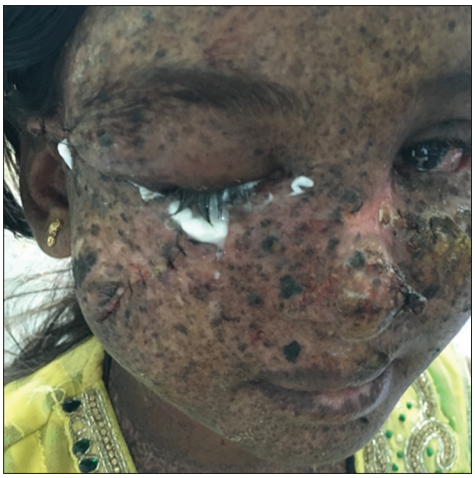

Figure 4: Post excision picture of the ulcero-proliferative lesion on histopathological examination revealed squamous cell carcinoma.

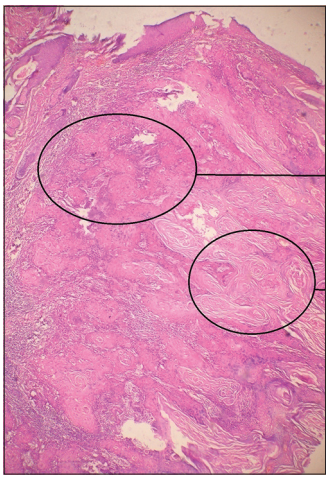

Figure 5: Epidermis and dermis, irregular rete ridges extending to dermis and atypicalcells. With well differentiated squamous cells, HPE:10X. In the order top to bottom: 1st arrow: well differentiated nests of malignant squamous epithelial cells. 2nd arrow: keratin pearls.

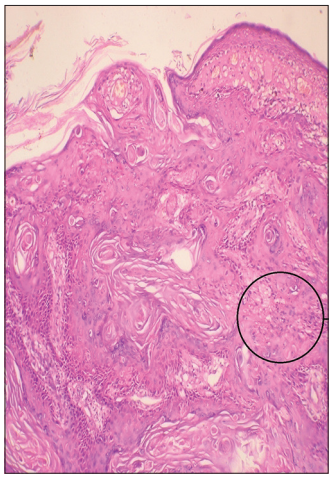

Figure 6: Epidermis and dermis, irregular rete ridges extending to dermis and atypicalcells. Chronic inflammatory cells composed of lymphocytes is seen, HPE:10X. Arrow: Chronic inflammatory cells composed of lymphocytes.

Thus the final diagnosis of xeroderma pigmentosus with squamous cell carcinoma was made.

DISCUSSION

XP is inherited as an autosomal recessive trait. It is a rare pigmentary atrophic disease that begins in childhood and progresses to early development of senile changes in sun-exposed skin [5]. Parental consanguinity is common. XP occurs with an estimated frequency of 1:250000 in the US and, according to Robbins et al., is more common in Japan. The incidence in the Indian population is insignificant. XP shows no sex preference [1,3,6].

The basic defect in XP is in Nucleotide excision repair, leading to deficient repair of DNA damaged by UV radiation. NER involves removal and the replacement of damaged DNA with new DNA. Two types of NER exist: global genome NER (GG-NER) and transcription-coupled NER (TC-NER). In addition to defects in the NER genes the immunosuppressive effects of UV-B radiation may also be involved in the pathogenesis of XP.

XP patients below 20 years of age have a >1000-fold increased risk of developing skin cancer [6,7]. The median age of onset of non-melanoma skin cancers reported in patients with XP is 8 years, compared to 60 years in the healthy population. Variations in the type of malignancies in XP appear to be related to the degree of sun exposure and genetic heterogeneity [8]. The two most common types of cancer found in XP patients are BCC and SCC, mainly occurring on the face, head, and neck. Melanomas occur in one-fourth of cases, and one-third of these occur in the head and neck [4]. Early detection of these malignancies is necessary because they are fast growing, metastasize early and lead to death. Two important causes of mortality are metastatic melanoma and SCC.

Most patients with XP do not live beyond the third decade because of the development of tumors [4]. Cutaneous neoplasms in XP patients cannot be prevented but early protection from UV radiation should be advised. Premalignant skin lesions may be treated with cryosurgery or topical antimitotic agents. Early removal of neoplasms should be accomplished with excision, chemosurgery or intralesional IFN-α. In selected XP patients, oral isoretinoin has been shown to significantly reduce the incidence of skin cancers [9].

A recent clinical trial by Yarosh et al. [10] found that

the enzyme T4 endonuclease V, applied regularly as a topical agent, significantly reduced the onset of both BCC and actinic keratoses, thus giving a little hope to XP patients, who suffer from much psychological trauma and face many socioeconomic problems. Although, early detection and treatment of cutaneous malignancies will reduce morbidity and mortality, genetic counseling remains the most important measure for preventing XP.

CONCLUSION

This case is being presented to highlight the rarity of a case of xeroderma pigmentosum with squamous cell carcinoma in a ten year old child. And need for early diagnosis and prompt treatment.

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Our Dermatology Online

Case Report

Multidermatomal nevus comedonicus: How dermoscopy aids diagnosis?

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ABSTRACT

Nevus Comedonicus (NC)is a rare subtype of epidermal nevus considered as a variant of adnexal hamartoma that presents as grouped hyperkeratotic papular lesions with appearance similar to comedones. Lesions usually present at birth but involvement can occur any time up to middle age. Here we report a case of NC in 16 year old female occurring after puberty. We report this case because of its multidermatomal involvement, strictly unilaterally distributed lesions with progressive pubertal enlargement showing characteristic features of NC on dermoscopy, thereby helping to differentiate it from other diseases with similar clinical presentation.

Key words: Epidermal nevus; Nevus comedonicus; Comedo like

INTRODUCTION

NC is uncommon anomaly affecting pilosebaceous unit in which follicular structure instead of forming hair shaft produces keratin plug occluding the follicle which presents as linear or grouped hyperpigmented comedo like papular lesions [1,2]. It most commonly involves face, head and neck region [3]. Incidence is equal in males and females with most of the cases occurring before 10 years of age [4]. Lesions can get secondarily infected or may develop pilar sheath acanthomas and trichofolliculomas [5]. In current scenario, dermoscopy being non-invasive and simple diagnostic tool plays pivotal role in diagnosing and ruling out clinically similar differentials.

CASE REPORT

A 16 year old female presented in the dermatology outpatient with asymptomatic multiple grouped hyperpigmented keratotic follicular lesions since the age of 11 years distributed strictly unilaterally over right side involving chest, mammary area, axilla, right arm spreading to involve back with multidermatomal distribution (cervical C4,C5 and thoracic T1-T7) (Fig. 1). Lesions were completely asymptomatic not associated with any complaint of itching, oozing or ulceration. Only concern of the patient was cosmetic appearance as these lesions progressively enlarged during last 3-4 years. No history of any lesions at time of birth, precipitating trauma or rash given by the patient. There was no similar family history and patient was born of non-consanguineous marriage. No other associated ocular, CNS or skeletal abnormalities were observed. All routine investigations were within normal limits. Lesions were examined using DermLite DL3 Dermoscope which revealed multiple dark brown areas studded with keratin plugs and numerous follicular openings showing characteristic dark colored plugging (Fig. 2). These changes were consistent with the clinical diagnosis of Nevus Comedonicus, in contrast to acne vulgaris where superficial circular areas of homogeneous hyperpigmention are usually seen. Skin biopsy done from the lesions present over the upper back revealed large dilated poorly formed follicles filled with lamellated orthokeratotic material (Fig. 3).

DISCUSSION

Nevus Comedonicus, also known as Zoniform Nevus is considered as adenexal hemartoma with developmental

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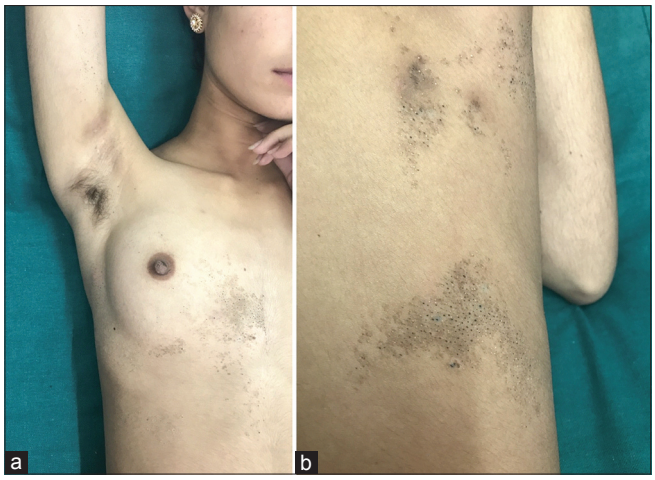

Figure 1: (a) Hyperkeratotic grouped comedo like papular lesions distributed unilaterally over right side involving chest, axilla and right arm. (b) Back of the patient showing keratotic plugging distributed unilaterally in a grouped pattern.

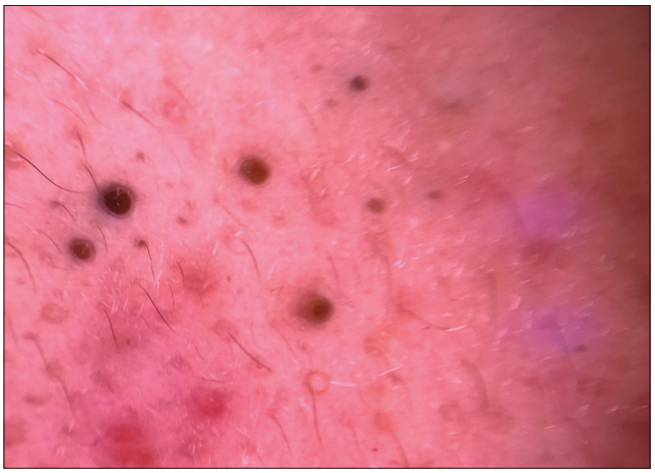

Figure 2: Dermoscopic picture of NC done from back of the patient showing multiple dark brown colored keratotic plugs.

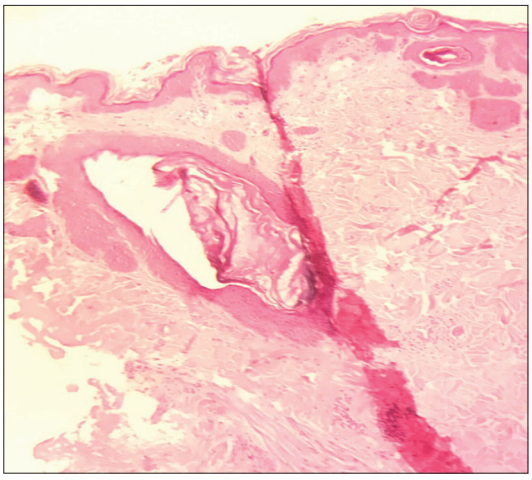

Figure 3: Histopathology of NC showing hair follicle filled with lamellated keratotic material with normal interfollicular epidermis.

failure of pilosebaceous unit. It is a misnomer as true comedones are absent [6]. Two types of NC have been reported [7]. First type is characterised by asymptomatic comedo like lesions distributed in various patterns like linear, interrupted, unilateral or bilateral which mainly is of cosmetic concern to the patients, where as second type is characterised by cysts, abscesses, fistulas and scarring mainly because of recurrent secondary infections and inflammation. The Case reported here is consistent with the presentation of first type with unilateral multidermatomal involvement.

Dermoscopy is a non invasive and confirmatory tool to rule out differentials of NC. It is recordable, repeatable and bed side investigation. Till now very few reports have been published in context of dermoscopic differentials of NC [8]. In acne, dermoscopic features depend upon comedone type whether open or closed. In contrast to NC, where follicular openings studded with keratin plugs are characteristic, in acne vulgaris circular homogeneous more superficially located areas of hyperpigmentation are seen. Other differentials includes sebaceous nevus where aggregated bright yellow dots without any hair follicles are seen. Rarely hair follicle nevus showing pseudo-pigmented network is seen on dermoscopy. In this case dermoscopic features characterically revealed keratin plugs consistent with NC.

Histopathologically lesions of NC typically reveal dilated hair follicles containing keratinous debris with acanthotic or hyperkeratotic interfollicular epidermis. In the current case inter-follicular epidermis is normal.

NC can be a part of epidermal nevus syndrome which is associated with several ocular abnormalities which includes cataract, corneal involvement, skeletal anomalies like hemivertebrae, absence of little finger, scoliosis and various neurological abnormalities. In long standing cases of NC follicular tumors have been reported [9]. Our patient did not have any such association thus it is not a part of nevus comedonicus syndrome.

Treatment is mainly aimed for cosmetic purposes. In this patient we tried 0.1% tretinoin cream for 4 weeks and two sittings with Fractional C02 laser were done, after which the patient was lost to follow up. Other topical modalities that can be used include adaplalene, tazarotene, calcipotriene alone or in combination with steroids [10]. In resistant cases mechanical removal of plugs using cosmetic strips or dermabrasion can

be tried. Various lasers such as Er-YAG laser, diode laser, CO2 laser have been therapeutically explored for treating NC with varied results [11]. Use of oral isotretinoin has been tried for treating few cases of extensive NC. Surgical excision has been kept as last resort or in cases of localised tumor growth or inflammation.

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Our Dermatology Online

Case Report

Angiolymphoid hyperplasia with eosinophilia – A report of three cases

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ABSTRACT

Angiolymphoid hyperplasia with eosinophilia (ALHE) is an uncommon, reactive vaso-proliferative disease, presenting with painless, vascular nodules in the dermal and subcutaneous tissues, sually seen in the head and neck region. It is characterized clinically by single to multiple red brown dome shaped papules or subcutaneous nodules. Its etiology is unknown and the histology is characterized by hyperplastic blood vessels lined by a hypertrophic endothelium with an inflammatory infiltrate rich in eosinophils. We report three cases of ALHE presenting over the ears and scalp.

Key words: Angiolymphoid hyperplasia with eosinophilia; Eosinophilia; Angioproliferative; Pseudopyogenic granuloma

INTRODUCTION

Angiolymphoid hyperplasia with eosinophilia (ALHE), also called as atypical or pseudo pyogenic granuloma, is characterized by solitary or multiple red to brown papules or nodules seen commonly in women between 20 and 40 years of age. The etiology of ALHE remains unknown, because it is not clear if it is primarily a vascular neoplasm, a lymphoproliferative process or a heterogeneous group of entities [1]. Trauma, infections and hyperestrogenic conditions (pregnancy or oral contraceptive agents) are considered to be the likely causes. ALHE usually appears in head and neck region, frequently in the auricular area and usually measures about 2-3 cm in size [2]. ALHE must be histologically and clinically differentiated from Kimura disease, which is a chronic inflammatory condition, characterized by large subcutaneous nodules in the head and neck region [3]. We report three cases of ALHE with characteristic clinical and histological features.

CASE REPORT

Case 1

A 45-year old male who presented to us with chief complaints of multiple, asymptomatic, gradually progressive reddish nodular lesions over the back of the head for the last three years. There was history of excessive bleeding from a few lesions even on minor trauma. The patient had consulted some physicians and had applied numerous medications without any relief. The patient had got a few bigger lesions removed surgically one year back, but they had recurred. On examination, there were multiple red, non-pulsatile smooth surfaced papules, over the occipital region (Fig. 1). There was no lymphadenopathy. Other systems were normal. Differential count showed 18% eosinophils and his IgE level was normal. An excisional biopsy of the lesions was done which revealed proliferation of small vessels lined by plump endothelial cells, surrounded by inflammatory cells including lymphocytes, mastocytes and eosinophils which confirmed the diagnosis of ALHE (Fig. 2). The bigger lesions were removed by radiofrequency but after a six month follow up, a few lesions had recurred.

Case 2

A 15 year old male presented to us with history of asymptomatic, gradually progressive nodular lesions over the meatus of the right ear. The lesions started at the age of 11 years and were a single nodule to begin with, but over the time, they had increased in size

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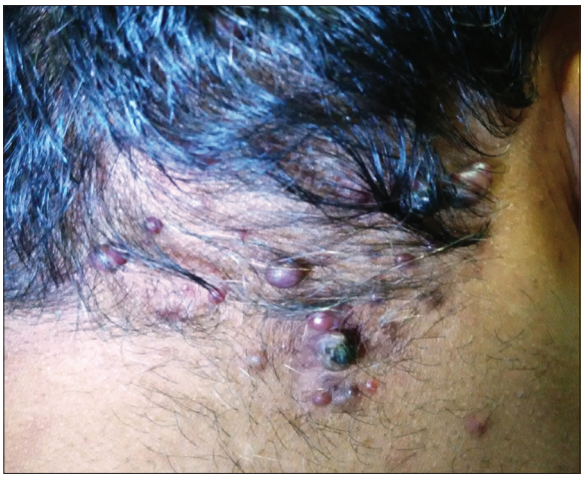

Figure 1 : Multiple reddish nodular lesions on the occipital region.

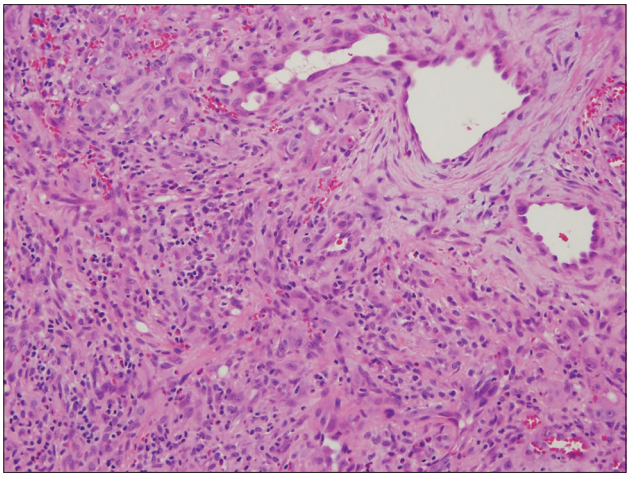

Figure 2: Proliferation of blood vessels in the dermis. The vessels are lined by plump epithelioid endothelial cells with surrounding eosinophilic infiltration.

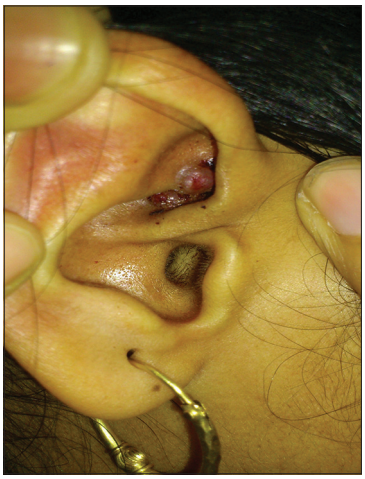

Figure 3: Multiple angiomatous nodules present over the meatus of ear.

and number to their present size, but had remained asymptomatic throughout. On examination, there were multiple red, non-pulsatile smooth surfaced papules, over the pre auricular region extending on to the crus of the helix, and into the meatus of the right ear (Fig. 3). There was no lymphadenopathy. The histology of the lesion was consistent with ALHE. The patient was advised radiofrequency removal of the lesion which was refused by the parents.

Case 3

A 27-year old female presented with multiple asymptomatic, reddish nodular lesions over the ear for the last four years. Examination revealed multiple red, non-pulsatile smooth surfaced papules, over the pre auricular region. The histology of the lesion confirmed the diagnosis of ALHE. The lesions were removed by radiofrequency and there was no recurrence of the lesions over a three month follow up period.

DISCUSSION

ALHE, also known as pseudo- or atypical pyogenic granuloma, subcutaneous angioblastic lymphoid hyperplasia with eosinophilia, and papular angioplasia, was first described by Wells and Whimster in 1969, who considered ALHE to be a late stage of Kimura's disease. It is usually seen in the third and fourth decade with a slight female predominance [3]. The pathogenesis of ALHE remains unclear. Some authors consider ALHE as a neoplasm developing from endothelial cells; others suggest that it is secondary to an inflammatory vascular reaction secondary to complex immunologic mechanisms. Many other hypotheses have been reported implicating environmental factors such as insect bite, trauma, and infections. Some authors consider arterio-venous shunt to be the main etiopathogenetic mechanism [3-5]. Cutaneous lesions consist of red papules or persistent subcutaneous nodules which may be associated with spontaneous bleeding, pain, pulsation and pruritus.

The diagnosis is based on clinical features and the histopathology which is characterized by a proliferation of small vessels lined by epithelioid endothelial cells and surrounded by inflammatory cells including lymphocytes, mastocytes and eosinophils. Serum hypereosinophilia is not a consistent feature, seen in around 20% of cases, and is not required to make the diagnosis. The various differential diagnoses include Kimura's disease, angiomatous neoplasias such as capillary hemangioma, granuloma pyogenicum with

satellite lesions, angiosarcoma of the face and scalp, and Kaposi sarcoma [2-5].

A variety of treatment modalities have been described for ALHE. Surgical treatment remains the treatment of choice, but recurrences are observed in 30% of the cases. Other described treatment modalities include electrodessication, cryotherapy, micrographic surgery, systemic corticosteroid treatment, intra-lesional injection of corticosteroids or sclerosing products, topical tacrolimus, imiquimod, phototherapy and alpha-2a interferon [2,3,5,6].

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Our Dermatology Online

Case Report

Pilomatrical carcinoma - a case report and review of the literature

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ABSTRACT

We report a case of an 85-year-old man, who presented with a polypoid skin tumor in the left lumbal region. Histology revealed a malignant tumor composed predominantly of pleomorphic basaloid cell population with a high mitotic and proliferative activity. This cellular component was admixed with aggregates of anuclear cells with eosinophilic cytoplasm reminiscent of „shadow" cells seen in pilomatrixoma, as well as with whorls of keratin material reminiscent of squamoid pearls. Immunohistochemically, the basaloid tumor part was positive for CD10, p120catenin and CD138 and very sporadically positive for cytokeratin 19 and BerEP4. Cytokeratin 20 was negative and epithelial membrane antigen labelled only eosinophilic squamoid structures. In some areas, numerous interspersed dentritic melanocytes strongly immunoreactive for S-100 protein were arranged singly and in larger expansile nests within basaloid tumor mass. Histopathology and immunoprofile of lesion favored a diagnosis of pilomatrical carcinoma with intratumorous melanocytic proliferation. To the best of our knowledge, only a few such cases have been described untill now.

Key words: Pilomatrical carcinoma; Pilomatrixoma; Adnexal tumors

INTRODUCTION

Pilomatrical carcinoma (PC) is a very rare low-grade cutaneous malignancy with matrical differentiation, considered to be the malignant counterpart of pilomatrixoma. It was first described in 1980 by Lopansri and Mihm [1] and until now, about 140 cases have been published in the English literature [2]. Given the rarity of this neoplasm, there is impossible to estimate its incidence, and knowledge on etiology, pathogenesis and biological behaviour is only sparse. Here, we report an additional case of PC with uncommon histological feature and provide an up to date review of the literature.

