Volume 7, Number 3, July 2016
p. 253 - 365
Issue online since Tuesday July 05, 2016

ISSN: 2081-9390
DOI: 10.7241/ourd

Our Dermatology Online

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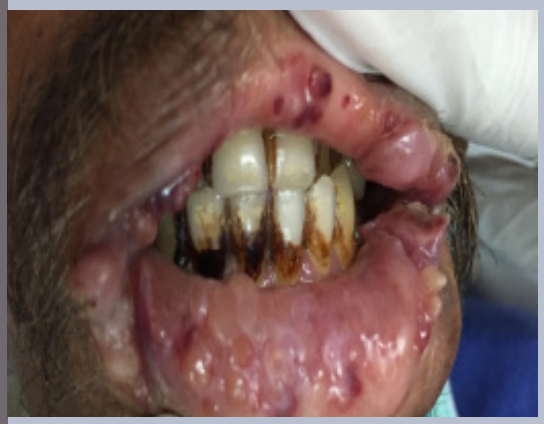

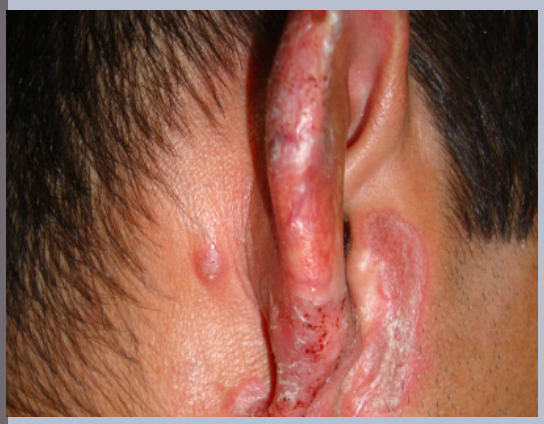

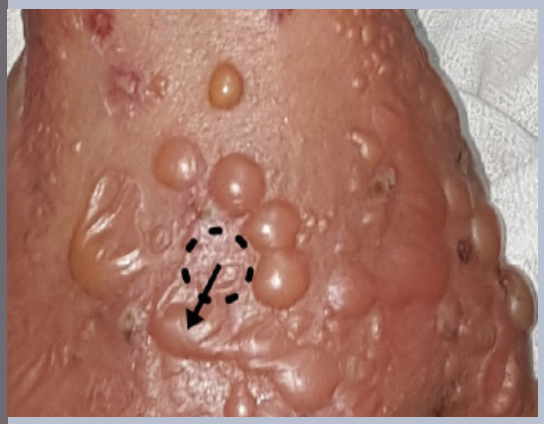

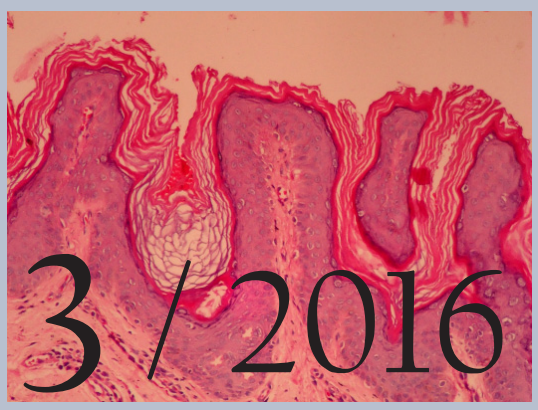

Editorial Pages

e-ISSN: 2081-9390
DOI: 10.7241/ourd

Quarterly
Our Dermatol Online

published since 01/06/2010 years

www.odermatol.com

Indexed in:

Universal Impact Factor for year 2012 is = 0.7319
system of opinion of scientific periodicals INDEX COPERNICUS (8,69)
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(Academic Search Premier) EBSCO
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	since issue 4.2011	www.odermatol.com
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Original Article

Topical therapy of molluscum contagiosum as a comparative therapeutic study using 15% phenol in lactic acid, 5% tincture iodine and pricking alone

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ABSTRACT

Introduction: Molluscum contagiosum is a common viral infection of the skin, caused by poxvirus, commonly affects young children. Although there was no specific treatment for molluscum contagiosum virus, many therapeutic modalities were used with different response rates. **Aims:** To evaluate the effectiveness of topical 15% phenol in lactic acid, 5% tincture iodine and pricking alone through comparative treatments of molluscum contagiosum. **Materials and Methods:** This randomized, therapeutic, single, blinded, comparative study was conducted in the Department of Dermatology–Baghdad Teaching Hospital Baghdad, Iraq, during the period from October 2007 to October 2008.Seventy-five patients with molluscum contagiosum were included in this study. They were diagnosed on clinical bases. Patients with prior treatment in the last 2 weeks and patients who had inflamed lesions were excluded. Full history and physical examination were done for all Patients. The patients were divided into 3 equal groups according to the mode of therapy (25 patients for each group): Group 1 patients treated by pricking the lesions with orange stick dipped in 15% phenol in lactic acid. Group 2 patients treated by pricking the lesions with orange stick dipped in 5% tincture iodine. Group 3 patients treated by pricking with orange stick alone. The treatment sessions were done at the time of presentation and then every week until complete cure was achieved, but not more than three sessions. The patients were followed up after one month from the last session to record the clinical cure and any local or systemic side effects. **Results:** Seventy five patients with molluscum contagiosum, 43 (57%) males and 32 (43%) females with male to female ratio 1.3-1, their ages ranged from 2-35 years(median:6 years). The most common age groups affected were below 10 years; 57 (76%) patients. The most common affected body sites were the face and neck 58 (77.3%) patients. Atopic diseases like atopic dermatitis, allergic bronchitis were found in 17 (22.7%) patients. After 3 weeks of treatment, the response rates were as follows: Group 1: 23/25 (92%) patients cured completely, 2 (8%) patients failed to achieve complete clearing, p-value was highly significant <0.000001. Group 2: 22/25 (88%) patients completely cured, 3 (12%) patients not responded completely, p-value was highly significant < 0.000001. Group 3: 21/25 (84%) patients completely cured, 4 (16%) patients continue to develop new lesions, p-value was highly significant < 0.000001. When these groups compared with each other there was no statistically difference (p-value>0.05).Scars were not reported in any group. **Conclusion:** Pricking molluscum contagiosum lesions by orange stick alone is safe, cost effective, highly effective and stimulating mode of therapy. Still topical 15% phenol in lactic acid and 5% tincture iodine are new and effective modes of treatment.

Key words: Molluscum contagiosum; Phenol in lactic acid; 5% tincture iodine; Pricking alone

INTRODUCTION

Molluscum contagiosum (MC) is a benign viral infection that generally affects young children and now is running an epidemic state in all over the country. It is characterized by smooth, dome-shaped discrete pearly papules that occasionally develop surrounding area of scale and erythema (molluscum dermatitis) [1].

How to cite this article: Sharquie KE, Noaimi AA, Ibrahim GA, Al-Husseiny AS. Topical therapy of molluscum contagiosum as a comparative therapeutic study using 15% phenol in lactic acid, 5% tincture iodine and pricking alone. Our Dermatol Online. 2016;7(3):253-257.

Submission: 08.01.2016; **Acceptance:** 01.04.2016

DOI: 10.7241/ourd.20163.70

Molluscum contagiosum caused by molluscum virus which belongs to the family poxviridae subgenus molluscipox virus, which comprises 4 genetically subdivided but clinically indistinguishable molluscum contagiosum viral types [2]. Generally, MC can occur on any part of the body surface including face, trunk, extremities, scalp, eyelids, lip, tongue and buccal mucosa [2,3]. The duration of both the individual lesion and the attack is very variable and although most cases are self-limiting within 6-9 months, some persist for 3 or 4 years [3].

Three groups are primarily affected: young children, sexually active adults, and immunosuppressed persons, especially those with HIV infection [4].

In many instances, therapy is not necessary and natural resolution can be awaited. This generally occurs without complications but often over a prolonged period of months to years in immunocompetent individuals. Curettage, cryosurgery, cantharidine, tretinoin and oral cimetidine and others were used in the treatment of molluscum contagiosum with different successes rate [5-9].

Phenol is also known as carbolic acid, is a toxic, colorless crystalline solid with a sweet tarry odor, it is used in, preparation of cosmetics including sunscreen, hair dyes, and skin lightening preparation. It is also used in cosmetic surgery as an exfoliant [10].

Lactic acid is a member of alpha-hydroxy acid, it is a colorless or slightly yellow, viscous hygroscopic organic acid liquid. Lactic acid works as an exfoliant, for treatment of warts, xerosis of the skin, icthyosis, follicular hyperkeratosis and seborrhoic keratosis [11-13].

Tincture iodine has been used as a topical therapy for treatment of vitiligo for many years and proved to be a successful topical therapy especially in patients with localized vitiligo [14].

Still the researcher search for other new, effective therapy, therefore the aim of the present study was to evaluate the effectiveness of topical 15% phenol in lactic acid, 5% tincture iodine and pricking alone in comparative treatments of molluscum contagiosum.

MATERIALS AND METHODS

This randomized therapeutic single blinded comparative study, was conducted in the Department of Dermatology and Venereology–Baghdad Teaching Hospital, during the period between October 2007 to October 2008.

A total number of eighty two patients with molluscum contagiosum were enrolled in this study. All patients were diagnosed on clinical basis.

Patients with prior treatment in the last 2 weeks, patients with inflamed lesions and lid margin lesions were excluded from the study.

Patients or their parents were fully questioned regarding: age, gender, site, duration, occupation, residence, associated symptoms (such as itching and pain), associated diseases like atopic dermatitis, diabetes mellitus, topical steroid usage and history of molluscum contagiosum in other family members.

Physical examination was carried out regarding site, size and number of lesions.

The procedure was fully described to the patients or their parents and the need for pre-and post treatment photographs explained for them and formal consent were obtained from all participants in this study after full explanation to each patient or their parent about the disease, course, prognosis and treatment.

Also this study was approved ethically by the Scientific Council of Dermatology and Venereology of Iraqi Board for Medical Specializations.

The patients had been equally divided into 3 groups depending on the mode of therapy (25 patients for each group).

Group 1: patients treated by pricking the lesion with orange stick dipped in 15% phenol in lactic acid.

The 15% phenol in lactic acid prepared by dissolving 15 grams phenol in 100 ml lactic acid (92%, pH 3.5).

Group 2: patients treated by pricking the lesion with orange stick dipped in 5% tincture iodine.

Five percent tincture iodine prepared by mixing iodine 25g, potassium iodide 25g, purified water 25g and alcohol (90%) up to 1000ml.

Group 3: patients treated by pricking the lesion with orange stick only.

The treatment sessions were done at the time of presentation and then every week until complete cure was achieved, but not more than three sessions.

The patients were seen regularly every week for 1 month during the treatment period, at each visit the response to treatment was assessed according to the change in size, number and development of new lesions. The side effects of treatment modalities were recorded such as burning, erythema, scar formation and signs of secondary bacterial infection like tenderness, erythema and swelling). Then the patients were followed after one month from the last treatment session for signs of relapse at the same location of treated lesions.

Statistical analysis was done by the EPI INFO system version 3.5.1.using chi-square. P-value of less than 0.05 was considered to be significant.

RESULTS

A total of eighty two patients were enrolled in this study, seven of them were defaulted after the first visit for unknown reason, while the remaining 75 patients completed the study.

Of these patients, 43 (57%) males and 32 (43%) females with male to female ratio 1.3:1. Their ages ranged from 2-35 years, the median were 6 years. The most common age group affected was from 2-10 years which comprised 57 (76%) patients.

The duration of the disease ranged from 7-30 days with mean ± SD of 19.5 ± 7.08 days. Thirty two (42.7%) patients had more than 2 anatomical sites involved. The most common involved sites were the face and neck 58 (77.3%) patients, trunk 29 (38.7%), extremities 8 (10.7%) and genital and perianal areas 6 (8%) patients.

Size of lesions ranged from few millimeters to one centimeter. A total number of 679 lesions with range from 2-26 with a mean ± SD of 9.05 ± 5.6 lesions/patient.

Twenty (26.6%) patients with molluscum contagiosum had associated diseases; 17 (22.7%) patients were atopic diseases like atopic dermatitis (asthma and hay fevers), diabetes mellitus 1 (1.3%) patient, alopecia areata 1 (1.3%) patient and chronic myelogenous leukemia 1 (1.3%) patient. One (1.3%) patient used topical steroid. Thirty four (45.3%) patients had history of contact with other infected persons or family members.

Group 1: Their ages ranged from 2-32 years, the median were 7 years, 14 (56%) males, 11 females with male to female ratio 1.3:1. A total number of treated lesions were 264, the duration of the lesions ranged from 10-30 days with mean ± SD of 21± 7.2 days. During the treatment period 13 (52%) patients were completely cured after the first session, 8 (32%) patients required another session and 2 (8%) patients were healed after the third session. Two (8%) patients still did not respond completely and continued to develop new lesions. So the total cure rate was 92%, p-value was highly significant < 0.000001, chi-square 53.33 (Table 1).

The reported side effects in this group were (Table 2): mild pain due to pricking occurred in 15 (60%) patients, mild burning sensation in 20 (80%) patients, transient erythema of the perilesional skin due to accidental contact with the medication in 5 (20%) patients and secondary bacterial infection in 6 (24%) patients.

Scarring was not reported in any patient. During the follow up period none of the cured patients developed new lesions.

Group 2: Their ages ranged from 2-35 years, the median were 5 years, 13 (52%) males, 12 (48%) females with male to female ratio of 1.1-1. A total number of treated lesions were 224. The duration of the disease ranged from 7-30 days with mean ± SD of 18.84 ± 7.9 days. After one session of treatment 11 (44%) patients completely cured, 8 (32%) patients needed another week of treatment, 3 (12%) patients were cured at the end of the third week. At the end of the treatment period 3 (12%) patients still developing new lesions. So the total cure rate was 88%, p-value was highly significant < 0.000001, chi-square 48.62 (Table 1).

Table 1: Rate of response during period of treatment of molluscum contagiosum

	1st week		2nd week		3rd week		p-value & Chi square
	No.	%	No.	%	No.	%	
Group 1 (n=25)	13	52	21	84	23	92	P<0.000001, Chi 53.33
Group 2 (n=25)	11	44	19	76	22	88	P<0.000001, Chi 48.62
Group 3 (n=25)	8	32	19	76	21	84	P<0.000001, Chi 48.78

Table 2: Side effects of treatment modalities of molluscum contagiosum

Side effects	Group 1 (%)	Group 2 (%)	Group 3 (%)	Total (%)
Pain	15 (60)	12 (48)	14 (56)	41 (54.7)
Burning	20 (80)	-ve	-ve	20 (26.7)
Erythema	5 (20)	-ve	-ve	5 (6)
Secondary bacterial infection	6 (24)	4 (16)	7 (28)	17 (22.7)

The side effects in this group were (Table. 2): mild pain occurred in 12 (48%) patients, secondary bacterial infection in 4 (16%) patients.

Scarring was not reported in any patients. During the follow up period none of the cured patients developed new lesions.

Group 3: Their ages ranged from 3-30 years, the median were 7 years, 15 (60%) males, 10 (40%) females with male to female ratio 1.5-1. A total number of treated lesions are 191, the duration of the disease ranged from 10-30 days with mean ± SD of 18.52 ± 5.99. After one session 8 (32%) patients cured completely, 11 (44%) patients required two sessions and 2 (8%) patients needed another session to accomplish complete healing, while 4 (16%) patients continued to develop new lesions. So the total cure rate was 84%, p-value was highly significant <0.000001, Chi-square 48.78 (Table 1).

The recorded Side effects were as follow (Table 2): mild pain in 14 (56%) patients, secondary bacterial infection in 7 (28%) patients.

Scarring was not reported in any patients. During the follow up period none of the cured patients developed new lesions.

When the results of treatment of the three groups compared with each other, there were no statistical differences p-value > 0.05, but the cure rate after the first session showed that phenol group had the best cure rate (52%) followed by the iodine (44%) and then the prick group (32%).

Ethics

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

DISCUSSION

Molluscum contagiosum is a common viral disease of the skin that mostly affects young children and could be seen in adults as part of sexually transmitted diseases [1-3].

The present work showed that male to female ratio was 1.3-1 and the disease was mainly affecting young children.

The disease is self-limited but this might takes several months-years [1]. Accordingly, the disease should be treated to clear the lesions as quick as possible due to bad cosmetic appearance and to limit the spread of the disease [7,8].

There is no specific treatment for MCV, although various surgical and medical strategies were used with different success rates and various side effects with longer duration of therapy such as curettage, cantharidine, 5% imiquimod cream and 10% KOH [15-18].

Curettage cure rate was 80%, used once and repeated as needed. Pain and scaring were common side effects [5].

Imiquimod cure rate was 75-82%, used 3-5 times/week for 5-9 weeks, it is expensive and associated with erythema, pruritus, post-inflammatory pigmentation and ulceration [17].

KOH 10% cure rate was 91.4%, used twice daily for 30 days, stinging and post-inflammatory pigmentation were reported side effects [18].

While the present study using 15% phenol in lactic acid, 5% tincture iodine and pricking only once weekly gave cure rates 92%, 88% and 84% respectively after 3 weeks of treatment. The reported side effects were mild pain due to pricking, mild burning sensation and transient erythema (phenol group) and secondary bacterial infection which required only topical antibiotic. Scaring were not reported. Although when these groups were compared with each other showed no statistically significant different response, but phenol and iodine groups when compared with prick group showed a quicker cure rate after one session (52%, 44% and 32%).

So this present work gave a high cure rates with shorter duration of treatment and mild side effects with low cost effectiveness.

But surprisingly, pricking of lesions by orange stick was as effective as other therapies (p>0.05) and this is very interesting and stimulating point in treatment as pricking only might change MC microenvironment and disturb architecture of the lesion and this was enough in clearance of lesions. Also we can speculate that pricking might stimulate the immune system through releasing relevant cytokines and other immune enhancers and thereby attraction of immune cells (T-cells) which are important in clearing of the lesions.

So the present work opened a new era of therapy that doesn't necessitate using any chemical agent or drug.

Statement of Human and Animal Rights

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

Statement of Informed Consent

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

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

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Original Article

Mucocutaneous malignancies in patients with AIDS. Report of 32 cases

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ABSTRACT

Background: The nature of the diseases that affect AIDS patients can be infectious, malignant, by HIV virus, a reaction to drugs, and regressional. The malignant type manifestations are not very widely known among our hospital population. **Aims:** To learn about the mucocutaneous malignancies in AIDS patients. **Materials and Methods:** This is a retrospective-prospective study of 32 mucocutaneous malignancies in patients with acquired immunodeficiency syndrome, AIDS, at the Infectology-Dermatology department of the Hospital General de Enfermedades IGSS (General Hospital of Common Diseases), during a 13-year period (2002-2014). The statistics books of those years were reviewed and the clinical and histological pictures of mucocutaneous malignancies in patients were revised as consulted to the Dermatology Department, for the periods from 2002 to 2009 and from 2009 to 2014. **Results:** The malignancies found were Kaposi's sarcoma, 19 cases (59.37%); non-Hodgkin's lymphoma, 6 cases (18.75%); basal cell carcinoma, 4 cases (12.5%); 1 (3.12%) epidermoid carcinoma; 1 (3.12%) patient with pubic sebaceous and left-ear epidermoid carcinomas; 1 case (3.12%) Queyrat's erythroplakia. Some of the 19 patients with Kaposi's sarcoma presented other conditions aside from that of the skin. 4 (21.05%) presented conditions in the oral cavity; 3 (15.78%) in the hard palate; 1 (5.26%) in the hard palate and gums; 1 (5.26%) in the hard palate and tongue; and 1 (5.26%) in the tongue. Six (18.75%) of the patients had non-Hodgkin's lymphoma: 3 (50.0%) at the hard palate level; 1 (16.66%) in the skin and oral cavity; 1 (16.66%) in the tongue; 1 (16.66%) in the hard palate and gums. The lymphomas were classified: 1 (16.66%) as large-cell lymphoma, 1 (16.55%) as non-classifiable, 1 (16.66%) as large-cell, high-grade B-phenotype lymphoma, and the remaining 3 (50.0%) as non-Hodgkin's. **Conclusions:** Mucocutaneous malignancies may occur in AIDS patients as the first manifestation of the disease, or during the evolution of the same. Therefore, it is crucial to learn about the different clinical aspects of these malignancies so an early diagnosis can be made and hence timely treatment can be provided.

Key words: Mucocutaneous malignancy; Kaposi's sarcoma; Basal cell carcinoma; Epidermoid carcinoma; Sebaceous carcinoma; AIDS, Non-Hodgkin's lymphoma

How to cite this article: Chang P, Machuca L. Mucocutaneous malignancies in patients with AIDS. Report of 32 cases. Our Dermatol Online. 2016;7(3):258-265.
Submission: 03.02.2016; **Acceptance:** 07.04.2016
DOI: 10.7241/ourd.20163.71

Our Dermatology Online

Original Article

Malignidades mucocutáneas en pacientes con sida. Reporte de 32 casos

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RESUMEN

Antecedentes: Las enfermedades que puedan afectar a pacientes con SIDA pueden ser de tipo infeccioso, maligno, por el virus del VIH, reacciones medicamentosas, de regresión, siendo las de tipo maligno una manifestación poco conocida en nuestra población hospitalaria. **Objetivos:** Conocer el tipo de malignidades mucocutáneas en pacientes con SIDA. **Material y Métodos:** Este es un estudio retrospectivo prospectivo sobre 32 casos de malignidades mucocutáneas en pacientes con el síndrome de inmunodeficiencia adquirida SIDA del departamento de Infectología-Dermatología del Hospital General de enfermedades IGSS, en un periodo de 13 años (2002-2014). Se estudiaron todos los pacientes con diagnóstico de SIDA que presentaron alguna manifestación mucocutáneo corroborada clínica e histológicamente de malignidad, sin importar edad ni sexo. Se revisaron libros de estadística de los años en mención revisando el cuadro clínico e histológico de los pacientes del 2,002 al 2,009 y del 2,009 al 2,014 se revisaron los pacientes con su cuadro clínico e histológico de malignidad muco cutánea que fue consultada al Departamento de Dermatología. **Resultados:** Las malignidades encontradas fueron el sarcoma de Kaposi con 19 casos (59.37%) linfoma No Hodgkin 6 casos (18.75%), carcinoma basocelular con 4 casos (12.5%), 1 caso de carcinoma epidermoide (3.12%), 1 paciente con carcinoma sebáceo pubis y epidermoide oreja izquierda (3.12%), eritroplasia de Queyrat 1 (3.12%). De los 19 pacientes con sarcoma de Kaposi 4 (21.05%) presentaban además de la piel afección de la cavidad oral, a nivel del paladar duro 3 (15.78%), paladar duro y encía 1 (5.26%), paladar duro y lengua 1 (5.26%), lengua 1 (5.26%) Seis de los pacientes presentaron linfoma No Hodgkin (18.75%) a nivel del paladar duro 3 (50.0 %), piel y cavidad oral 1 (16.66%), lengua 1 (16.66%), paladar duro y encía (16.66%). Los linfomas fueron clasificados como 1 linfoma de células grandes (16.66%), 1 no clasificable (16.55%), y 1 linfoma de células grandes alto grado fenotipo B (16.66%) y los 3 restantes como NO Hodgkin (50.0%). **Conclusiones:** Las malignidades mucocutáneas pueden presentarse en pacientes con SIDA como primera manifestación de la enfermedad o bien durante la evolución de la misma, por lo que es importante dar a conocer los diferentes aspectos clínicos de las mismas para poder hacer un diagnóstico temprano y por ende un tratamiento oportuno.

Palabras claves: Malignidad mucocutánea; Sarcoma de Kaposi; Carcinoma basocelular; Carcinoma epidermoide; Carcinoma sebáceo, SIDA, Linfoma No Hodgkin

INTRODUCCION

Las diferentes afecciones de piel y mucosas pueden verse en pacientes con infección VIH, las cuales pueden ser la primera manifestación de la enfermedad, marcador de la progresión de la misma y causa alta de morbimortalidad. En este tipo de pacientes puede haber malignidades mucocutáneas como el sarcoma de Kaposi, carcinoma epidermoide, epitelioma basocelular y de las extra cutáneas tenemos al cáncer del ano, cérvix, linfoma Hodking No Hodgking, cáncer de hígado y otros. La inmunosupresión del virus parece jugar un papel importante en su aparición, evolución y exacerbación de la misma, el conteo de CD4 está relacionado con la presentación de las malignidades y un conteo de ≤200 células/mL representa mayor incidencia de desarrollar enfermedades oportunistas.

How to cite this article: Chang P, Machuca L. Malignidades mucocutáneas en pacientes con sida. Reporte de 32 casos. Our Dermatol Online. 2016;7(3):258-265.
Submission: 03.02.2016; **Acceptance:** 07.04.2016
DOI: 10.7241/ourd.20163.71

El presente trabajo da a conocer las malignidades mucocutáneas en una población de pacientes con Sida atendidos en el departamento de Infectología-Dermatología del Hospital General de Enfermedades IGSS periodo de 13 años (2002-2014).

MATERIAL Y MÉTODOS

Este es un estudio retrospectivo prospectivo sobre 32 casos de malignidades mucocutáneas en pacientes con el síndrome de inmunodeficiencia adquirida SIDA del departamento de Infectología-Dermatología del Hospital General de enfermedades IGSS, en un periodo de 13 años (2002-2014).

Se estudiaron todos los pacientes con diagnóstico de SIDA que presentaron alguna manifestación mucocutáneo corroborada clínica e histológicamente de malignidad, sin importar edad ni sexo.

Se revisaron libros de estadística de los años en mención revisando el cuadro clínico e histológico de los pacientes del 2,002 al 2,009 y del 2,009 al 2,014 se revisaron los pacientes con su cuadro clínico e histológico con malignidad mucocutánea.

RESULTADOS

Se estudiaron 32 casos de malignidades mucocutáneas con correlación clínica y patológica con diagnóstico de SIDA, 31 correspondieron al sexo masculino (96.87%) y 1 caso del sexo femenino (3.12%).

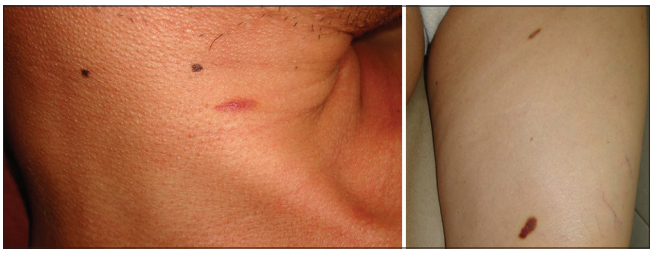

Figure 1: Lesiones maculosas tempranas del S. Kaposi.

En el grupo etario de 0-20 años no se estudió ningún caso, 21-40 años se estudiaron 15 casos (46.87%), 41-60 13 (40.62%), 61-80 años 4 casos (12.50 %).

Las malignidades encontradas fueron el sarcoma de Kaposi con 19 casos (59.37%) (Figs. 1-5), linfoma No Hodgkin 6 casos (18.75%) (Figs. 6 and 7), carcinoma basocelular con 4 casos (12.5%), 1 caso de carcinoma epidermoide (3.12%) (Fig. 8), 1 paciente con carcinoma epidermoide oreja izquierda y sebáceo pubis (3.12%) (Fig. 9), eritroplasia de Queyrat 1 (Fig. 10) (3.12%) (Cuadro 1).

De los 19 pacientes con sarcoma de Kaposi 4 (21.05%) presentaban además de la piel afección de la cavidad oral, a nivel del paladar duro 3 (15.78%), paladar duro y encía 1 (5.26%), paladar duro y lengua 1 (5.26%), lengua 1 (5.26%) (Fig. 11) (Cuadro 2).

Seis de los pacientes presentaron linfoma No Hodgkin (18.75 %) a nivel del paladar duro 3 (50.0%), piel y cavidad oral 1 (16.66%), lengua 1 (16.66%), paladar duro y encía (16.66%) (Fig. 12). Los linfomas fueron clasificados como 1 linfoma de células grandes (16.66%), 1 no clasificable (16.55%), y 1 linfoma de células grandes alto grado fenotipo B (16.66%) y los 3 restantes como NO Hodgkin (50.0%). (Cuadro 3).

DISCUSIÓN

Se sabe que el VIH destruye los linfocitos T CD-4, los cuales son esenciales para que el sistema inmunológico nos defienda contra infecciones y otras enfermedades. Cuando el VIH debilita el sistema inmunológico, presenta una oportunidad para que ciertos tipos de cáncer se desarrollen y crezcan rápidamente, especialmente aquéllos causados o asociados con virus [1].

Las personas con el virus de inmunodeficiencia humana (VIH) o que padecen SIDA tienen un riesgo mayor de padecer ciertos tipos de cáncer como sarcoma de

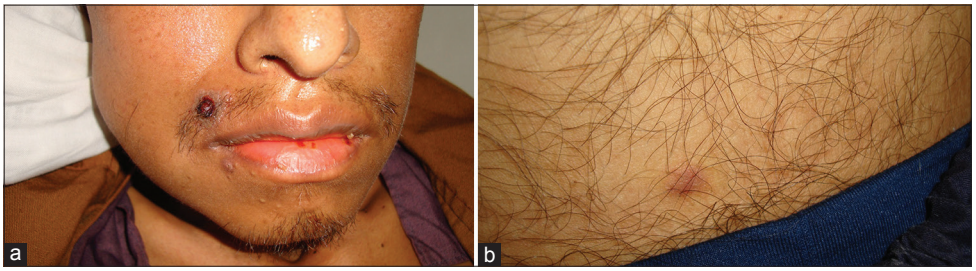

Figure 2: (a,b) Lesión nodular peri labial y maculosa en abdomen.

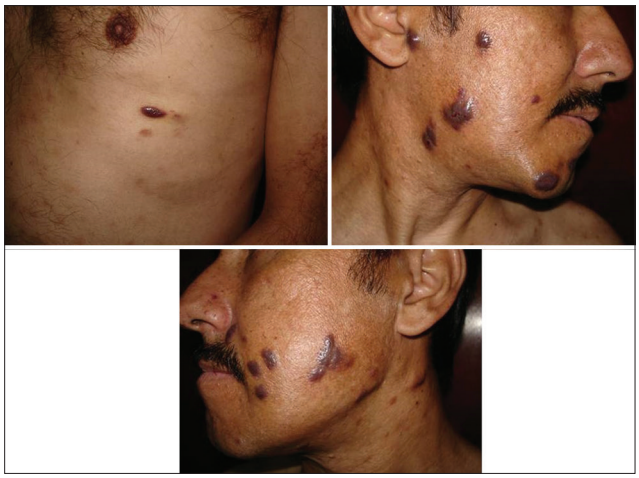

Figure 3: Placas y nódulos eritematovioláceos en diferentes localizaciones.

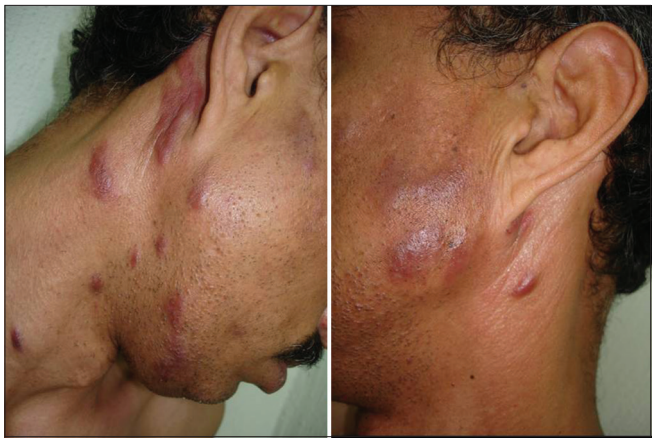

Figure 4: Placas y nódulos eritematovioláceos en diferentes localizaciones.

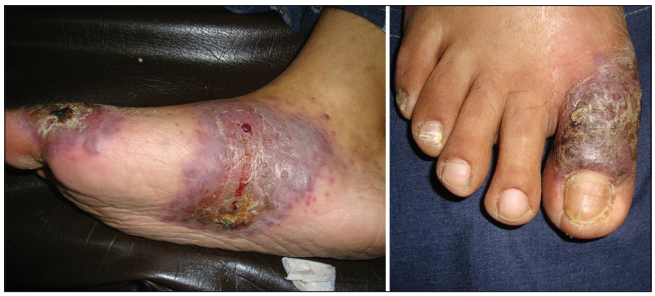

Figure 5: Placas y nódulos violáceos a nivel podal.

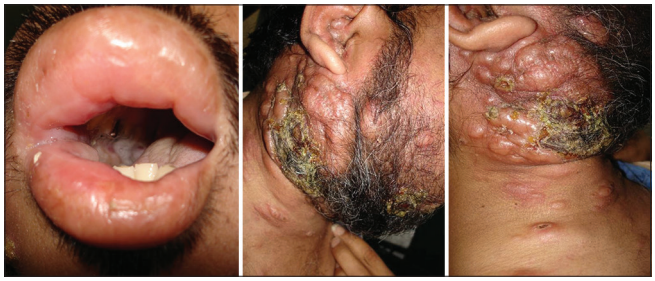

Figure 6: Neoformaciones por linfoma No Hodgkin.

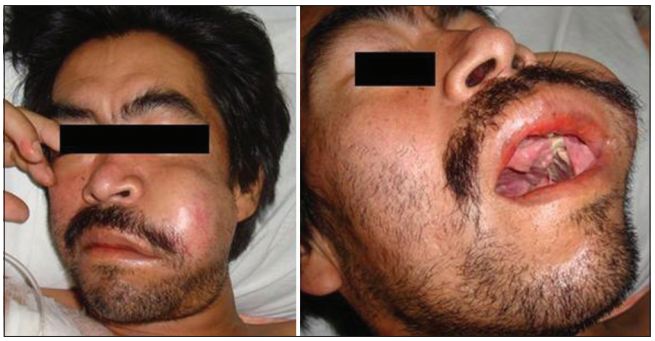

Figure 7: Neoformaciones por linfoma No Hodgkin.

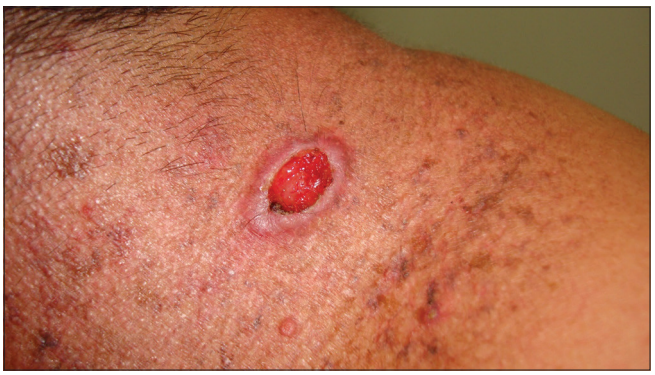

Figure 8: Carcinoma epidermoide ulcerado.

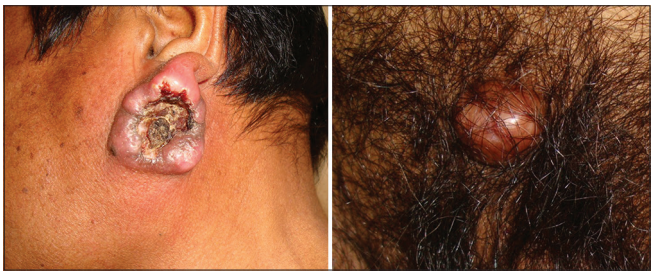

Figure 9: Carcinoma epidermoide oreja y sebáceo en pubis.

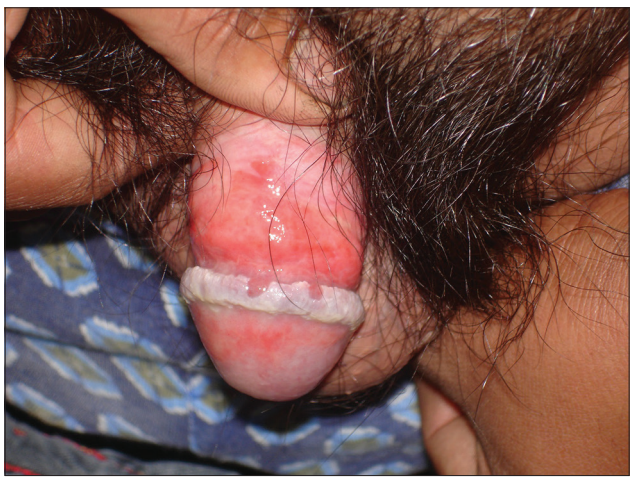

Figure 10: Eritroplasia de Queyrat.

Kaposi con un riesgo de miles de veces más, linfoma no Hodgkin con 70 veces más de riesgo, el cáncer de

cérvix al menos 5 veces mayor y el cáncer del ano con 25 veces mayor de riesgo [2].

Cuadro 1: Malignidades mucocutáneas en pacientes con SIDA

Tipo de malignidad	No. de casos	%
Sarcoma de Kaposi	19	59.37
Linfoma NO Hodgkin	6	18.75
Carcinoma basocelular	4	12.50
Carcinoma epidermoide	1	3.12
Carcinoma Sebáceo y epidermoide	1	3.12
Eritroplasia de Queirat	1	3.12

Cuadro 2: Afección de la cavidad oral en pacientes con Sarcoma de Kaposi y SIDA

Sitios afectados de la cavidad oral por Sarcoma de Kaposi	No. de casos	%
Paladar duro	2	10.52
Paladar duro y lengua	1	5.26
Paladar duro y encía maxilar	1	5.26

Cuadro 3: Linfomas NO Hodgkin cutáneo-mucosos en pacientes con SIDA

Afección cutáneo mucosa de linfomas NO Hodgkin	No.	%
Paladar duro	3	50.00
Lengua	1	16.66
Piel y cavidad oral	1	16.66
Paladar duro y encía	1	16.11

El riesgo de padecer los siguientes tipos de cáncer también aumenta en las personas con VIH/SIDA, el linfoma de Hodgkin el riesgo es 10 veces mayor, cáncer de pulmón 3 veces mayor riesgo, cáncer de hígado 5 veces mayor riesgo.

El SIDA no es una sola enfermedad, sino la etapa final de la infección por VIH, caracterizada por un conjunto o "síndrome de enfermedades potencialmente fatales". Si bien existen medicamentos para ayudar a las personas con SIDA, todavía no hay una cura y la mayoría de las personas mueren entre 1 y 3 años después de haber diagnosticado la enfermedad.

El SIDA es una pandemia caracterizada por una inmunodepresión profunda que conduce al desarrollo de infecciones oportunistas, neoplasias secundarias y manifestaciones neurológicas [3].

Existen 3 condiciones malignas que en la actualidad son consideradas como definitorias del SIDA, ellas son: el sarcoma de Kaposi, el linfoma no Hodgkin (LNH) y el cáncer cérvico uterino. Dentro de los no definitorios tenemos al cáncer anal, enfermedad de Hodgkin, leiomiosarcoma (pediátrico), carcinoma escamoso oral, carcinoma de células de Merkel, hepatocarcinoma.

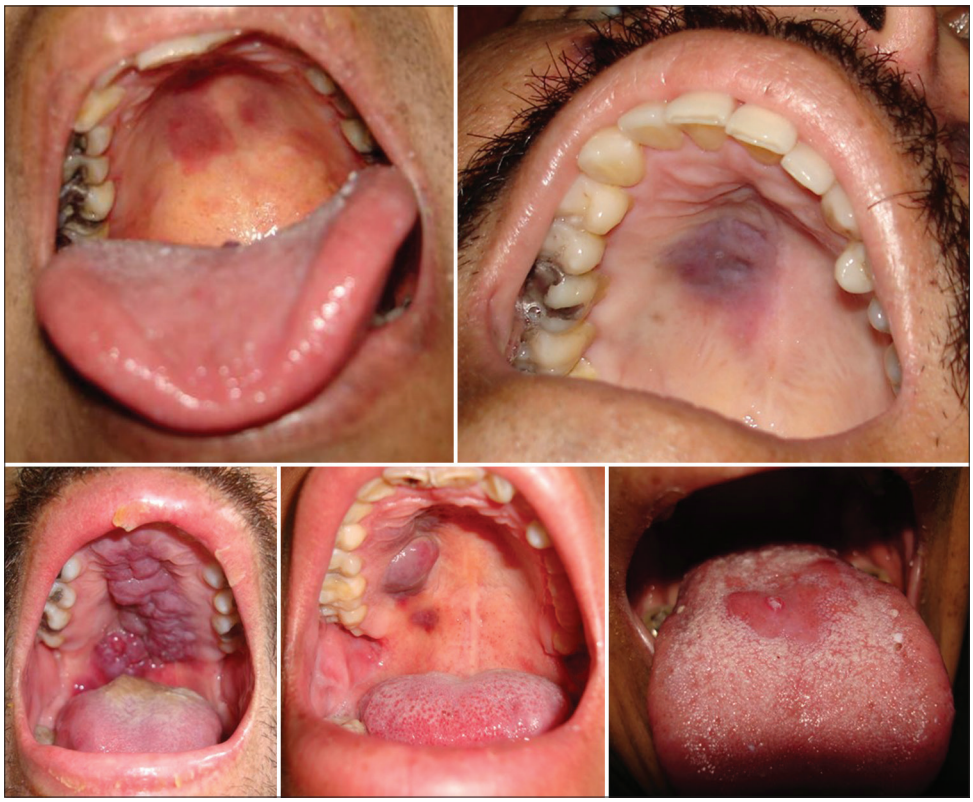

Figure 11: Lesiones maculosas y tumorales por sarcoma de Kaposi en paladar duro y lengua.

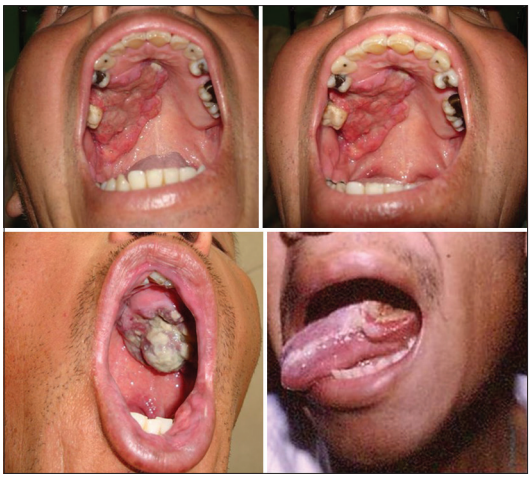

Figure 12: Linfoma No Hodgkin en paladar duro y lengua.

Además puede verse el angiosarcoma, el mieloma múltiple y linfoma de Hodgking [4,5].

Varios estudios muestran que el número de casos en otros cánceres, como linfoma no Hodgkin (NHL) se mantiene igual o incluso ha aumentado. Dichos cánceres pueden ocurrir incluso cuando los conteos de células T son altos, y por eso que la terapia anti-VIH no ayuda a reducir estos cánceres. De hecho, puede que se vean más casos de estos cánceres debido a que las personas con VIH viven más tiempo. Otra consideración es que las personas con VIH/SIDA parecen estar a mayor riesgo de desarrollar cierto tipo de cánceres no relacionados con el SIDA, como enfermedad de Hodgkin, cáncer en el labio, en los testículos, en el ano o el recto, de piel y de pulmón [6].

El sarcoma de Kaposi (SK) está asociado con el virus herpes humano 8 (HHV-8), se presenta en más del 35% de los casos por cada cinco lesiones cutáneas, se encuentra una lesión interna [7].

Es un tumor vascular multifocal que puede afectar piel o vísceras, se caracteriza por una neoformación capilar y proliferación de tejido conjuntivo peri vascular, en piel las manifestaciones son leves predominan en extremidades sobre todo en las inferiores caracterizadas por manchas rojas múltiples que se elevan y se tornan violáceas u ampollas hemorrágicas luego se convierten en placas y tumores semiesféricos como "nódulos" de 2 a 3 mm incluso pueden medir hasta 12 cm y ulcerarse dejando atrofia. En órganos internos afecta un 10 a 15% especialmente tubo digestivo, linfáticos, hígado y pulmones manifestando melena, rectorragias y diarreas [8].