CASE PRESENTATION

An 85-year-old man with a history of multiple cutaneous basal cell carcinomas presented with a polypoid skin tumor in the left lumbal region. He visited a dermatology outpatients' department. On physical examination, the lesion was sharply demarcated, dark-brow in color and measured 12x8x6 mm. A presumptive clinical diagnosis of BCC was made. The patient was reffered to the hospital, when a total surgical excision of tumor had been performed.

Histopathology and Immunohistochemistry

Biopsy specimen was completely processed using standard hematoxylin and eosin stained paraffin sections along with a wide spectrum of immunohistochemical markers, summarized in Table 1. Histology revealed solid tumor formations, which were predominantly composed of pleomorphic basaloid cell population with frequent mitoses. This cellular component was admixed with multiple aggregates of anuclear cells with eosinophilic cytoplasm reminiscent of „shadow" cells seen in pilomatrixoma, as well as with whorls of pink keratin material reminiscent of squamoid pearls (Figs. 1 and 2). There was very high mitotic and proliferative activity. The

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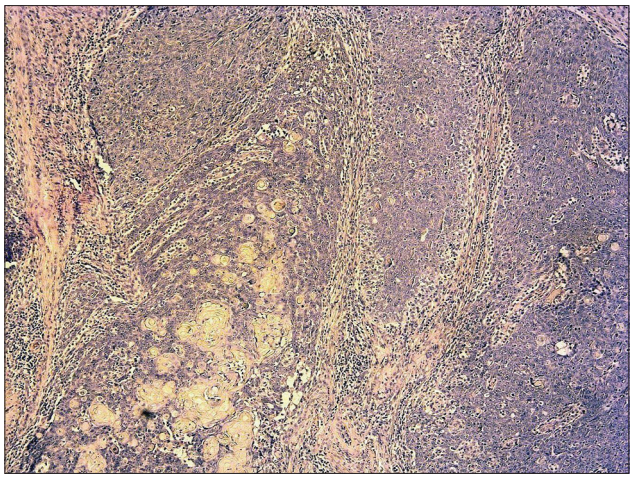

Figure 1: Predominant pleomorphic basaloid tumor cell population admixed with aggregates of eosinophilic anuclear cells and whorls of pink keratin material. (H&E, original magnification 40x).

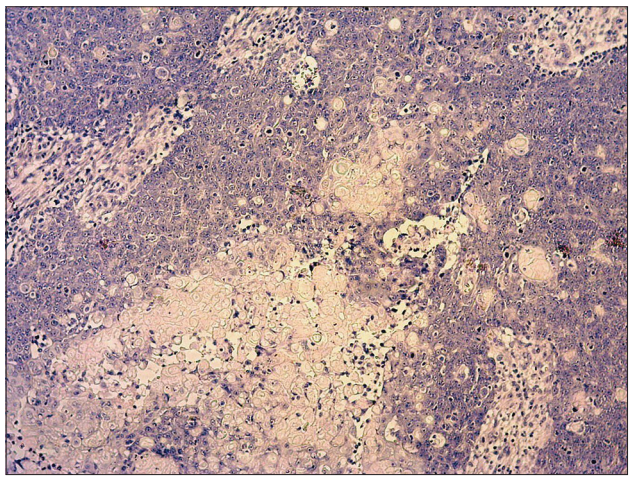

Figure 2: Detail on interface between basaloid tumor part and anuclear eosinophilic cells reminiscent of „shadow" cells, as well as whorls of keratin squamoid structures. (H&E, original magnification 200x).

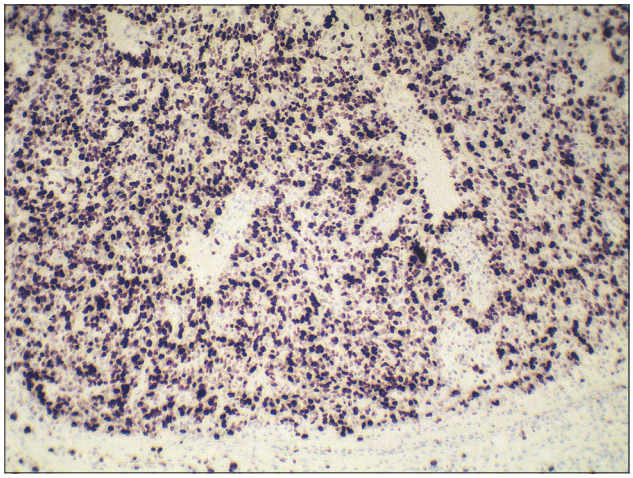

Figure 3: High proliferative activity (Ki-67 index) of basaloid cell population. (original magnification 200x).

Ki-67 index reached 90% (Fig. 3) and more than 40 mitotic figures per 10 high power fields were counted. Immunohistochemically, the basaloid tumor part was apparently positive for CD10, p120catenin and CD138 and very sporadically positive for cytokeratin 19 and BerEP4 (both < 5% of cells). Cytokeratin 20 was negative and epithelial membrane antigen (Fig. 4) labelled only eosinophilic squamoid structures and „shadow" cells. Interestingly, in certain areas, numerous interspersed dentritic melanocytes strongly immunoreactive for S-100 protein were arranged singly and in larger expansile nests within basaloid tumor mass (Fig. 5). The tumor extended into the epidermis, resulting in focal ulceration. At the base, it grew invasively with infiltration of the dermis. Neither lymphovascular nor perineural tumor invasion was found. A spectrum of histomorphological findings along with immunophenotype favored a diagnosis of pilomatrical carcinoma with intratumorous melanocytic proliferation. Resection margins were free of tumor and a minimum of 5-mm clearance was achieved. The patient continued to be under close follow-up and at the time this report was written (16 months after operation), no evidence of local recurrence or distant metastasis have been found.

DISCUSSION

PC is exceedingly rare cutaneous malignancy derived from the hair follicle matrix. It shows a predilection for middle aged and elderly individuals [3,4], but the cases involving the young people have also been described [5-7]. PC has no specific anatomic location and may arise anywhere in the body. Although it has been shown a slight predilection for the head and neck region [3,4,8-12], various other sites affected have been reported to date, such as the back [3,13], chest [3,4], upper limbs [3,7,14], lower limbs [2,15], buttock [3] or vulva [5].

Table 1: Details of the primary antibodies and the corresponding detection systems used in the present case

Antigen	Antibody	Source	Dilution
Ki-67	monoclonal, clone MM1	Leica	1 : 200
CD10	monoclonal, clone 56C6	Dako	ready to use
p120catenin	monoclonal, clone MRQ-5	Dako	1 : 25
CD138	monoclonal, clone MI15	Dako	ready to use
cytokeratin 19	monoclonal, clone RCK108	Dako	ready to use
cytokeratin 20	monoclonal, clone Ks20.8	Dako	1 : 50
epithelial antigen	monoclonal, clone BerEP4	Dako	ready to use
epithelial membrane antigen	monoclonal, clone E29	Dako	1 : 100
S-100 protein	monoclonal, clone 15E2E2	BioGenes	1 : 100

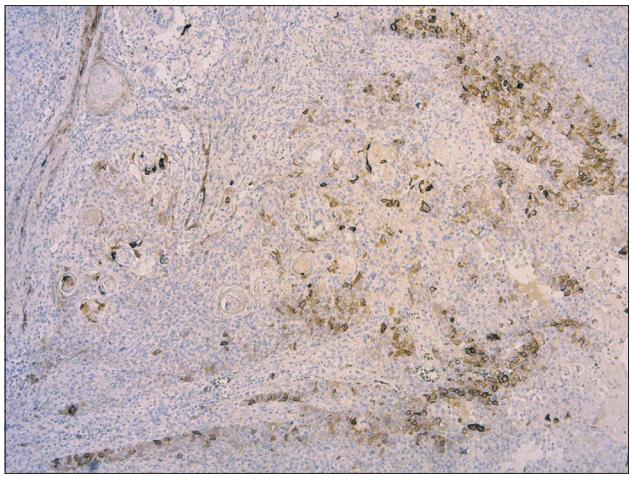

Figure 4: Positivity for EMA in eosinophilic tumor component, while basaloid tumor population is negative. (original magnification 100x).

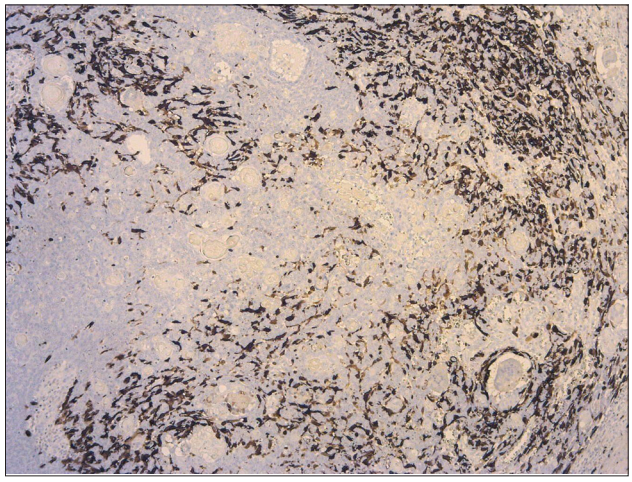

Figure 5: Strong positivity for S-100 protein in dentritic melanocytes interspersed within basaloid tumor mass. (original magnification 100x).

The origin of PC still controversial. Based on the literature research, the vast majority of the cases have been shown without histologic association with its benign counterpart, the finding supporting the development *ab initio*. Nevertheless, the literature notes some case reports of patients in whom a biopsy specimen first identified the tumor as a benign lesion that underwent carcinomatous changes after a certain period of latency [6,10,15]. In the present case, we did not observe any benign tumor component within the tumor mass, so it was probably malignant from the onset. In this regard, it should be mentioned that although PC is a malignant counterpart of pilomatrixoma, and both tumors share common histopathologic and immunophenotypic features, their epidemiology is different. While benign pilomatrixomas are seen more often in women (female/male ratio 3:1), PCs are seen in the opposite ratio (female/male ratio 1:3-5) [3,4,14]. Further, pilomatrixomas typically occur in children and young adults, but PCs usually occur in older people in the 5th and 6th decades of life [4,14].

A definite diagnosis of PC and in particular, its differentiating from pilomatrixoma remains based only on the histomorphological features. However, much of the histopathology of PC resembles its benign counterpart and the criteria of malignancy have not been well established. In general, these include tumor asymmetry, poor circumscription, marked anaplasia of basaloid neoplastic cells with vesicular nuclei and prominent nucleoli, frequent atypical mitoses, areas of necrosis, infiltrative growth pattern, sometimes ulceration and vascular and/or perineural invasion [4,12,15]. In our case, the vast majority of them have been clearly visible confirming a malignancy. PC possess no specific immunoprofile and immunohistochemistry studies have not yielded markers that may consistently and definitively confirm a diagnosis. Moreover, many case reports published until now have not performed special immunoanalyses. In general, benign and malignant cutaneous adnexal tumors of follicular origin variably express the „hair differentiation" keratins, such as cytokeratin 7, 8, 18 and 19 [16]. In both, pilomatrixoma and PC, the basaloid cells showed a nuclear and cytoplasmic expression of β-catenin [17] and in pilomatrixoma, they showed strong positivity for CD138 and CD10 [18]. Two papers [7,12] also demonstrated a reactivity for BerEP4 in PC.

An interesting feature in our case was a remarkable population of dentritic melanocytes within a basaloid tumor tissue. In the last years, there were reported the cases of PCs accompanied by marked intratumorous melanocytes colonisation [2,11,13,19]. The terminology of this entity is inconsistent, since it has been entitled as melanocytic pilomatrix carcinoma [13], pilomatrix carcinoma with intralesional melanocytes [11], matrical carcinoma with melanocytic proliferation [2], or matrical carcinoma with prominent melanocytic hyperplasia [19]. Nevertheless, they are likely the same tumor entity under the different names. We believe that our current lesion represents an additional case of this unique variant. This histologic feature probably recapitulates the intimate relationship existing between matrical epithelium and melanocytes in the embryonal hair follicle or in the anagen stage of the hair cycle. Theoretically, a presence of numerous melanocytes in PC should not be surprising, but is in fact a very unusual feature. In the series of 20 PCs published by

Sau et al. [3], immunohistochemical stain for S-100 protein was negative in all tumors.

From the clinical point of view, PC usually behaves like a low-grade malignancy, but it possess a high propensity for local recurrence. In the study of Sau et al. [3], approximately half of the cases investigated relapsed. A few PCs with regional and/or visceral metastases have been published until now [3,8,9], some of which have led to death within a relatively short period from time of diagnosis [8,9]. Although PC metastasizes in only about 10% of the cases, when it becomes metastatic, a mortality rate approaches nearly 100% [14]. Therefore, a close clinical follow-up after diagnosis is a priority for further managment of the patient.

In conclusion, we described a unique case of PC accompanied by intratumorous melanocytic proliferation. To the best of our knowledge, only a few such cases have been published untill now. The biologic significance of melanocytic proliferation in this rare cutaneous neoplasm is uncertain and requires further study. It would be interesting to analyze larger number of the cases to elucidate, whether they may be considered a distinct histologic variant of PC with possibly different biological behaviour.

Consent

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

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Our Dermatology Online

Case Report

Eruptive syringomas in Down's syndrome

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ABSTRACT

Down's syndrome is a common chromosomal anomaly characterized by mental retardation and distinct physical appearance. It is associated with a large variety of dermatological disorders like anetoderma, cheilitis, cutis marmorata, elastosis perforans serpiginosa, fissured and geographic tongue, onychomycosis, palmoplantar hyperkeratosis, psoriasis, syringomas, alopecia areata and vitiligo. Syringomas are benign adnexal tumors which have been reported to occur with higher frequency among patients with Down's syndrome. We report a case of eruptive syringomas over face and hand in an 8-year old child with Down's syndrome.

Key words: Syringomas; Trisomy 21; Down's syndrome

INTRODUCTION

Down's syndrome is a common chromosomal anomaly associated with multiple malformations, medical conditions, and cognitive impairment because of the presence of extra genetic material from chromosome 21. The typical physical findings include hypotonia, brachycephaly, epicanthal folds, flat nasal bridge, upward-slanting palpebral fissures, small mouth, small ears, excessive skin at the nape of the neck, single transverse palmar crease, and short fifth finger with clinodactyly and wide spacing between the first and second toes [1]. Down's syndrome is also associated with accelerated aging and an increased incidence of a variety of dermatological disorders like anetoderma, cheilitis, cutis marmorata, elastosis perforans serpiginosa, fissured and geographic tongue, onychomycosis, palmoplantar hyperkeratosis, pityriasis rubra pilaris, psoriasis, seborrheic dermatitis, syringoma, alopecia areata, vitiligo and xerosis [2,3].

Syringomas, benign adnexal neoplasia derived from eccrine ductal elements, are found in a higher frequency among patients with Down's syndrome. We report a case of eruptive syringomas over face and hand in an 8-year old child with Down's syndrome.

CASE REPORT

An 8-year old male with Down's syndrome was brought to us with the complaints of gradually progressive asymptomatic skin lesions over the face and hands for the last six months. The lesions started over the face and gradually increased in number and over the course of time appeared over dorsa of hands too. On cutaneous examination, numerous skin colored papules were present over the cheeks, forehead, periorbital area, bridge of nose and dorsa of hands (Fig. 1). There was no associated hypohidrosis and the patient's nails, hair and teeth were normal. The patient was advised a skin biopsy which was refused by the parents owing to the asymptomatic nature of the lesions. The patient was advised topical tretinoin 0.025% gel application over the lesions but there was no improvement in the lesions after three months of application.

From the history and clinical examination, a diagnosis of eruptive syringomas was made owing to the increased prevalence of syringomas in Down's syndrome patients.

DISCUSSION

Down's syndrome is one of the most common autosomal chromosomal disorders with an incidence

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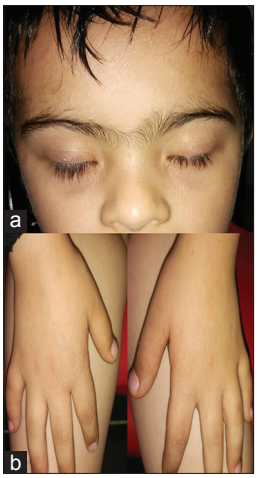

Figure 1: Multiple skin colored papules over the a) face and b) dorsa of hands.

of around 1 in 800 live births. Apart from numerous physical traits, a large variety of dermatological conditions with increased prevalence or increased severity are seen in patients with Down's syndrome [1]. Various authors have reported an increased prevalence of skin conditions like anetoderma, cutis marmorata, cheilitis, palmoplantar hyperkeratosis, pityriaisis rubra pilaris, xerosis and syringomas [2,3]. A higher incidence and severity of alopecia areata, vitiligo and atopic dermatitis has also been observed which has been attributed to immunological deficiency in T-cell function seen in patients with Down's syndrome. The B- and T- cell function and number is also reduced in these patients leading to a higher risk of infections and malignancies [2,4].

Syringomas are benign appendageal tumors of eccrine origin, most typically found in the periorbital region. Syringomas have been classified into four groups, according to their clinical features and associations, namely localized form, generalized/eruptive form, a form associated with trisomy 21 and familial form [5]. Clinically, they appear as small skin colored papules, rounded or flat-topped with angular margins, varying in size from 1 to 5mm. The front of the chest, face and neck are the main sites of involvement. Eruptive syringomas may appear on the neck, chest, abdomen and buttocks. The typical histological features include multiple small ducts lined by rows of flattened epithelial cells with epithelial strands within the dermis, giving rise to the characteristic tadpole appearance.

Syringomas are more common in patients with Down's syndrome and twice as common in female patients. The incidence of syringomas in Down's syndrome has been reported to be approximately 30 times greater than in the general population and syringomas of the eyelids have been reported exclusively in Down's syndrome. Studies have reported an incidence varying from 18% to 39% in patients with Down's syndrome [2,3,5].

Clinically, syringomas resemble trichoepitheliomas but syringomas tend to be smaller, rather more flat-topped and disposed more evenly over the cheeks and eyelids, rather than favoring the nasolabial creases. The treatment is mainly for cosmesis and various treatment modalities like surgical excision, electrocautery, cryotherapy, dermabrasion, trichloroacetic acid and carbon dioxide laser ablation have been used.

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Our Dermatology Online

Case Report

Caesarean section scar endometriosis: A case report and review of the literature with special emphasis on malignant transformation

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ABSTRACT

Implantation endometriosis following caesarian hysterectomy is not an uncommon entity. Seeding of endometrium within the peritoneum and pelvic organs and the Pfannenstiel incision commonly occur. Post caesarean implantation endometriosis occurrence is determined by several factors. While endometriosis is a morbid disease, which causes pain, dysfunctional uterine bleeding and infertility, it is also a precancerous condition, and efforts should be made to avoid implantation endometriosis during uterine surgery. The object of this article is to present a 30 year old female with implantation endometriosis in a Pfannenstiel abdominal scar and to review the diagnostic facilities available and highlight the premalignant potential of endometriosis.

Key words: Endometriosis; Caesarean scar; Malignant transformation

INTRODUCTION

Sampson 1927,postulated that retrograde flow of endometrium during the menstrual cycle, seeded the Fallopian tube and pelvic and abdominal organs causing endometriosis to occur in these sites under favorable conditions. This he referred to as implantation endometriosis. In recent times another form of implantation endometriosis has been reported post caesarian section, and following surgery on the uterus. Implantation often occurs within the Pfannenstiel scar and also within the pelvis and peritoneum. The case in question presents a 30-year-old female who suffered pain and discomfort within her pelvis and abdominal scar for four years following caesarian section. Her pain and discomfort continues for she most likely has implantation endometriosis within her pelvic organs as a result of implantation during caesarian section. The article focuses on the malignant transformation of endometriosis a phenomenon that is often forgotten as part of the natural sequel of endometriosis. In recent times cases of caesarian scar endometriosis with malignant transformation [1-4] have been reported in the literature indicating that malignant transformation of endometriosis can occurs at every site where it is deposited. The true incidence of malignant transformation of endometriosis is unknown.

Efforts should be made to avoid implantation endometriosis during surgical procedures of the uterus, and also to diagnose and treat this entity promptly to avoid the risk of its malignant transformation.

CASE REPORT

A 30 years old female, G2P1+1 who had a LSCS in the year 2009, presented to the Gynecology Outpatient Clinic in March 2013 with complaints of a lump on the right end of the LSCS scar. The lump had been present for over one year. She also complained of severe pain and swelling on the same side of the lesion, and in her pelvis, during and after her menstrual period.

She had her menarche at age 15years. She admitted to using oral contraceptive for 2years. She had no history

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of inter-menstrual bleeding, dyspareunia, post-coital bleeding or sexually transmitted disease.

She had an excision biopsy of the lump under general anesthesia.

The tissue received in pathology was a 4 x 2.5 cm^2 fibroadiopose mass admixed with old hemorrhages. Histological examination of the scar revealed endometriosis (Fig. 1).

DISCUSSION

Endometriosis is classically defined as the presence of endometrial glands and stroma in ectopic locations, primarily the pelvic peritoneum, ovaries, and recto-vaginal septum. The manifestation of endometriosis is not straightforward and does not occur in each individual during retrograde menstrual flow, for there seems to be interplay between immune, hormonal, genetic and environmental factors in its pathogenesis [3,4].

Sampson's implantation theory of endometriosis is the most accepted [5]. Here retrograde menstrual flow seeds the Fallopian tube, the ovaries and the peritoneum and pelvic organs with endometrial tissue, and with the right milieu, endometriosis is generated. This mechanism of endometriosis has been demonstrated both in humans and in laboratory animals [6].

Sampson 1927, proposed a second theory of endometriosis. He postulated that endometriosis occurred as a result of dissemination of endometrial tissue to other organs via the blood stream and lymphatic [7].

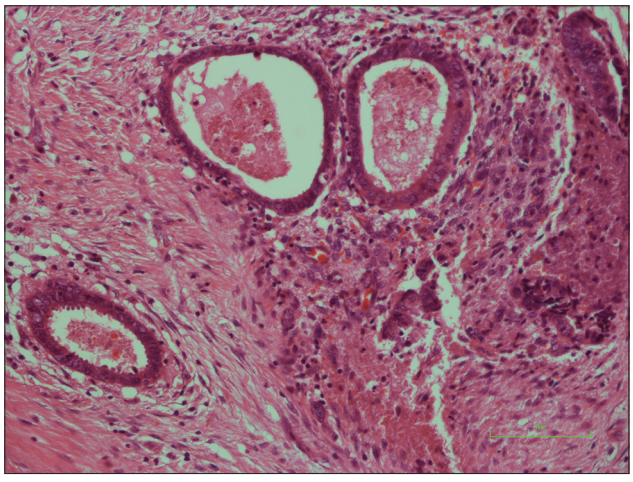

Figure 1: Endometrial glands and stromal cells within scar tissue. The glands are non-secretory.

A third hypothesis is the coelomic metaplastic theory, which postulates that undifferentiated coelomic cells are transformed into endometrium-like tissue under favorable conditions [8].

The case in question is one of implantation endometriosis, namely Caesarean section scar implantation endometriosis [9,10] and is another mechanism for the pathogenesis of endometriosis. The abdominal incision was seeded with endometrial tissue during the caesarian section and it is also possible that the peritoneum and other pelvic organs were similarly implanted at surgery, for her abdominal pains and discomfort continued after excision of the endometrial deposit in the surgical scar. The frequency with which endometriosis post-uterine surgery takes place is yet unknown and may very well be the major contributing factor to the causation of present day endometriosis.