Los pacientes con infección por el VIH-SIDA presentan una frecuencia mayor de carcinoma de células escamosas, basocelular, de células de Merkel y melanoma maligno. Los factores de riesgo son los mismos que en los pacientes sin infección por el VIH e incluyen la exposición al sol, la raza caucásica y antecedentes familiares de cáncer de piel. Los pacientes con infección por el VIH tienden a ser más jóvenes y presentan un mayor número de lesiones, más frecuencia de metástasis, más de complicaciones, mayor recurrencia y peor pronóstico [9].

El carcinoma basocelular se presenta en cabeza, cuello o tronco, puede manifestarse como una lesión tumoral hemisférica, de 1 mm a 10 mm de diámetro, con superficie nacarada, lisa y brillante, surcada por telangiectasias. No se ha podido correlacionar un valor de CD4 ni una mejor evolución con el TARGA [10,11].

Los diferentes tipos de carcinoma Basocelular pueden presentarse como el tipo nodular que es una neoformación de color piel con telangiectasia; y si forma úlceras fagedénicas, se torna en una lesión destructiva llamada ulcus rodens, esta aparece cuando se produce destrucción del tejido subyacente, se manifiesta en forma de ulceración de tendencia serpiginosa y se ubica por lo general en rostro, orejas y en el dorso de la mano.

El tipo quístico que es raro y difícil de distinguir de la nodular excepto por tener una pequeña cavidad llena de líquido, pigmentada con una variante de la nodular, a menudo confundida con un melanoma, la forma esclerosante/cicatrizante que es una lesión similar a una cicatriz, también llamada plano cicatricial y el tipo superficial que es un placa rojiza escamosa, en pacientes con SIDA las formas de manifestarse pueden ser atípicas debido a la inmunosupresión que presentan [11].

El riesgo de desarrollar un LNH (Linfoma no Hodgkin) en pacientes con SIDA, depende del grado y la duración de la inmunosupresión. Pueden presentar múltiples factores de mal pronóstico [12]. La posibilidad de ocurrencia de LNH a 2 años de diagnóstico de SIDA *vs* 3 años, es de 8 % contra 29%. Se ha planteado por algunos investigadores que este riesgo es independiente de la forma de haber adquirido la infección por el virus [13].

La segunda neoplasia en frecuencia en pacientes con enfermedad HIV/sida, luego del sarcoma de Kaposi es el linfoma No Hodgkin (LNH) [14].

El VIH puede aumentar el riesgo de desarrollar una neoplasia linfoide ente 100 a 200 veces más.

Una de las características clínicas salientes de los linfomas en pacientes HIV positivos es su presentación en etapas avanzadas de la enfermedad neoplá- sica y su localización extraganglionar en el 70% a 80% de los casos [15,16]. Entre los sitios extranodales más comunes se citan el tracto gastrointestinal, incluyendo la boca y glándulas anexas, la piel, el SNC y la médula ósea [17,18].

Estos linfomas suelen presentarse en su mayoría de células B con histología de alto grado, principalmente del subtipo difuso de células grandes, con compromiso extraganglionar frecuente y principalmente en el sistema nervioso central (SNC) [19].

En el paciente con SIDA puede presentarse en cualquier estadio de la enfermedad, pero por lo general aparece cuando este presenta un conteo de células (linfocitos T CD4 en el momento del diagnóstico del LNH) inferior o igual a 200 células/mL3 que es ahí donde la inmunosupresión da lugar a la presencia de enfermedades oportunistas [20].

Aunque el carcinoma epidermoide representa el 4% de la incidencia total de cáncer en el hombre, su relación con la presencia de virus de papiloma humano (VPH) es cada vez más frecuente, de hecho existen estudios que hablan de una incidencia de alrededor del 50%, en la presencia de VPH de cavidad oral y de tracto respiratorio alto, con un pronóstico sombrío para aquellos tipos llamados de alto riesgo (HPV-16 y HPV-18) [16].

En lo que respecta al SIDA no se han documentado reportes de especial aumento de la incidencia del carcinoma epidermoide. La comparación entre sexo sobre la incidencia de esta malignidad muestra aumento en las mujeres especialmente relacionado con cáncer cervico uterino y su relación con el VPH.

El servicio de Infectología del Hospital General de Enfermedades del IGSS empezó a tratar pacientes con VIH/SIDA desde el año 1,986, en la actualidad se atienden 2,500 pacientes en forma bimensual diagnosticándose de 16 a 18 casos nuevos por mes. Por lo que se considera importante dar a conocer este tipo de malignidades ya que en algunos casos es la primera manifestación de su infección VIH/SIDA así como la progresión de la misma para hacer un diagnóstico y tratamiento oportuno.

Statement of Human and Animal Rights

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

Statement of Informed Consent

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

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

Our Dermatology Online

Original Article

Manifestations and intensity of indirect self-destructiveness in patients with psoriasis vulgaris

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ABSTRACT

Introduction: Psoriasis is a chronic systemic disease which often significantly reduces the quality of life in extreme situations can provide to severe depression and even suicide. Indirect self-destructiveness is a generalized trend of behavior consisting of taking steps to increase the likelihood of negative and reduce the likelihood of positive consequences for the entity within a general manifestations such as transgression of norms and risk, addictions, poor health maintenance, personal and social neglect, lack of planfulness, helplessness and passiveness. Polish and world literature has no publications on indirect self-destructiveness in psoriasis nor in any skin diseases. The main aim of this study was to investigate the intensity and symptoms of indirect self-destructiveness in population of patients with psoriasis vulgaris **Material and methods:** The material consisted of 82 patients with psoriasis vulgaris hospitalized in the Department of Dermatology, Pediatric Dermatology and Oncology in 2013-2014. For the achievement of the objectives of the research socio-demographic questionnaire (own authorship) and Indirect Chronic Self-Destructiveness Scale by Kelley in Polish adaptation of Suchańska (version for men and women - each containing 52 issues) was used. **Results:** The results showed that in a population of people with psoriasis overall severity of indirect self-destructiveness was in the lower range of the average results. The dominant class of indirect self-destructiveness was helplessness and passivity that preceded the poor health maintenance and lack of planning. **Conclusions**: The results will enrich the existing knowledge about the harmful conduct of psoriasis and a better approach to the patient.

Key words: Psoriasis; Indirect self-destructiveness; Symptoms; Stress; Addictions

INTRODUCTION

Psoriasis Vulgaris and its Psychosocial Aspects

Psoriasis is a common chronic inflammatory disease, whose main manifestation are skin lesions such as hyperkeratotic papules and plaques covered with a layer of scales. The disease affects approximately 2-3% of the world's population. In addition to skin psoriasis also affects joints and cardiovascular system [1-5].

Psoriasis significantly impairs the quality of life of people affected by it, and the degree of reduction in quality of life is similar to that of diabetes, cancers or acute myocardial infarction [6-11].

Psoriasis also contribute to the formation of secondary changes in the psyche of a person suffering from this condition, they are responsible for depressed mood, depression, anxiety, the social isolation and withdrawal. Severe itching that often accompanies psoriasis very significantly promotes sleep disorders, especially an insomnia. About 5% of patients with the most severe forms of psoriasis (eg. erythrodermic psoriasis) attempt suicide. Psoriasis also affects social functioning, is primarily the cause of stigmatization of people suffering from this disease, it contributes to worse functioning in the family and at work. It has been proven also that the fact of having psoriasis favors addictions (smoking, drinking, drug use) that may be a mechanism of escape from a difficult situation resulting from the disease.

How to cite this article: Bienias W, Kaszuba A. Manifestations and intensity of indirect self-destructiveness in patients with psoriasis vulgaris. Our Dermatol Online. 2016;7(3):266-270.

Submission: 17.02.2016; **Acceptance:** 05.04.2016

DOI: 10.7241/ourd.20163.72

People with psoriasis often statistically don't join in the formal and informal relationships and divorce more often. Finally, the presence of psoriasis impairs the sexual sphere [12-29].

Susceptibility to the development of psoriasis is genetically conditioned, and the inheritance of susceptibility is poligenic. In addition to genes the occurrence or exacerbation of psoriasis is also influenced by environmental factors, among which the most important are stress, addictions (smoking and alcohol abuse) and infections, injuries or certain drugs [1-5].

In this situation stress and addictions are often both one of the causes and main effect of psoriasis.

Indirect Self-Destructiveness- Characteristics of the Concept and its Relationship with Psoriasis

Indirect self-destructiveness, also called chronic, hidden or latent is defined most often as a tendency to engage in behavior that increase the likelihood of negative and reduce the likelihood of positive consequences for the entity [30-33].

Indirect self-destructiveness is often called "suicidal lifestyle." What differentiates indirect self-destructiveness from its direct form (which include suicide and self-mutilations) is mainly the result of the action which is unnecessary and away in time.

The main manifestations of indirect self-destructiveness include: intentional suffering and defeat, helplessness, passivity, social and health neglects, addictions, transgression and violation of social norms and heedlessness and inability to plan and succumbing to temptation. According with Suchańska and Kelley we can distinguish 5 classes of indirect self-destructiveness designated A1-A5 (A1- transgression of norms and risk, addictions, A2-poor health maintenance, A3 - personal and social neglect, A4- lack of planfulness and A5 - helplessness and passiveness in the face of the problems). These classes are not separate entities but mutually overlapping, so that it is often difficult to classify the exact manifestation of a particular class [30-37].

After characterization of psoriasis vulgaris and indirect self-destructiveness it is well obvious that although this notions are seemingly quite far apart, they have several common aspects.

Addictions and neglect of health are manifestations of indirect self-destructiveness which are certainly associated with psoriasis, as evidenced by numerous publications devoted to this subject [13,15,17,19].

Indirect self-destructiveness determine unfavorable lifestyle, a lifestyle is according to the paradigm of Lalonde the major determinant of human health, and also the only one for which the subject has any impact thus linking frequent dermatological disease- psoriasis with indirect self-destructiveness, in the absence of any existing so far publications this topic seems to be well reasoned and practical at the same time [38].

The aim of the study was to determine the intensity and major symptoms of indirect self-destructiveness in the population of patients with psoriasis.

MATERIALS AND METHODS

The study included 82 adult patients with psoriasis vulgaris hospitalized in the Department of Dermatology, Pediatric Dermatology and Oncology in Lodz in 2013-2014 (27 women and 55 men, mean age 46.6 years). The study excluded patients suffering from other serious chronic diseases, especially neurological and psychiatric diseases. The subjects completed a questionnaire, patients demographic and composed of 52 sentences questionnaire investigating indirect self-destructiveness in adaptation of Suchańska, separate version for men and women. At each of the 52 statements patient chose an option answers - from A (strongly agree) on E (strongly disagree), on the basis of assigned each response scores determined the overall rate of intensity of indirect self-destructiveness (minimum score 52 points, the maximum 260 points) in turn, each related to one of 5 classes of indirect self-destructiveness, allowing also to define the dominant symptoms of a patient. The study was approved by the Bioethics Committee and participation in the survey was voluntary.

All calculation and graphs were made using Statistica 11PL and Ms Office 2007.

RESULTS

The average score of indirect self-destructiveness in the population studied was 117.61 ± 21.97 (median = 114.5).

The average score of class A1 "Transgression risk" of indirect self-destructiveness was in the examined population 41.40 ± 10.39 (median = 40.00).

The average score of class A1a "Addictions" of indirect self-destructiveness was in the examined population 39.11 ± 17.36 (median = 36.67).

The average score of class A2 - "Poor health maintenance" of indirect self-destructiveness was in the examined population 48.89 ± 12.66 (median = 47.50).

The average score of class A3 "Social neglect" of indirect self-destructiveness was in the examined population 39.15 ± 9.40 (median = 39.09).

The average score of class A4 " lack of planfulness" of indirect self-destructiveness was in the examined population 48.63 ± 10.59 (median = 48.89).

The average score of class A5 "helplessness and passivity" of indirect self-destructiveness was in the examined population, 58.68 ± 13.65 (median = 60.00). The data shown in Fig. 1 and Table 1.

DISCUSSION

The average score of indirect self-destructiveness in patients with psoriasis vulgaris was in the lower range of average results.

The most expressed class of indirect self-destructiveness in patients with psoriasis was A5 "Helplessness and passivity" in second place of intensity there were almost simultaneously class A2" Poor health maintenance" and class A4 " Lack of planfulnes". The less expressed class of indirect self-destructiveness was in this population class A1 "Transgression of norms and risk" and its subclass A1a "Addiction".

As we can see passive forms of indirect self-destructiveness strongly dominate in patients with psoriasis vulgaris over its active forms.

As it turns out, the biggest problem of people with psoriasis is their overwhelming helplessness, passivity and resignation. People with psoriasis, especially with its most severe forms are resigned, don't believe in the therapeutic success, don't believe in success in life are pessimistic for the future, have a great sense of injustice in the world and have feelings of lack of control.

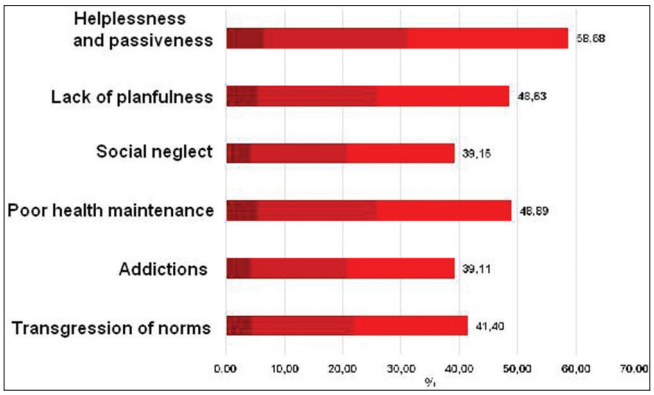

Figure 1: Intensity of classes of chronic self-destructiveness in patients with psoriasis.

Table 1: Intensity of chronic self-destructiveness in patients with psoriasis – the overall score

Chronic self-destructiveness	
Number of patients	82
Minimal score	73,00
Maximal score	167,00
Median score	114,50
Average score	117,61
Standard deviation	21,97
Asymmetry index	0,31

This passivity and helplessness can be a cause of a poor health maintenance, with a predominance of non adherence to medical recommendations and leaving the check-ups.

Thus smoking and drinking alcohol is not a leading problem in population of psoriatic patients and it seems that it can be an ineffective and desperate form of escaping from the psychosocial problems caused by psoriasis.

CONCLUSIONS

The conclusions from this study allow to optimize the approach to a patient with psoriasis. In everyday contact with the patient dermatologist must first activate and motivate a person, strengthen his motivation and educate and explain that it is possible to achieve the remission with the appropriate involvement of the patient and his positive thinking so needed in the therapeutic process. The attitude of forbidding of smoking and drinking alcohol is then ineffective and destroys the therapeutic relationship as well as may contribute to even greater helplessness.

Chronic self-destructiveness is a quite enigmatic psychological issue that in dermatology has been

never investigated or reported. Thus, it is the first study integrating chronic self-destructiveness with skin disease. Until now indirect self-destructiveness was studied in field of psychiatry in patients with schizophrenia in people after suicide attempts and in drug addicts [39-41].

The relationship of chronic self-destructiveness with psychiatry seems obvious, however, as shown in this work, because of bilateral interaction between the skin and the psyche, the study of indirect self-destructiveness in psoriasis has its strong justification and practical implications.

Statement of Human and Animal Rights

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

Statement of Informed Consent

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

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

Our Dermatology Online

Original Article

The limberg flap reconstruction - the optimal surgery for pilonidal sinus disease

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ABSTRACT

Introduction: Pilonidal sinus disease is a common condition usually seen in young adults causing significant morbidity. Many methods have been employed for its management but are also associated with high complications. Our study analyzes the efficiency of rhomboid excision of sinus and its tract, and reconstruction with Limberg flap in the management of pilonidal sinus disease. **Materials and Methods:** From July 2013 to May 2015, 46 patients were operated for pilonidal sinus disease by rhomboid excision Limberg flap reconstruction in two surgical units. Duration of operation, postoperative pain, duration of hospitalization and postoperative complications were noted. The follow up was done on an out-patient basis, every month for first three months. **Results:** Out of 46 patients, 40 were male (86.9%) and 6 were females (13.04%). The mean age of presentation was 28 years (range 16–50 years). The operative time ranged from 40 to 75 minutes, mean operation time being 50 minutes. Hospitalization ranged from 3-5 days, drains were removed on postop day 2/3. The stitches were removed after 12–14 days. Four patients developed mild infection. One patient developed necrosis at the tip of the flap. One patient developed recurrent lesion after 4 months, which may be due to improper excision of the sinus. The time off-work ranged from 12 to 22 days. The mean follow-up period was 3 months. **Conclusion:** Limberg flap is very effective for pilonidal disease with comparatively fewer complication, short hospitalization, lesser rates of reoccurrence, early healing and lesser time off-work. The technique can be mastered easily and provides an effective procedure for primary as well as recurrent disease. The results of this study support the wide excision and Limberg flap rotation as a preferred treatment of the disease.

Key words: Limberg flap; Pilonidal; Surgery

INTRODUCTION

Intergluteal pilonidal disease is a commonly encountered condition in adult primary care, and it causes significant morbidity. The estimated incidence is 26 per 100 000 people affecting men twice as often as women [1,2]. Pilonidal disease generally presents as a cyst, abscess, or one or more sinus tracts with or without discharge in the upper part of the natal cleft [3,4].

Etiology is uncertain but relates to the implantation of loose hair into the depth of natal crease. Other factors associated, are increased sweating with sitting and friction, poor personal hygiene, obesity, local trauma, narrowness of natal cleft [5,6]. Implantation of hair leads to infection and abscess formation later leading to discharging sinus.

The management of pilonidal sinus disease remains controversial, and gold standard treatment modality has yet to be established [7]. There has been a debate regarding the best treatment for pilonidal diseases for many years. The choice of ideal therapy should be guided by several principles - Simple to perform, Limited inpatient stay, Low recurrence rate, Minimal postoperative pain, Limited wound care, Early return to activity, Cost effective, tailored to the patient and the extent of the disease [8].

How to cite this article: Srihari RS, Naveen AM, Sreekar H. The limberg flap reconstruction - the optimal surgery for pilonidal sinus disease. Our Dermatol Online. 2016;7(3):271-275.

Submission: 12.11.2015; **Acceptance:** 02.02.2016

DOI: 10.7241/ourd.20163.73

In 1946, Limberg first described a technique for closing a 60° rhombus-shaped defect with a transposition flap. It meets the entire requirement for being the ideal procedure for sacrococcygeal pilonidal sinus if performed according to appropriate surgical principles [6,9]. The advantage of this reconstruction is that it is very easy to perform and design. It flattens the natal cleft with a wide and well vascularized pedicle that can be sutured without tension. That eventually helps in maintaining local hygiene, avoids hair insertion by reducing the friction between buttocks, reducing humidity, maceration, erosions and scar formation at the natal cleft [9].

Limberg procedure is a safe and reliable technique in the treatment of sacrococcygeal pilonidal sinus disease, with low complication and recurrence rates if performed according to appropriate surgical principles [10]. This study was carried out to evaluate the advantages, results of rhomboid excision and Limberg flap reconstruction in the management of pilonidal sinus disease.

MATERIALS AND METHODS

This prospective study was conducted from July 2013 to May 2015 in two surgical centers. It includes 46 patients who were treated for pilonidal sinus disease by sinus excision and Limberg flap procedure. Incision and drainage was done for Patients who had pilonidal abscess, later underwent definitive surgery. Surgery was performed under general anesthesia or regional anesthesia. After adequate skin preparation, Patients were placed in prone jack-knife position with buttocks strapped with adhesive tapes for wide exposure. The extent of the sinus was assessed by injecting methylene blue into all sinuses to outline its cavity. The rhomboid area to be excised was marked and flap lines were mapped on the skin (Fig. 1).

The rhomboid skin incision (with each side equal in length) was deepened to the pre-sacral fascia centrally and to the gluteal fascia laterally (Fig. 2).Under the guidance of methylene blue, all sinuses were excised en-bloc without incising outpocketings of the sinus cavity.

After excision, the Limberg fasciocutaneous flap was prepared by extending the incision laterally down to the fascia of the gluteus maximus muscle (Fig. 3). The size of the prepared flap was equal to that of the rhomboid area.The fasciocutaneous flap was transposed medially to cover the rhomboid defect created by excision of the sinus without any tension.

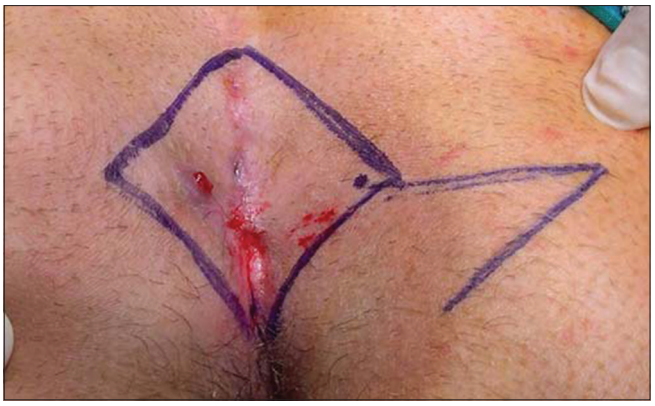

Figure 1: Marking of skin for excision and rhomboid flap.

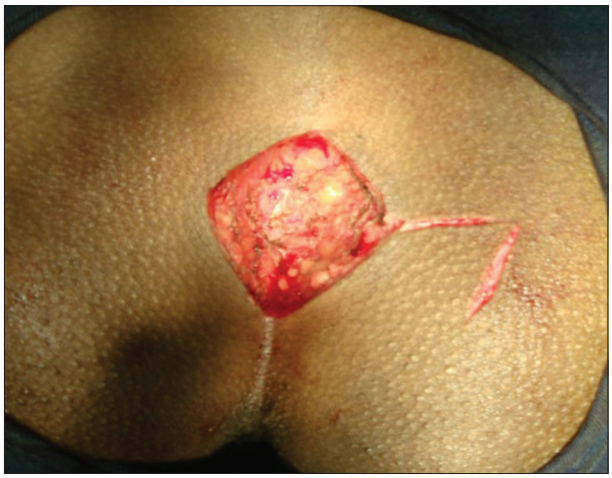

Figure 2: Flap with sinus excised.

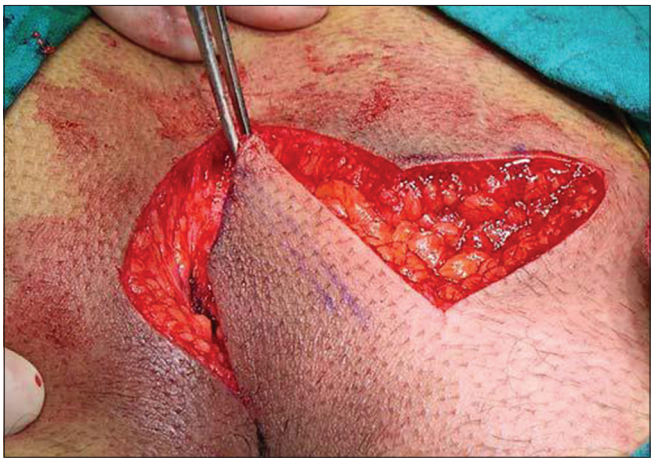

Figure 3: Fasciocutaneous flap raised.

Hemostasis was achieved by the use of electrocautery. A single multiple-hole, closed suction drain was inserted underneath the flap. Subcutaneous tissue was approximated with interrupted vicryl 2-0 suture. The skin was closed with interrupted prolene 3-0 suture (Fig. 4). Antibiotics are given for 5 days initially intravenous subsequently oral. Drain was removed

after 48–72 hours. Alternate sutures were removed on 10th postoperative day (POD).Rest of the sutures were removed on the 12th POD.

Postoperatively patients' were advised, to avoid prolonged sitting or exercise for two weeks. Hair removal either by shaving or by hair removal cream was advised for at least 1 month. Patients were followed up in OPD monthly for 6 months.

The duration of operation, postoperative pain, length of hospital stay, duration of incapacity for work, postoperative complications (infection, flap edema, wound dehiscence), and postoperative recurrence were recorded. Duration of operation was defined as the length of time between the first incision and placement of the last suture. Postoperative pain was assessed according to a visual analogue scale (VAS) from 0 (no pain) to 10 (worst pain imaginable) on the first postoperative day. Duration of incapacity for work was defined as Length of hospital stay. Duration of inability to work is defined as the time from the date of surgery to the date on which patient returned to normal activities, including employment and leisure activities.

Ethics

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

RESULTS

The study consisted of 40 males (86.96%) and 6 females (13.04%), with a mean age of 28 years (range 16–50 years). Mean operative time was about 50 minutes (range 40-50 minutes). The mean age of presentation was 28 years old (range 16–50 years old) Table 2. 29 patients (63.05%) presented with discharge, 11 presented with pain (23.9%), 4 with infection (8.6%) and 2 (4.3%) with pilonidal abscess Pain score, as calculated by VAS, has a mean of 3.65 (range 3-5). The mean length of hospital stay was 3.5 days (range 3 - 4 days) and most patients returned to work within 3 weeks. The distribution of patients according to demographic characteristics, history of the disease, operative time, pain score and duration of hospital stay are given in Table 1 and Figs. 5 and 6.

Three patients developed mild infection which was treated with antibiotics. One patient developed necrosis at the tip of the flap. One patient developed recurrent lesion 6 months postoperatively, which may be due to improper excision of the sinus (Table 2).

The stitches were removed after 12–14 days (Table 3). The time off-work ranged from 12 to 22 days. The time to walk without pain ranged from 10 to 16 days. The mean follow-up period was 4 months.

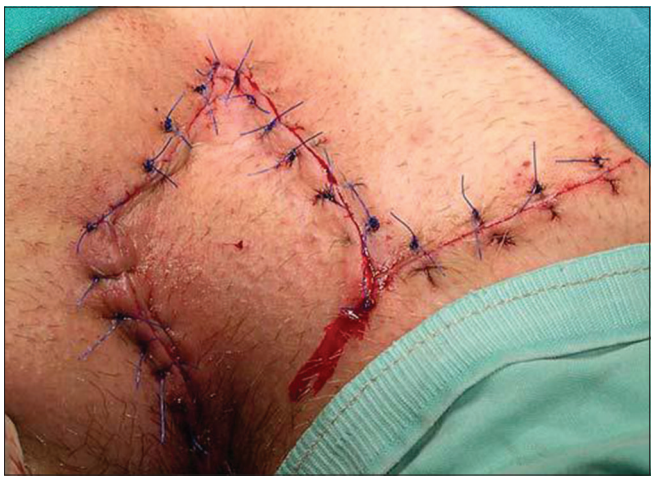

Figure 4: Flap stitched.

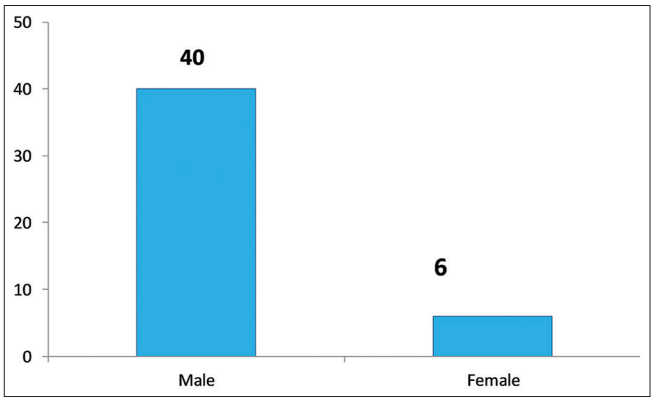

Figure 5: Sex distribution

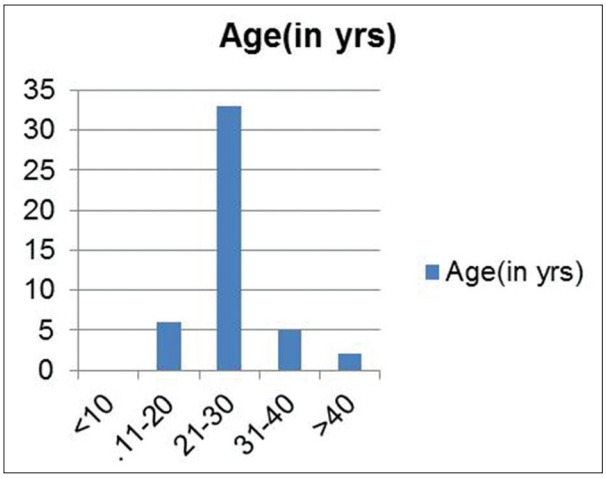

Figure 6: Age distribution

DISCUSSION

Multiple subcutaneous sinuses or abscesses containing hair characterize the disease [11]. Pilonidal sinus disease is an acquired condition affecting young adults. The etiology and pathogenesis of pilonidal sinus is still a matter of debate. Three main factors play a role in embedding of hair: Invaders formed by free hair (H-hair), the force that provides hair embedding the (F-force), and the vulnerability of the skin that lets the embedding of the hair deeper in the gluteal region (V-vulnerability) [12]. Other factors affecting the incidence are increased sweating associated with sitting and buttock friction, poor personal hygiene, obesity, and local trauma, Increase depth, narrowness of the natal cleft and the friction movements of the buttocks paves the way for loose hair to collect and insert in deep cleft [13].

The varied surgical techniques proposed for the treatment of pilonidal sinuses are evidences of the lack of an ideal procedure of management, reflecting the need for a safe and efficient surgical method for the same. The hair is perceived as a foreign body, initiates an inflammatory response and can then lead to a pocket of infection leading to abscess or sinus formation [5].

The surgical treatment should intend towards removing all the sinus tracts as well as the predisposing factors that contribute in the formation of pilonidal sinus [14].

Recurrence is the main problem associated with all surgeries described which ranged from 21.4% to 100% for incision and drainage, 5.5%–33% for excision and open-packing, 8% for marsupialization, 3.3%–11% for Z plasty [14]. The Limberg flap procedure is a safe choice for the surgical treatment of sacrococcygeal recurrent pilonidal sinus disease due to its low complication rate, short length of hospital stay, early return work, low VAS score, high patient satisfaction and shorter complete healing duration [15]. With the Limberg flap technique, internal flap cleft can be flattened and tissue can be approximated without tension.

In this study, 46 patients with sacrococcygeal pilonidal disease were managed with rhomboid excision and Limberg flap reconstruction. Recurrence was noted in one patient (2.1). Akin et al [5]. Operated on 411 patients and reported recurrence rates of 2.91%, so results were comparable to them. Superficial necrosis was seen in one patient (2.1%), which may be due to the design of the long flap or fault technique. El-khadrawy [16] operated on 40 patients and had superficial necrosis at the tip of the flap in four patients (10%). 12-18 days was Time off-work in this study which is comparable to that reported by Karaca AS et al (group II) [17].

Several studies have been reported till date and results of this study are in accordance with them (Table 4) in terms of hospital stay, complication and recurrence rate.

Exercise or sitting down on the wound to be avoided for two weeks and the patient has to return slowly to normal activities. Hair removal either by shaving the edges of the wound is mandatory [14].

The advantages of Limberg flap reconstruction are:

- Easy to perform, learn and design
- Flattens the natal cleft with a large well-vascularised pedicle that can be sutured without tension. Midline dead space and scar is avoided. Useful in complex sinuses with multiple pits where radical

Table 1: Clinical presentation

Complaint	Number	Percentage
Discharge	29	63.10
Pain	11	23.9
Infection	4	8.6
Pilonidal abscess	2	4.4
Total	46	100

Table 2: Early postoperative data

	Range
Operative time	40-75 minutes
Hospital stay	3-4 days
Healing (removal of stitches)	10-12 days
Drain removal	48-72 hours
Pain – VAS	3.6

Table 3: Postoperative complications

Complications	Number	Percentage
Infection	3	6.5
Necrosis at tip of the flap	1	2.1
Gaping	1	2.1
Recurrence	1	2.1
Total	7	12.8

Table 4: Comparison of results with other studies

Author/s	Patients (no.)	Hospital stay (days)	Complication (%)	Recurrence (%)
Katsoulis et al.[18]	25	4.0	16	-
Akin et al.[5]	411	3.2	15.75	2.91
Mentes et al.[13]	353	2-19	11	3.1
Aslam et al.[6]	110	3.0	5	1
El-khadrawy et al.[18]	40	5-11	40	10
Jethwani et al.[9]	67	2-3	11.94	1.49
Our study	46	3-4	12.8	2.1

excision leaves large defect.
- Useful in recurrent pilonidal disease.
- Reduces hospital stay and time to resume normal activities.

CONCLUSION

The study infers that "wide local excision and Limberg flap reconstruction is an effective procedure for primary and recurrent pilonidal sinus disease with low complication rates, short hospitalization, low recurrence rates, earlier healing and shorter time off-work. The surgery can be mastered easily.

The results of this study favor rhomboid excision and Limberg flap reconstruction for pilonidal disease.

Statement of Human and Animal Rights

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

Statement of Informed Consent

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

Our Dermatology Online

Original Article

Serum levels of homocystiene, vitamin B_{12} and folic acid in Indian patients with psoriasis: results of a pilot study

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ABSTRACT

Introduction: Hyperhomocystienemia has emerged an independent risk factor for cardiovascular diseases with an implication for cardiovascular morbidity in psoriasis patients. Both vitamin B_{12} and folic acid influence homocystiene metabolism as cofactors. **Aim:** To study the serum levels of homocystiene, vitamin B_{12} and folic acid in patients with chronic plaque psoriasis and age matched controls. **Methods:** 55 males aged 22-66 years with chronic plaque psoriasis of 6 months to 20 years and 55 healthy male controls aged 20-65 years were studied. **Results:** Body surface area involvement was <10% in 38 (69%), between 10-20% in 10 (18.2%) and >20% in 7 (12.8%) patients, respectively. The PASI was <6 in 41 (74.5%), 6-12 in 10 (18.2%) and >12 in 4 (7.3%) patients, respectively. The serum homocystiene levels of >12 μmol/L were higher than normal (5-12 μmol/L) in all patients and 11 (22%) controls and the difference was statistically significant. The serum vitamin B_{12} levels of <150 to 513pg/ml were on the lower side of the normal (174-878pg/ml) in all patients. The serum folic acid levels varied between 5.65 and >24ng/ml and elevated levels of 17.83 to >24ng/ml (normal 3-17 ng/ml) were noted in 17 (30.9%) patients. Except for elevated serum homocystiene in 11 (22%) controls, other biochemical parameters were within normal range. **Conclusions:** Implications of hyperhomocystienemia for cardiovascular comorbidities in psoriasis patients and whether supplementing vitamin B_{12} and folic acid will prevent comorbidities by normalizing homocystiene metabolism needs evaluation by large well designed studies.

Key words: Hyperhomocystienemia; Psoriasis comorbidities; Psoriasis

INTRODUCTION

Psoriasis is a common inflammatory dermatosis with epidermal hyperproliferation in the basal layer. Both genetic and environmental influences (trauma, infection, drugs, alcohol, smoking, metabolic factors, psychological stress) are considered important in its pathogenesis. The disease has a significant impact on health related quality of life because of lifelong chronicity, extent of severity, periodicity of flares, and more importantly, from associated comorbidities. Elevated plasma homocystiene has been widely studied as an independent risk factor for atherosclerotic disease involving the coronary, peripheral, and cerebral circulations that may result in early death from myocardial infarction, pulmonary embolism or stroke [1,2]. Psoriasis patients reportedly have significantly higher plasma homocysteine levels corresponding with severity of disease than control subjects [3]. They also demonstrate significantly lower levels of vitamin B_{12}, folate, and tissue plasminogen activator than controls. Reduced plasma folate and vitamin B_{12} levels in psoriasis patients have been attributed to their increased utilization in the skin, reduced absorption from the gut, or as an adverse effect of systemic medications like methotrexate (folate

How to cite this article: Khatri G, Mahajan VK, Mehta KS, Sharma KK, Bhushan S, Chauhan PS. Serum levels of homocystiene, vitamin B_{12} and folic acid in Indian patients with psoriasis: results of a pilot study. Our Dermatol Online. 2016;7(3):276-279.

Submission: 15.11.2015; **Acceptance:** 29.02.2016

DOI: 10.7241/ourd.20163.74

antagonist) [3,4]. Deficiencies in vitamin B_{12} and folate have been associated with increased levels of plasma homocystiene, and supplementation is shown to decrease plasma homocystiene levels [1,3]. It is possible that higher levels of plasma homocysteine perhaps contribute to the increased risk for cardiovascular morbidity observed in patients with psoriasis [5]. We studied serum levels of homocystiene, vitamin B_{12} and folic acid in Indian patients with psoriasis and age matched controls.

MATERIAL AND METHODS

Serum levels of homocystiene, vitamin B_{12} and folic acid were studied in 55 consecutive adult males having chronic plaque psoriasis for at least 6 months after written/informed consent during Jan-Dec 2012. The study was approved by the Institutional Protocol Review Board and Institutional Ethics Committee (Rgn no ECR/490/Inst/HP/2013). Patients were instructed to stop taking alcohol, coffee or any topical/systemic treatment for 1 week, 2 weeks and 4 weeks, respectively, and consumption of animal protein 24 h prior to blood sampling. Only topical emollients and oral anti-histaminics were allowed. Patients having palmoplantar psoriasis, psoriatic arthritis, systemic diseases (thyroid, hepatic or renal disease, hematologic disorders diabetes mellitus, coronary heart disease, stroke, peripheral vascular disease, systemic lupus erythematosus), on antifolate medications (anticonvulsants, penicillin, levodopa, cyclosporine, isoniazide), or drugs that cause hyperhomocystienemia (phenytoin, carbamazapine, theophylline, oral contraceptives, azathioprine, thiazide diuretics, metformin), were excluded from the study. Patients with history of substance (opium) abuse and current smokers were also excluded. A detailed demographic profile, medical history and clinical details of psoriasis were recorded. Body Surface Area (BSA) involvement was calculated as per 'Rule of Nines' and the Psoriasis Area-and-Severity Index (PASI) score was determined as suggested originally by Fredriksson and Pettersson [6]. Fifty-five age-matched males with minor dermatoses (scabies, dermatophytoses) were enrolled from the outpatient clinic after informed written consent as controls for serum sampling in a similar manner.

Venous blood (5ml) samples were collected after overnight fasting between 8.00 and 10.00 AM for complete blood count including platelets, fasting blood glucose, urea, creatinine, bilirubin, serum glutamic oxaloacetic transaminase and serum glutamic pyruvate transaminase, alkaline phosphatase and thyroid functions tests. Quantitative estimation for serum homocystiene vitamin B_{12} and folic acid levels was performed in institutional biochemistry laboratory by standard chemiluminescence enzyme immunoassay (CLIA) method [7] and as per manufacturer protocol using Immulite® ready to use *in-vitro* kits purchased from Siemens Healthcare, Diagnostic Products Ltd, United Kingdom. Results were analyzed using unpaired student's *t*-test and standard deviation for mean. A '*p*' value <0.05 calculated at 5% level (95% confidence limits) was considered statistically significant.

Ethics

This study was performed on human subjects; thus, all patients were aware of the presence of the study and they were fully informed about study parameters.

RESULTS

These 55 male patients aged between 22 and 66 years had 25 (45.5%) patients between 22 and 40 years and comprised the majority. Twenty-three (41.8%) patients were aged 41 - 60 years, and 7 (12.7%) patients were >60 years of age. They had psoriasis for 6 months to 20 years at the time of visit. In 20 (36.4%) patients, the psoriasis was present for >5 years while 35(63.6%) patients had the disease for <5 years. Body surface area involvement was <10% in 38 (69%), 10-20% in 10 (18.2%) and >20% in 7 (12.8%) patients, respectively. PASI score was <6 in 41 (70%), 6-12 in 10 (18.2%) patients and >12 in 4 (7.3%) patients, respectively. The controls comprised 55 males aged between 20-65 years.

Biochemical Parameters

None of the patients or controls showed any alteration in routine hemogram, serum biochemistry or thyroid function tests. Other studied biochemical parameters of 55 patients and 55 controls are tabulated (Table 1). Elevated homocystiene levels (normal 5-12 μmol/L) in the range of 12.8 to >50 μmol/L (mean 31.49 ± 9.99 μmol/L) were seen in all patients while in controls the values ranged between 5.1 and 22.3 μmol/L (mean 9.99 ±3.71 μmol/L). The difference was statistically significant when compared with controls. However, the elevated serum homocystiene levels did not vary with BSA/PASI score (Figs. 1a,b,c and 2a,b). The vitamin B_{12}

levels in patients ranged from <150 to 502 pg/ml (mean 230.74 ± 64.09 pg/ml) and were on the lower side of the normal range (174-878 pg/ml). Serum vitamin B_{12} measuring between <150 and 156 pg/ml were lower than normal in 6 (10.9%) patients and controls each. Mean serum folic acid levels in all the patients were 14.42 ± 5.21 ng/ml and ranged from 5.65 to >24 ng/ml (normal 3-17 ng/ml). The mean value of serum folic acid levels of >17ng/ml (17.83 - >24ng/ml) in 17 (30.9%) patients was above the normal range. The mean value of serum folic acid levels of controls was 13.51±4.64 ng/ml (range 5.9-25ng/ml). Overall, except for slightly elevated serum homocystiene levels (12.1-24 μmol/L) in 11 (22%) controls, all parameters were within normal range and the difference was not statistically significant (Table 2).

DISCUSSION

High plasma homocystiene is considered an independent risk factor for coronary artery disease, stroke, peripheral vascular disease and possibly Alzheimer's disease especially in patients with homocystienuria [8]. A non-linear and inverse association between plasma homocystiene concentration, vitamin B_{12} and plasma folate concentration has been well documented and attributable to either deficient absorption or excessive utilization of folic acid, vitamins B_6 and B_{12} [3]. All the 55 (100%) patients in our study had high serum homocystiene levels (mean 31.49 ±9.99 μmol/L, range 12.8 - >50 μmol/L) as compared to the controls (mean 9.99 ± 3.71 μmol/L, range 5.1-22.3 μmol/L) and the difference was statistically significant (p <0.05). The levels of serum vitamin B_{12} were at the lower levels (mean 217.7±46.22 pg/ml, range 150-310 pg/ml) of the normal in 26 (86.6%) patients while in 6 (10.9%) patients the values were lower (<174 pg/ml) than the normal.

Table 1: Biochemical parameters of patients and controls

Range	Patients (%) n=55	Controls (%) n=55
Serum homocystiene levels (μmol/L), normal 5-12 μmol/L		
0-12	0	44 (80)
12.1-24	12 (21.8)	11 (20)
24.1-36	27 (49.1)	0
36.1-48	12 (21.8)	0
>48	04 (7.3)	0
Serum vitamin B12 levels (pg/ml), normal 174-878 pg/ml		
1-173	6 (10.9)	6 (10.9)
173.1-275	38 (69.1)	38 (69.1)
275.1-374	9 (16.4)	7 (12.7)
374.1-474	2 (3.6)	4 (7.3)
Serum folic acid levels (ng/ml), normal 1.9-25 ng/ml		
3-10	14 (25.5)	15 (27.3)
10.1-17	24 (43.6)	33 (60)
>17	17 (30.9)	7 (12.7)

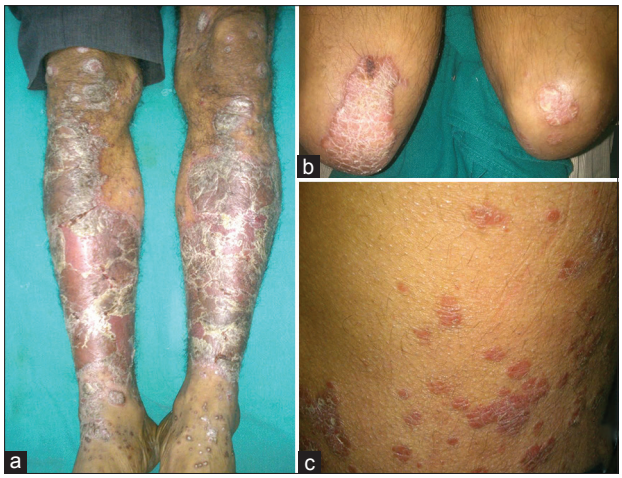

Figure 1: (a) A patient of psoriasis with BSA-30% and PASI 8.4 had Serum homocystiene 25.6μmol/L, Serum vitamin B_{12} 156pg/ml, and Serum folic acid 15.9ng/ml. (b and c) This patient had BSA-10%, and PASI 4.9, and Serum homocystiene 36 μmol/L, Serum vitamin B12 <150 pg/ml, and Serum folic acid 7.7ng/ml.