Endometriosis is an inflammatory estrogen dependent condition associated with pelvic pain and infertility. Classically it is diagnosed by identifying endometrial tissue that is glands and stromal in extra uterine sites; the primary sites being the rest of the internal genital like the Fallopian tubes and ovaries, other pelvic organs, the peritoneum, the gastrointestinal tract. Endometriosis has been reported to involve even the mouth [11], probably as a result of the metaplastic theory or Sampson's or dissemination theory via lymphatic or bloodstream.

Endometriosis affects approximately 10% of women in their reproductive lives [12]. The diagnosis is not often readily made. Many biochemical markers have been investigated for the diagnosis of endometriosis but they lack specificity [13], although the evaluation of CA125 might me a useful marker in some forms of endometriosis and might be used to monitor improvement in endometriosis when therapy is instituted. Maiorana et al 2007 [14], found CA 125 serum levels were related to endometriosis and R-AFS score, in their study of patients with endometriosis. Because of the alarming statistical bias of endometriosis transforming into adenocarcinoma (79%)within the ovary, it would be useful to do early screening for ovarian cancer in patients with proven endometriosis [15].

Endometriosis is not only to be considered as a benign painful, morbid, condition but; a disease entity which undergoes malignant transformation [16].

Function and dysfunction of the female immune system play important roles in the initiation and progression of the disease and its relation to infertility and cancer [17]. In 79% of cases, the ovary has been recorded as the most common site where malignant transformation of endometriosis has taken place [18,19]. Melin et al [20] found an increased risk of some types of malignancy, above all ovarian cancer, in women with endometriosis, and postulates that the risk of transformation of endometriosis deposits in the ovary has been underestimated. Given the fact that ovarian cancers represents gynecological cancers with the worst prognosis [21], and that 79% of malignant transformation of endometriosis takes place in the ovaries, it would be prudent to avoid ovarian implantation endometriosis during uterine surgery.

Adenocarcinoma developing in extra-ovarian endometriotic sites have been recorded namely in the colon [22], symphysis pubis [23], the urethro-vaginal septum [24] and many other sites including the caesarian scar (1,2,9). The evidence supports the fact that endometriosis is yet another hyperestrogenic condition that has a propensity for the development of carcinoma, and should be considered to be a precancerous condition. Melin et al [20] states that approximately 1.0% of women with endometriosis have lesions that undergo malignant transformation.

Because of its troublesome clinical features of pain and infertility, its impact on the quality of life of the patient [25] and its propensity for malignant transformation [9], implantation endometriosis during gynecological surgery and caesarian sections should be avoided and early detection and treatment of endometriosis with a view to its eradication should be the goal.

CONCLUSION

Implantation endometriosis within Caesarean section is not an uncommon occurrence.

This malady needs to be diagnosed and treated early for endometriosis is a premalignant condition.

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Our Dermatology Online

Case Report

Punctate porokeratosis: Case report and review of the literature

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ABSTRACT

We present the case of a 63-year-old woman with a one-year history of punctiform, hyperkeratotic lesions on the palms of both hands. The histologic examination showed the presence of a cornoid lamella and allowed the establishment of the diagnosis of punctate porkeratosis. Porokeratosis is a clonal disorder of keratinization that exhibits multiple forms of presentation, with punctate porokeratosis being an infrequent form. The differential diagnosis of palmo-plantar punctiform, hyperkeratotic lesions is composed of porokeratosis punctata, the spiny keratodermas, other punctate keratodermas, and other pathologies with distinctive characteristics. In the review of literature of this article we have focused on explaining and clarifying the historical problem concerning the terminology used for these pathologies.

Key words: Porokeratosis; Cornoid lamella; Disorders of keratinization; Punctate keratoderma; Spiny keratoderma

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Our Dermatology Online

Case Report

Poroqueratosis punctata: reporte de un caso y revisión de la literatura

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RESUMEN

Presentamos el caso de una paciente de 63 años con lesiones puntiformes hiperqueratósicas de un 1 año de evolución en palmas de ambas manos. El estudio histológico demostró la presencia de una lámina cornoide y permitió establecer el diagnóstico de poroqueratosis punctata. La poroqueratosis es un desorden clonal de la queratinización que exhibe múltiples formas de presentación, siendo la variante punctata una forma infrecuente. El diagnóstico diferencial de lesiones puntiformes hiperqueratósicas palmo-plantares está compuesto por la poroqueratosis punctata, las queratodermias espinosas, otras queratodermias punctatas y otras patologías de características distintivas. En la revisión del presente reporte nos hemos enfocado en exponer y clarificar la histórica problemática que ha existido en la terminología empleada para estas patologías.

Palabras clave: Poroqueratosis; Laminilla cornoide; Desorden de la queratinización; Queratodermia punctata; Queratodermia espinosa.

INTRODUCTION

La poroqueratosis es un desorden clonal de la queratinización que exhibe múltiples formas de presentación. La característica común a este grupo heterogéneo de manifestaciones clínicas es el hallazgo histológico denominado lámina o laminilla cornoide [1]. Reportamos un caso de poroqueratosis punctata, variante inusual caracterizada por lesiones puntiformes hiperqueratósicas de ubicación palmo-plantar [2].

CASE REPORT

Paciente de sexo femenino, de 63 años de edad, con antecedentes de hipertensión arterial, hipotiroidismo, depresión, fibromialgia y cáncer de mama operado. Usuaria de ácido acetil salicílico, levotiroxina, escitalopram, carbamazepina, pregabalina y duloxetina. Consultó por un cuadro de 1 año de evolución caracterizado por lesiones puntiformes en palmas de ambas manos, generalmente asintomáticas, con episodios de dolor ocasionales. Sin antecedentes de exposición a arsénico ni relato de casos familiares. Al examen físico se constataron lesiones de 1-2 milímetros de diámetro en ambas superficies palmares y en zonas flexoras de todos los dedos a excepción de ambos pulgares (Fig. 1). Las lesiones eran espiculadas, hiperqueratósicas, algunas de color marrón y otras de coloración similar a la piel, levemente sensibles a la palpación.

Se realizó una biopsia de una lesión en palma izquierda. La muestra obtenida permitió evidenciar histológicamente la presencia de una columna paraqueratósica compacta, subyacente a la cual se evidenció un marcado adelgazamiento del estrato granular. Al observarse uno de los bordes basales de la columna paraqueratósica, en donde comenzaba a

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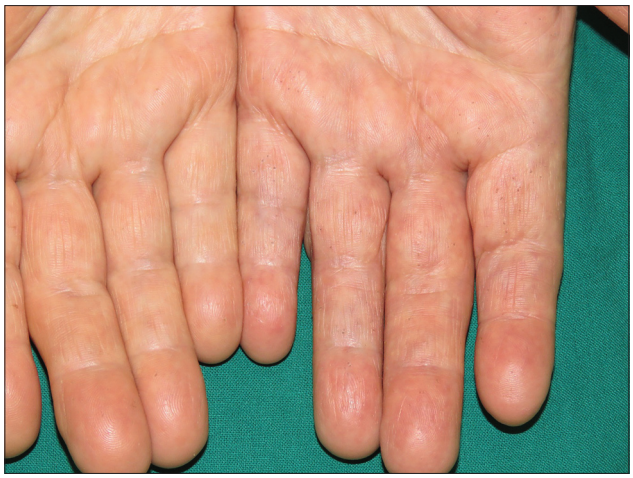

Figure 1: Lesiones puntiformes hiperqueratósicas en palmas y zonas flexoras de los dedos. [Hyperkeratotic punctiform papules involving the palms and the ventral surface of digits]

contactar a la epidermis adyacente, se logró observar un adelgazamiento casi total del estrato granular, con algunos queratinocitos vacuolados en el estrato espinoso aledaño (Figs. 2a and 2b). Además de esto, se encontró una discreta acantosis y discreta papilomatosis de la epidermis. A nivel de la dermis papilar se observó un escaso infiltrado inflamatorio mononuclear con escasos linfocitos de ubicación perivascular. Los hallazgos fueron compatibles con la presencia de una lámina cornoide y fue establecido el diagnóstico de poroqueratosis.

Se inició tratamiento con tretinoína crema al 0,1% y emulsión fluida queratorreguladora con lactato de amonio al 15%, citándose a control para luego de 1 mes. Sin embargo, la paciente acudió a la consulta dermatológica 7 meses después. Durante la segunda visita se constató que las lesiones seguían de igual aspecto. La paciente relató que controlaba sus lesiones utilizando una lima y retirándolas con pinza.

DISCUSSION

Las variantes más comúnmente descritas de poroqueratosis son la forma clásica de Mibelli, la poroqueratosis diseminada superficial actínica (DSAP, de su nombre en inglés, *disseminated superficial actinic porokeratosis*), la poroqueratosis diseminada superficial, la poroqueratosis linear, la poroqueratosis palmaris et plantaris disseminata y la poroqueratosis punctata (PP) [3]. Las dos primeras mencionadas son las formas más frecuentes, observándose en ellas las lesiones típicas de este desorden: pápulas queratósicas que evolucionan de forma centrífuga

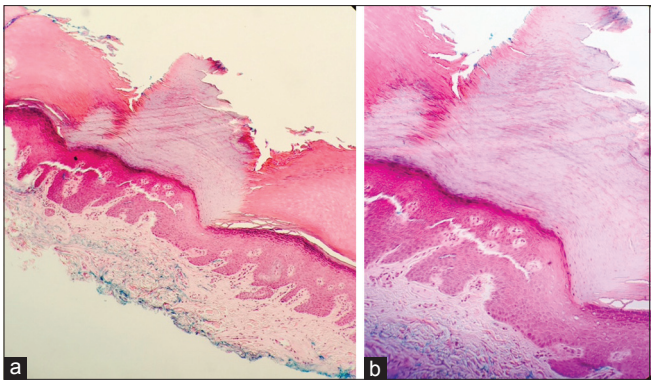

Figure 2: Biopsia obtenida de lesión hiperqueratósica en palma izquierda. (a) Se distingue una columna paraqueratósica compacta con un estrato granular subyacente adelgazado. (b) Se observa en un borde de la columna paraqueratósica un adelgazamiento casi completo del estrato granular, con algunos queratinocitos vacuolados en el estrato espinoso adyacente, [Biopsy of a hyperkeratotic papule on the left hand. (a) Histology reveals a compact parakeratotic column with an underlying decreased stratum granulosum. (b) The periphery of the parakeratotic column reveals an almost absent stratum granulosum along with the presence of some vacuolated keratinocytes].

hasta constituir lesiones anulares, demarcadas, de centro atrófico y con una periferia hiperqueratósica solevantada, pudiendo ser asintomáticas o asociadas a prurito [2]. Al obtener una biopsia de la región periférica, la histología revela el hallazgo común a todo tipo de poroqueratosis: la lámina cornoide, una columna compacta paraqueratósica que deprime un área epidérmica caracterizada por un adelgazamiento o ausencia del estrato granular. Para completar el cuadro histológico, queratinocitos vacuolados y/o disqueratósicos deben observarse en el estrato espinoso subyacente [1]. Junto a esto se ha descrito también un compromiso inflamatorio de la dermis de tipo linfohistiocitario [1-3].

La lámina cornoide representa una forma anormal de queratinización. A pesar de ser el elemento unificador de la poroqueratosis, no constituye un hallazgo patognomónico; es posible observarla en otros trastornos cutáneos inflamatorios o hereditarios, siendo en ocasiones un hallazgo incidental [1]. Descrita por primera vez en 1893 por Mibelli [4], el nombre "poroqueratosis" surge ante la idea errónea de que las láminas cornoides pudiesen tener su origen en las glándulas ecrinas [1,5,6]. Wade y Ackerman fueron los que describieron por primera vez en 1980 la participación de esta lámina en múltiples patologías, considerándola más bien una "reacción tisular menor" en lo que corresponde a la aproximación diagnóstica de las patologías inflamatorias de la piel basada en patrones [7]. Patologías como psoriasis, queratosis liquenoide crónica, dermatomiositis, corresponden

a algunos ejemplos en los que la histología puede también revelar una lámina cornoide; sin embargo, son otros los elementos clínico-patológicos los que guían el diagnóstico de aquellos cuadros [1].

En cuanto a la morfogénesis de la lámina cornoide, se han observado clones mutantes de queratinocitos predispuestos genéticamente con alteraciones en la ploidía del ADN. Una apoptosis prematura con alteración de la diferenciación terminal explicaría la queratinización desorganizada [1]. Se ha reportado también una queratinización rápida asociada a una descamación defectuosa y se ha demostrado con inmunohistoquímica la sobreexpresión del producto del gen P53 en los queratinocitos subyacentes a la lámina cornoide [1,8]. La asociación entre poroqueratosis y malignidades cutáneas ha sido descrita por múltiples autores [2,9,10]. Esta asociación puede comprenderse a través de la morfogénesis señalada: los queratinocitos comprometidos corresponderían a un estadío celular intermedio entre células normales y las células halladas en la enfermedad de Bowen. La transformación maligna se estima en un 7.5% de los casos, con predominio de la variante linear y con el carcinoma espinocelular (CEC) siendo el tumor principalmente asociado [1,2].

La patogenia de la poroqueratosis ha sido principalmente estudiada en relación a sus dos formas más frecuentes, destacando un componente genético, un rol de la luz ultravioleta (si bien existe un caso reportado en que un paciente con DSAP presentó mejorías al tratarse con PUVA [11]) y un rol también de la inmunosupresión [2,12]. Asimismo, infección por virus papiloma humano y traumatismos han sido considerados como elementos con un posible rol patogénico [2].

No existen estudios aleatorizados sobre el tratamiento de la poroqueratosis y las respuestas suelen ser impredecibles: ningún tratamiento ha demostrado una eficacia consistente y de largo plazo [13,14]. Sin tratamiento, las lesiones persisten indefinidamente [10,13]. La regresión espontánea, si bien descrita, es rara [15]. Finalmente, dado el carácter premaligno de este desorden, es el seguimiento de los pacientes lo que resulta esencial. La única variante que constituye la excepción es la PP, sin casos de malignidades asociadas reportadas [10].

La PP es una variante infrecuente de poroqueratosis que, a diferencia de las variantes más usuales, tiene una presentación clínica que se caracteriza por lesiones puntiformes hiperqueratósicas localizadas en palmas y/o plantas [5]. En el caso de nuestro paciente, el compromiso fue exclusivamente palmar. Clásicamente las lesiones se describen en "forma de semilla" (*seed-like)*, pudiendo ser solevantadas de forma espicular o más bien deprimidas en forma de *pits* [1,2]. Las lesiones suelen ser asintomáticas, pero pueden también ser sensibles a la presión [2]. Es notoria la existencia de una gran confusión respecto a la terminología a emplear para describir a la gama de patologías que cursan con estas lesiones palmo-plantares como manifestación clínica [3]. El diagnóstico diferencial ante este cuadro, utilizando la nomenclatura que nos parece actualizada y correspondiente, gira entorno a cuatro grandes grupos: la PP, las queratodermias espinosas, otras queratodermias punctatas y otras patologías de características distintivas [2,13,16,17]. Para comprender la confusión y los términos planteados, resulta imperiosa una revisión histórica de tales descripciones. De lo contrario resulta difícil tener una visión íntegra de la problemática.

En 1923, Sweitzer publica un artículo titulado "Keratoderma punctatum" [18]. En este, el autor describe un cuadro de hiperqueratosis punctata palmo-plantar y considera que ninguna de aquellas lesiones puede ser considerada una "poroqueratosis" hasta demostrar una relación con las glándulas ecrinas, característica considerada aún esencial en aquel entonces. En su revisión de la literatura, destaca como primera descripción de un cuadro similar un artículo de 1879, de Davies-Colley, donde se describe un cuadro clasificado como "*disseminated clavus of hands and feet*" ("clavos diseminados de manos y pies") [19]. Besnier, comenta el autor, describe un caso con lesiones hiperqueratósicas punctata en palmas, a lo que denominó "queratodermia eritematosa simétrica de las extremidades: forma punctata" [20]. Sweitzer termina su análisis con un comentario personal: expresa haber recopilado 18 casos, ninguno de ellos idénticos entre sí, evidenciando una nomenclatura que alternaba entre poroqueratosis y queratodermias. Algunos de esos casos, considera, probablemente eran verrugas o incluso liquen plano. El artículo de Sweitzer corresponde al primer incentivo explícito hacia la comunidad científica para indagar en estas manifestaciones clínicas difusamente comprendidas en su época.

Muchos años después, en 1971, Guss describe un "tercer tipo de poroqueratosis" [21], diferente a las descripciones hechas por Mibelli y por Chernosky (quien describe en 1967 la variante DSAP [22]). El

cuadro de Guss terminaría correspondiendo a la variante palmaris et plantaris disseminata. La define como un cuadro autosómico dominante, de inicio alrededor de los 20 años y de compromiso tronco-palmo-plantar, morfológicamente similar a la DSAP. También en 1971, Brown publica su artículo titulado "Punctate keratoderma" ("Queratodermia punctata") [23]. Este último ha sido considerado por diversos autores como el inicio de la confusión terminológica [3,16,17]. Brown comienza su artículo estableciendo que "la queratodermia punctata es un diagnóstico descriptivo que indica la presencia de pequeñas excreciencias puntiformes, cornudas, dispuestas irregularmente sobre palmas, plantas y la superficie flexora de los dedos". A la histología destaca una hiperqueratosis con acantosis; de haber paraqueratosis, dice, es mínima e incidental. Brown define su cuadro como una patología hereditaria autosómica dominante y reporta un caso del San Diego Naval Hospital: hombre de 20 años, con lesiones palmo-plantares, con claro componente familiar. Brown comenta que fue sugerido el diagnóstico tentativo de "poroqueratosis punctata", pero considerando la historia familiar, la edad de aparición y la falta de extensión periférica, decide finalmente considerar el caso como alguna forma de queratodermia punctata. Comenta no haber encontrado en la literatura ningún caso similar al que él se encontraba reportando. El mismo Brown relata que histológicamente le parece ver estructuras similares a la lámina cornoide de Mibelli.

En 1974, Herman expresa la necesidad de un mayor reporte de casos que permita esclarecer las diferencias observadas en estos cuadros descriptivamente denominados queratodermias punctatas [24]. Comenta casos con predominio de hiperqueratosis y otros de paraqueratosis, así como también casos tanto hereditarios como esporádicos.

En 1977, Rahbari postula de manera decisiva una nueva variante de la poroqueratosis de Mibelli: la poroqueratosis punctata, nombre ya utilizado en reportes previos, pero sin concretarse en un diagnóstico oficial [25]. A pesar de incorporar esta variante, destacando las lesiones de tipo *seed-like*, sus dos casos reportados no eran de ubicación exclusiva palmo-plantar. El primer reporte correspondía a una mujer de 16 años que presentaba lesiones en dedos y palma de la mano derecha y en axila derecha; el otro caso, un paciente de 59 años, presentaba lesiones en codo, muñeca y dedos. El autor considera que el caso de Brown correspondía a una poroqueratosis, y que el nombre no fue empleado tan solo por la falta de similitud clínica con la forma clásica de Mibelli, tal como el mismo Brown permite ver en su publicación. Rahbari considera que el diagnóstico diferencial de mayor dificultad radica en diferenciar tres entidades, en sus palabras: la PP, la queratodermia punctata y la queratosis arsenical.

Himmelstein publica el año 1984 lo que considera el quinto caso reportado de PP [26]. Paciente de origen hispano, de 26 años, con lesiones recurrentes puntiformes de 1-2 milímetros, de tipo *seed-like*, en palma y planta izquierda. A la histología se describe una columna paraqueratósica considerada como lámina cornoide. Himmelstein realiza una revisión de la literatura que evidencia el ya presente problema terminológico: considera que el primer caso reportado de PP correspondería al reporte de Herman de 1973 titulado "Queratodermia poroqueratósica punctata" [27], y se suma a Rahbari en considerar que el caso de Brown probablemente correspondía a una poroqueratosis.

Sakas publica en 1985 otro caso de PP, en un paciente de 60 años con ascendencia coreana, considerándolo el sexto caso reportado de contabilizarse al de Brown como el primero [28]. Sakas nota, no obstante, que de los 6 casos de PP, solo 3 fueron exclusivamente palmo-plantares. Por esta razón extiende el nombre de su diagnóstico a PP palmaris et plantaris. De gran relevancia nos resulta el hecho de que Sakas es uno de los primeros autores en destacar activamente la presencia de queratinocitos vacuolados en la histología de su paciente. En la revisión bibliográfica realizada por el autor, destaca el compromiso tanto de hombres como de mujeres, la tendencia a no ser hereditaria (con la excepción de 1 caso), y la ausencia de algún predominio racial. La edad tendería a la pubertad, pero no sería exclusiva de ese rango etario.

Friedmann comenta en 1988 que aún era necesaria una clasificación adecuada para estos casos [29]. El autor diferencia clínicamente las lesiones acuminadas, de tipo "caja musical", y las lesiones deprimidas, denominadas *pits*. En el caso de las lesiones solevantadas palmo-plantares, considera solo 3 o posiblemente 4 casos previos. A pesar de los reportes anteriores, Friedmann apoya la categoría de Wolff-Schreiner, de 1987, en la que no consideran a la PP como una entidad por sí sola, sino una variante de la poroqueratosis linear o de la forma clásica de Mibelli [30]. A los dos casos de su publicación se les atribuye histológicamente la presencia de lámina cornoide, pero en ausencia

de queratinocitos vacuolados y/o disqueratósicos a la microscopía de luz y electrónica, como ya se había descrito para las láminas cornoides de la forma clásica de Mibelli y de la DSAP [31,32]. Es por esto que Friedmann considera que sus reportes no son verdaderas poroqueratosis, catalogándolas más bien como queratodermias poroqueratósicas punctata, parte del grupo de lo que se denominó como dermatosis en caja musical (en inglés, *music box spine dermatoses)* [33]. Estas entidades que no satisfacían los criterios histológicos para ser denominadas verdaderas poroqueratosis tuvieron también sus respectivos cambios en términos de nomenclatura. Zarour realiza una clasificación en 1992 agrupándolas en lo que denomina queratosis filiformes [34] y luego McGovern y Gentry, en 1994, cambian aquel término por el de queratodermias espinosas (SK, del inglés *spiny keratoderma)*, diferenciando los cuadros paraqueratósicos de los ortoqueratósicos [35].