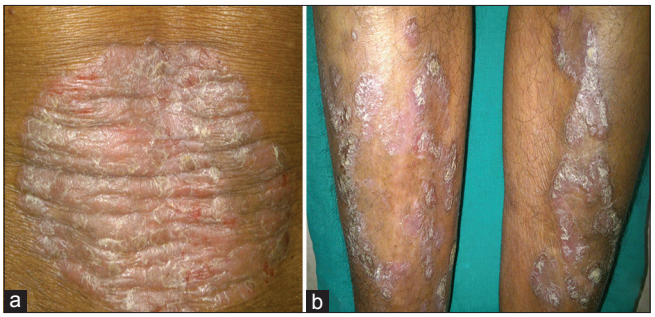

Figure 2: (a) A patient of psoriasis with BSA-5% and PASI 4 had Serum homocystiene >50 μmol/L, Serum vitamin B_{12} <150 pg/ml, and Serum folic acid >24ng/ml. (b) This patient had BSA-3% and PASI 4.1, and Serum homocystiene >50μmol/L, Serum vitamin B_{12} 156pg/ml, and Serum folic acid >24ng/ml.

Table 2: Significance of the results

	Serum homocystiene (N=5-12 μmol/L)		Serum vitamin B_{12} (N=174-878 pg/ml)		Serum folic acid (N=3-17 ng/ml)	
	Patients	Controls	Patients	Controls	Patients	Controls
Range	12.8- >50	5.1-22.3	150-502	150-513	5.65- >24	5.9-25
Mean	31.49±9.99	9.99±3.71	230.74±64.09	235.1±77.63	14.42±5.21	13.51±4.64
p value	<0.00001		0.357		0.166	
	Significant		Not significant		Not significant	

Note: p value<0.05 was considered statistically significant

The difference was not statistically significant when compared with controls. Serum folic acid levels varied between 5.65 and >24 ng/ml (mean 14.42±5.21 ng/ml) and 5.9 and 25 (mean 13.51±4.64 ng/ml) in all the patients and controls, respectively, and were within normal range. Brazzeli et al [9] also made similar observations in a cohort of 98 patients with chronic plaque psoriasis and 98 healthy controls. They observed significantly higher prevalence of hyperhomocystienemia and low serum vitamin B_{12} levels in psoriasis patients as compared to healthy controls but not for serum folic acid. In a similar study, Malerba et al [3] noted higher plasma homocystiene levels and lower folic acid levels in 40 chronic plaque psoriasis patients without known risk factors for acquired homocystienemia than 30 age-matched controls. Although the plasma homocystiene levels in patients with psoriasis also correlated directly with disease severity and inversely with folic acid levels, no abnormalities were detected in plasma vitamin B_6 and B_{12} levels. In a similar study by Cakmak et al [10] serum homocystiene levels inversely correlated with serum folic acid levels but not with serum vitamin B_{12} both in 70 patients with psoriasis and healthy controls. However, authors did not find any difference between their serum levels in patients and controls. Trends towards low serum folic acid and hyperhomocystienemia in patients with psoriasis were also observed by Tobin et al [11] but they did not study serum vitamin B_{12} in their patients. Our observations of hyperhomocystienemia, serum vitamin B_{12} at lower end of the normal and variable serum folic acid levels in all patients are suggestive of some significance of hyperhomocystienemia and low serum vitamin B_{12} and folic acid levels. However, a possibility of avoiding non-vegetarian food altogether by these patients subsequent to development of their disease leading to serum vitamin B_{12} at lower end of the normal range cannot be ruled out entirely. Hyperhomocystienemia in patients with psoriasis as in our 100% patients has been documented previously [3,9-11]. Contrarily, Uslu et al [12] in a recent study of 50 patients with psoriasis and 48 healthy controls found no statistically significant differences between the patients and the control group in terms of age, sex, body mass index (BMI), plasma homocystiene folic acid, and vitamin B_{12}. They attributed this variation to the differences in the genetic pool of the studied Turkish population.

CONCLUSIONS

Hyperhomocystienemia, lower than normal levels of vitamin B_{12} and variable serum folic acid levels in all our patients suggests their possible dysregulation in psoriasis patients. Implications of hyperhomocystienemia for cardiovascular comorbidities in psoriasis patients and whether supplementing vitamin B_{12} and folic acid will prevent comorbidities by normalizing homocystiene metabolism needs further evaluation by large well designed studies in different ethnicities.

Statement of Human and Animal Rights

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

Statement of Informed Consent

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

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

Our Dermatology Online

Brief Report

Breast rhagades in nursing and non lactating women: A gordian knot to loose by the aid of a phytocosmetical formulation

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ABSTRACT

We have tried to solve an everlasting problem inherent the treatment of breast rhagades in woman, since mammary gland proposes several troubles: 1) drug penetration through the mammal areola epidermis is the quickest, after the one through eye conjunctive and so whichever chemical or natural substance (for instance triterpenes or cyanhydrin or amygdalin present in sweet almond oil) may be retrieved in human milk and blood after few minutes) 2) Milk is suckled by the bébé and it is supposed that whichever remedy against breast rhagades should be applied topically before and after each feeding and thus most of the substances included in the remedy passes into human milk. 3) Mammary gland is victim of the aggression of manifold germs and bacteria from the bébés' mouth. There is an avalanche of cosmetic and pharmaceutical commodities that always fail in some of the aforesaid corollaries. Our gélée tends to comply all the prerequisites and we have demonstrated this hypothesis.

Key words: Breast density; Cukjati-Rebersek's method; The Clock method; Paeonia albiflora root extract; Vernier calliper

INTRODUCTION

We are deeply persuaded that breast rhagades do not represent an exclusive burden of competence of obstetricians but may be studied and analysed even from a dermal cosmetological point of view. It is a matter of fact that breast rhagades whenever untreated may lead to infective mastitis and seldom to severe abscesses, fibrosing adenosis, idest the proliferation of myothelial and intralobular connective tissue, that is the chief responsible of the building up of the whole gland that constitutes breast itself. It is well documented that between 10% and 33% of breast-feeding women develop lactation mastitis [1,2], moreover that the incidence is highest in the first few weeks postpartum, decreasing gradually afterwards [3] and that however, abscesses and patent infective mastitis may occur as long as the woman is breast-feeding.

Rhagades are painful fissures of the nipple and areola, which appear after 2-3 days after delivery and are usually caused by incorrect nursing. They very often have an infective basis, causing the entry of germs that may cause mastitis (and too often they are bacteria deriving from the baby's mouth entering the milk duct through a crack in the nipple) Obstetricians of all over the world suggest to prevent the occurrence of rhagades by always cleaning the nipple and areola carefully before and after feeding.

No specific drug treatments are available hitherto, in order to avoid the incidence of rhagades, and thus mastitis and finally fibroadenomas. It is suggestive to introduce that breast density is of relevant importance in dealing this concern:

Breast density is a measure used to describe the proportion of the different tissues that make up a

How to cite this article: Martini L. Breast rhagades in nursing and non lactating women: A gordian knot to loose by the aid of a phytocosmetical formulation. Our Dermatol Online. 2016;7(3):280-283.

Submission: 10.11.2015; **Acceptance:** 05.01.2015

DOI: 10.7241/ourd.20163.75

woman's breasts. Breasts are made up of fat and breast tissue (the milk ducts and lobules, which may be called glandular tissue). It is actually connective tissue which helps hold everything place. Breast density is not a measure of how the breasts feel, but rather how the breasts look on a mammogram. It compares the area of breast and connective tissue seen on a mammogram to the area of fat.

High breast density means there is a greater amount of breast and connective tissue compared to fat. Low breast density means there is a greater amount of fat compared to breast and connective tissue. The American College of Obstetricians and Gynecologists of New Jersey (5) draws the following classification:

Almost entirely fatty - 10% of women
Scattered areas of fibro-glandular density - 40% of women
Heterogeneously dense - 40% of women
Extremely dense -10% of women

Breast infections most commonly occur one to three months after the delivery of a baby, but they can occur in women who have not recently delivered as well as in women after menopause.In healthy women, mastitis is rare. However, women with diabetes, chronic illness or an impaired immune system may be more susceptible.

It has been referred too that Asian women are less prone to breast cancer, since their absolute area breast density (as Breast density is associated with breast cancer risk) is minor than that of White women and Black/African-American ones [4,5], anyway it is still unclear if Asian women are less prone to breast rhagades than White or Black ones.

And is this the principal intent of our study.

MATERIALS AND METHODS

We have recruited nine women, three Black/Africans, three Asians, three White/Europeans and we have had the fortunateness to dispose of three classes of subjects:

a) (1,2,3) three women, one Asian, one White and one Black 54-59 years old, all three in menopause
b) (4,5,6) three girls, one Asian, one White and one Black 27-32 years old, all three nursing
c) (7,8,9) three girls, one Asian, one White and one Black 19-27 years old, which put out to nurse their babies and use to pull the milk with a breast pump 3-6 times a day.

Each of everyone is affected by severe nipple fissures and in two cases (2 and 9) by evident abscesses.

The cosmetic ingredients of the gélée we have prayed the individuals to spread 6 times a day for three weeks belong to the following botanical categories:

One anti-oedema herb: idest Paeonia albiflora root ethanol extract

Some essential oils endowed by carminative and anti-borborygmus functions: caraway, cumin, anis, meant, fennel oils [6].

One anti-inflammatory plant: Achillea millefolium (entire plant) tincture.

The gélée is made of magnesium stearate and ethanol.

The choice of the essential oils is due to the fact that the essential oils themselves are capable to perform digestive and carminative function when, once passed into human milk, this is suckled by the newborn, so that he has less probabilities of meteorism and diarrhetic phenomena.

Moreover, it must be asserted that Hippocrates of Chios used to advice nannies and puerperas to spread fennel oil onto their breast, since it acts as lactopoietic and lactogenic.

There is no evidence of any surreptitious toxic effect of Poenia albiflora root extract and/or Achillea millefolium tincture [7] on human organism, when ingested, indeed their flavour is gentle and appetizing, enev when passed into human milk for the bébé.

Diagnostic and analytical methods employed to determine the final recovery are the following:

α) The Cukjati-Rebersek's method, that indicates the capacity of wound healing and expresses a dimensionless value obtained calculating the greatest average wound margin distance from the wound centre divided by the time in weeks necessary to the complete wound closure. The standard reference parameters, idest topical applications of emulsions of Neomycin sulphate or Hypericum perfoliatum oil, are for instance 0.062 after 4 weeks, 0.484 after 5 weeks and 0.900 after 6 weeks.
β) The Clock method, that indicates the Decrease of periareolar erythema and consists in employing

acetate sheets (6X6 cm) to lay onto the area afflicted by the erythema and measuring across the surface by the aids of a fine-point transparent film-making pen from the 12:00 to the 06.00 position to the outer margin of the periwound erythema and then from the 03.00 to the 09.00 position and to the same outer margin and multiplying the values obtained.

The score is simply the result of the multiplication of the mathematical differences between the two distances in the two cases (idest the distance in cm from 12:00 to 06:00 and from 03:00 and 09:00).

Measurements are carried out every three days, and it is evident that the dimensionless score decreases and a significant plot can be drawn.

γ) Measurement with a vernier calliper that gives reason of the percentages of fat present in the gland, and scores are calculated by the Regnault's algorythms. The values obtained do represent the nipple and surrounding areola elasticity obtained after the treatment.

In Table 1 it is possible to observe the range of standard values obtained by pinching with a regular vernier calliper the nipples and areolas of healthy girls and women, depending on the subject's age, that indicate the percentage of fat in the entire gland, and the increments of percentage do show the elasticity reconvery of the nipple and areola of the individuals, subjects of our experimentation.

Ethics

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

RESULTS AND DISCUSSIONS

All the tables recorded below refer to the ameliorating of parameters during the three weeks of topical application of our gélée (Tables 2-4).

It is suggestive to notice that as far as the capacity of wound healing of the breast rhagades, the percentage of wound healing (the complete recovery during the three weeks of experiences) corresponds to the average value of 96.16%.

Table 1: Percentages of breast fat in healthy woman scored by the use of a generic skinfold vernier calliper

Percentages of breast fat in healthy woman scored by the use of a generic skinfold calliper (according to Regnault's classification based on algorithms) (LMS scale)			
% Fat for average value determined by pinching nipple and areola			
Average value in cm	Age		
	16-29	30-49	50-59
1.4	9.4	14.1	17.0
1.6	11.2	15.7	18.6
1.8	12.7	17.1	20.1
2.0	14.1	18.4	21.4
2.2	15.4	19.5	22.6
2.4	16.5	20.6	23.7

Table 2: Values of increment of the recorded percentages of complete wound healing of breast rhagades during the three weeks of treatment with our gélée

Case	Cukjati-Rebersek's score		Percentage of recovery (complete wound healing)
	Initial	Final	
1	0.03	0.74	95.95
2	0.02	0.81	97.54
3	0.01	0.66	98.49
4	0.04	0.59	93.23
5	0.02	0.89	97.76
6	0.05	0.74	93.25
7	0.01	0.52	98.08
8	0.01	0.83	98.80
9	0.06	0.79	92.40

As far as the erythemal regression is concerned, it can be observed that a decrement of 32.90% is achieved.

The scoring of percentage of fat in the entire gland that is recovered by the use of the géléee is more complicated, since it must be considered that Case 1, 4 and 7 are represented by the Asian individuals that present a minor percentage of breast fat, when safe and healthy, and Case 3,6 and 9 correspond to the Black women that present the highest percentages of breast fat.

Notwithstanding all nine start from low values of breast fat, recorded at the very beginning of the experience, since rhagades compromise the breast density, the final scores demonstrate an elevate average increment of breast fat, even if it is not valuable (we deem) to declare the general increase, as every case is to be regarded as a single fact.

And, as announced in the introduction, the amount of the breast fat is synonym of elasticity recovery of the nipple and areola.

Table 3: Values of decrease of the recorded percentages of erythema in the surface afflicted by breast rhagades during the three weeks of treatment with our gélée

Case	Value of clock method's score at 3rd day	Value of clock method's score at 6th day	Value of clock method's score at 9th day	Value of clock method's score at 12th day	Value of clock method's score at 15th day	Value of clock method's score at 18th day	Value of clock method's score at 21st day	Percentage of erythemal regression
1	44	42	38	32	29	23	12	27.27
2	51	50	46	36	38	29	18	35.29
3	47	45	43	37	33	25	16	34.04
4	39	31	30	27	25	23	17	43.58
5	58	54	52	51	48	34	14	24.17
6	38	37	31	29	27	24	13	34.21
7	50	48	46	41	42	38	18	36.00
8	33	30	30	29	27	25	11	33.33
9	46	44	41	35	33	28	13	28.26

Table 4: Values of increase of the recorded percentages of breast fat in the glands afflicted by rhagades during the three weeks of treatment with our gélée

Case	Initial value by vernier calliper	Final value by vernier calliper	Percentage of increment of breast fat
1	16.7	25.1	66.50
2	18.8	24.7	76.11
3	17.5	27.4	63.80
4	21.9	32.1	73.81
5	14.1	19.9	68.58
6	13.7	28.2	48.58
7	21.8	29.3	74.40
8	12.4	20.9	59.50
9	16.1	31.0	51.93

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

Our Dermatology Online

Brief Report

A precious chance for muslim hijab women of all the world to keep their hair scalp safe and not to incur pracox alopecia

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ABSTRACT

The scope of this research is to develop a revolutionary formula for a shampooing apt only for Muslim Hijab girls and women who, because of the fact that are forced to cover their hair scalp, are more prone than others to baldness, excess of sebum production and hair brittleness and dandruff. We have created a special shampoo that could remedy some incongruities remarkable in hair scalp of females who never have contact with air, sun or revitalizing physical agents. We have stated that after 7 daily applications of the cosmetic product onto hair scalp, before to go to bed, a suggestive lowering of sebum production may be recorded, and sebum is the chief cause of baldness and itching and dandruff, especially when hair is covered and cannot transpire at all. Hijab girls and women may remedy their own problems, due to religious tenets, by the daily usage of a shampooing, as it is recognized that Muslim females have more time for themselves, than other women of other cultures and traditions.

Key words: Hijab woman; Sebum production; Cholecalciferol; Squalene; Musa sapientum peel extract

INTRODUCTION

Muslim hijab women and girls represent a huge amount of individuals that are forced and/or like, for sake of their religious credos, to cover their face and head when in public spaces, throughout the entire world so that a neologism has been created, that is Eurabia.

This coinage indicates the complete fusion between Moohummudan Culture and Occidental Traditions, and thus the cult of Hijab is rather common and widespread.

It is indeed curious to stress that some countries exist which could be declared "burqa-ban", for instance, Albania where Government has introduced a draft legislation to ban the hijab in public schools, Belgium where several by-laws have been draft on face-covering clothing to ban public wearing of the niqab and burqa, Denmark, where the need for passengers to show their faces in airports is mandatory, Estonia, where the Ministry of Justice is going to prepare a bill that would ban wearing a burqa or niqab in public spaces, such as state offices, schools and hospitals, France, where the 2004 French law on secularity and conspicuous religious symbols in schools bans all clothing which constitutes an ostensible religious symbol from government-operated schools (according to the holy principle of Laïcité), Germany, where the states of Baden-Württemberg, Bavaria, Hesse, Lower Saxony, Saarland, Bremen, North Rhine-Westphalia contain restrictions on wearing the hijab by female teachers and the city-state of Berlin that banned all religious symbols in public institutions, including the Christian crucifix and the Jewish kippah,Italy where Law 152/1975 – which prohibits the use of motorcycle helmets to evade identification – cannot be extended to cover the veil or burqa even if Region Lombardy has banned facial veils for security reasons in government buildings and hospitals, in December 2015, Kosovo, where headscarves in schools

How to cite this article: Martin L. A precious chance for muslim hijab women of all the world to keep their hair scalp safe and not to incur pracox alopecia. Our Dermatol Online. 2016;7(3):284-287.

Submission: 19.01.2016; **Acceptance:** 29.03.2016

DOI: 10.7241/ourd.20163.76

were banished since late 2009 because Kosovo is proclaimed secular territory, The Netherlands, where Government Parliament in January 2012 enacted a ban on face-covering clothing, popularly described as the "burqa ban" and where offenders can be fined up to 390 euro even if the prohibition does not apply to face covering that is necessary for the health, safety or the exercise of a profession or practicing a sport, although some events such as Sinterklaas, Carnival, Halloween or when a mayor granted an exemption for particular kermises are excluded, Switzerland, where in September 2013, by popular referendum approved by 66% of the voters, the canton of Ticino prohibited to hide the face in a public area, and finally Norway, Spain, Sweden and United Kingdom where this matter is controversial till today, due to politic, more than religious, diatribes.

Here follows a quotation referred by a woman speaking Urdu (that is the lingua franca of Pakistan and other Indian states) that has been retrieved in a forum [1] where Muslim women and girls argue about the concern of the pro and contra of wearing hijab: "If your hair is covered all day it can cause sweat and sebum to build up and deposit on your scalp and cause itching and dandruff…".

Other cybernauts decree the following assertions: Tying up wet hair is already known to be damaging to the roots of the hair, but it can even worse for hijabis because the hair doesn't get to naturally dry and stays tied up for hours. Tying up wet hair adds another element to the dreaded condition of 'hijab-hair' since the hair ends up drying flat against the scalp so that when you remove your hijab, probably hours later, your hair is flat and limp.

And moreover:

Given the fact that you wear hijab pretty much all day, chances are your hair and scalp probably don't see much sunlight. And depending on what kind of neighbourhood you live it, it might not be feasible for you to just sit outside in your background with your hijab on.

So we suggest opening up in the blinds and curtains in your home and sitting in the sunniest part of the house for at least half an hour a week. Vitamin D is essential for the hair and scalp, and just because you keep your hair covered doesn't mean that your hair can't get any! So let in the sunlight and soak up some rays.

And furthermore:

When you come home, take your hijab off! Open up your hair, take out that hair tie, and run your fingers through the hair and scalp to get some circulation going. Some of us wear hijab for several hours a day, depending on our lifestyle, and our hair doesn't really get to breathe. So maximize those hours when you're home and don't have to cover up- let your hair loose and let your scalp breathe.

The chief problem of hijab women is represented by flourishing of dandruff, sebum and by a very scarce microcirculation, due to the fact that transpiration (the so-called breathing) of hair scalp is foreclosed [2-6].

A shampooing mousse to suggest to keep off Muslim hijab girls and women from hair damage, shall keep on account of the hypersecretion of sebum and dandruff, the quasi-total absence of incidence of sun rays (that represent the source of life and indisputed benefit for hair and skin as well) and the scarce or reduced microcirculation.

A shampooing product apt to loose these problems shall contain:

1) A surrogate of human sebum, apt to avoid sebaceous glands producing too much sebum (by means of the phenomenon of the pharmacological negative feedback, similarly to the action displayed by phytosterols which are able to low the LDL in blood serum). Sebum contains a variety of different lipids, including cholesterol, glycerides, fatty acids, squalene, and wax and cholesterol esters, even if the exact composition of sebum varies with a person's age.
2) A natural antidandruff agents (Musa sapientum peel and pulp extract and brown rice vinegar) [7]
3) Cholecalciferol (vitamin D3), that recently has achieved resounding success for topical use in case of xerotic stratum corneum and keratinocytes in general [8], even if in absence of sun rays incidence. It must be considered that Cholecalciferol is prohibited as cosmetic ingredient, even though cholecalciferol-PEG 12 ether is admitted.
4) An activator of microcirculation, and for this we have chosen caffeine derived from Theobroma cacao.

MATERIALS AND METHODS

We have created the formula of a shampooing mousse that contains the following ingredients: Aqua; Potassium oleate; shea butteramide/castoramide dea; Sorbitol; Caprylic/capric glycerides; Squalene; Cholesterol; Cholecalciferol peg 12 ether; Musa sapientum peel extract; Musa sapientum pulp extract; Brown rice vinegar; Cyamopsis tetragonoloba gum; Caffeine; Preservatives and Antioxidants q.s.

We have moreover selected nine volunteers (hijab women and girls, aged from 18 to 60 y.) who had previously declared to wear hijab more than 10 hours pro day and to wash their hair scalp twice a week, using a normal shampooing product.

Seven of the nine individuals suffer from excessive dandruff and sebum hypersecretion (A,C,D,E,G,H,I); two of them,(B,F) indeed, were prone to alopecia praecox.

It is utterly arguable if the baldness would have occurred in these two individuals, whenever they had never worn hijab, albeit the two cases assert they have no family history of alopecia, of whichever type.

The shampoo we have ideated was consigned to the nine individuals, which were prayed to use it every day for one week, each afternoon before to go to sleep, rigorously their hair loose.

We have been able to detect the decrease of the spontaneous sebum production by the aids of a Sebometer (SM 810. Courage- Khazaka electronic) apt to measure the sebum quantity in μg/ft2 and we have the starting measure made the first day (before the first application of our cosmetic system) and afterwards the second, the third, the fourth, the fifth, the sixth and the final evaluation every morning, applying the sebometer onto the scalp of each of every volunteer.

We plotted the results in the following Table 1 and commented the success of the activity of the shampoo by calculating the percentages of the decrease of sebum production.

In Table 2 the percentages of decrease of sebum production recorded for every volunteer are plotted.

It is remarkable that in case F, the individual most prone to praecox alopecia, the initial production of sebum is the most consistent, before treatment, but the percentage of decrease of sebum secretion is evidently drastic.

Two are the plausible theories to explain this phenomenon in women (idest in the Female pattern alopecia).

The former is based on the fact that the angle of follicles and resultant sebum building-up in women is different respect to men: effectively, the angle at which hairs come out of a female scalp allows the sebum to "run off", whereas the angle of hairs on a male scalp can lead to oil blockage.

The latter hypothesis is that scalp skin contains more clusters of inflammatory cells than

almost all other areas of non-scalp skin. These cell clusters are often very close to follicles, and they contain immune cell types that are not normally present in healthy skin and do contain mastzellen, lymphocytes and neutrophils.

The increasing of the amount of inflammatory cells usually indicates a response to some odd that may occur to damage scalp skin, and in this case the reactions are represented by an excessive production of sebum followed by a progressive hair loss.

Albeit, when sebum production decreases, due to the negative feedback evoked by the surrogate of human sebum applied onto hair scalp, even hair loss must diminish as well.

Table 1: Values of sebum production detected during the treatment by means of the Sebometer

Progressive day of evaluation	A	B	C	D	E	F	G	H	I
1st day	89801.09	87564.30	91896.21	87965.23	88635.22	86915.45	89991.07	88673.28	92785.40
2nd day	79905,90	76854.22	80988.72	82117.04	86342.50	80177.76	84768.39	81243.56	89553.28
3rd day	71581.79	70785.39	69997.87	72451.02	71425.68	72463.28	79265.34	77354.68	78635.37
4th day	58101.56	57356.20	58739.22	57979.89	56342.10	55472.19	54879.33	57362.85	57999.98
5th day	54868.53	53681.29	54687.33	56397.60	55315.54	52314.98	51888.97	53672.30	54712.55
6th day	47139.00	46377.98	44978.33	46891.32	45389.99	47325.11	46983.25	47320.21	46782.41
7th day	43097.72	41223.89	42561.89	40897.24	41667.23	42576.78	41889.99	41342.87	40235.48

Table 2: Percentages of decrease of sebum production recorded for every volunteer

Volunteer	Percentage of lowering of sebum production
A	47
B	47
C	46
D	46
E	47
F	48
G	46
H	46
I	43

CONCLUSION

It is noticeable that a stratagem of a daily usage of a shampooing product apt to reduce sebum hypersecretion and consequently to avoid dandruff, itching and praecox alopecia in women who cannot let their hair scalp "breathe", could be forecasted as a Columbus' egg.

It is notorious that the majorpart of all the trademark shampoos of all over the world do contain salts (designed as soaps) of sodium or other alkaline or alkaline earth metals with aliphatic acids,

(they represent the surface active agents tout court), and especially sodium lauryl sulphate has been pointed since decades as a progressive hair damaging agent [9-11].

So, a shampooing that contains Potassium oleate as detergent, idest Savon de Castille (or Marseille soap) and other ingredients apt to decrease the sebum production is really welcome especially by women and girls who has not the chance to let their hair to "transpire", because their hair is almost always covered and suffocated by fabric and veils.

Statement of Human and Animal Rights

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

Statement of Informed Consent

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

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

Our Dermatology Online

Case Report

Refractory lymphedema of the hand: an unusual presentation of recurrent herpes simplex virus infection

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ABSTRACT

Introduction: Herpes Simplex Virus (HSV) infection of the hand resulting in lymphatic complications such as lymphangitis and lymphedema is exceedingly uncommon. Although these complications typically resolve in 21 days, they can be persistent and may not resolve even with antiviral use, thereby mimicking dyshidrotic eczema or a bacterial event and often being misdiagnosed and inappropriately treated as such. We report a case of frequently recurring HSV infection of the hand over a long period of time resulting in refractory lymphedema which did not resolve with antiviral treatment. We further endeavor to raise awareness about this highly unusual presentation of HSV infection. A comprehensive review of the literature was conducted for similar cases using PubMed and Medline. **Case Report:** This is the first reported case with nearly a decade-long interval between the onset of primary HSV infection and the development of chronic lymphedema. Although valacyclovir significantly reduced the episodic aggravation of the lymphedema, it did not entirely resolve it. Similar cases of persistent lymphedema also included a long history of untreated and recurrent HSV infection of the hand, suggesting that this lymphatic outcome may be circumvented by prompt treatment with antivirals. **Conclusion:** This case report not only presents a highly uncommon lymphatic manifestation and unusual timeline of exacerbation of the very common HSV infection, but also highlights the importance and benefits of early initiation of antiviral therapy and the prevention of reactivation.

Key words: Herpes simplex; Lymphedema; Communicable diseases; Viruses

INTRODUCTION

Herpes simplex virus (HSV) infection usually presents with grouped vesicles on an erythematous base. The two serotypes of this virus, HSV-1 and HSV-2, most commonly affect the labial mucosa, and the genital mucosa, respectively, in addition to the surrounding skin. While both serotypes can result in infection in different body sites, recurrent infection of the hand is more commonly caused by HSV-2 and the majority of these cases are seen in the age range of 20-40 years, with a notable preponderance of women [1]. HSV infection of the hand has a variety of symptoms and manifestations, yet lymphangitis and lymphedema of the hand are infrequently observed as a consequence [2]. Furthermore, recurrent HSV infection of the hand resulting in persistent lymphedema that is resistant to antiviral medicines is highly uncommon with only two cases reported in the literature [2,3].

We report a case of unresolved HSV-induced lymphedema of the hand which developed almost one decade following the onset of primary HSV infection and following frequent episodes of reactivation throughout this period. Such prolonged duration between the onset of primary HSV infection and presentation of chronic lymphedema has not previously been reported. We further review other comparable

How to cite this article: Majdzadeh A, Al Jasser MI, Kalia S. Refractory lymphedema of the hand: an unusual presentation of recurrent herpes simplex virus infection. Our Dermatol Online. 2016;7(3):288-290.

Submission: 01.12.2015; **Acceptance:** 25.01.2016

DOI: 10.7241/ourd.20163.77

cases in the literature and highlight the similarities between cases of resolved and unresolved lymphedema of the hand.

CASE REPORT

A 33-year-old Caucasian female presented to our clinic with a two-and-a-half-year history of persistent swelling of the left hand and forearm. She further revealed that there was a history of painful episodic vesicular eruptions that occurred on the dorsal aspect of her left middle finger. Her first presentation of the vesicular eruption alone occurred at the age of 21. Since the development of the left hand and forearm swelling, the episodes recurred every one to two months. Each episode lasted approximately one week with an exacerbation of the swelling occurring within two days of vesicle formation. The patient has no known family history of any relevant skin disease. She denied any history of confirmed oral or genital herpes. Lymphatic massage drainage and compression bandages were previously recommended for managing the swelling, but worsened the condition.

Physical examination revealed non-pitting edema of the left hand and forearm (Fig. 1). During her subsequent clinical visit, grouped vesicles on an erythematous base involving the left middle finger were seen. No axillary or cervical lymphadenopathy was noted. Viral culture acquired from the base of a vesicle was positive for HSV-2. A prophylactic oral treatment with valacyclovir 500mg daily resulted in rapid and significant reduction of the periodic vesicular eruption and the associated transient aggravation of her persistent swelling. However, the chronic lymphedema persisted. After a 6 month follow-up, the patient reported only two recent milder episodes attributed to non-adherence to valacyclovir.

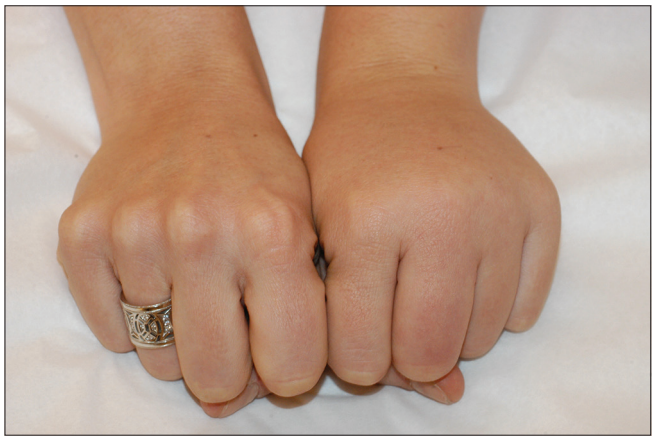

Figure 1: Chronic lymphedema involving the left hand and forearm with swelling and erythema of the left middle finger.

DISCUSSION

Uncomplicated HSV infection of the hand is marked by the recurrence of painful vesicles, with occasionally systemic symptoms such as malaise and fever [3]. Furthermore, lymphatic complications, such as lymphangitis and lymphadenopathy in axillary and/or epitrochlear lymph nodes, are well described and consist of proximal swelling, pain, and erythematous streaking [3,4]. More severe cases, lymphedema of the hand and/or forearm may rarely develop a few days after the onset of vesicles, and such complications can be misdiagnosed as cellulitis or dyshidrotic eczema [2,3,5]. These lymphatic events typically resolve within 21 days, but sometimes persist indefinitely [5].

Persistent lymphedema of the hand secondary to HSV infection despite antiviral treatment is exceedingly rare and has been reported only twice in the literature [2,3]. Furthermore, although there is initially involvement of lymph nodes and the spleen as early as 24 hours following infection with HSV [6], as well as immediate activation of tissue-resident CD8αα$^+$T cells in containment of HSV-2 [7], the majority of patients with prolonged HSV-associated lymphedema of the hand did not present with any lymphadenopathy [2-4]. This clinical presentation adds further difficulty in arriving at HSV infection as a differential diagnosis. The repeated occurrence of HSV infection over long periods of time is a common element in all reported cases of unresolved and persistent lymphedema, including ours, and is likely the cause of the unresolved lymphedema. Interestingly, those who developed lymphedema which ultimately resolved in response to antiviral treatment experienced episodes of reactivation which were shorter in duration and less frequent than those with unresolved lymphedema.

Table 1 lists all five reported cases in the literature of lymphedema secondary to HSV infection of the hand [2-5,8]. Our case and those reported in the literature demonstrate favorable outcomes with antiviral use, including suppression of the recurrence of vesicular eruptions and the episodic aggravation of the persistent lymphedema. The presentation of lymphedema of the hand as a complication of recurrent HSV infection nearly one decade following the primary infection and its persistence with antiviral treatment is of great clinical interest and didactic value. We hope

Table 1: Reported cases of resolved and unresolved lymphedema of the hand secondary to herpes simplex virus infection

Reference	Gender	Age	Type of HSV	Site of HSV	First episode of HSV	Frequency of episodes	Sites of LE	Duration between development of vesicles and LE	Duration of LE	Effect of treatment on LE	Lymphangitis/ lymphadenopathy
Howard et al 1979	Female	32	HSV-2	Left middle finger	6 years	NA	Left hand and forearm	NA	≥2 years	No treatment. Persisted and worsened during acute HSV episode	Present/absent
Sands et al 1988	Male	52	NA	Left index finger	3 years	< 1/year	Left forearm	1 week	10 days	Resolved	Present/present
Butler et al 1999	Female	28	HSV-2	Right middle finger	21 months	NA	Right Hand and forearm	NA	18 months	Significantly improved but persisted	Absent/absent
Bowling et al 2005	Female	32	HSV-2	Left middle finger	2 years	2/year	Left forearm	NA	10-21 days	Resolved (prophylaxis also helped)	Present/present
Fletcher et al 2005	Female	33	HSV-2	Right hand	3.5 years	Monthly exacerbation of symptoms	Right hand and forearm	1 week	3.5 years	Prophylactic acyclovir ineffective. LE persisted.	Absent/absent

Legend: LE: Lympedema; HSV: Herpes simplex virus; NA: Not available

that this case report will increase awareness of the abnormal manifestations and complications of HSV infection of the hand, and help clinicians avoid the unnecessary use of antibiotics, surgical manipulation, and hospitalization. The reviewed cases also strongly suggest that the prompt provision of antiviral treatment can be quite effective and may prevent the emergence of irreversible, chronic and antiviral-resistant lymphedema.

CONCLUSION

In conclusion, although uncommon, cases of episodic lymphedema in the hand with or without associated lymphangitis and lymphadenopathy should always take into account the possibility of HSV infection as a primary cause, and antiviral treatment should be promptly pursued if HSV infection is confirmed. This approach would help avoid inappropriate and unnecessary medical treatment and surgical manipulation.

Consent

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

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

Our Dermatology Online

Case Report

Eyelid nonbullous impetigo

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ABSTRACT

We report an unusual case of nonbullous impetigo of the eyelid. A 21-year-old woman had a painless eruptions right eyelid and around nostril. Clinical examination revealed eyelid nonbullous impetigo. The patient was otherwise healthy. Any minor trauma or skin problems there were not founded. Staphylococcus aureus was isolated from the lesions. Complete resolution was achieved with oral cephalexin an ointment tobramycin. Impetigo nonbullous occurs commonly on the face of children but it might be developed in healthy adults even those was not necessary evidenced previous trauma or dermal infiltrate with pus or any other active cutaneous injury as this case is demonstrated. The treatment of impetigo around the eye included a careful examination of the segment anterior and the application de topical measures with oral and topical antibiotics. The case is being reported in view of its rarity and unusual location.

Key words: Impetigo; Staphylococcus aureus; Eyelid; Streptococcus pyogenes

INTRODUCTION

Impetigo is a highly contagious bacterial skin infection affecting children [1-4] worldwide that is caused by Gram-positive bacteria *Staphylococcus aureus* [5-7] *Streptococcus pyogenes* [2,4] or both [2,8-10].

The face, especially the perioral region, is one of the most frequently involved areas [2]. The infection usually begins with an outbreak of vesicular lesions on the arms and legs and on occasion they are seen around the nose, mouth and scalp [11]. Here we report an unusual case of nonbullous impetigo in a woman who presented crusting, vesiculobullous skin lesion in the right upper eyelid.

CASE REPORT

A 21-year old woman present with a lesion in right eyelid. It began a week earlier as an erythematous macule that soon becomes vesicular that rupture, ooze, and lead to the layer of crusting. There was no history of ocular trauma or history of any surgery performed in the past. No family or personal history of skin problems. The clinical appearance was scattered, painless, slightly pruritic reddened sores with honey–yellow exudates crusting on them, localized in upper eyelid (Fig. 1). Satellite lesion was observed around the nose (Fig. 2). There was region auricular lymphadenopathy. The eyelid culture was positive for *S. aureus*. The remainder of the ocular and general physical examination was normal. Treatment was initiated with oral cephalexin mono hydrate 500mg per each 12 hours, ointment tobramycin four times daily. The patient responded well to the antibiotic, and the denuded area showed slow epithelialization and complete healing of the skin within 3 weeks.

DISCUSSION

Impetigo is a common localized crusting, vesiculobullous skin lesion resulting from superficial infection by bacteria. The bacteria usually infect skin that has been damaged by scratching an insect bite or picking a scab. The lesions may cause mild soreness an itching, but are typically painless. Impetigo was described in 1864 by Tilbury–Fox. The isolated micro-organism usually identified in cultures of lesions is a *S. aureus* [5-7] although *S. pyogenes* infection is also common [2,4]. Many lesions represent a mixture of the two infections [2,8-10].

How to cite this article: Corredor-Osorio R, Corredor-Osorio M, Corredor-Osorio A. Eyelid nonbullous impetigo. Our Dermatol Online. 2016;7(3):291-293.
Submission: 06.01.2016; **Acceptance:** 01.03.2016
DOI:10.7241/ourd.20153.78

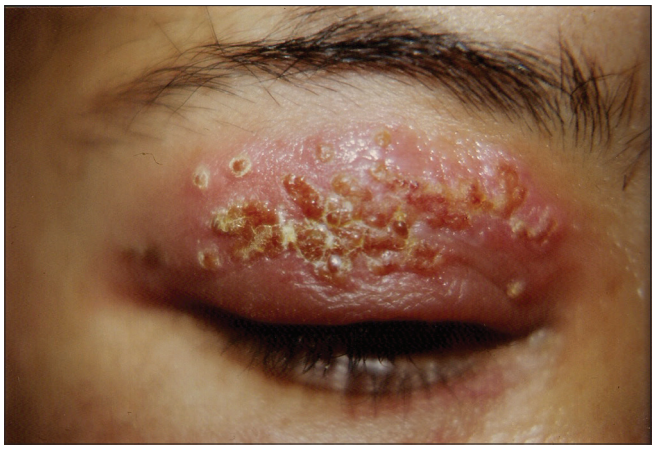

Figure 1: Round blisters and erosions with clear borders, many of which are crusted.

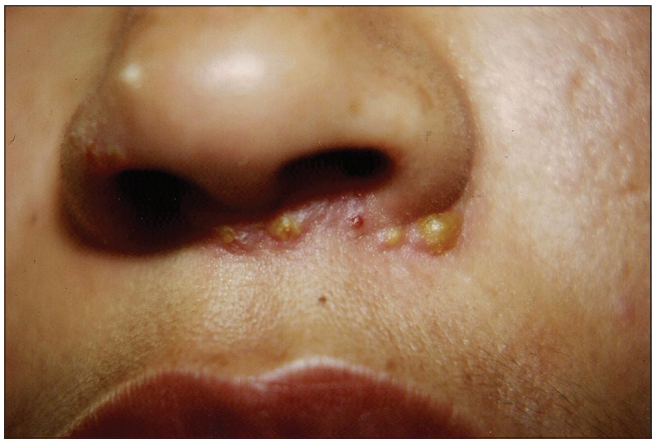

Figure 2: Lesion satellite around the nose.

Susceptibility to these infections depends on host immune factors, as well as virulence of the organism. It is most likely to occur under conditions of crowding, poor hygiene, and hot, humid climate, and it can spread rapidly between members of a household, school. Impetigo can occur as a primary infection or secondary to pre-existing skin conditions, such as eczema or scabies [3]. Impetigo can be divided on clinical and bacteriological grounds into basic forms: Impetigo non-bullous (contagiosa)and bullous based on the presence or lack large blisters, called bullae [2,4,9,10]. Both forms involve only the most superficial layers of the skin [10]. Impetigo nonbullous is the most common form and is likely to be caused by a mixed staphylococcal and streptococcal infection [2,9,10] depending on geographical variations, with the streptococcal form being more prevalent in warmer and humid climates[8], however, *S. aureus* in the main cause. The early lesion of impetigo non-bullous begins as a small, 2-4 mm, erythematous macule which soon becomes vesicular or pustular [12].

The vesicle is a very thin-walled hardly noticeable, as it soon ruptures, leaving an exudate [4,12]. The purulent exudates dries and form the classic thick yellow-brown crust (meliceric)[3,4]with a honey color [2,8,9]. The crusts lesion, may show no residual or active vesicles and measures 1-3 cm, in greatest diameter. It may be mildly pruritic. Satellite lesions occur in the vicinity due to autoinoculation [2]. When removed, the crust quickly reappears [4]. Removal of the crust results in the reaccumulation of fresh exudates. The crusts eventually dry separate and disappear, leaving an area of erythema that heals without scarring.

The lesions are painless and usually localized, often occur in the face around the mouth and nose. The trunk and limbs can also be affected. Systemic signs are usually not present however with extensive impetigo, fever and regional lymphadenopathy may occur [2].

Bullous impetigo is almost always caused by *S. aureus* [2,4,5,9] is characterized by erythematous macule that progress to bullae, large thin-walled blister that contain clear or cloudy yellow fluid [2,4], and measure less that 5cm in diameter [10]. Bullae are caused by staphylococcal infections. Staphylococcally produced epidermolytic toxin has been recovered from the blister fluid in some cases [9,13]. These blisters easily rupture and leave behind a moist area of eroded skin surrounded by a thin ring of the remaining blistered skin [2]. The lesion dries and crust over, creating a light brown appearance that resembles "varnish" which may be white or grey in color [11]. These lesions are discrete, with little redness or inflammation surrounding them [2]. Its usual distribution involves the face, buttocks, truck and perineum [3]. The condition might be associated with fever, diarrhea and weakness [10]. Although the nonbullous impetigo is a dermatological disease may be associated with conjunctivitis [14]. Most cases resolve without sequelae in 2 to 3 weeks. Complications of impetigo include cellulitis, lymphangitis, supurative lymphadenitis, nephritis and sepsis. The differential diagnosis of nonbullous impetigo should include herpes simplex viral infections, herpes zoster, candidiasis, atopic contact dermatitis, seborrheic dermatitis, insect bites, varicella, scabies and burns thermal [4,10]. The aim of treatment is to clear the eruption and prevent the spread of one infection to other. Nonbullous impetigo can be treated with either oral or topical antibiotics. At the initiation of treatment the crusty scabs should be removed after softening with wet compresses.

The lesion should be washed with an antibacterial soap [4] or povidine-iodone shampoo two or three times daily [3]. Initial cultures are usually not necessary, since health care professional commonly prescribe erythromycin [2-4,8], amoxicillin/clavulanate [2,3], cephalexin [2,3,10], vancomycin [1,2], azithromycin, oxacycline [4], dicloxacillin [14] and clindamycin [2]. The course of treatment is 7 days.