Basta con estos artículos señalados para comprender la dificultad histórica de la nomenclatura utilizada y la gran cantidad de términos propuestos. Consideramos que la revisión realizada por Urbani en 1998 ofrece un análisis comprensivo e integrativo de estos cuadros, plasmando y resumiendo un esquema aún vigente [36]. En su publicación diferencia categóricamente las PP de las SK en relación a la presencia o ausencia de queratinocitos vacuolados y/o disqueratósicos, respectivamente, utilizando como referencia el trabajo de Wade y Ackermann de 1980. Las SK además, a diferencia de las PP, no presentarían compromiso inflamatorio de la dermis, pero sí compartirían la disminución del estrato granular comprometido. Urbani comenta que en la literatura científica probablemente hay casos de SK descritos como PP y viceversa. Plantea además la hipótesis de 2 categorías de SK: una forma hereditaria que sería siempre benigna y una forma adquirida o idiopática que puede ser paraneoplásica o estar asociada a trastornos metabólicos. La asociación con distintas malignidades internas ha sido reportada particularmente en los casos idiopáticos de queratodermia poroqueratósica punctata (que pertenece a la categoría de SK paraqueratósica). Existen casos de asociación con carcinoma bronquial, cáncer de ovario, leucemia mieloide crónica, entre otros [13].

Tal como ya se ha expuesto, son varios los trabajos que describen casos de hiperqueratosis palmo-plantar puntiformes en los que histológicamente se ha descrito la presencia de una lámina cornoide, pero en ausencia del resto de los elementos encontrados en la poroqueratosis. Algunos, por el contrario, se limitan a describir una columna paraqueratósica sin una denominación específica [16]. En este aspecto consideramos relevante exponer la extensa revisión realizada por Biswas el año 2015. En esta, el autor establece que para denominarse lámina cornoide propiamente tal, 3 características se deben encontrar: 1) presencia de una columna vertical paraqueratósica, 2) pérdida o disminución del estrato granular en el punto donde la paraqueratina indenta la superficia epidermal, y 3) disqueratosis y/o vacuolización de células en el estrato espinoso que subyace a la columna paraqueratósica. Siguiendo este criterio, la presencia de lámina cornoide queda excluida de las SK y sigue constituyendo el elemento unificador, pero no patognomónico, de las poroqueratosis [1].

Ya hemos mencionado previamente la patogenia de la poroqueratosis; en el caso de las SK, Hashimoto realiza en 1999 un análisis de 6 casos utilizando anticuerpos antiqueratinas AE13 y AE14, además de apoyarse con microscopía electrónica. Las muestras obtenidas fueron positivas para AE13, marcador de queratina inmadura del pelo. Los resultados sugieren que las SK pueden constituir enfermedades de formación ectópica abortiva de pelos en palmas y plantas [37].

A partir del año 2000 se ha vislumbrado una clara tendencia a ocupar la nomenclatura actualizada de PP y SK, en especial destacándose la preferencia por el término SK por sobre otros términos históricamente utilizados [17]. Sin embargo, aún no existe una evidente homogeneización de la terminología [33]. Es importante recordar que ambas patologías son muy poco frecuentes y escasamente reportadas [1,17].

CONCLUSION

En conclusión, y utilizando la nomenclatura que consideramos apropiada, el diagnóstico diferencial de lesiones puntiformes hiperqueratósicas palmo-plantares, ya sea acuminadas o deprimidas, en forma de caja musical o en forma de semillas, lo constituyen los 4 grandes grupos que hemos mencionado con anterioridad: 1) la PP, 2) las SK, 3) otras queratodermias punctatas [36], y 4) otras patologías de características distintivas [1,16]. Ya hemos argumentado la nomenclatura de las PP y las SK. En el caso del grupo 3, recalcamos la importancia de utilizar el concepto de queratodermia punctata solamente a modo descriptivo, como fue

concebido originalmente, y no como una patología o una entidad en sí misma. En este grupo, otras queratodermias punctatas incluyen patologías como la queratosis arsenical, la enfermedad de Buschke-Fisher-Brauer y la acroqueratoelastoidosis liquenoide [16]. En cuanto a otras patologías de características distintivas, se puede señalar la enfermedad de Darier, la enfermedad de Cowden, verrugas, queratolisis punctata (*pitted keratolysis)* y el síndrome del carcinoma basocelular nevoideo [1,16].

La PP y las SK son patologías poco frecuentes con múltiples modalidades de tratamientos descritos para ambas, sin alguno particularmente establecido [13,14]. Más allá de las posibles molestias cosméticas o sintomáticas, resulta esencial el estudio histológico que permita un diagnóstico específico para el subsecuente seguimiento del paciente: en el caso de las PP por el riesgo teórico de CEC, y en el caso de las SK por las malignidades internas que se han visto asociadas.

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Our Dermatology Online

Case Report

Unusual presentation of psoriasis

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ABSTRACT

Psoriasis is a widely prevalent disease, a chronic disorder with genetic predisposition and many environmental triggering factors. It has various clinical presentations. Typical lesions are recurring, chronic, scaly erythematous papules and plaques. Clinical presentation differs among patients, from those with only few localized plaques over extensors to those with generalized skin involvement. We report a 28-year-old man who presented with unusual clinical presentation of psoriasis.

Key words: Psoriasis; Unusual presentation; Whorled presentation

INTRODUCTION

Psoriasis is an autoimmune polygenic skin disorder. Many triggering factors, e.g. infection, trauma or drugs, may elicit the disease in susceptible individuals [1].

The classic lesion is a well demarcated erythematous scaly plaque; these plaques can be limited or widespread in distribution. It is a dynamic disease; associated morphological changes and the development of a new lesion into an advanced plaque can slowly enlarge or remain static [2].

Here we discuss an unusual presentation of psoriasis that was managed as inflammatory linear verrucous epidermal nevus for years before reaching the diagnosis.

Case Presentation

A 28 -year-old Saudi male, without notable medical history, presented in 2008 with a chronic itchy scaly cutaneous lesion involving the trunk in whorled pattern for eight years. The lesion was stable with no changes for several years, not associated with joint pain or other systemic symptoms. Patient has a positive family history of psoriasis. He refused to have a skin biopsy at that time and diagnosed as inflammatory linear verrucous epidermal nevus (ILVEN)based on the clinical presentation, with no scalp or nail changes.

In 2013, patient presented again to dermatology clinic with new skin lesions involving extremities. Skin punch biopsy taken from the left side of the trunk and labs were done. Biopsy result showed mild acanthosis with epidermal spongiosis, hyperkeratosis with parakeratosis, elongated rete ridges and neutrophilic infiltrate in stratum corneum. In the dermis, the capillaries are elongated and tortuous, extending upward into dermal papillae. A minimal perivascular infiltrate is seen that consist primarily of lymphocytes. All Labs results were within normal range.

Patient managed as case of psoriasis, started on medium potent topical corticosteroid over active lesions (mometasone furoate ointment, 1 % twice daily)and topical calcipotriol (Diavonex cream twice daily). Narrow-band ultraviolet B light was offered and discussed with the patient but he refused.

Patient missed the follow up and presented again in 2016 reporting new skin lesions over the lower limbs.

By examination there are multiple erythematous whorled and linear scaly plaques involving both trunk and limbs respectively (Figs. 1 and 2). Scalp has

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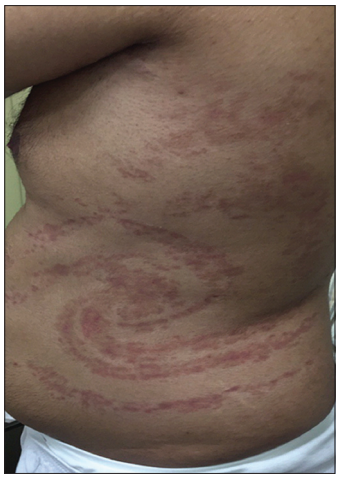

Figure 1: Note the whorled pattern of psoriatic plaques.

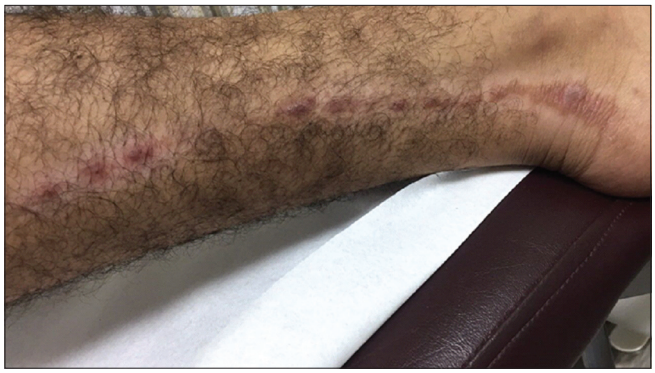

Figure 2: Linear arrangement of psoriatic papules and plaques that mimic ILVEN.

multiple yellowish well adherent scaly plaques over erythematous base mainly involving hair line. Nails have multiple deep irregular pitting and no Joints involvement.

Started on narrow-band ultraviolet B light three times \ week plus the topical medications.

Now he is following up in the dermatology clinic regularly with good subjective and objective improvement.

DISCUSSION

Psoriasis is a chronic inflammatory disease characterized by altered immune responses, with dysregulated production of proinflammatory and anti-inflammatory cytokines [3,4].

Typically has a chronic, persistent course with a reported incidence of 2-3% worldwide [5]. It can occur at any age; majority of cases occur before 35 years, 10-15% of new cases begin in children younger than 10 years. Psoriasis is characterized by well-defined scaly, erythematous cutaneous plaques that vary in extent from few patches to generalized involvement. Psoriasis impact on patient's life is significant [6].

The International Psoriasis Council identifies four main types of psoriasis: plaque-type, guttate, generalized pustular Psoriasis, and erythroderma. Other several subphenotypes according to distribution (localized vs. widespread), onset (early vs. late), size (large vs. small) and thickness (thick vs. thin)of plaques, anatomical localization (flexural, scalp, palms/soles/nail), and disease activity (active vs. stable) [2].

Plaque psoriasis is the most common form (around 90% of patient with psoriasis). Characterized by erythematous, well defined plaques with silvery micaceous scales. Plaques generally located on the extensor surface of knees and elbows, scalp, and in the lower back, but can affect any area of the body.

Guttate psoriasis is characterized by multiple to numerous scaly papules and plaques involving mainly the trunk and upper limbs. It has a sudden onset, within 2–4 weeks after a bacterial upper respiratory tract infection, mostly beat hemolytic streptococcal pharyngitis in pediatrics and adolescence age groups [2].

Generalized Pustular Psoriasis is a relatively rare but can be life-threatening disease. Characterized by widespread cutaneous involvement in form of pustules admixed with the psoriatic plaques and systemic manifestation like fever, lethargy, and neutrophils leukocytosis. Acute attacks can be triggered by infection, pregnancy, hypocalcemia, exposure to or withdrawal of medication.

Erythrodermic psoriasis represents one of the rarest form and it carries substantial morbidity and mortality [7].

Characteristic nail findings occur in 10-80% of psoriatic patients and are more common in people with psoriatic arthritis [8].

Typical histologic findings include hyper- and parakeratosis, acanthosis with regular elongated rete ridges, hypogranulosis, dilated blood vessels in papillary dermis and a perivascular infiltrate of lymphocytes with neutrophils singly or within aggregates in the epidermis [6].

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Our Dermatology Online

Case Report

Ichthyoses: Case series

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ABSTRACT

Ichthyoses are disorders of cornification in which abnormal differentiation and desquamation of the epidermis result in a defective epidermal barrier. They can be inherited or acquired. Skin changes are clinically characterized by hyperkeratosis or scaling or both. In this series we present 2 cases of collodion baby and 3 cases of lamellar ichthyosis.

Key words: Ichthyoses; Collodion baby; Lamellar ichthyosis; Acitretin

INTRODUCTION

Ichthyoses and erythrokeratodermas are disorders of cornification in which abnormal differentiation and desquamation of the epidermis result in a defective epidermal barrier. Ichthyoses represent a large clinically and etiologically heterogeneous group of conditions that feature generalized scaling of the skin. There are congenital and acquired forms of the disease. The congenital forms include lamellar ichthyosis (LI), non-bullous congenital ichthyosiform erythroderma, and Harlequin ichthyosis; the acquired forms include ichthyosis vulgaris and X-linked ichthyosis.

CASE REPORTS

Case 1

A term newborn infant presented after delivery due to the detection of peeling on his skin. The patient's prenatal and natal history was insignificant. The parents were 2nd degree relatives.

On Examination

Taut, shiny collodion membrane covering the baby all over the body with erosions and fissuring at the flexures (Fig. 1). Ectropion, eclabium, sausage shaped digits and flattened ears was also seen. Other examination findings were normal.

Diagnosis

Collodion baby.

The neonate was admitted in NICU and managed with IV fluids and prophylactic antibiotics. Topical emollients were regularly applied. Fucidic acid cream was applied over the erosions. Artificial tears were applied to prevent drying of eyes.

Case 2

A term newborn infant presented after delivery due to the detection of peeling on his skin. The patient's prenatal and natal history was insignificant. There was no history of consanguinity in the family and their other child was healthy. However there is history of 2 intrauterine deaths previously.

On Examination

Taut, shiny, membrane was present covering the baby all over the body (Fig. 2). Fingers, knuckles and feet showed blanching and pallor due to taut membrane. Erosions and fissuring was seen over the flexures. Severe ectropion and eclabium was present. Other examination findings were normal.

Diagnosis

Collodion baby.

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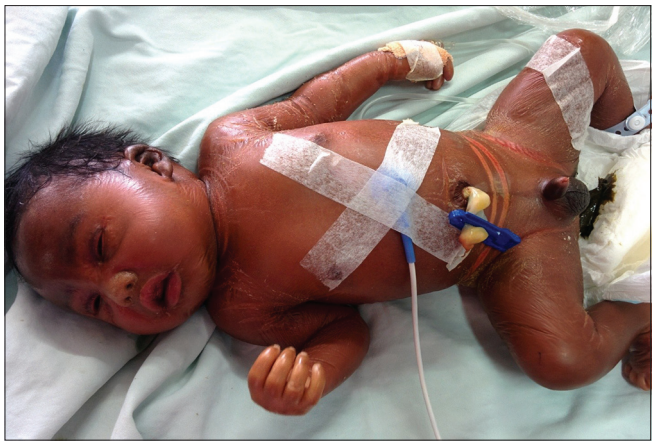

Figure 1: Taut, shiny collodion membrane covering the baby all over the body with erosions and fissuring at the flexures. Ectropion, eclabium, sausage shaped digits and flattened ears is also seen.

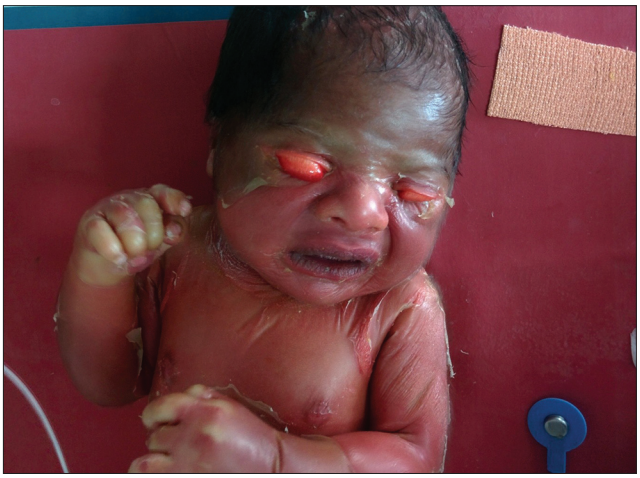

Figure 2: Taut, shiny, membrane seen covering the baby with severe ectropion and eclabium.

The neonate was admitted in NICU and managed with IV fluids and prophylactic antibiotics. Topical emollients were regularly applied. Fucidic acid cream was applied over the erosions. Artificial tears were applied to prevent drying of eyes.

Case 3

A 1 year old boy was brought to us with complaints of adherent scaling all over the body since 1 month after birth. The patient's prenatal and natal history was insignificant, with no history suggestive of collodion baby presentation at birth. Grade 4 consanguinity present in the parents. Patient's siblings are healthy.

On Examination

Multiple hyper pigmented, thick, adherent, quadrilateral scaly plaques were present all over the face, trunk, both upper limbs and lower limbs (Figs. 3a and 3b). Diffuse hyperkeratosis of the palms and soles was also noted along with generalized Xerosis. Rest of the findings were normal.

Diagnosis

Lamellar ichthyosis

The patient was treated with liberal application of emollients thrice daily. Fucidic acid cream was applied over erosions after some of the scales peeled off. Urea containing creams were applied to the scaly areas.

Case 4

A 24 year old women presented to us with complaint of dark scales all over the body since birth. History of periods of remission and aggravation of the disease was present. Her mother gives history suggestive of collodion membrane presentation in the patient at birth. There is 2nd degree consanguinity in the parents and grandparents as well.

On Examination

Hyper pigmented, brownish-black, thick, adherent scales all present over the trunk, scalp, palms, bilateral upper limb and lower limb (Figs. 4a and 4b). Scarring alopecia was noted over the frontal and occipital region with tufts of hair left in the vertex region. Rest of the examination was normal.

Diagnosis

Lamellar ichthyosis, confirmed with biopsy.

The patient was treated with oral Acitretin 25mg once daily to which the patient responded very well with near complete resolution of lesions 1 month after initiation of treatment. Supportive management in the form of emollients with urea combination for application on the trunk, twice daily. Glycolic acid 6% cream application over the face at night was also advised.

Case 5

An 8 year old girl presented to us with complaints of dark scales over both legs since 1 year. She also complained of generalized dryness of the skin and scaling over the scalp. There was history of 2nd degree consanguinity in the parents as well.

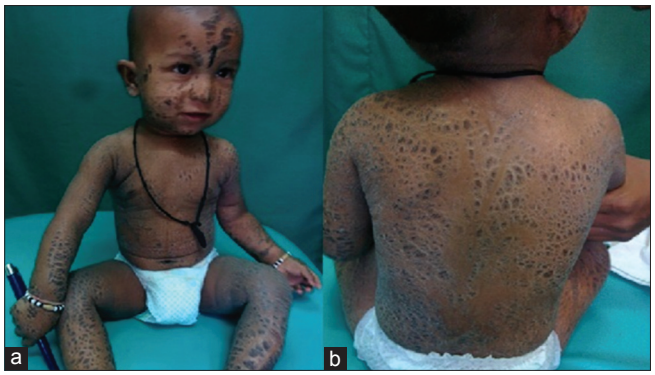

Figure 3: (a-b) Multiple hyper pigmented, thick, adherent, quadrilateral scaly plaques seen all over the face, trunk, both upper limbs and lower limbs.

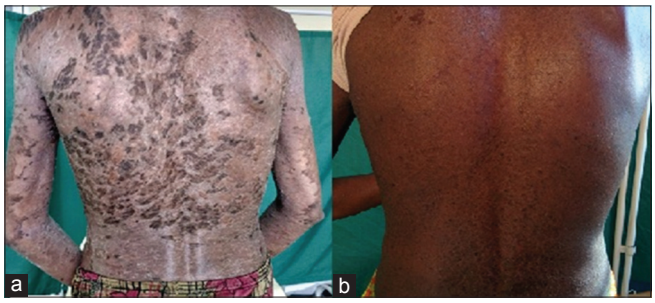

Figure 4: (a-b) Hyper pigmented, brownish-black, thick, adherent scales which cleared completely on treatment with Oral Acitretin.

On Examination

Thick, dark, brownish, adherent scales present over anterior and posterior aspect of legs bilaterally. Minimal scaling with diffuse hair loss over the scalp. Generalized xerosis and exfoliation present all over the body. Rest of the examination findings were normal.

Diagnosis

Lamellar ichthyosis, confirmed with biopsy.

The patient was treated with oral Acitretin 10mg once daily to which the patient responded very well. Liberal application of bland emollients thrice daily was also advised.

DISCUSSION

The term ichthyosis is derived from the Greek word 'Ichthys' meaning "fish" and refers to the similarity in appearance of the skin to fish scales. Early reports of ichthyosis in the Indian and Chinese literature date back to several hundred years. The ichthyoses form part of a large, clinically and etiologically heterogeneous group deshmukh4of Mendelian disorders of cornification and typically involve all or most of the integument [1].

Hallopeau and Watelet were the first who gave the term Collodion baby (CB). The skin of the newborn is replaced by a cornified substance, which gives the body a parchment like appearance or a varnished appearance. This condition is inherited primarily as autosomal recessive ichthyosis either lamellar ichthyosis (LI) or Non-bullous congenital ichthyosiform erythroderma (NBCIE). CB is an extremely rare dermatological condition with an estimated incidence of 1 in 50,000 to 100,000 birth. A new form of the disease has been notified as "self-healing collodion syndrome' in these cases newborn completely recovers within few months after birth [2].

These infants are born with a shiny, taught, transparent collodion membrane. The membrane starts to desquamate within one to two weeks. There is usually no erythroderma, and there is no involvement of the mucosa. Due to thickened skin structure and pulling of the soft tissues around the lips and conjunctivae, ectropion and eclabium develop, which are the common diagnostic symptoms in almost all cases [3]. In our case, ectropion and eclabium were both observed.

Due to impaired skin integrity, the most significant problems for which these infants are at risk are hyperthermia, increased insensible fluid loss and electrolyte disorders, skin infections, and sepsis [4]. Sucking and pulmonary ventilation may be hindered.

It is seen that the collodion membrane sheds off in next 2-4 weeks after birth revealing the underlying skin disorder. In long term course, approximately 75% of collodion baby cases will develop an AR Congenital ichthyosis (LI or NBCIE). In just 10% of these cases the membrane sheds off and underlying skin is normal for rest of the life termed as "Self-healing collodion baby". In rest 15% cases association with various entities is seen like ichthyosis vulgaris, trichothidystrophy, metabolic and endocrinal disorders which involve keratinization disorders gives rise to various ichthyosiform syndromes. The exact cause of the CB syndrome is not known but in most of the cases autosomal recessive inheritance pattern is seen and they are very rare and may be associated with consanguinity [5].

The first line of management is moisturizers and topical keratolytic agents, they enhances skin barrier and facilitate desquamation. Sodium chloride, urea, vitamin E acetate, glycerol and petroleum jelly are

various agents available as moisturizers and lubricants. In severe cases with marked hyperkeratosis keratolytic agents like lactic acid, glycolic acid, salicyclic acid, N- acetyl- cystine, and glycol can be used. Ectropion is managed by application of artificial tears and eye lubricants. In cases with severe ectropion surgical correction is done [6]. In severe cases Acitretin 0.5-0.75 mg/kg/day can be given.

LI follows autosomal recessive transmission; its frequency is 1/100,000. There is often a mutation in the gene encoding keratinocyte transglutaminase type 1 (TGM 1). TGM 1 encodes the TGAase 1 enzyme, which is one of the three TGAase enzymes found in the epidermis. This enzyme participates in the cornified envelope. In patients with TGM 1 mutation, the cornified envelope is missing, and TGAase 1 activity is reduced or nonexistent. More than 110 mutations have been reported [3]. Some of the other mutations include ABCA12, NIPAL4, ALOX12B, ALOXE3, CYP4F22, PNPLA1 (OMIM 615024) [7], and CERS3 (OMIM 615023).

Clinically patients with LI present as large dark brown firmly adherent scales over the scalp, limbs and abdomen mainly, but may involve other sites also. In severely affected patients, the thick rigid scales are intermittently shed causing deep painful fissures especially around the flexures and on the digits, palms and soles. Other features include limitation of joint movements, flexion contractures, digital sclerodactyly, palmoplantar keratoderma, scarring alopecia and persistent ectropion, congenital hypoplasia of nasal and aural cartilage and impaired sweating. The child has a normal growth and intellect. Severe forms of LI seldom improve with age and psychological problems resulting from cosmetic effects and limited mobility can lead to isolation, depression and poor school performance [7].

Treatment includes emollients, keratolytics, topical calcipotriol, 10% urea, topical N-acetyl cysteine and topical tazarotene 0.05% gel. In severe cases Acitretin 0.5-0.75 mg/kg/day.

Oral retinoids have keratolytic effects that help eliminate scales and prevent excessive hyperkeratosis. Acitretin is a synthetic analog of retinoic acid. Publications regarding its successful use in ichthyosis treatment have increased in recent years. Although its mechanism of action is not precisely known, it is believed that it controls the differentiation and proliferation of the keratinized epithelium. In order to prevent relapse, treatment can take several years for some patients. Long-term use of systemic retinoids has been reported to induce teratogenic and toxic effects in the bone tissue. Bone mineralization may also be affected, and calcification frequently develops in osteophytes and ligaments. Other side effects of oral retinoids include cheilitis, dryness in the mucous membranes, mild hair loss, and itching [8].

Through starting the 0.5 mg/kg/day dose of Acitretin in our cases, the skin symptoms were significantly improved during the second week of the treatment, and by the end of the third week, the patient's skin was close to normal. The patient's tolerance for the drug was good. Side effects were not observed.

CONCLUSION

These cases are being reported for their rarity and also to highlight the association of consanguinity in them and their excellent response to oral retinoids.

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Our Dermatology Online

Case Report

Pityriasis folliculorum colocalized with vitiligo: An example of immunocompromised cutaneous district

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ABSTRACT

The concept of immunocompromised cutaneous district suggests that different types of skin injuries can render the affected areas vulnerable to subsequent opportunistic infections, tumors, and immune reactions. Herein a 44-year-old woman with facial vitiligo associated with pityriasis folliculorum, a type of demodicosis, is reported. The right cheek with a large patch of vitiligo showed clinical, dermoscopic and microscopic evidents of demodicosis, while the other facial areas without vitiligo were devoid of Demodex mites. This case is another example of immunocompromised cutaneous district, probably developed due to immunologic alterations and increased ultraviolet light exposure caused by vitiliginous skin.

Key words: Demodex; Demodicosis; Pityriasis folliculorum; Rosacea; Vitiligo

INTRODUCTION

Pityriasis folliculorum (PF) is characterized by hyperkeratotic follicles filled with *Demodex* mites, which results tiny facial spicules [1,2]. Vitiligo is an acquired depigmentation disorder caused by complex influences on melanocytes, including genetics, autoimmunity, cytotoxic and oxidant-antioxidant mechanisms [3]. Herein a case of PF colocalized with facial vitiligo is described and the probable mechanisms of this association are discussed.

CASE REPORT

A 44-year-old woman complained of a milky white patch, dry skin and redness on the right cheek, present for 1 year. During the last 6 months white spots had developed also on the genital area and around the eyes. She had been suffering from panic attack disorder since 2009, and had treated with venlafaxine and olanzapin. Dermatological examination showed bilateral depigmented patches involving periorbital and malar regions, in addition to a large depigmented and erythematous patch on the right cheek. In this latter area follicular minute spicules were evident, causing a sandpaper feeling on palpation (Fig. 1) Examination under Wood lamp revealed accentuation of whitening over depigmented areas confirming the diagnosis of vitiligo (Figs. 2a and 2b). On the left cheek no depigmentation, erythema, or follicular spicules were noted, besides no accentuation with Wood light (Figs. 3a and 3b). Dermoscopy showed mites protruding from follicular orifices (Fig. 2c). Standardized skin surface biopsies taken from the areas showing PF revealed ample amount of Demodex mites, either as follicular bunchs of 4-7 mites, or freely floating isolated mites (Figs. 2d and 2e).

Laboratory investigations including complete blood cell count, TSH, anti-TPO, serum vitamin B12 level, and fasting glucose were within normal limits.

DISCUSSION

Demodex mites (*D. folliculorum and D. brevis*) are common commensals of the skin, typically found on the face. They are usually resident in the pilosebaceous unit at a density of ≤ 5cm^2. An increase in the number

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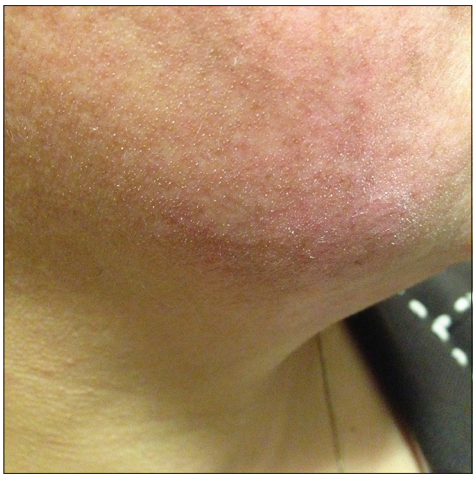

Figure 1: Follicular spicules and erythema overlying the depigmented patches

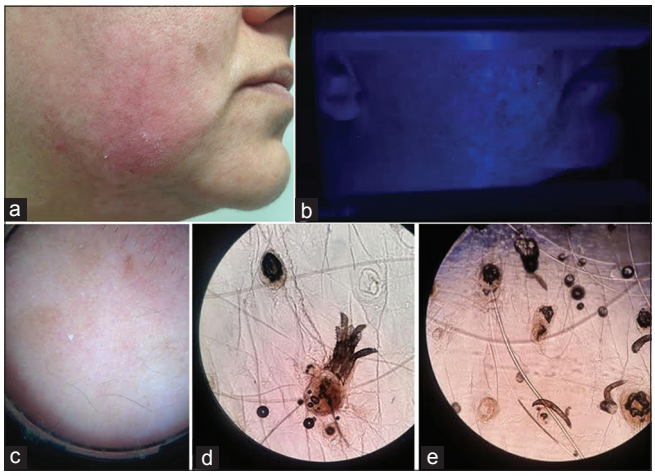

Figure 2: (a,b) White patches showing bright, blue-white fluorescence with sharp borders under Wood light; (c,d,e): Dermoscopy of the lesional area showing Demodex mites protruding from the follicular orifices. Superficial skin biopsy revealing profuse amount of Demodex mites.

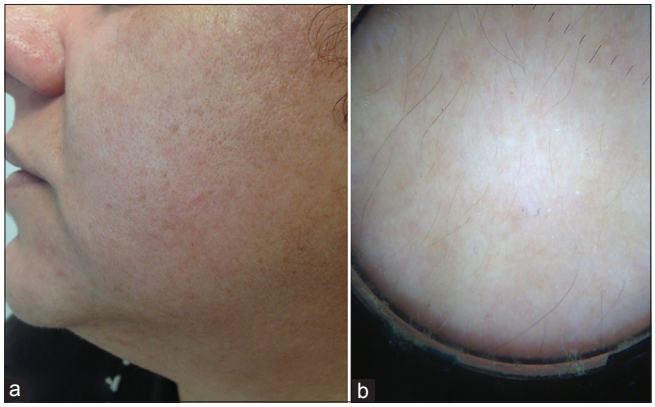

Figure 3: (a,b) Left cheek free from vitiligo and PF

of mites in the pilosebaceous unit, or less commonly, penetration of mites into the dermis is considered pathogenic [1]. Demodicosis is a term refers to chronic skin disorders induced by *Demodex spp.*, including pustular folliculitis, PF, rosacea-like demodicosis, and demodicosis gravis [2].

In PF the mites proliferate profusely with no, or a low immune response from the host. They plug the hair follicles and cause erythema and a prominent, sandpaper-like dryness. Tiny spicules composed of mite bunches are seen with naked eye and are very characteristic for this entity [2].

As few individuals develop symptoms in contrast to high prevalance of mites in general population, demodicosis can be considered as a multifactorial disorder, induced by external and internal factors. Immunosuppression may play a role on transition from an asymptomatic infestation to the development of clinical features. *Demodex* density has been reported to be high in immunosuppressive conditions such as HIV infection, chronic renal disease, diabetes mellitus, malnutrition, leukemia, cancer, corticosteroid or cytostatic therapy [2,4,5].

The presented patient had no history of systemic or local immunosuppresive treatment, including topical calcineurin inhibitors and corticosteroids. Her past medical history was unremarkable except vitiligo, emotional stress and psychiatric problems.

Vitiligo colocalized with PF has not been reported previously. This concurrence seems to be more than a simple coincidence, as the areas of the left cheek lacking vitiligo lesions were devoid of mites either.

The exact mechanisms of this colocalization are not clear yet. There is plenty of evidence for altered immunological processes in vitiligo, involving both innate and adaptive immune system. Oxidant stress and inadequate antioxidative defences leading to accumulation of reactive oxygen species also play important roles in the development of vitiligo [3,6]. It has long been known that ultraviolet (UV) radiation, in particular the UVB range, suppresses the immune system in several ways [7]. Loss of melanocytes in vitiligo increases the detrimental effects of UV. On the other hand, exposure to UV is an external stimuli that may trigger rosacea lesions [8]. In our patient increased UV exposure and insufficient defences due to vitiligo may have augmented the negative effects of UV on *Demodex* control [3,6]. Furthermore, an old hypothesis "locus minoris resistentiae (LMR)" has recently gained a new popularity by the concept of

immunocompromised cutaneous district proposed by Ruocco et al. It suggests that different types of skin injuries including UV radiation, can render the affected areas vulnerable to subsequent opportunistic infections, tumors, and immune reactions, due to a local immune imbalance [9].

CONCLUSION

In this context, vitiligo may be considered a LMR due to its susceptibility to UV damages, and immunologic alterations, leading to increased proliferation of mites.

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Our Dermatology Online

Case Report

Dermoscopic findings in extragenital lichen sclerosus

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ABSTRACT

Lichen sclerosus (LS), also known as lichen sclerosus et atrophicus is a chronic inflammatory dermatosis of unknown aetiology. It has both genital and extragenital presentations, nevertheless genital forms significantly outnumber extragenital LS. Dermoscopy is noninvasive diagnostic tool traditionally employed in pigmented lesions, however its usefulness in inflammatory skin conditions is becoming continuously more meaningful. Although the clinical diagnosis of fully developed LS rarely causes difficulties, unusual presentations require differentiation from the diseases such as lichen planus, morphea, extramammary Paget disease, SCC and others. In these cases histopathology contributes to the diagnosis. Studies on the use of dermoscopy in LS are sparse, nevertheless some dermoscopic features of LS has been described.

Key words: Extragenital lichen sclerosus; Dermoscopy; Lichen sclerosus

INTRODUCTION

Lichen sclerosus (LS)is a chronic, usually asymptomatic, inflammatory dermatosis that results in epidermal atrophy and scarring. It is prevalent in females with bimodal onset in prepubertal and postmenopausal age group [1,2]. It most often affects genital and perianal areas of postmenopausal women, however it can affect men and pre-pubertal children. LS is ten times more common in women than in men. Extragenital LS affects 10 % of women with vulval disease [3]. Penile LS is the leading cause of the phimosis in adult men [1]. Genital LS has been associated with a certain risk of squamous cell carcinoma (SCC), whereas extragenital LS does not appear to predispose to cancer. Cancer is estimated to affect up to 5 % of patients with vulval, penile or anal LS [3].

The exact aetiology of LS has not been ascertained yet, however, evidence points to an increased likelihood of autoimmune and genetic component. The most common autoimmune diseases associated with LS are autoimmune thyroiditis, alopecia areata, vitiligo and pernious anemia [3].

CASE REPORT

A 32-year-old woman with no previous medical history of autoimmune diseases, with multiple itchy, hypopigmented, atrophic, well-demarcated, brightening round and oval papules and plaques on the upper back since 1,5 year (Fig. 1). Hair, nails, oral mucosa and anogenital region were unaffected. Blood investigations did not show any abnormalities. The patient has not been treated yet.

The dermoscopic examination revealed white-yellowish structureless areas with comedo-like openings and sparse linear vessels in the centre of each lesion and an erythematous halo, which is a marker of activity in LS (DermLite DL4)(Fig. 2).

The histopathology of the lesion showed epidermal atrophy, hyperkeratosis, follicular plugging and basal vacuolization. The dermis showed oedema, initial homogenization of collagen, interstitial and perivascular lymphocytic infiltration (Figs. 3 and 4).

DISCUSSION

The clinical differentiation of LS and morphea, especially in extragenital regions, is a diagnostic challenge. White structureless areas and comedo-like openings are typically seen in LS [4], whereas fibrotic bands are characteristic of morphea. Nevertheless, comma shaped vessels, hairpin like vessels and dotted

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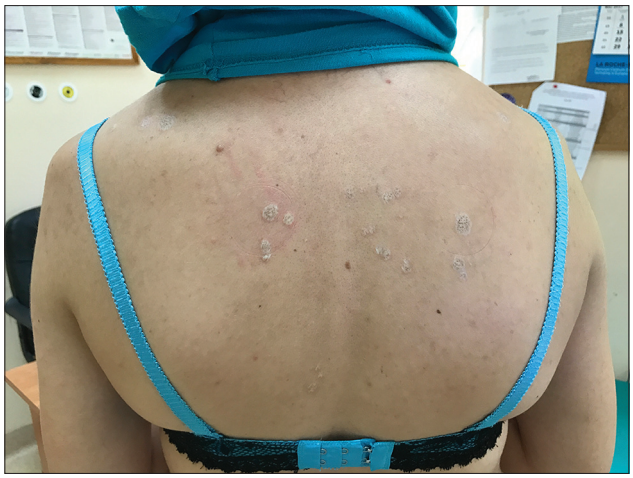

Figure 1: Multiple brightening hypopigmented round and oval papules and plaques on the upper back.

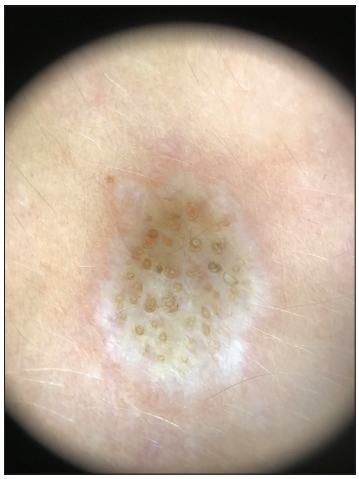

Figure 2: White-yellowish structureless area with comedo-like openings and sparse linear vessels in the centre and an erythematous halo.

vessels are usually absent in morphea and are seen only in LS [5].

Comedo-like openings in LS are predominat in early lesions, whereas white chrysalis like structures suggest homogenization of collagen in the dermis and can be seen only in late lesions. Long persisting lesions appear atrophic [2,6].

Dermoscopic structures in LS correlate with histopathology: white structureless areas are representing epidermal atrophy, whereas comedo-like openings are representing follicular plugging in histopathology [5].

Histopathology of morphea shows the continuity of the basal membrane zone (BMZ), whereas in LS numerous invaginations are present in BMZ [7].

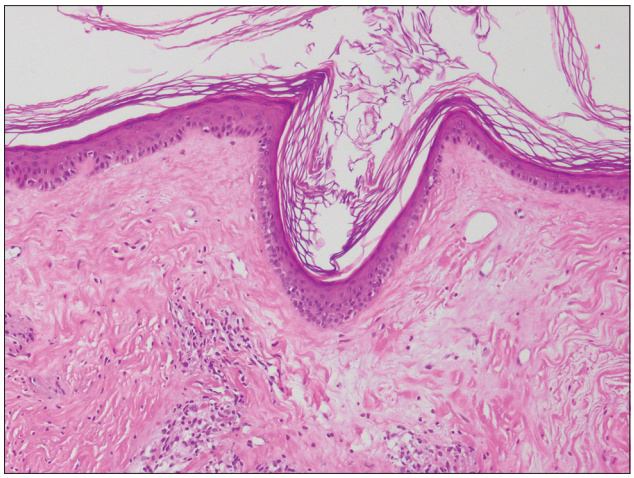

Figure 3: Lichen Sclerosus- epidermal atrophy, follicular plugging and basal vacuolization, initial homogenization of collagen in the dermis [HE x 20].

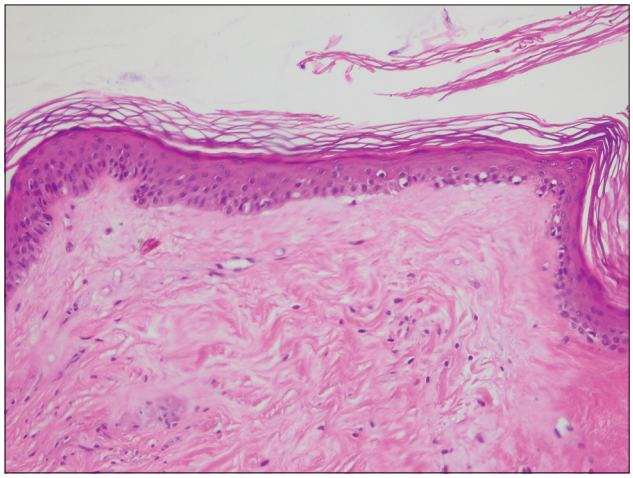

Figure 4: Lichen Sclerosus - epidermal atrophy, follicular plugging and basal vacuolization [HE x 20].

CONCLUSION

Although the clinical diagnosis of fully developed LS rarely causes difficulties and the disease is usually recognized by its appearance, early forms or unusual presentations require differentiation from other diseases including morphea, lichen planus, SCC and others. In these cases histopathology contributes to the diagnosis and is mandatory for any clinical situation in which co-existing SCC cannot be ruled out. Early diagnosis and treatment play a substantial role in patient's prognosis and result in decreased risk of malignancy and scarring.

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Our Dermatology Online

Case Report

Tattoo reaction: Case series

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ABSTRACT

Tattoo is going to be a very common practice especially among young people and we are witnessing a gradual increase of numerous potential complications to tattoo placement which are often seen by physicians, but generally unknown to the public. The most common skin reactions to tattoo include a transient acute inflammatory reaction due to trauma of the skin with needles and medical complications such as superficial and deep local infections, systemic infections, allergic contact dermatitis, photodermatitis, granulomatous and lichenoid reactions, and skin diseases localized on tattooed area (eczema, psoriasis, lichen, and morphea). In this series we present three cases of tattoo reaction.

Key words: Tattoo; Lichenoid reaction; Pseudolymphomatous reaction

INTRODUCTION

Tattoos are defined as the intentional or accidental deposit of pigment into the skin. Inadvertent use of variety of dye materials in the tattoos has led to a spectrum of histological reactions. It has tremendous religious and spiritual significance. In addition, tattooing for cosmetic purposes has become quite popular in recent times. With this increasing trend, there is also an increased risk of adverse effects. Cutaneous reactions to tattoos are uncommonly reported in literature. They are generally attributed to the metallic salts used in the preparation of the pigment. Allergic reactions to a particular pigment can manifest in several ways including allergic contact dermatitis and photoallergic dermatitis.

CASE REPORTS

Case 1

Thirty two year old male presented with raised lesions over his left hand one month ago which appeared five days after he got a tattoo which was associated with itching. Red tattoo pigment was used. He gives no history of pus exudation, fever or pain.

On Examination

A star shaped erythematous plaque with scaling over tattoo was seen. Mantoux test was negative (Fig. 1).

Biopsy was done and and the histopathological examination (Fig. 2) showed hyperkeratosis, acanthosis, hypergranulosis, and mild spongiosis in epidermis. Papillary dermis shows non specific mixed inflammatory cell infiltrate. Features were of pseudoepithelomatous hyperplasia.

A final diagnosis of Reaction to Tattoo pigment was given.

Patient was started with intralesional Triamcinolone injection which was given once in three weeks.

Case 2

Twenty seven year old female presented with a raised lesion over a six month old tattoo on her right hand which presented two months after she got a tattoo done. The tattoo was of red pigment and the lesion was associated with itching. She gave no history of fever, pain,cough or exudation of material from the lesion.

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On Examination

A raised erythematous plaque over the contour of the tattoo was present (Fig. 3). No discharge was observed. Mantoux was negative.

Biopsy was done and epidermis showed hyperkeratosis and acanthosis. Subepidermis showed round cell infiltrate with lakes of extracellular dye particles. The inflammatory cell infiltrate composed of lymphocytes and few macrophages with dye particles within. Features are suggestive of tattoo reaction.

A final diagnosis of Reaction to tattoo pigment was given.

She was started with intralesional Triamcinolone injection which was given once in three weeks.

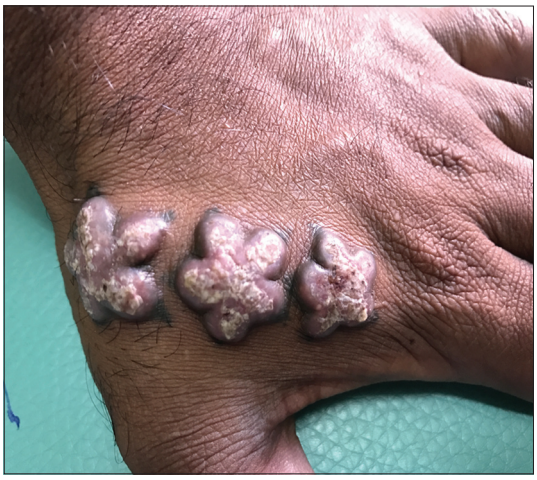

Figure 1: Star shaped erythemathous plaque with scaling over tattoo present over left hand.

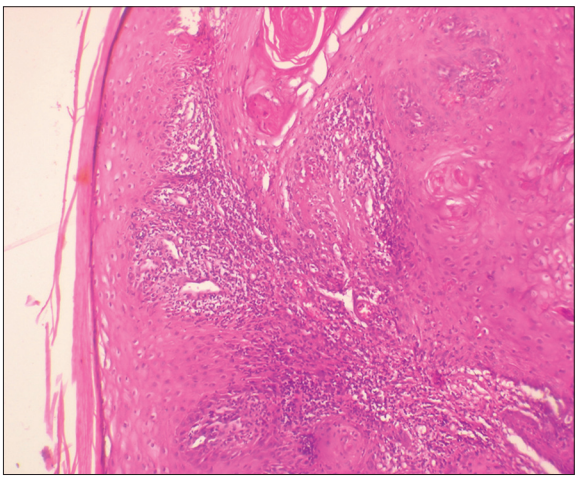

Figure 2: Epidermis- shows hyperkeratosis, acanthosis, hypergranulosis, and mild spongiosis. Papillary dermis shows non specific mixed inflammatory cell infiltrate.

Case 3

A twenty two year old male presented with redness and itching over the tattoo site since one month. Tattoo was done three month back over the right side of neck.

On Examination

A single star shaped erythematous plaque present over the right side of neck with minimal scaling was present (Figs. 4a and 4b). Mantoux was negative.

Biopsy was done and epidermis was unremarkable. Upper dermis shows dense lymphohistiocytic infiltrate around the blood vessels extending in to the deep dermis. The histiocytes contain pigment in the cytoplasm. Features are suggestive of tattoo reaction.

A final diagnosis of reaction to tattoo pigment was given.

Patient was started on intralesional triamcinolone injection which was given once in three weeks.