For mild infections, a topical antibiotic efficacious against gram-positive bacteria, especially *S. aureus* and S. *pyogenes* is the preferred first-line therapy. Topical treatment may be selected if area of impetigo is localized. An antibiotic ointment or cream should be then applied, such as mupirocin 2% ointment [1,4,8], neomycin, bacitracin ointment [2,3], chloramphenicol ointment [14] fusidic acid cream 2% [1,4,6], gentamycin ointment [3], retapamulin 1% ointment [1,4,6], NVC-422 topical gel [1], sulphadiazine cream [11]. This approach is sufficient to clear mild to moderate cases. It is of interest that the patient developed localized crusting, vesiculobullous skin lesion, clinically similar a nonbullous impetigo characterized for several adjacent clusters of small clear vesicles and whitish pustules which are partly confluent and is yellow-brown break open early to release a clear, oozing secretion which quickly becomes crusted on the surface. The crusted lesion is honey color. This lesion is localized in the right upper eyelid, although the upper lip, labial commisure, suborbital skin are affected more often. For widespread, localized infections, such as this patient, require oral and topical antimicrobials to be sure that infection has not worsened and developed into skin infection called cellulitis.

In summary, impetigo non bullous is a common, vesiculopustular, crusting skin lesion resulting from superficial infection by bacteria. Typically affects children but it can affect age group. Intact, healthy skin seems largely immune to the disease, as most lesions occur in areas of dermatitis or previous trauma and, is usually transmitted direct skin contact or indirect contact with the liquid ooze of an lesion active. When impetigo non bullous occurs around of the eye especially in the eyelids, a careful examination of the anterior segment of the eye is necessary and, if are signs of increased redness, or pain in the skin surrounding of the lesion, oral and topical antibiotics should be used for avoid potential complications. Ophthalmologists, dermatologists, and physicians should be familiar with this entity of impetigo and consider it in the differential diagnosis of the spectrum of the vesicobullous disorders in the eyelid.

Consent

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

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

Our Dermatology Online

Case Report

Primary idiopathic systemic amyloidosis – Rare classical cases with fatal outcome

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ABSTRACT

Primary systemic amyloidosis is a rare condition. We report two cases of primary systemic amyloidosis. Both the cases were without any hematological abnormality. Cutaneous features were the predominant presenting symptoms in these patients. The patients presented with typical waxy lesions on face and macroglossia. Diagnosis was confirmed by histopathology with haematoxylin and eosin staining and Congo red staining.

Key words: Primary systemic amyloidosis; Macroglossia; Congo red stain

INTRODUCTION

Amyloidosis is a generic term originally coined by Rudolf Virchow in 1854, which denotes extracellular deposition of a proteinaceous substance composed of one of a family of biochemically unrelated proteins depending on the underlying condition, and which is associated with considerable tissue dysfunction [1]. Amyloid deposition may be localized to one organ system or it may involve multiple organs depending on which amyloidosis is classified as localized or systemic. Localized cutaneous amyloidosis, which includes macular amyloidosis and lichen amyloidosis, is a benign common disease without systemic involvement. Skin may also be involved in systemic amyloidosis. Systemic amyloidosis is classified into primary, secondary and familial [2]. Primary systemic amyloidosis may be idiopathic or myeloma-associated. Secondary systemic amyloidosis is associated with many chronic inflammatory disorders. Skin involvement is common in primary amyloidosis whereas secondary amyloidosis rarely involve skin [1]. Systemic amyloidosis is usually a disease of the elderly.

CASE REPORTS

Case 1

The first case of 61 year old male presented with features of asymptomatic waxy papules on the face especially involving the periorbital areas (Fig. 1) and features of macroglossia (Fig. 2) were seen along with purpuric lesions (Fig. 3) involving the face, neck and groin. Thickened palms and fingers associated with burning sensation, difficulty in moving fingers.

Other changes seen were Hepatomegaly, left ventricular hypertrophy and 10% plasmacytoma was seen on bone marrow biopsy. Evidence of renal failure like raised blood urea and serum creatinine along with urine albumin was present. Because of the typical presentations, diagnosis of primary systemic amyloidosis, was suspected clinically and confirmed by histopathology. Histopathological features noted were epidermis showing hyperkeratosis and focal atrophy; dermis showing diffuse deposits of eosinophilic hyaline material extending into subcutaneous tissue (Fig. 4).

Case 2

36 year old male presented with Weight loss, weakness, fatigue, and asymptomatic raised lesions on face since 3 months.

On examination Macroglossia with lateral teeth indentations and fissures on tongue (Fig. 5).

How to cite this article: Patil S, Chinthala S, Lunge S. Primary idiopathic systemic amyloidosis – Rare classical cases with fatal outcome. Our Dermatol Online. 2016;7(3):294-298.

Submission: 02.11.2015; **Acceptance:** 28.01.2016

DOI: 10.7241/ourd.20163.79

Multiple waxy papules, nodules and tumefactive lesions in oral mucosa (Fig. 6), periorbital and perioral areas (Fig. 7). Ecchymotic patches were seen on face and trunk. Investigations revealed a low complete blood count with Haemoglobin of 7 gm/dl. Blood urea of 89 mg/dl, Serum creatinine.

Figure 1: Periorbital pigmented waxy papules.

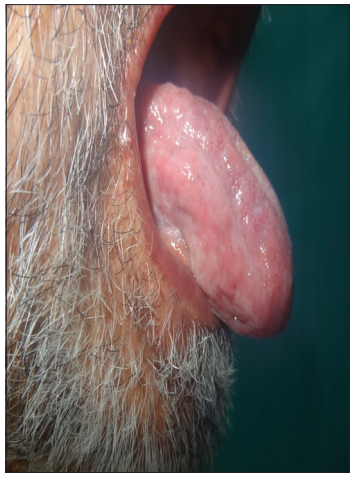

Figure 2: Macroglossia.

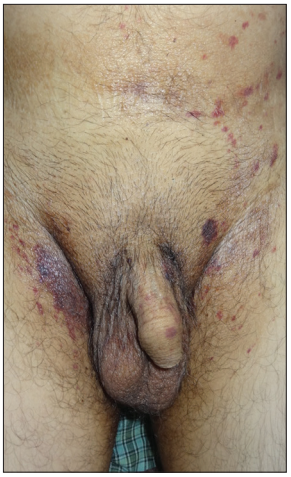

Figure 3: Multiple areas of purpura in the inguinal region.

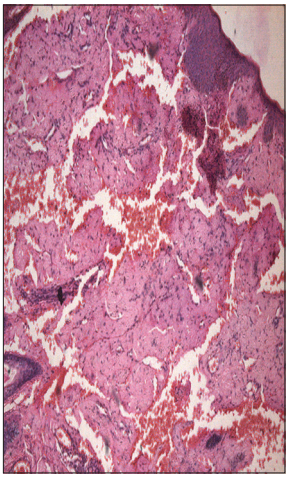

Figure 4: Dermis with diffuse deposits of eosinophilic hyaline material extending into subcutaneous tissue. H&E – 40x.

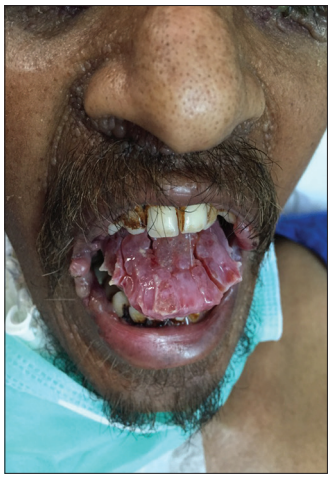

Figure 5: Macroglossia with fissures and lateral teeth indentations.

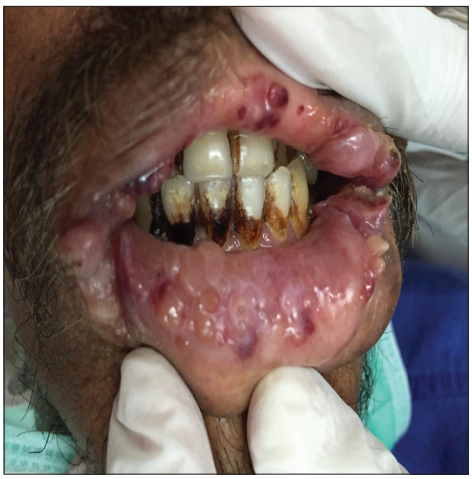

Figure 6: Multiple waxy papules and purpuric lesions on the oral mucosa.

2.8 mg/dl and Urine albumin - 3+, serum and urine electrophoresis was normal. USG abdomen revealed grade 1 renal parenchymal disease. Biopsy H and E staining –Eosinophilic amorphous material. Congo red staining - brick red staining of amyloid (Fig. 8). Bone marrow examination was normal.

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

DISCUSSION

Amyloidosis is a group of diseases characterized by extracellular deposition of beta-sheet fibrils. In the systemic forms, the amyloid causes progressive organ dysfunction leading to death of the patients. Over 20 proteins capable of amyloid formation have been identified. They include immunoglobulin (Ig) light chains in primary systemic amyloidosis (AL), Ig heavy chain (AH), amyloid A in secondary amyloidosis (AA), beta2-microglobulin in dialysis-associated arthropathy (Ab2M) and amyloid beta protein (Ab) in Alzheimer's disease and Down's syndrome. There are also hereditary forms that include transthyretin (ATTR), apoli-poprotein A-I (AApoAI) and A-II (AApoAII), gelsolin (AGel), lysozyme (ALys), fibrinogen A-alfa chain (AFib) and others. Another amyloidogenic protein is leukocyte chemotactic factor 2 (LECT2) [3].

Inherited amyloidosis is due to mutation in certain precursor protein, which makes them susceptible to mis- folding. In case of primary systemic amyloidosis, the amyloid is derived from monoclonal immunoglobulin light chain and is called as AL amyloid where L stands for light chain of immunoglobulin molecule. In case of secondary amyloidosis which is associated with many chronic inflammatory diseases, amyloid fibrils are derived from cleavage fragment of the circulating acute phase reactant serum amyloid A protein (SAA), hence called as AA amyloid. Serum amyloid A protein is synthesized in liver during inflammation [2]. It has been proposed that in macular and lichen amyloidosis, focal epidermal damage and filamentous degeneration of keratinocytes is followed by apoptosis and conversion of filamentous masses (colloid bodies) into amyloid material in the papillary dermis [1]. The reason that many diverse conditions are associated with amyloidosis may be because each of these conditions results in excessive production of proteins that are prone to mis-folding [2] In multiple myeloma-associated AL amyloidosis, precursor light chains of immunoglobulin (Bence Jones protein) are produced in large quantity by malignant plasma cell clone and can be detected in serum or urine by electrophoresis. Multiple myeloma is a malignancy of plasma cell. Amyloidosis develops in about 15% of patients of myelomatosis [1] Majority of patients of AL amyloidosis do not have obvious B-cell/plasma cell neoplasm (idiopathic). These patients might have underlying B-cell dyscrasia in which production of abnormal protein, rather than production of tumor masses, is the predominant manifestation [2]. In one study from Lebanon, of 39 cases of systemic amyloidosis, 21 were of AL type and out of these 21 cases of AL amyloidosis, 9 (43%) were associated with multiple myeloma and 12 (57%) were idiopathic [4].

Cutaneous involvement is seen in 40% patients with AL amyloidosis. Cutaneous manifestation depends

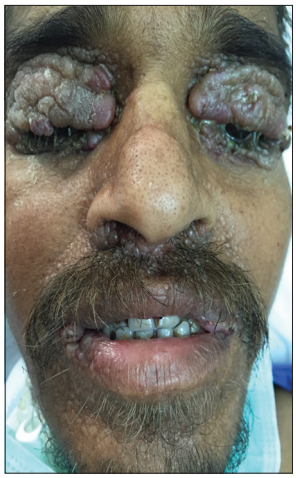

Figure 7: Multiple waxy papules and tumefactive plaques on periorbital and perioral areas.

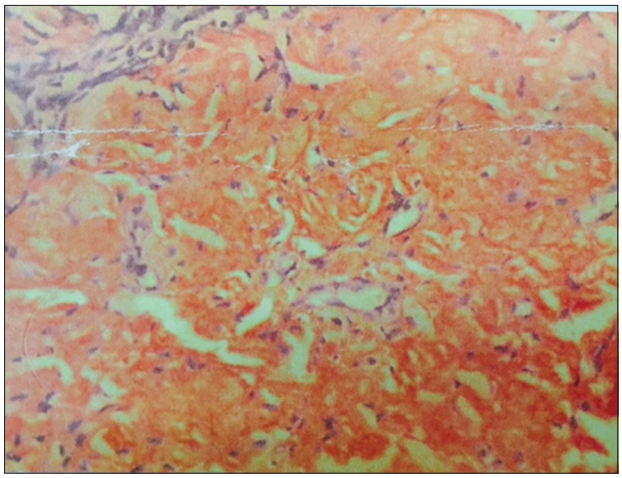

Figure 8: Special stains for amyloid deposits in the form of Congo red are positive for amyloid.

upon the site of amyloid deposited. Amyloid deposition in superficial dermis produces shiny waxy translucent papules and common sites for predilection are eyelids, retro-auricular areas, neck, axilla. Infiltration of nail matrix by amyloid may produce ridging, splitting and brittleness of nail plate [1]. Similar lesions and distribution was noted in both our patients and nails were found to be dystrophic. Amyloid deposited around pilosebaceous unit leads to the destruction of hair, producing alopecia. Diffuse infiltration of scalp skin results in the enlargement of skin which gets thrown into longitudinal folds resembling cutis vertices gyrata. Diffuse infiltration of large area of skin may simulate scleroderma [1].

Amyloid infiltration of vessel wall causes capillary wall fragility, which leads to purpura and ecchymosis after a minor trauma or even spontaneously. Periorbital area is one of the common sites of expression of purpura. The capillary fragility may be demonstrated by pinching the skin. Ecchymotic lesions were present in both cases. Purpuric lesions with normal platelet count and normal coagulation profile should suggest the possibility of capillary fragility [1].

Amyloid deposition in tongue leads to macroglossia. Tongue is diffusely enlarged and firm and there may be tooth indentation along its lateral border. Amyloidosis is the commonest cause of macroglossia in adults [1]. Macroglossia if severe might lead to dysphagia. Macroglossia with tooth indentation was present in second case.

Hepatomegaly occurs in 50% of patients and splenomegaly in 10%. Hepatomegaly was present in our case 1. Cardiac involvement leads to conduction defects, arrhythmias, congestive cardiac failure and may account for 40% of deaths. Our first patient had left ventricular hypertrophy and second patient had no cardiac involvement as indicated by normal ECG and X-ray chest.

Carpal tunnel syndrome is seen in up to 25% of patients of primary systemic amyloidosis [1], as was present in our case 2. Renal involvement presents with proteinuria and renal failure. It is one of the bad prognostic indicator and was present in our case 2 as indicated by proteinuria and USG findings.

Both the patients were diagnosed as having primary amyloidosis on clinical ground. Diagnosis was confirmed by demonstration of amyloid in skin biopsy. Clinically, it is difficult to distinguish primary, secondary or familial form of amyloidosis. Immunohistochemical staining using commercially available antisera is useful for classifying the type of amyloid deposited in tissues [5], which was not done in our patients, this being a resource poor setting. Biopsy is very important for the diagnosis. Hematoxylin and eosin staining suggests the possibility of amyloidosis but Congo red staining confirms the diagnosis. Congo red staining results in a brick red color of amyloid when seen under ordinary light and under polarized light shows classical green birefringence. Unfortunately, polarized microscopy is not easily available in developing country like India. In systemic amyloidosis, amyloid deposits are seen in dermis, subcutaneous tissue and blood vessels, where as in localized cutaneous amyloidosis, deposits are seen only in papillary dermis; subcutaneous tissues and blood vessels are not involved.

Prognosis in AL amyloidosis is poor and major causes of death are cardiac and renal failure. The median survival of patients with myeloma-associated amyloidosis is five months and 2.1 years for patients with primary systemic amyloidosis [1]. Prognosis depends upon the extent of involvement. Treatment of amyloidosis is aimed at reducing the supply of precursor proteins [1]. In AL amyloidosis, the precursor is immunoglobulin light chain produced by B lymphocytes/plasma cells hence treatment with cytotoxic agents like melphalan and prednisolone that reduces plasma cell proliferation is useful [1]. Chemotherapy will be useful only when precursors are supplied by plasma cells like AL amyloidosis.

In localized cutaneous amyloidosis, such as lichen amyloidosis and macular amyloidosis, where precursors are derived from keratinocytes and not from plasma cells, alkylating agents or any other chemotherapeutic agents will not be beneficial and may be harmful [6].

These cases of systemic amyloidosis are presented for its rare occurrence. High index of suspicion is necessary for the diagnosis of such rare cases.

Consent

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

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

Our Dermatology Online

Case Report

A rare case of solitary trichoepithelioma treated with carbon dioxide laser

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ABSTRACT

Trichoepithelioma (TE) is a benign skin neoplasm derived from the hair follicle. It is commonly located on the face and hairy skin. We report a rare and interesting case of a solitary trichoepithelioma (sTE) treated with a carbon dioxide laser. This report shows a positive therapeutic effect of a carbon dioxide laser on a patient with sTE located on the medial angle of the eye region. The carbon dioxide laser may be one of the therapeutic options in the case of sTE.

Key words: Solitary trichoepithelioma, Benign skin neoplasm, Carbon dioxide laser

Abbreviations: TE: Trichoepithelioma; sTE: Solitary trichoepithelioma; BCC: Basal cell carcinoma

INTRODUCTION

Trichoepithelioma (TE) is a benign skin neoplasm derived from the hair follicle. It is commonly located on the face and hairy skin. Malignant transformation is quite rare. TE affects adults and more frequently women. The most common presentations are noted in the 4th decade [1]. There are three variants of TE: solitary TE, multiple TE and desmoplastic TE [2-4]. The solitary TE is non-familial. The multiple TE is familial. The gene for the development of familial trichoepithelioma encodes of chromosome 9 [5]. We report a rare and interesting case of solitary trichoepithelioma (sTE) treated with carbon dioxide laser.

CASE REPORT

We present the case of a 47-year-old female patient, who was hospitalized in January 2009, for the presence of a skin tumor, 2 cm in diameter, brownish color, asymptomatic, located on the medial angle of the eye region (Fig. 1). The patient had past surgical history. The tumor appeared two years ago and it was gradually increasing in size. The patient was treated by surgery. Twice she had undergone ineffective surgical excisions (March 2007 and September 2008). A biopsy initially found nodular basal cell carcinoma (BCC).

There was no significant family history. The patient's medical history included hypertension. Systemic examination and a general physical revealed no abnormality. Results of a routine laboratory studies were normal. During hospitalization, we performed a tumor biopsy. The histopathological examination, using Hematoxylin and Eosin (H+E)-stained paraffin cross-sections, showed microscopic structure of trichoepithelioma (Fig. 2). According to medical history, clinical features and histopathologic findings, a diagnosis of solitary trichoepithelioma (sTE) was made. We decided to avoid surgical procedures. The patient was treated with 10,600 - nm carbon dioxide laser CO_2 excision. We performed two procedures with 3 weeks interval. Pretreatment anesthesia of 1% lidocaine was

How to cite this article: Borowska K, Borzęcki A, Szponar-Bojda A, Raszewska-Famielec M. A rare case of solitary trichoepithelioma treated with carbon dioxide laser. Our Dermatol Online. 2016;7(3):299-301.

Submission: 03.03.2016; **Acceptance:** 11.04.2016

DOI: 10.7241/ourd.20163.80

used. The tolerance of the treatment was very good with a total absence of clinical adverse events (Fig. 3). This complete remission was maintained after 7 years of follow-up.

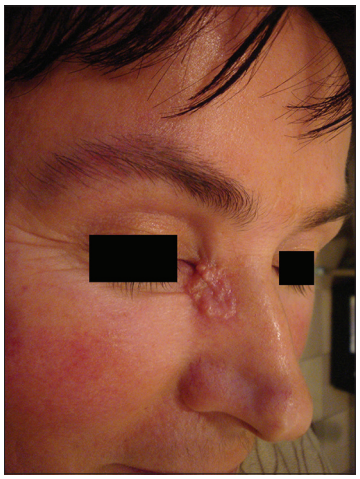

Figure 1: The patient before treatment.

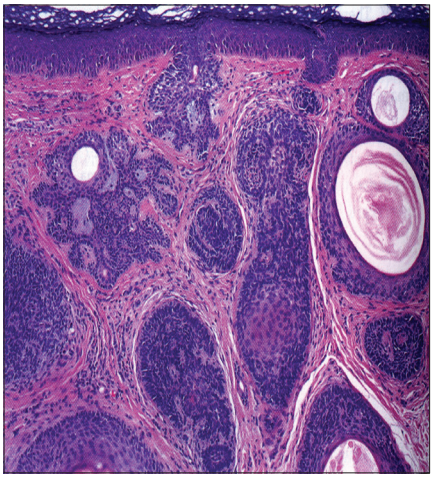

Figure 2: Microscopic picture of trichoepithelioma, H+E staining, 100x.

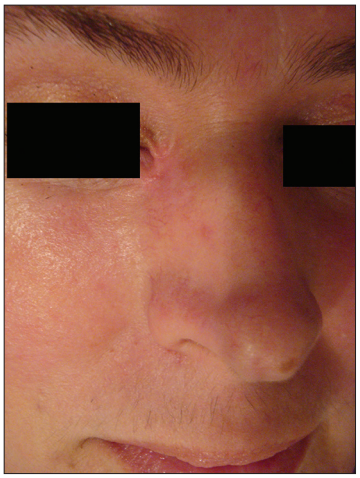

Figure 3: The patient after treatment.

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

DISCUSSION

Trichoepithelioma (TE) was first described by Brooke as epithelioma adenoides cysticum, in 1892 [6]. Differential diagnosis for TE includes BCC. It is important to accurately differentiate between the two neoplasms. In certain cases, it may be difficult to clinically distinguish TE and BCC, particularly solitary trichoepithelioma (sTE). The distinction between BCC and TE on a histopathological basis is also quite difficult. BCC is the most prevalent cutaneous tumor. In contrast to BCC, TE is a benign tumor with clear follicular differentiation. This case highlights the importance of recognizing the solitary trichoepithelioma. The carbon dioxide laser is the most versatile laser used in the treatment of cutaneous tumors. The carbon dioxide laser constitutes an alternative to surgery in the treatment of large solitary nasal tip trichoepithelioma [7] and multiple trichoepithelioma [8,9]. This report shows a positive therapeutic effect of carbon dioxide laser on a patient with solitary trichoepithelioma located on the medial angle of eye region. Treatment option for patients with TE is treatment with carbon dioxide laser. It gives excellent cosmetic result and a high degree of satisfaction among treated patients. The carbon dioxide laser may be one of the therapeutic options in the case of solitary trichoepithelioma.