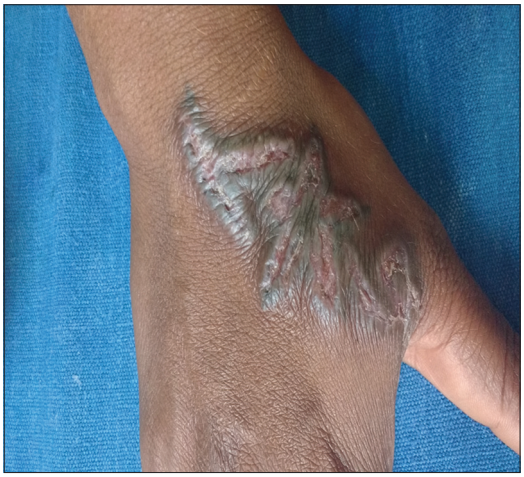

Figure 3: Raised erythematous plaques over the contour of the tattoo over right hand.

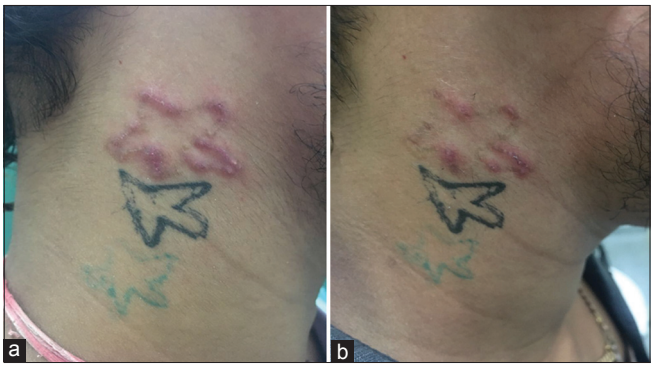

Figure 4: (a) Single star shaped erythematous plaque present over the right side of neck with minimal scaling over red colored tattoo. (b) Shows improvement after 3 sittings of Intra lesional steroid.

DISCUSSION

Tattoos are defined as the intentional or accidental deposit of pigment into the skin [1]. Inadvertent use of variety of dye materials in the tattoos has led to a spectrum of histological reactions. They are also plagued by complications like bacterial, viral, mycotic infections, allergic disorders as well as risk of tumours like lymphoma, basal cell carcinoma, squamous cell carcinoma, keratoacanthoma among others. Tattoo colors consist of inorganic pigments, organic dyes, or a combination of both [2]. In the past, it appears that heavy metals, that were the backbone of tattooing for decades, have been replaced by organic colorants. Tattoo artists use various pigment compounds to create different colors and hues. Depending upon the compounds used and the color of the tattoo, a variety of cutaneous reactions can be expected. The composition of ink used for professional and amateur tattoo differs significantly. For amateur tattoos, carbon particles are used, while for professional tattoos, a mixture of insoluble metals with organic dyes is used. Histological reactions to tattoo ink have been described including pseudolymphomatous, lichenoid, granulomatous, scleroderma or morphea-like, sarcoidal, pseudoepitheliomatous hyperplasia, allergic contact dermatitis and photoallergy. Lichenoid and sarcoidal reactions are both less common than eczematous reactions [3]. Pathogenic mechanisms implicated in reactions to tattoo pigments include a localized, T-cell mediated, delayed hypersensitivity response (lichenoid and sarcoidal reaction). In addition, allergic reactions have been observed in the form of type I and III reactions, according to Coombs and Gell classification. Cutaneous hypersensitivity reactions, although most common with red (mercuric sulphide) tattoos, have also been reported with other colors like yellow (cadmium sulphide), brown (iron oxide), blue (cobalt), purple (manganese), green (chromium), and black (carbon)tattoos.

Allergic reactions are more frequently seen to red tattoos than other colors. This is reflected in a literature review of 17 case reports by Aberer et al., which showed that red ink was responsible for 11 out of 26 reactions reported. These reactions may occur within days of the tattoo application or up to 17 years later. Lichenoid reactions are more frequently reported with red pigment tattooing that contains mercury. Clinically, verrucous papules or plaques characteristic of hyperkeratotic lichen planus are usually seen. Currently data is lacking regarding the safety of tattoo pigment ingredients. Also, none of the tattoo ink or additives are FDA approved [4].

Steroids, laser therapy, and excision are the backbone of treatment for allergic reactions to tattoos. Other methods are mechanical dermabrasion, cryosurgery and application of caustic chemicals.

Various lasers used are Argon, carbon dioxide and Q switched lasers.

CONCLUSION

These cases are being reported owing to various reactions occurring to tattoo and fewer availability of study on the same.

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Our Dermatology Online

Case Report

A crushing ulcerous lesion of the internal angle of the right eye: Cutaneous leishmaniasis is one of the causes

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ABSTRACT

Niger is a tropical country where leishmaniasis is endemic. The first case was reported in 1911. *Leishmania major* is practically the pathogen found in Niger, a country lying between 8° and 20° north latitudes from the Atlantic to the Chadian border. We report a case of cutaneous leishmaniasis in the type of ulcerative lesion, characterized by its localization at the internal angle of the right eye posing a diagnostic problem. The diagnosis of leishmaniasis was made by parasitological examination. Anatomipathologic examination eliminated cutaneous tuberculosis, pyogenic granuloma, molluscum contagiosum and basal cell epithelioma. Metronidazole management has accelerated healing. Thus in a tropical country, in front of any chronic, painless ulcerative lesion and resistant to all therapeutics, the diagnosis of cutaneous leishmaniasis must be evoked, for a consequent management after confirmation.

Key words: Cutaneous leishmaniasis; Internal right eye angle; Niger

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Case Report

Lésion ulcérocroûteuse de l'angle interne de l'œil droit: la leishmaniose cutanée en est une cause

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RÉSUMÉ

Le Niger est un pays tropical où la leishmaniose sévit de façon endémique. Le premier cas a été rapporté en 1911. Leishmania major est pratiquement l'agent pathogène retrouvé au Niger, pays situé dans une bande comprise entre les 8° et 20° latitudes nord allant de l'Atlantique à la frontière tchadienne. Nous rapportons une observation de leishmaniose cutanée à type de lésion ulcérocroûteuse, caractérisée par sa localisation à l'angle interne de l'œil droit posant un problème de diagnostic. Le diagnostic de leishmaniose était posé par un examen parasitologique. L'examen anatomopathologique a permis d'éliminer la tuberculose cutanée, le granulome pyogénique, le molluscum contagiosum et l'épithélioma basocellulaire. La prise en charge par le métronidazole a permis d'accélérer la guérison. Ainsi dans un pays tropical, devant toute lésion ulcérocroûteuse chronique, indolore et résistante à toute thérapeutique, le diagnostic de la leishmaniose cutanée doit être évoqué, pour une prise en charge conséquente après confirmation.

Mots clés: Leishmaniose cutanée; Angle interne œil droit; Niger

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INTRODUCTION

Le Niger est un pays tropical où la leishmaniose sévit de façon endémique. C'est une parasitose transmise à l'homme et à certains animaux par la piqure d'un insecte hématophage, le phlébotome. Les formes cutanées sont fréquemment vues en consultation surtout à type de lésions ulcérées ou ulcérocrouteuses [1]. Le parasite quand il est mis en évidence est le *Leishmania major* qui également fait parti de ceux qui sont en cause de la leishmaniose cutanée dans le Mahgreb tels que: le *Leishmania infantum* et le *Leishmania tropica* [2]. Nous rapportons une observation d'une localisation au niveau de l'angle interne de l'œil droit.

OBSERVATION

Il s'est agit d'un garçon de 7 ans qui présentait une lésion ulcérocroûteuse de l'angle interne de l'œil droit évoluant depuis plus de 6 mois, sans notion de traumatisme. Cet enfant vivait dans un des quartiers périphériques de la communauté urbaine de Niamey (capital du Niger) où les patients atteints de leishmaniose cutanée nous parviennent. Devant cette évolution chronique et le caractère réfractaire aux traitements locaux et par l'antibiothérapie générale (amoxicilline en particulier), nous avons évoqué le diagnostic clinique de la leishmaniose cutanée. C'était une lésion de 1 cm de diamètre, à toit ulcérocrouteux reposant sur un nodule inflammatoire (Fig. 1). Cette lésion était indolore mais inquiétante, car augmentant de taille progressivement. Sur le plan général, il n'y avait pas de fièvre, ni d'altération de l'état général. La recherche d'adénopathie ou autre organomégalie était négative. La numération formule sanguine était normale. L'examen parasitologique mettait en évidence la présence de leishmanies amastigotes sans spécifier l'espèce. La recherche des BAAR était négative. L'examen anatomopathologique montrait un infiltrat inflammatoire granulomateux épithéloïde et gigantocellulaire sans nécrose caséeuse, ayant permis d'éliminer: une tuberculose cutanée, un granulome pyogénique, un molluscum contagiosum et autre processus malin. Le traitement par métronidazole en raison de 25 mg/Kg/jour en deux prises, pendant 8 semaines a accéléré la guérison en laissant une cicatrice indélébile (Fig. 2).

DISCUSSION

La leishmaniose est une maladie parasitaire caractérisée sur le plan clinique par des lésions papulonodulaires

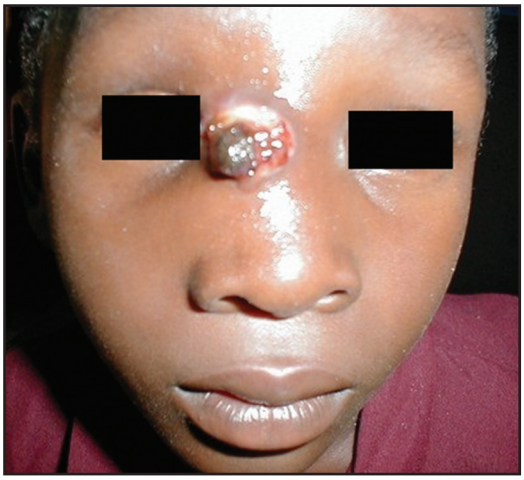

Figure 1: Lésion ulcérocroûteuse avant le traitement.

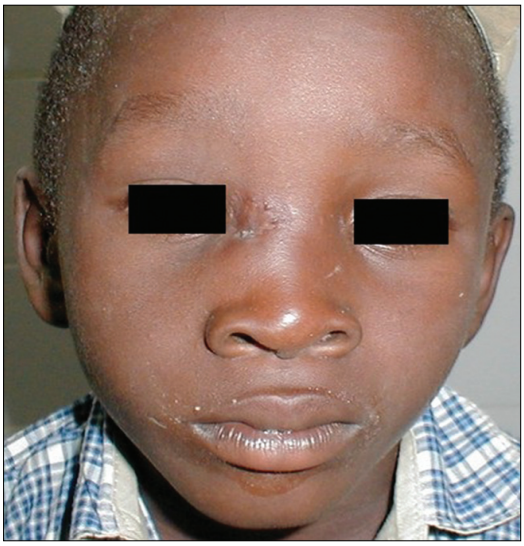

Figure 2: Guérison avec cicatrice indélébile après le traitement.

qui évoluent le plus souvent vers une ulcération ou une ulcération crouteuse [3,4]. Le caractère clinique de cette lésion observée chez notre patient à type d'ulcération crouteuse est classique de celui des données de la littérature dans leur nombre et leur localisation [5-7]. Cependant cette localisation au visage et précisément à l'angle interne de l'œil droit est rare, faisant évoquer d'autres pathologies telles que: lymphome cutané, la tuberculose cutanée et le granulome pyogénique. Les espèces des parasites mis en évidence sont variables [8,9]. Dans cette observation l'espèce n'a pas été déterminée; Il ressort cependant des travaux de certains auteurs [1,10] que les espèces de leishmanies isolées seraient de type *Leishmania major*, du fait que le Niger est inclus dans le foyer ouest-africain où les rongeurs constituent le réservoir à cet agent responsable de cette zoonose. L'effet du

métronidazole est attesté depuis la première étude mondiale au Mexique sur *Leishmania mexicana*, malgré que ce produit ne constitue pas une indication dans le traitement de la leishmaniose [11]. Les résultats obtenus à partir de cette étude mexicaine ont incité d'autres travaux sur le *Leishmania major* avec des résultats variables [11-14]. Certains auteurs défendent l'abstention thérapeutique, étant donné l'évolution vers la guérison spontanée dans plus de la moitié des cas en moins de 3 mois [15]. De nouveaux traitements ont été rapportés mais avec une variabilité des résultats et une taille petite des séries [16,17]. Différentes études au Niger utilisant le kétoconazole [10] l'antimoniate de méglumine [1] ont montré leur efficacité. Dans cette observation tout comme dans un travail que nous avons effectué [18], nous avons obtenu une accélération de guérison avec le métronidazole en 8 semaines. Le métronidazole accélère la guérison dans certains cas, après l'échec de traitement par l'antimoniate de méglumine [19].

CONCLUSION

La localisation à l'angle interne de l'œil d'une lésion de leishmaniose est rare; dans tous les cas toutes lésions ulcérocroûteuse indolore, chronique et résistante aux soins locaux et traitement général par antibiothérapie, doit faire évoquer une leishmaniose. Le métronidazole fait partie des molécules qui accélèrent la guérison.

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Our Dermatology Online

Opinion Article

How to defeat male pattern alopecia in a trompeur de femmes, who loves to abuse of libido boosters?

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Sir,

No particular expertise is required to recognize the alopecia androgenetica in male [1], for the recession of the hair line, loss of hair from the crown and the bald pate are more than familiar. In the affected areas the hairs become steadily shorter and finer and finally cosmeticalally useless.

Almost a third of the follicles may disappear at all and the reduction in the lenght of the growing periods reflected in the increased ratio of telogen to anagen in samples of plucked hair.

Alopecia androgenetica is commonly inherited, as an autosomal dominant trait, but always it manifests only in presence of male hormons.

Eunuchs retain their scalp hair even when they have a family history of baldness, unless they are treated with testosterone [2].

Though a correlation with hairiness of the chest has been suggested [1], baldness does not seem to be associated with other indices of masculinity, such as sebum secretion, muscle size and body hai in general [3].

It is suggestive to notice that bald scalp has a greater capacity to convert 5α dehydrotestosterone than non-bald scalp [4], anyway the key to understand the phenomenon of baldness lies in the field of steroid metabolism, even if hypothesis of administering female hormons to men suffering from male pattern alopecia is not plausible.

Manifold trials with 5 alpha reductase inhibitors have been proposed even most recently (it is supervacaneous to stress that the route of administration should be the topical one), for instance:
Zinc hydrosoluble salts [5]
Riboflavin (vitamin B2) [6]
Azelaic acid
β-sitosterol (and many other phytosterols as well)
Polyphenols [7]
Alizarin (from Rubia tinctoria)
Curcumin
Green tea catechins, including (-)-epicatechin-3-gallate, and (-)-epigallo-catechin-3-gallate (EGCG) [8]

Valoneic acid dilactone and gallagyldilactone are two hydrolysable tannin polyphenols isolated from the heartwood of Shorea laeviforia [9] and oaks species such as the North American white oak (*Quercus alba*) and European red oak (*Quercus robur*) are inhibitory [10].

Angelica koreana

Garden Balsam or Rose Balsam (*Impatiens balsamina*)
Pollen of Turnip, turnip rape, fast plants, field mustard, or turnip mustard (*Brassica rapa*)

Dodder (*Cuscuta reflexa*)
Euphorbia jolkinii
Lingzhi mushroom or Reishi mushroom (*Ganoderma lucidum*)
Ganoderic acid or Ganoderol B are thought to be the compounds in the mushroom that are specifically active.
Chinese Knotweed (*Polygonum multiflorum*), contains resveratrol-like Stilbenoids.
Black Pepper leaf extract (*Piper nigrum*)
Red Stinkwood (*Pygeum africanum*)
Saw Palmetto (*Serenoa repens*, active substance possibly lauric acid [11].

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The berries of saw palmetto (Serenoa repens), a small palm native to the south east United States, possess a dual 5a-reductase inhibition activity, due to their high content of phytosterols: β-sitosterol, stigmasterol, lupeol, lupenone, and cycloartenol. Permixon® was launched in Europe in 1984 but has no FDA approval. The lipido-sterol extract markedly inhibits both the human isoenzymes. Type 1 isoenzyme is noncompetitively (Ki = 7.2 μg/mL) and type 2 isoenzyme uncompetitively (Ki = 4.9 μg/mL) inhibited.

Pine (*Pinus sp.* resin, active substance abietic acid)
Japanese hedge parsley (*Torilis japonica*)
Eastern Arborvitae, Northern Whitecedar (*Thuja occidentalis)*
Spore of Japanese climbing fern (*Lygodium japonicum*)

Certain unsaturated fatty acids keeping on account that the relative inhibitory potencies are, in decreasing order: Gamma-Linolenic acid, alpha-linolenic acid, linoleic acid, palmitoleic acid, oleic acid, and myristoleic acid [12].

Medium chain fatty acids such as those found in coconut and the kernel of many palm fruits.

We have recruited a man (49 y. old) who likes to be defined a "latin lover" as he has been always a trompeur de femmes since he was a pre-pubertal child and observed the very first manifestations of alopecia androgenetica when he was 21 y. old.

He uses all the types of possible aphrodisiacs to induce in himself an increasing appetite for sex intercourses (yohimbine, damiana turnera extract, sildenafil and tadalafil compounds at massive dosages that may permit him to have 4 or more sexual encounters for day) with diverse women.

He is accustomed too to spread onto his poor hair scalp hairdressing gels and brilliantines, that are considered [13] exceptional inducers of baldness in man.

We prayed him to massage onto his scalp affected by baldness twice a day a cosmetic containing four 5 alpha reductase inhibitors, dispersed in the Macassar oil (an ancient formula by Dr. Rowland), that foresees the presence of essential oils of cloves, cynnamon, rose églantine and alkanna concrete in olive oil.

The 5 alpha reductase inhibitors were:
- betasitosterol (1%)
- green tea catechins (2%)
- black pepper oil (piper cubeba seed oil) (1.5%)
- curcumin (0.5%).

The experimentations lasted three months, and I have always wanted to be sure that he used the remedy We gave him everyday.

Amongst the manifold ways to clinically state the degree of hair re-growth, We have choosen the histological H&E test, efefctuated by the aids of a microscope.

It is known that Hematoxylin dyes the cellular nuclei blue, while eosin dyes the non-nuclear cells and structures pink/orange, such as cytoplasm and collagen. Combined, these dyes assist in recognizing the basal cells or germ layer: Cells rest on the dermis; blue nuclei are visible and this is where mitosis occurs and cells divide.

This clearily represents the phenomenon of the restoring of matrix of the hair follicle.

It is mandatory to effectuate a previous histological test, and to repeat it picking some hair from the scalp every 2 weeks for all the period of the experimentation.

The very first microscopic evaluation on hair revealed that, due to testosterone, numerous hair follicles were in the telogen phase as they showed characteristics of telogen follicles i.e. being short and hollow, presence of necrosis, more destroyed follicles, follicle shrinkage means diameter decreases and not deeper. Just after the first week the number of follicles in anagen phase was considerably increased and the number of follicles in telogen phase was decreased because surely the cosmetic item has inhibited the action of testosterone on hair follicles. The increase in the number of hair follicles was also noted. At the evaluation of the second week follicles showed characteristics of anagen follicles i.e. longer follicle hair and follicles were dense, less cell necrosis, and present deeper.

It can be asserted that the number of follicles in hair growth phase increased with duration of treatment. The hair follicle density and anagen/telogen ratio (A/T ratio) was calculated and plotted in the following Table 1.

RESULTS

It is evident that after only two weeks of treatment one may assert that hair restoration is almost complete, as far as the anagen/telogen ratio is concerned.

Table I: Values scored after one and two weeks of experimentations by H and E test on hair picked from hair scalp of the volunteer

Day of the performing of H and E tests	Hair follicular density (no./mm)	Anagen to telogen
First evaluation (at day 0)	1.5±0.90	1:3.16
After 1 week (7th day)	3.3±0.77*	1.22 :1
After 2 weeks (14th day)	2.75±0.75**	1.12:1

The Savin scale is a objective method of investigation on the hair re-growth (it is used generically for testing the hair re-growth women) but I attempted to use it even for my volunteer.

The scale ranges from degree III (complete baldness of the pate) to II-2, II-1, I-4, I-3, I-2 and finally I-1 (complete restoration of hair scalp).

We could affirm that after two weeks the degree II-1 is reached and after two months (8 weeks of treatment) degree I-2 is achieved completely.

As far as the blood testosterone, the first measurements reveal an amount of 1103 ng/dl meanwhile at the end of the experimentations, this value decreased to 256 ng/dl

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Our Dermatology Online

Review Article

Aesthetic practitioner as a physician and businessperson – Is it achievable?

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ABSTRACT

Aesthetic medicine subspecialty is no longer limited to the fields of plastic surgery and dermatology, as many specialties are offering aesthetic medical procedures to better accommodate their patients' aesthetic needs. During the last decade there is an enormous increase of cosmetic treatments worldwide and the absence of regulations in aesthetic practice has become more noticeable. This article illustrates the challenges that every aesthetic physician must overcome daily to maintain high ethos and make decisions according to patient's best interest and safety. Aesthetic practitioner should be more than a businessperson with a medical degree. If they place their integrity and moral standards to the test, then they will continue to be judged by their colleagues and society. Is it possible to maintain the same moral obligations during cosmetic procedures as with any other medical treatments?

Key words: Aesthetic dermatology; Cosmetic dermatology; Ethics; Malpractice; Safe practice; Good clinical practice

INTRODUCTION

"A physician shall be dedicated to provide competent medical service in full professional and moral independence, with compassion and respect for human dignity" as it has been described by Word Medical Association International Code of Medical Ethics [1]. This article will explain the challenges that every aesthetic physician must overcome daily to maintain high ethos and make decisions according to patient's best interest and safety. Aesthetic medicine subspecialty is no longer limited to the fields of plastic surgery and dermatology, as many specialties are offering aesthetic medical procedures to better accommodate their patients' aesthetic needs [2]. How difficult is for all the aesthetic practitioners to set their priorities straight? People are willing to spend a lot of money for staying forever young having the perfect facial skin and body. Therefore, in the western world, financial incentives for the aesthetic practitioners can be dangerous against ethical practices [3].

In UK, cosmetic interventions are booming business, which worth of £2.3 billion in 2010, and it was estimated to rise to £3.6 billion by 2015 as it was reported by Professor Sir Bruce Keogh KBE [4]. On the other side of the Atlantic Ocean, American society for aesthetic plastic surgery reported more than $13.5 billion spent for the first time ever in the year of 2015, with surgical procedures accounted for 58% of the total expenditures, and nonsurgical procedures accounted for 42% [5]. A comprehensive analysis, which was published in USA in August 2016, indicated that the current medical aesthetic market was worth $52,405 million for both service and product revenue, which over the next five years was expected to grow at 5.5% Compound annual growth rate (CAGR) and to hit $69,786 million in 2021. The invasive aesthetic procedures were set to grow at a CAGR of 4.3%, while the non-invasive ones were supposed to reach a CAGR of 4.5% over the coming five-year period [6]. Therefore, due to this increase of cosmetic treatments worldwide, the absence of regulations in aesthetic practice has become more noticeable. In fact, in the review document regarding the regulation of cosmetic interventions in U.K., the committee was surprised to discover that non-surgical interventions, which can have major and irreversible

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adverse impacts on health and well-being, are almost entirely unregulated [4].

THE ROLE OF AESTHETIC PRACTITIONER AS A PHYSICIAN

Physicians, who choose to work in aesthetic areas, should maintain a high standard of safety. Patients are more demanding and treatments are embellishments, so there is little tolerance for poor results and complications are less acceptable [7]. Prendergast mentioned in his book that there is no "medical" indication for the procedure and the physician is only responsible for proposing a method by which the patient's goal should be achieved [1]. Patients must be able to trust that their physicians are always acting in the patients' best interests [7]. The Council of Medical Speciality in U.K. states that "*physicians should treat each patient with honesty, compassion, dignity, respect for individual autonomy and educate them about health issues*" [8].

The role of physician demands on their cautious judgment guide about the different aesthetic treatments that can be provided to patients, rather than simply yielding to patients' demands [7]. Unfortunately, the patient's conception depends to media advertising resulting to exaggeration of patient's expectations on aesthetic results of procedures. Therefore, legal claims are higher than other surgical operations, especially if physicians do not respect patient's autonomy by not explaining treatment process and outcomes [9]. The initial consultation must not serve the purpose of "selling" an aesthetic procedure, but placing the interests of patients above those of the physician is one of the fundamental principles of professionalism [1,10].

The principle of informed consent tends to reflect the concept of autonomy of the person requiring and requesting medical and/or surgical interventions [11]. Seven criteria relate to an informed consent: (i) competence to understand and to decide, (ii) voluntary decision making, (iii) disclosure of material information, (iv) recommendation of a plan, (v) comprehension of terms and (vi) decision in favour of a plan, and (vii) authorization of the plan [9]. A person, who is a candidate for an aesthetic procedure, must complete all the criteria in order to give an informed consent, and every aesthetic physician must make sure patients are given enough time and information before they decide whether to have an intervention [12]. If the patients ask for a cosmetic treatment that the physician considers unnecessary and/or dangerous, then it is physician's obligation to discuss the issues and explore the reasons for their request [13]. Always, it is important during the first consultation to consider the patients' psychological needs and if necessary to seek expert advice from colleagues prior to any intervention [12]. It is extremely challenge for every aesthetic practitioner to go ahead with an unnecessary treatment since the financial incentives outweigh his/her decision.

Aesthetic practitioners should be open and honest about their skills, experience, fees and conflicts of interests [12]. Attendance only at a training course is insufficient to become competent in a procedure, whether this is a surgical or a non-surgical procedure. As it is mentioned in the Professional standards for cosmetic practice, direct practice is also necessary. Therefore, it is highly recommended that all practitioners undertake a period of formal or informal mentorship and they must keep their skills up to date by undertaking relevant continuing professional development [13].

THE ROLE OF AESTHETIC PRACTITIONER AS A BUSINESSPERSON

A businessperson's main objective is to maximize profit by increasing productivity and minimizing costs, but an aesthetic practitioner must not follow this goal, as this would have negative implication on the quality of the service offer [14,15]. The role of the dermatologist, for example, is transforming from a traditional healer into one of a businessperson, who will promote "big ideas". "Bluffing, puffing, and spinning", which were described by Illingworth [16], are acceptable marketing practices for businesses but are those behaviours acceptable for physicians as well?

In microeconomics, the profit maximization rule states that if a company chooses to maximize its profits, it must choose that level of output where marginal cost (MC) is equal to marginal revenue (MR) [14]. In more simple terms, profit maximization is an advantage for a company, but it can be a disadvantage for consumers if the company starts to use cheaper products/services or decides to raise prices especially when the firm has a monopoly power in the market [17]. If this concept is applied in the health care sector then the outcome can be dangerous and unethical for the patients. According

to Sidorsky, it may be argued, that healthcare providers should not function merely as profit-maximizing entities since they principally promote patient health [7]. Unfortunately, in the free market economy due to the minimum government interference through insufficient laws and regulations, this might not always be achievable.

Firstly, most aesthetic practitioners want to maximize their profit by increasing provision of services in their clinics. Recently, this has been achieved by hiring an aesthetician to perform a variety of skin-care procedures like mild chemical peels, microdermabrasion etc. However, it is important to remember that aestheticians treat "clients," not "patients" [18]. In the USA, there are many aesthetic practices, where "dermatology physician assistant (PA)" assists during the cosmetic procedure and "physician extenders" (PE) are employed for basic dermatological evaluation and treatments [19]. Therefore, as correctly stated by Slade and Grant-Kels [18], it would be appropriate only if the aesthetician, PA or PE are employed in a cosmetic practice to receive only patients, who are first evaluated by a physician and are referred to them for specific treatment part of the medical plan of care. In addition, they recommended that aesthetic practitioners need to monitor the scope of practice, delegate appropriately, and manage patient expectations [18]. Secondly, aesthetic practitioners in order to maximize their profit they might try to minimize their costs by cheaper unauthorized products for their treatments.

Recently, aesthetic physicians have achieved profit maximization by allying with some major cosmetic companies [15]. However, as it was mentioned in the editorial note of Kassirer and Angell [3] "it is one thing to recommend health-related products based on careful scientific scrutiny; it is another to enter into an exclusive marketing arrangement with a single company in which royalties are linked to sales". In addition, some physicians become a paid spokesperson at scientific and professional meetings, for off-label cosmetic products or aesthetic uses of products and procedures, without any mention of the risks or actual approved indications in order to promote them [15]. Are these ethical behaviours? The statement of American Academy of Dermatology's on office dispensing, for example, emphasises that "Dermatologists should not dispense or supply drugs, remedies or appliances unless it is manifestly in the best interest of their patients" [7]. Often dermatologists do not only sell cosmetic products in their practices, but in order to maximize their profits they have produced their own skincare products, sometimes devoid of evidence based support. Cantor mentioned in her article that every product should be evaluated by randomized, double-blinded and placebo controlled trials regardless the overall cost burden for the physicians [20].

Furthermore, information asymmetry, one type of market failure in the free market, is a challenging issue in aesthetic practice. According to definition, asymmetric information is a situation in which one party in a transaction has more or superior information compared to another [21]. In aesthetic practice, this often happens when the doctor knows more than the patient. Potentially, this could be a risky situation because one party can take advantage of the other party's lack of knowledge [21]. Information must be communicated clearly and respectfully to the patient and a shared understanding of expectations and limitations that consent to intervention is sought by the doctor [12]. However, few practitioners' desire to make profit might carry on unnecessary treatments hiding important information.

Aesthetic physicians, who want to be good businesspersons, need to apply a business plan consisting of their financial, marketing, and operational strategies and actions of their practice. It is a demanding, time-consuming process to apply a business plan as it requires setting accountable, realistic and measurable objectives that should be achieved in the future. These would be even more difficult for a practitioner of no business background. The purpose of advertising is to increase the demand of services and products and building brand loyalties to make their demand more inelastic. This would allow practitioners to charge higher prices for their services without considerable fault in their quantity demanded raising their total revenues and enhance their profits [22]. However, spending on advertising is highly expensive to carry out and in many cases, it might be misleading to the consumers due to false claims regarding the advertising product or service. If aesthetic practitioners, for example, market their services irresponsibly by making unjustifiable claims about interventions, trivialising the risks involved, or using promotional tactics might encourage people to make "ill-considered" decisions [12]. Ethical dilemmas related to the commercialization of medicine have been a challenging matter in the western world [23]. White –coat endorsement is a marketing tool that has been used in aesthetic medicine for promotion cosmetic products as Cantor said "Great products need no MD endorsement, the market will find them" [20].

DISCUSSION

Aesthetic medicine makes profit from the ideology of a society that serves only vanity, youthfulness and personal success, and one which is losing sight of the real values [24]. In fact, "a person having a non-surgical cosmetic intervention has no more protection and redress than someone buying a ballpoint pen or a toothbrush", as it was mentioned by Professor Sir Bruce Keogh KBE [4]. The tricky question is "How do we approach this issue?" Actions should be taken for enhancing patients' safety. The first step in every solution and decision-making process is to identify and define the problem [25]. In this situation, a worldwide concern must be confronted.

An informed and empowered public is crucial to driving up the standards of aesthetic practice and the quality of care provided [14]. Today, it is believed that the public is more informed through social media and advertisements, but they are not better or correctly informed. Therefore, aesthetic practitioners have an obligation to educate their patients using up to date evidence that will favour patient's best interest reducing asymmetry information.

In addition, a certification system should be introduced or a governmental body should be assigned to monitor the quality and safety of products that are used by the practitioners [12]. Dermal fillers, for example, are used by healthcare and non-healthcare practitioners with no requirement for previous experience, training or even basic knowledge of the product. For example, the scope of the EU Medical Devices Directive, in the European Union, should be extended to include all cosmetic implants including dermal fillers, and legislation should be introduced to classify fillers as a prescription-only medical device [12]. All practitioners must be registered and the register should be independent of professional groups or commercial bodies. Entry to the register, for example, should be subject to: (i) achievement of accredited qualification; (ii) premises meeting certain requirements; (iii) adherence to a code of practice that covers handling complaints and redress, insurance requirements, responsible advertising practice and consent practices; and (iii) continued demonstration of competence through an annual appraisal [26].

The initial question of this article was whether an aesthetic practitioner can be a physician and a businessperson at the same time. Both of their roles are discussed above, but my own opinion regarding the subject would be supported by the following example. The scope of dermatologic practice is broadening owing to recent advances in cosmetic services; these services reduce the dermatologic workforce resources that can be devoted to medical dermatologic patients [27]. It is not illegal or unethical when aesthetic practitioners, like dermatologists, decide to restrict their everyday practice to only cosmetic or procedural dermatology [19]. However, what will it happen if most dermatologists have no available time to treat complex dermatological conditions or follow up suspicion malignant skin lesions? Tsang and Resneck [28] stated in their article that even for patients willing to pay for face dermatology appointment due to changing pigmented lesions, the mean wait-time was 38 days (median, 26 days) in the metropolitan areas in the United States. However, Meffert and Villegas mentioned that the waiting time for a botulinum toxin injection appointment also varied widely with geographic location (6–32.5 days) but had a relatively short median wait of 8 days [19]. Therefore, this argument raised an important issue that there is an increase shift from classical medical dermatology to the more cash–driven cosmetic procedure and sometimes due to shortage of dermatologists in certain areas several dermatological conditions remain underdiagnosed

In conclusion, I agree with the statement first mentioned by Cantor in one of her articles "Ethics requires doctors to be more than just salespeople with an M.D" [20] I strongly believe that aesthetic practitioners, especially physicians, must set their priorities straight. If they decide to practice aesthetic medicine, they need to appreciate that they have the same moral obligations to the cosmetic patients as with any other patients. Cosmetic dermatologist should follow the good medical practice guidance as General Medical Council in U.K. states: 'Good doctors make the care of their patients their first concern: (i) they are competent, keep their knowledge and skills up to date, establish and maintain good relationships with patients and colleagues, they are honest and trustworthy, and act with integrity and within the law' [29]. If they continue to place their integrity and moral standards to the test, they will continue to be judged by their colleagues, society [30].

Finally, every aesthetic practitioner should remember that pull of profit cannot be a primary motivation like in the case of businessperson. Instead, they should remember what Sir William Osler advised the 'Students of Medicine' : "*Seek your own interests, make of a high*

and sacred calling a sordid business, regard your fellow creatures as so many tools of trade, and, if your heart's desire is for riches, they may be yours; but you will have bartered away the birthright of a noble heritage, traduced the physician's well deserved title of the Friend of Man, and falsified the best traditions of an ancient and honourable Guild" [21,30,31].

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Clinical Image

Staphylococcal sycosis of pubis in a young girl: Exceptional location

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Staphylococcal sycosis is a rare variety of deep folliculitis caused by *Staphylococcus aureus* [1]. It is presented in the form of pustular inflammatory cupboards, maintained by shaving and poor hygiene. It mainly affects the hairy regions of the face [1]. We present a rare localization of staphylococcal sycosis in the pubis and vulva, in a teenage girl. A 15-year-old girl, consulted for painful, pubic and vulvar skin lesions, evolving for two months. The dermatological examination showed an erythematous-oedematous and infiltrated plaque, at the pubis and the vulva, with pustules and crusts, associated with partial depilation (Fig. 1). Dermoscopy was non-specific, showing follicular pustules and pubic anisotrichy (Fig. 2).

A pus collection with bacteriological study showed a staphylococcus aureus infection, while the mycological examination was negative.

The diagnosis of a pubic staphylococcal sycosis was retained.

The patient was put under fucidic acid 1g per day 10 days, with local treatment with an antiseptic and cream based on copper zinc. The evolution was favorable (Fig. 3). The pubis and the vulva constitute an exceptional location of the staphylococcal sycosis. Pediatric cases are rare [2]. It is seen mainly in man after puberty at the level of the beard.

It is often secondary to inoculation by the razor [3].

The diagnosis of sycosis of the pubis can sometimes be difficult to establish with other diagnoses such as

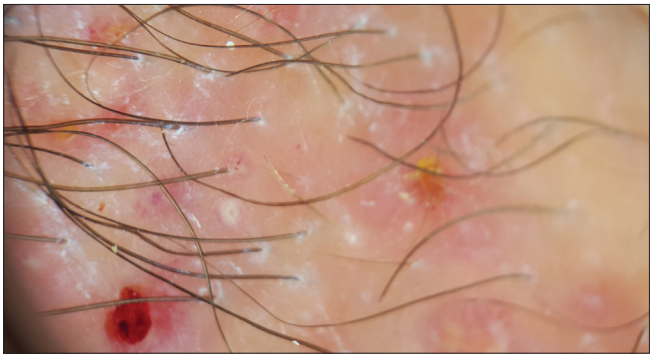

Figure 2: Non-specific dermoscopy follicular pustules and pubic anisotrichy.

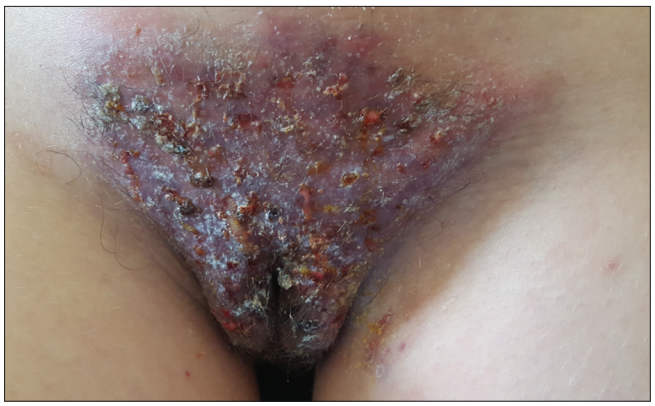

Figure 1: Erythematous-edematous placard, surmounted by pustules and croutons, with pressure pus, and partial depilation.

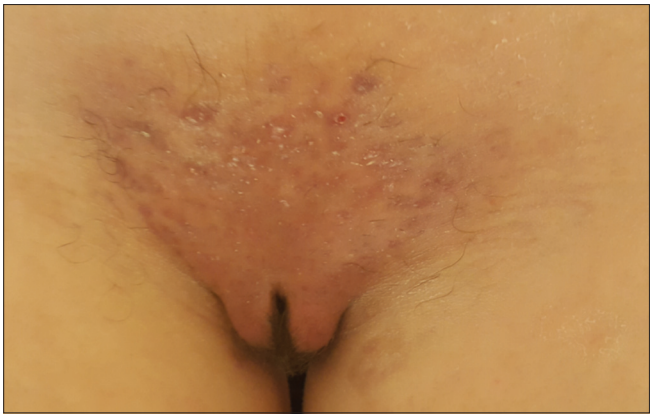

Figure 3 : Good progress under anti-staphylococcal treatment.

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a pubic celiac kerion, a pustular psoriasis, or even a cutaneous manifestation of a crohn.

The anti-staphylococcal treatment of the pubis appears effective and sufficient. The course under treatment is generally performed without sequelae.

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Clinical Image

Infantile perianal (perineal) pyramidal protrusion

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A 4-month-old infant was referred from surgery clinic for evaluation of *perianal warts* noticed recently by her mother. Her medical history was unremarkable and there was no suggestion of constipation, diarrhea, sexual abuse or maternal anogenital warts. Examination showed a solitary, small, fleshy, smooth surfaced, painless protrusion in the midline at superior aspect of anal verge (Fig. 1). No bleeding, excoriations, anal fissure, hemorrhoids or rectal prolapsed was noted. With a diagnosis of infantile perianal pyramidal protrusion the parents were counseled about its benign nature.

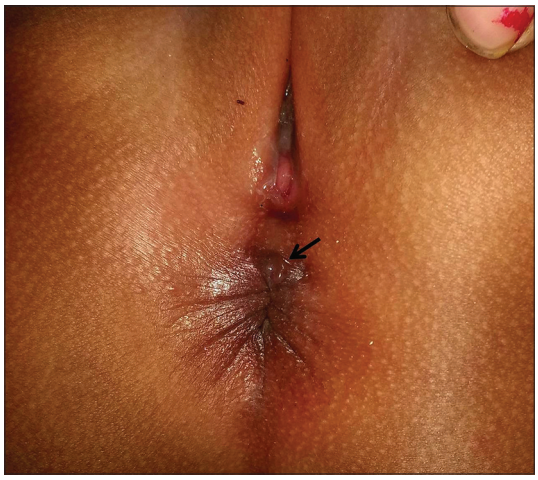

Figure 1: Infantile perianal pyramidal protrusion, a small, fleshy, smooth surfaced protrusion (arrow) at its classic location in a female infant.

Infantile perianal (perineal) pyramidal protrusion (IPPP), a relatively newly described entity, is reported primarily in female infants. Its exact pathogenesis remains unknown but is considered as (i) embryologic remnant of urogenital septum or a congenital anatomical weakness of median raphe, (ii) functional or acquired secondary to mechanical irritation from wiping, constipation/diarrhea, or fistulas/anal fissures, and (iii) lichen sclerosus et atrophicus (LSA)-associated that results from post inflammatory rearrangement of fibrous tissue [1,2]. Resolution is spontaneous especially in functional variety once the primary trigger is removed [3,4]. Topical corticosteroid may help early resolution of LSA-associated IPPP [5]. The histologic features of PPP are not defined but will exclude other simulators [4]. Nevertheless, it is imperative to differentiate the condition from simulating lesions affecting perineum/perianum such as hemorrhoids, prianal warts, condyloma, mollascum contagiosum, sentinel tag, granulomatous lesions of Crohn's disease, perineal midline malformation, rectal prolapse, infantile hemangioma and most importantly from signs of sexual abuse to avoid unnecessary treatment and parental anxiety.

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Letter to the Editor

Subungual hematoma treated successfully with 2940 nm erbium YAG laser

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Sir,

Subungual Hematoma (SUH) is the most common nail injury. It presents as a collection of blood beneath a finger or toe nail that causes severe pain and changes the nail color due to pressure [1].

SUH may be a result of acute or repeated trauma, and the most common cause of nail darkening [2]. many treatment modalities have been prescribed for treatment of SUH like heated paper clips, heated needles, nail avulsion, dental burrs, ne-pointed scalpel blades, drills, cautery devices, and carbon dioxide lasers for trephining [3].

Here, we describe three cases of SUH treated successfully with 2940 nm Er: YAG laser.

Case 1

A 21 year old female came to the clinic complaining of severe pain confined to the distal part of the middle finger for 3 days after trauma of variable pressure associated with change in the nail color.

Upon examination, there was a purple- black discoloration of the nail plate, sparing the nail fold. We decided to treat her with 2940 nm Er: YAG laser Lutronic action II, Korea, by making a hole in the nail plate. We used the following Parameters: spot size = 1 mm, fluence: 50 J/cm^2.One shot done only. Patient was evaluated directly after the procedure and showed immediate pain relief and disappearing of the dark color. No complications have been reported (Figs. 1A and 1D).

Case 2

A 37 year old female present to clinic complaining of severe pain localized to the distal left big toe for 2 days after nail trauma associated with change in the color of the nail.

Upon examination, there was a purple- bluish discoloration of the nail plate and sparing nail fold. We treated the patient by the same parameters and technique used in the case1, 2 shots hit in the same site due to thickening of the big toe nail. Immediate relive of pain and color was noticed by the patient and the physician (Figs. 1B and 1E)

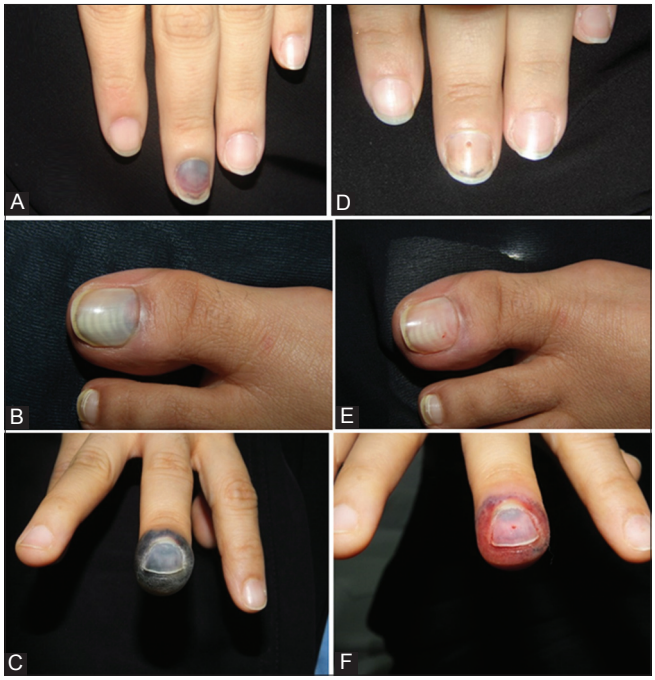

Figure 1: Before treatment (A-C), and after treatment (D-F).

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Case 3

A 28 year old female came to the clinic complaining of severe pain confined to the distal part of the middle finger in the right hand for 1 day after trauma of variable pressure associated with change in the nail color. Upon examination, there was a black discoloration involving almost the whole distal phalanges of the right middle finger including the subungual area. We used the same parameters mentioned in case 1. Immediate relive of pain and color noticed by the patient and the physician (FigS. 1C and 1F).

CONCLUSION

Treatment of subungual hematoma with 2940 nm Er: YAG laser to drain the collected blood beneath the nail plate is an excellent tool. It is an easy, simple, safe, and painless procedure done in the clinic. Pain relief and improvement in the color is resulted immediately. The cases that respond better should be within few days after nail trauma. We encourage considering this modality for treatment of subungual hematoma.

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Letter to the Editor

Eruptive porokeratosis in an 80-year-old immunocompetent man

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Sir,

Porokeratosis is typically an asymptomatic or mildly pruritic disorder of atrophic epidermal lesions surrounded by a hyperkeratotic border, histologically representing cornoid lamella. The most common clinically distinctive forms include classic porokeratosis of Mibelli, disseminated superficial porokeratosis (DSP), disseminated superficial actinic porokeratosis (DSAP), linear porokeratosis, dissiminated plantar and palmar porokeratosis, and punctate porokeratosis. Here we report a case of intensively pruritic, eruptive disseminated porokeratosis with acute onset, another subset that is rarely reported.

An 80-year-old immunocompetent Caucasian male with a several decade diagnosis of DSAP presented with pruritic, burning, painful, erythematous plaques located on the arms, hands, legs, and trunk. He was being treated for DSAP with doxepin but was still having intensely pruritic outbreaks of lesions twice a year that would last for 6 to 8 weeks and then dissipate. The lesions developed suddenly and acutely worsened. Brightly erythematous and edematous 5-10mm atrophic scaly papules with elevated borders and a thin ribbon of scale involved all extremities and his chest (Figs. 1 and 2). The palms, soles, head, and mucosal surfaces were spared. Punch biopsies were taken and revealed parakeratotic columns with loss of the granular layer and presence of scattered necrotic keratinocytes, compatible with cornoid lamellae and suggestive of porokeratosis (Fig. 3). There was mild epidermal spongiosis with focal spongiotic microvesicle formation (Fig. 4). Within the dermis, there was a perivascular lymphohistiocytic inflammatory infiltrate with scattered eosinophils (Fig. 5). PAS stain was negative for fungal organisms. Histology and clinical symptoms suggested the diagnosis of eruptive disseminated porokeratosis. The patient was treated with methylprednisolone 5 mg tablets in a dose pack and triamcinolone cream. His symptoms of intense pruritis were improved upon follow-up two weeks later. All lesions were healed except for a few remaining lesions on the legs.

DSAP is a variant of porokeratosis that presents with nonpruritic, nonpainful, scaly, erythematous papules/plaques on sun-exposed skin areas. A rare subset of this disease is eruptive disseminated porokeratosis (EDP), which presents with acute onset of intensely pruritic erythematous papules [1]. EDP differs from DSAP in that it has an acute onset of plaques that are typically pruritic. DSAP develops over years and is typically

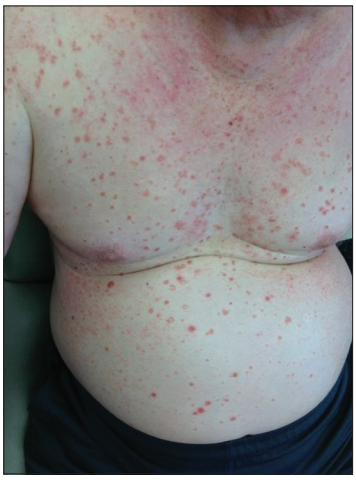

Figure 1: Brightly erythematous and edematous 5-10mm atrophic scaly papules with elevated borders and a thin ribbon of scale on the chest and abdomen.

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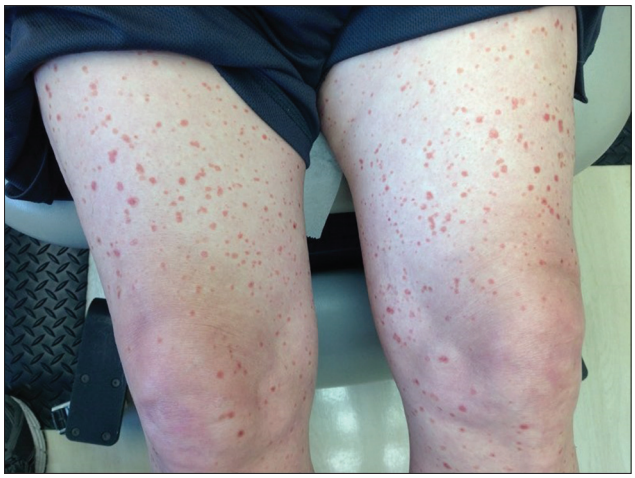

Figure 2: Brightly erythematous and edematous 5-10mm atrophic scaly papules with elevated borders and a thin ribbon of scale on bilateral legs.

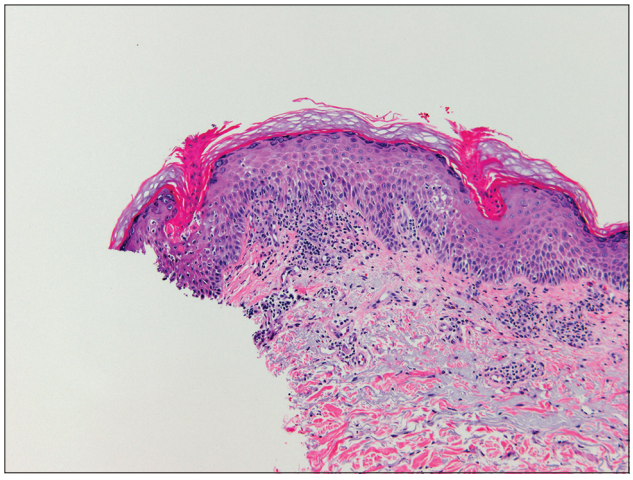

Figure 3: Angled columns of parakeratosis with loss of granular layer and necrotic keratinocytes in the subjacent epidermis, consistent with cornoid lamellae.

cosmetic with little or no symptoms. Only around 10 cases of EDP have been previously reported [2]. Histology typically demonstrates cornoid lamellae, epidermal spongiosis, and a perivascular inflammatory infiltrate of eosinophils and lymphocytes [1,3]. The course of EDP is similar to that of the presented patient, with repeated flares of intensly pruritic lesions and eventual regression. Our patient had hypertension but was an otherwise healthy 80-year-old man. Although initially diagnosed with DSAP, his clinical course and histologic findings are more consistent with the diagnosis of eruptive or inflammatory porokeratosis. Several terms have been proposed to describe this rare, atypical variant, including inflammatory DSP, eruptive pruritic papular porokeratosis, and eruptive disseminated porokeratosis [4]. It has been proposed that the inflammatory change leading to the intensely

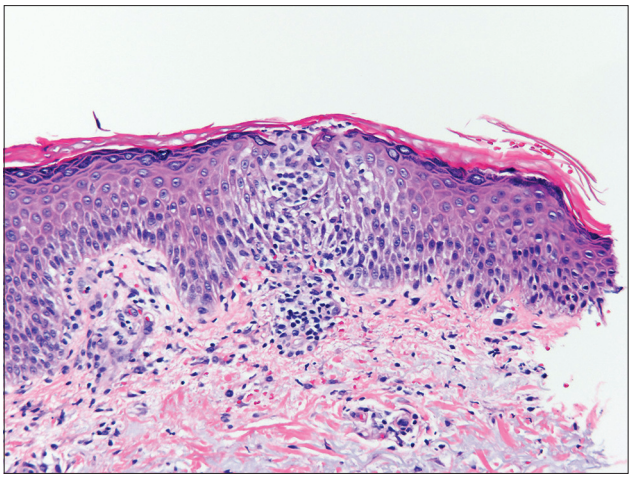

Figure 4: Epidermal spongiosis with spongiotic microvesicle formation.

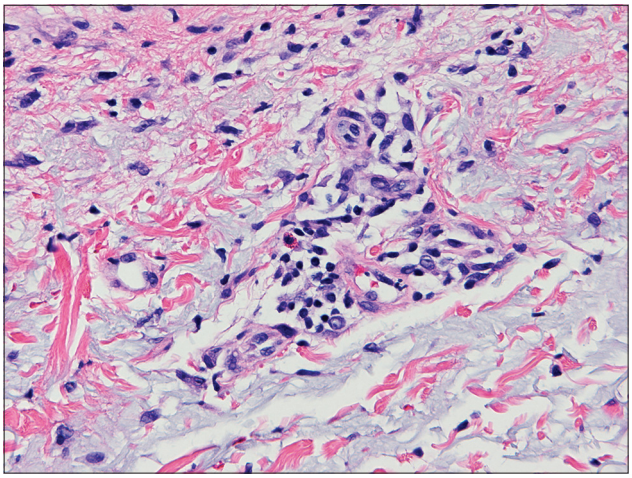

Figure 5: Perivascular inflammatory infiltrate with eosinophils.

prurutic erythematous papules may result from an immunological reaction against the abnormal clones of DSP, suggesting that porokeratosis is a precursor skin manifestation of malignant tumors [3,5]. Tanaka et al. postulated that regression of lesions was due to a CD4+ T cell-mediated immune reaction against the abnormal clones that resided in the epidermis [3]. Although the lesions typically resolve within several weeks, topical, intralesional and/or systemic steroid treatment results in improvement of symptoms. Use of topical 5-fluorouracil ointment has also been reported [1,3] Although EDP is considered a relatively rare disorder, it is imperative for dermatologists to be aware of this disease and consider it in their differential diagnosis.

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Letter to the Editor

Bulky cutaneous metastasis from lung cancer

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Sir,

A 65-year-old woman presented with a fungating tumor of her right arm that appeared three weeks prior and was rapidly enlarging. Her past history was unremarkable, she took no medications including over-the-counter medications before the illness, had no known allergies, and did not drink alcohol or use illicit drugs.

Examination revealed an alert, afebrile, normotensive woman. She appeared mildly ill. A bulky, fixed, ulcerated, slightly friable, and easily bleeding mass with a size of 12 cm in diameter and 8 cm in height in its greatest dimension was seen over the lateral surface of her right arm (Figs. 1a and 1b). The patient fully moved the arm and did not complain of pain or reduced strength. Inspiratory crackles were present in both lungs, there was no asymmetry on neurologic strength testing and the remainder of the examination was normal. Laboratory test results were normal except for mild anemia (Hb 10.5 g/dl) and elevated C-reactive protein (11.3 mg/dl). A total-body computed tomography performed with the administration of contrast material disclosed a cancer of the right lung with brain and liver metastases and raised concern that the mass represented a metastasis to the skin. A biopsy of the skin lesion revealed a poorly differentiated squamous-cell carcinoma. The patient was discharged to a hospice facility and died one month later.

Autopsy series have detected metastases to soft tissues, including skeletal muscle, subcutaneous tissue and skin, in up to 9% of patients who died from metastatic lung carcinoma [1]. Lung cancer and malignancies of kidney, colon, breat and ovary are most commonly associated with metastases to the skin and other soft tissues [1,2]. Approximately

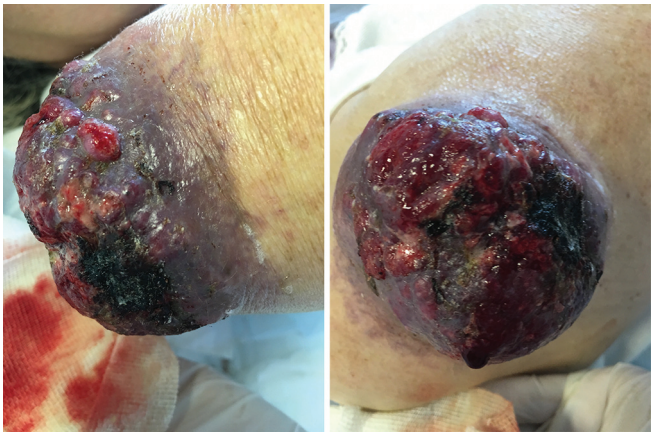

Figure 1: The patient had a bulky, fixed, ulcerated, slightly friable, and easily bleeding mass over the lateral surface of her right arm.

10 percent of patients with lung cancer develop metastases to the skin at any point in the course of the disease with a mean time of 6 months after the initial diagnosis [2,3]. Cutaneous metastases appear before or synchronously with the diagnosis of the primary tumor in about half of cases [3]. In our patient this was the first manifestation of an otherwise occult and advanced lung cancer. Lung cancer often presents in unusual ways but this is a very uncommon presentation.

All histological types of lung cancer may metastasize to the skin. Adenocarcinoma and squamous-cell carcinoma are more often implicated than other types and large-cell carcinoma is the less represented type [3]. A wide morphologic spectrum of clinical appearances has been described in cutaneous metastases from lung cancer. This variable morphology includes, papules, plaques and ulcers with sizes ranging from 2 mm to 6 or more cm in diameter that can be single or multiple in the same site. The nodules may be mobile or fixed and hard or flexible with color ranging from flesh-colored to red, pink, purple, or

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bluish black. The chest, abdomen, head and neck are most commonly involved [3]. Alopecia could be seen if the scalp or a burn scar is involved [4]. Cutaneous lymphangitis carcinomatosa could mimick a skin infection presenting as an eresypela-like or an eczematiform itchy rash due to invasion of the lymphatic vessels or from seeding after a surgical procedure [5]. This variable appearance could explain the failure in recognizing metastases to the skin at their first presentation as well as the delay in diagnosing the primary malignancy.

Bleeding may occur and it could be severe when masses are greater than 5 cm. Patients rarely complain of pain at the site of the metastasis, however severely painful zosteriform lesions could result from penetration of the dorsal root ganglions [6].

Why tumors in the upper lobes of lungs have a greater probability to metastasize to the skin than tumors of the middle and lower lobes is unclear [7]. No other features of the primary tumor have been associated with an increased probability of metastasizing to the skin.

Surgery alone or combined with chemotherapy and/or radiation is the mainstay of treatment of patients with cutaneous metastases from lung cancer. However, most of them have end-stage disease with a poor outcome and high rates of short-term mortality.

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Our Dermatology Online

Letter to the Editor

Uniform faint reticulate pigment network - A dermoscopic hallmark of nevus depigmentosus

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Sir,

Nevus depigmentosus (ND) is a localized hypopigmentation which most of the time is congenital and not uncommonly a diagnostic challenge. ND lesions are sometimes difficult to differentiate from other hypopigmented lesions like vitiligo, ash leaf macules and nevus anemicus. Among these naevus depigmentosus poses maximum difficulty in differentiating from ash leaf macules because of clinical as well as histological similarities [1]. Although the evolution of newer diagnostic techniques like dermoscopy has obviated the need for invasive diagnostic procedure, evidence and literature is yet to build on different hypopigmentary conditions. We herein describe the dermoscopic features of nevus depigmentosus and rightfully compare it with the features of its close contenders namely vitiligo and ash leaf macules to establish its distinguishing dermoscopic patterns.

A 9 month old male child, born of non consanguineous marriage, presented with a white patch on the abdomen since birth. The mother had brought the patient for evaluation owing to the cosmetic concerns. History and physical examination of the child was non contributory. Local examination of the skin revealed a well defined hypo- depigmented macule with no evidence of leucotrichia located on the left side of the abdomen 1.5cm away from the umbilicus. Diascopy test was negative and revealed feathering of margins and accentuation of the hypopigmentation. Wood's lamp evaluation showed an off white color. We went on to do the dermoscopy of the lesion to establish its pattern. Dermoscopy revealed uniform faint reticular melanocytic/pigment network throughout the lesion without any evidence of total loss of pigmentary network (Fig. 1).

ND is a form of cutaneous mosaicism with functionally defective melanocytes and abnormal melanosomes. Histopathologic examination shows normal to decreased number of melanocytes with S-100 stain and less reactivity with 3,4-dihydroxyphenylalanine reaction and no melanin incontinence [2]. Electron microscopic findings show stubby dendrites of melanocytes containing autophagosomes with aggregates of melanosomes.

For ease of understanding the pigmentary network patterns will be compared with that of normal skin will be provided herewith (Fig. 2). Normal reticulate pattern of pigmentation seen over normal skin corresponds to the pigmentation of the keratinocytes along the rete ridges while the pale area in between corresponds to the papillary dermis [3].

Vitiligo is an autoimmune disease of unknown etiology characterized by destruction and absence of melanocytes. Pigmentary network changes, and perifollicular and perilesional hyperpigmentation on polarized light examination, and a diffuse white glow on ultraviolet light examination were noted in evolving vitiligo lesions [4]. Established lesions of vitiligo demonstrate complete absence of reticulate pigmentary network as opposed to the normal skin which corresponds to the destruction of melanocyte and absence of melanin histologically (Fig. 3a).

Ash leaf spots (ALS) form an important part of the diagnostic criteria of tuberous sclerosis complex (TSC) although less than 2 ALS can be seen in normal individuals without TSC. On dermoscopy ash leaf macules are characterized by areas of faint reticular

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Figure 1: Nevus depigmentosus showing uniform faint reticulate pigmentary network pattern.

Figure 2: Normal skin showing reticulate pigmentary network (for comparison).

pigmentation and zones of total loss of any pigmentary pattern (Fig. 3b).

Hypopigmentation in ND is due to defect in transfer of melanosomes from melanocytes to keratinocytes rather than total loss of pigment [5]. Melanocytes usually are normal in number and size. Melanosomes are usually normal in size, shape, and internal structure, but can be diminished in number, heteromorphic, aggregated in melanocytes, or located in membrane bound aggregates [6]. This pathology manifests itself dermoscopically as the presence of faint reticular pigment pattern as opposed to total loss of pigment (due to absent melanocytes) as in vitiligo (Fig. 1).

To summarise, dermoscopy forms an essential non invasive diagnostic tool for hypopigmented lesions.

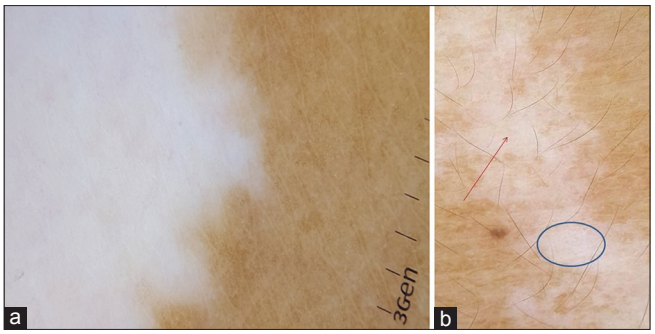

Figure 3: (a) Vitiligo showing loss of reticulate pigment network. (b) Ash leaf spots showing areas of faint reticular pigmentation and zones of total loss of any pigmentary pattern.

Differentiation of Nevus Anemicus from Nevus Depigmentosus is done by diascopy and firmly stroking lesional area. Vitiligo shows absence of melanocytic network.

Nevus Depigmentosus reveals uniform faint reticular pigment pattern whereas faint melanocytic network interspersed with total loss of the network characterizes ALS dermoscopically. To the best of our knowledge this is the first ever description of dermoscopic findings in nevus depigmentosus in the skin of color.

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Our Dermatology Online

Letter to the Editor

Striae distensae over scalp: A trichoscopic revelation

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Sir,

Striae distensae is a common disfiguring cutaneous condition which is characterized by linear, smooth and atrophic bands over the skin. It is a reflection of breaks in the connective tissue and occurs at the areas of dermal damage. The formation of striae distensae has been attributed to underlying mast cell degranulation with subsequent damage of elastin and collagen [1].

On one hand, striae can be the end result of various physiological conditions like pregnancy, adrenocortical excess and change in the body habitus, while on the other hand they are indicators of excessive topical steroid use [2,3].

Topical corticosteroid is considered as the first line therapy in treating conditions like alopecia areata and psoriasis. While its appropriate and accurate use can provide satisfactory results, its over-usage can lead to inadvertent effects such as development of erythema, telangiectasia, acneiform eruptions, striae and eventually atrophy and scarring. The misuse of steroid on the face can be picked up easily by the signs mentioned afore. However on the scalp, due to the presence of hair, these signs can easily be missed. The role of trichoscopy holds importance especially in treating scalp lesions as these signs can be identified much before they are appreciated clinically.

We herein report 3 cases of striae distensae over the scalp detected incidentally while examining the scalp of patients on topical steroids.

A 35-year old female presented with patchy hair loss and was applying topical beclomethasone prescribed by her previous Dermatologist. On trichoscopy, there was presence of exclamation mark hair and yellow dots confirming the diagnosis of alopecia areata. Trichoscopy also demonstrated linear, atrophic bands on her scalp; striae distensae along with telangiectasia (Fig. 1). On taking a detailed history, the patient confessed to applying topical steroid for a long duration.

Keeping this trichoscopic finding in mind we subsequently started looking for similar linear atrophic bands on scalp of patients who were treated with topical steroids.

A 17-year old male having patchy hair loss for 6 months who was applying a lotion, bought over the counter, reported to us. He complained of itching at the site of application of the lotion. Trichoscopic evaluation confirmed him to be a case of alopecia areata (Fig. 2). He also had features of steroid misuse like telangiectasia and linear white atrophic bands suggestive of striae distensae. The lotion was later on identified to be fluocinolone acetonide 0.01 %.

A 28-year old female presented with hair loss for two years. She had observed widening of her central parting and thinning of her pony tail. She visited a general practitioner who prescribed beclomethasone lotion, which she had been using for the past one year with no significant improvement. After taking detailed history to rule out chronic telogen effluvium, we proceeded for trichoscopy and observed hair diameter variability and multiple vellus hair at the frontal region which confirmed the diagnosis of female pattern hair loss. Trichoscopy also demonstrated the presence of telangiectasia with few, linear, white streaks which were arranged in parallel to form a band indicating striae distensae.

We would like to highlight that a tool as simple as a dermoscope can help us identify the steroid overuse. Periodic monitoring of the patient on topical corticosteroids can reveal subtle changes of dermal

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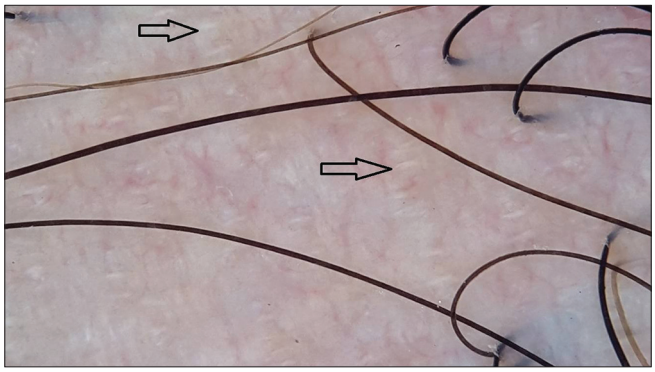

Figure 1: Linear atrophic bands (straie distensae) and telangiectasia over scalp on a patient with alopecia areata.

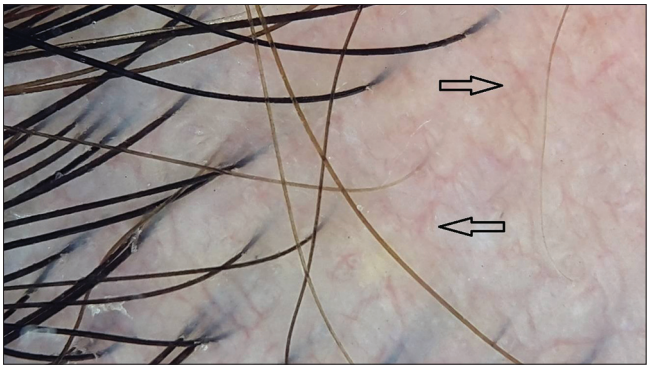

Figure 2: Telangiectasia over the scalp seen after steroid misuse.

damage. There might be a need to revise the treatment options in such cases and switch them to non-steroidal medications. An observant physician with minimal effort and time can easily recognize complications of steroid misuse like scarring and telangiectasia in patients and correct the damage done. Thus, trichoscopy is not only a diagnostic tool but also a monitoring tool wherein it can reveal the tell-tale signs of inadvertent topical therapy.

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