Consent

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

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

Our Dermatology Online

Case Report

Ulcerative giant solitary trichoepithelioma of scalp: a rare presentation

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ABSTRACT

Trichoepithelioma is a trichogenic tumor which arises from the inferior segment of hair follicle epithelium as hamartoma. Giant solitary trichoepithelioma (GST) has been defined as a solitary trichoepithelioma with a diameter greater than 2 cm. A 49-year-old female presented with a slow growing skin coloured swelling on the scalp of 8 years duration with recent history of ulceration and occasional bleeding. The local examination revealed a single well defined nodular swelling which was irregular in shape measuring approximately 2 x 2.5 cm. Histopathology from biopsy specimen revealed dark basaloid cells with scanty cytoplasm and darkly stained nucleus arranged in nests with horn cysts lacking high-grade atypia and mitosis, which was consistent with features of trichoepithelioma. Giant solitary trichoepithelioma of scalp is itself a rare entity and the present case is being reported with the additional component of ulceration in the lesion.

Key words: Trichogenic; Tumour; Scalp; Ulcerative; Trichoepithelioma

INTRODUCTION

Trichoepithelioma is a trichogenic tumor which arises from the inferior segment of hair follicle epithelium as hamartoma. Giant solitary trichoepithelioma is a distinct variant of trichoepithelioma. It has been defined as a solitary trichoepithelioma with a diameter greater than 2 cm [1]. It arises in elderly individuals and occurs mostly on the face, thigh and peri-anal region [2,3]. These features are in contrast to the conventional trichoepithelioma, which presents as multiple small translucent circumscribed papules of 2–4 mm in diameter, in children and young adult on the face. To the best of our knowledge no case of ulcerative giant solitary trichoepithelioma on scalp has been reported previously.

CASE REPORT

A 49-year-old female presented with a slow growing skin coloured swelling on the scalp of 8 years duration with recent onset of ulceration and occasional bleeding. The local examination revealed a single well defined nodular swelling which was irregular in shape measuring approximately 2 x 2.5 cm (Fig. 1). On palpation it was a firm, mildly tender with no increase of local temperature. The lesion bled on manipulation. There were no signs of infection at the site. Routine blood investigations were within normal limits. Touch impression smear showed few neutrophils with no organism. The general physical and systemic examinations did not reveal any abnormality. There were no palpable lymph nodes in head and neck area.

Histopathology from incisional biopsy specimen revealed dark basaloid cells with scanty cytoplasm and darkly stained nucleus arranged in nests with horn cysts, lacking high grade atypia and mitosis which was consistent with trichoepithelioma (Fig. 2). There was loss of epidermis suggesting ulcerative nature of the lesion (Fig. 3).

How to cite this article: Chowdhry S, Jaiswal P, Dhali TK.Ulcerative giant solitary trichoepithelioma of scalp: a rare presentation. Our Dermatol Online. 2016;7(3):302-304.

Submission: 26.11.2015; **Acceptance:** 01.02.2016

DOI: 10.7241/ourd.20163.81

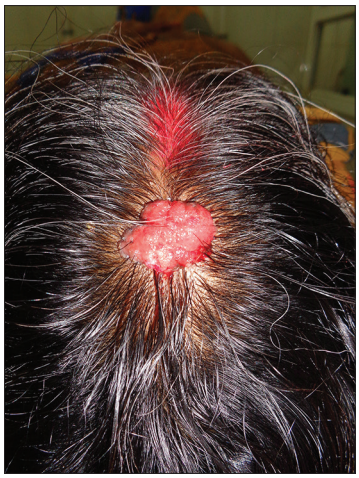

Figure 1: Single well defined nodular swelling on scalp.

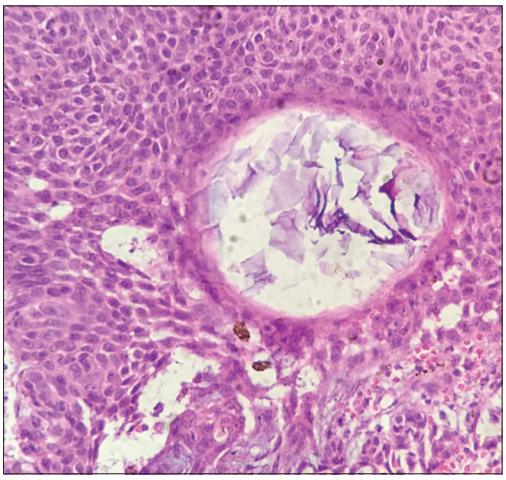

Figure 2: Dark basaloid cells with scanty cytoplasm and darkly stained nucleus arranged in nests, (H&Ex400).

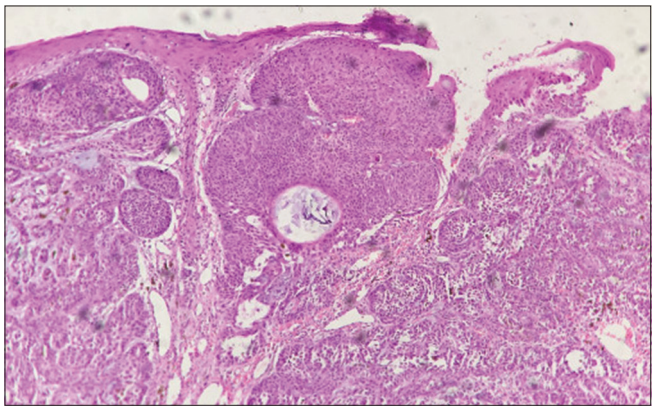

Figure 3: Loss of epidermis, (H&Ex100).

The patient was referred to the Department of Surgery for total excision and repair.

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

DISCUSSION

Trichoepithelioma (TE) was first described by Brooke in 1892 as Epithelioma Adenoides Cysticum [4]. Clinically, this tumor occurs either as a solitary lesion without familial association or as multiple lesions in multiple familial trichoepithelioma [5].

Since trichoepithelioma is inherited in an autosomal dominant fashion, males and females receive the gene equally. Because of lessened expressivity and penetrance in men, most patients are women [6]. A giant solitary trichoepithelioma is a distinct variant of trichoepithelioma that may have a predilection for the perianal region. At this site, this rare tumor must be distinguished from basal cell carcinoma of the perineum and from malignant basaloid (cloacogenic) carcinoma of the anal canal [2].The mean age of the presentation is 60 years with a predilection for the older age group [7]. The majority of the GST has been subcutaneous but pedunculated [8] and ulcerated [6] appear as well.

The histopathology shows islands of uniform basaloid cells with scanty cytoplasm and darkly stained nucleus arranged in nests and adenoid pattern with epithelial islands. The epithelial islands may not connect to the overlying epidermis. The stroma may be fibromyxoid or fibrocellular [8,9]. Immature hair appears as keratinous cysts. These horn cysts are the characteristic features in most GST. They consist of fully keratinized center surrounded by basophilic cells that lack high-grade atypia and mitosis. The keratinization is abrupt and complete – the so called trichilemmal keratinization, which differentiates it from squamous cell carcinoma having gradual and incomplete keratinization in horn pearls [10]. GST presents as a pigmented lesion because of the increase activity of melanocytes or increased retention of pigment in the basal keratinocytes [11].

Treatment for multiple TEs include excision, electrodessication, dermabrasion, cryotherpy and radiotherapy, Argon, Carbon dioxide, erbium-YAG lasers. Surgical excision with or without flap is the standard treatment for most of GSTs. Radio-surgical ablation can be considered for cosmetic reasons when the tumor is situated over face. It helps in accurate removal of the tumor with minimal bleeding without destroying the underlying structures like cartilage. Malignant transformation into BCC after surgical excision requires adjuvant radiotherapy [12]. Recurrence and possible transformation into BCC necessitates patient follow up at regular intervals.

Consent

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

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

Our Dermatology Online

Case Report

Chromoblastomycosis

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ABSTRACT

Chromoblastomycosis is a subcutaneous, chronic, granulomatous mycosis that occurs more frequently in tropical and subtropical countries. We report a case of chromoblastomycosis of the earlobe due to *Fonsecaea sp* in a male patient of 34 years old, due to its uncommon localization.

Key words: Chromoblastomycosis; Fonsecaea pedrosoi; Fonsecaea compacta; Cladosporium carrionii; Fumagoid cells

INTRODUCTION

The chromoblastomycosis is a sub cutaneous mycosis in tropical and subtropical areas considered as an American disease, the main agents are *Fonsecaea pedrosoi*, in endemic areas of tropical and subtropical environments; *Fonsecaea compacta*, *Cladosporium carrionii*. The diagnosis of the disease is through the presence of fumagoids cells.

In our environment, chromoblastomycosis is the third most common subcutaneous mycosis. It predominates in the lower limbs in warty form and F pedrosoi is the most frequent etiological agent.

CASE REPORT

Male patient, 34 years, who consulted a private doctor because of a dermatological condition in his right ear. A biopsy was performed and the report indicated a squamous cell carcinoma. The patient was sent to the oncology outpatient clinic for treatment, but as there was doubt about the histological diagnosis and the clinical picture, a second biopsy was performed that reported a granulomatous-type lesion. An inter-consultation was held at the Dermatology Service for reevaluation. Upon examination, a dermatosis was found in the ear and right facial region, made up of an atrophic squamous plate, hematic crusts and one retroauricular nodule with slightly warty appearance (Figs. 1 and 2). The rest of the physical exam was within normal limits.

The patient says that his disease started 3 years ago with a small asymptomatic "pimple" in his right ear that slowly increased its size until he decided to consult. In the last 6 months he had an occasional itch and was prescribed different antibiotics and non-specific creams. He does not remember bruising the area.

Three clinical diagnosis were made based on the clinical data: chromoblastomycosis; leishmaniasis; and sporotrichosis fixed plaque. A new skin biopsy was performed, along with a direct exam and a culture for fungus and PCR for leishmania.

The PCR for leishmania was negative; the direct exam of the biopsy showed presence of fumagoid cells (Fig. 3), and the biopsy showed a granulomatous infiltrate with giant cells and fumagoid cells (Figs. 4 and 5). *Fonsecae sp* was isolated from the fungus culture (Figs. 6 and 7).

DISCUSSION

Chromoblastomycosis is a disabling granulomatous dermatomycosis of chronic evolution that affects

How to cite this article: Chang P, Arana E, Arenas R. Chromoblastomycosis. Our Dermatol Online. 2016;7(3):305-308.
Submission: 19.11.2015; **Acceptance:** 30.01.2016
DOI: 10.7241/ourd.20163.82

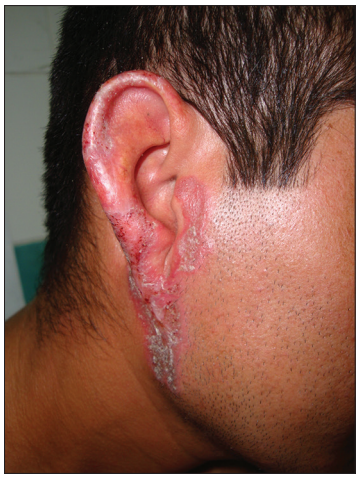

Figure 1: Panoramic view of the lesion.

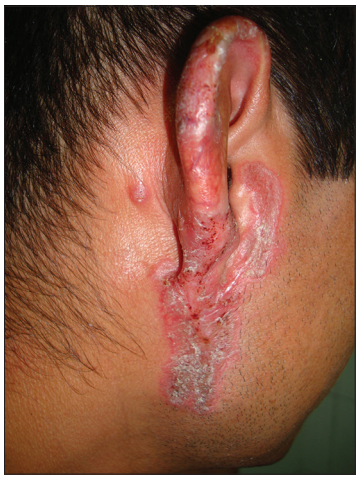

Figure 2: Erythematous scaly plaque, ear lobe, pre nodule region and retroauricular.

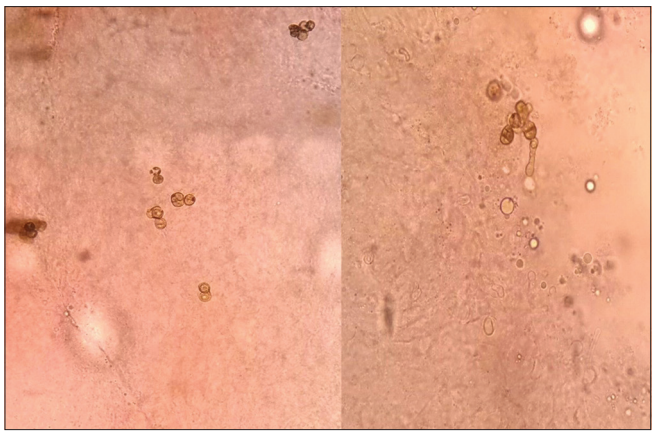

Figure 3: Fumagoid cells on direct exam of skin biopsy.

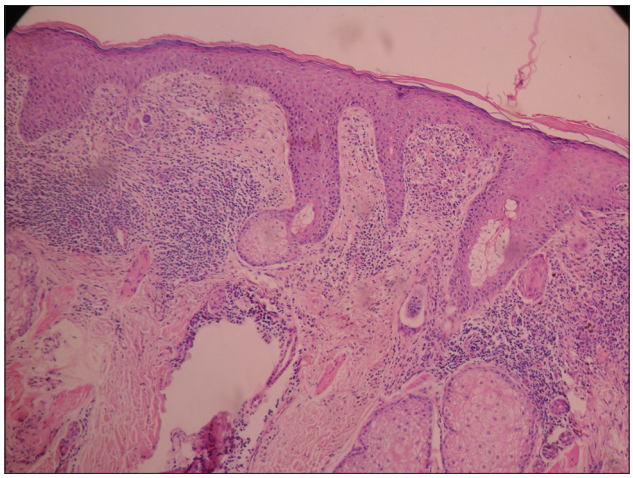

Figure 4: Panoramic view of skin biopsy.

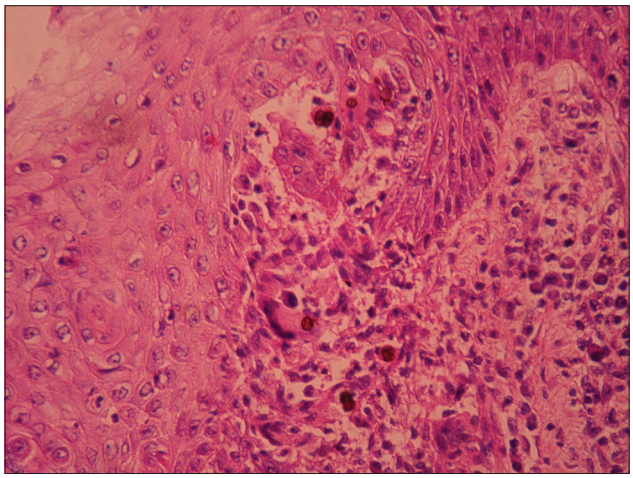

Figure 5: Granulomatous infiltrate with giant cells and presence of fumagoid cells.

the skin and subcutaneous tissue and localizes in the inferior limbs after a trauma, with formation of nodules and warty plates that can become ulcerated and give place to tumor masses, caused by a variety of black or dematiaceous fungus. It is observed in people who work barefoot [1,2]. It predominates in tropical and subtropical regions particularly in Latin America, although there have been cases in the five continents and therefore is considered of cosmopolite character. It has been called chromomycosis, Fonseca disease, Pedroso and Lane disease [2].

The main agents are *Fonsecaea pedrosoi*, in endemic areas of tropical and subtropical environments; *Fonsecaea compacta*, *Cladosporium carrionii*, in dry climates, in a lesser proportion *Phialophora verrucosa*, *Rhinocladiella aquaspersa*, *Fonsecaea monophora*, *Fonsecaea nubica*, *Exophiala dermatitidis*, *Exophiala spinifera*, *Rhinocladiella richardsiae*, *Cladophialophora yegresii* [3,4].

The term chromoblastomycosis was first used by Terra in 1922 to designate a deep subcutaneous mycosis in the lower limbs, especially feet [5]. The first case was described in 1911 by Pedroso y Sao Paulo, Brazil; in

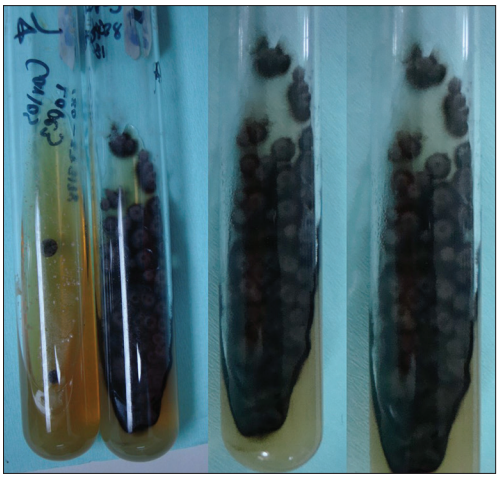

Figure 6: Fungi culture, presence of blackish colony.

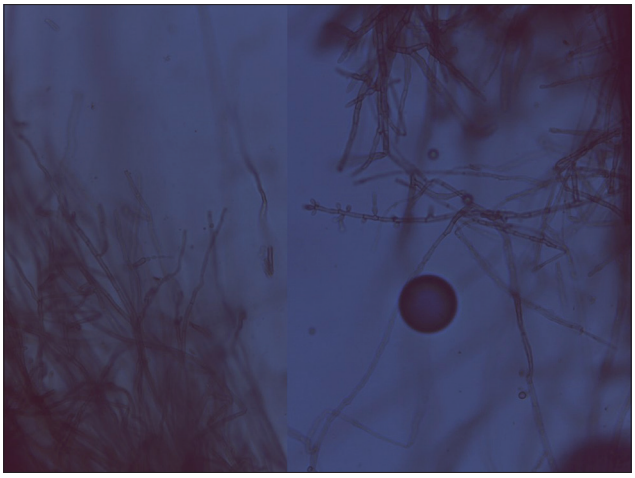

Figure 7: From the skin culture *Fonsecae sp* was isolated.

1935, Morales publishes the first chromoblastomycosis case in Guatemala, and successively, multiple chromoblastomycosis cases have been reported in the world [2].

It is a mycosis in tropical and subtropical areas considered as an American disease. However, because of the many cases published in the world, is considered cosmopolite with predominance in the American continent. The country with the highest number of reported cases is Brazil, although the country with the highest per capita proportion is Costa Rica, which has one case per 24,000 habitants. Dominican Republic, Cuba and Puerto Rico follow by frequency order. Other countries are: México, Guatemala, Honduras, Venezuela, Colombia, Madagascar, Australia, Congo and the south of China [2,5].

In Dominican Republic more than 500 cases have been reported and all the clinical forms reported in the literature have been studied, as well as other clinical varieties [1].

The fungus live in the soil and in vegetables, in humid and hot climates with a temperature range between 25°C and 30°C [2].

It is frequent in farmers and lumberjacks. There is a traumatic inoculation of the etiological agent. It is located in the limbs -95%-, and the lower limbs -75%-, with clear predominance in the dorsal part of the foot. The remainder cases appear in the trunk, and rarely in the face or other regions of the body [3].

These fungus are dimorphic and in its parasitic phase manifest as fumagoid cells called muriform cells or sclerotic bodies; these in turn are called Medlar sclerotia.

Clinically the disease can manifest in a warty or vegetative form, as nodules, tumors, in superficial plate or as psoriasis, scars, tumors and elephantiasis. The warty form is the most frequent -53% [1,2].

The initial lesion can appear as a papule that extends in the surface and forms well-limited erythematous-scaly plates, asymmetric and unilateral. They grow slowly in the term of months; generate erythematous nodules that get covered in scales. Approximately one year after appear as extended warty or vegetative plates covered abundantly in scales, ulcers, bloody scars [6]. The size varies from one millimeter up to big plates of several centimeters. Symptomatology of the disease is variable. As the disease advances, it becomes chronic and leaves scars. Most of the patients consult one to five years after the onset of the disease, and there have been cases with 40 years of evolution [1,4].

Among the differential diagnosis are leishmaniasis, cutaneous tuberculosis, psoriasis, sporotrichosis, coccidioidomycosis, blastomycosis, squamous cell carcinoma.

Complications of the disease are added bacterial infection. In very chronic cases, warty lymphostasis and degeneration to squamous cell carcinoma and melanoma, due to the chronic swelling and fibrosis [6].

The easiest way to diagnose is direct examination. The parasite elements must be sought in fragments of the tissue and most of all in scales presenting "black dots." Potassium hydroxide is used 10-40% in places

where the fumagoid cells are in groups of two or more. These are spherical or ovoid, measure 4 to 8 microns in diameter, are yellowish/brownish in color, have a thick membrane, and on occasions have divisions or filaments. Can be compared to coffee grains. Their culture allow confirmation of the causing agent [1,2].

Histopathology is very useful in diagnosis. Sometimes it can show a suppurating granuloma that sometimes is tuberculous. At skin level there is hyperkeratosis with parakeratosis, marked irregular acanthosis that sometimes form a pseudoepitheliomatous hyperplasia. In the superficial and medium skin a granulomatous reaction with lymphocytes, plasma cells, leukocytes, giant epithelioid cells of Langhans type, and the presence brown fumagoid cells [1,2].

Treatment has been a clinical challenge in spite of modern systemic antifungals. In small lesions with little evolution, cryosurgery is recommended as long as they are not located in bending sites, as well as surgery. Some authors recommend itraconazole alone in combined with 5-fluorocistine [7].

When lesions are extended or spread 5-fluorocistine, amphotericin B itraconazole, fluconazole or terbinafine can be used, the last 3 for 6 to 12 months.

Electrodessication, radiotherapy, local heat and vitamin D are other modes of treatment.

In lesions small and of short evolution medical surgical treatment is recommended such as: itraconazole, fluconazole or terbinafine for 3 months, with cryosurgery.

Therapeutic success can be related to the etiological agent (*C. carrionii* is more sensitive than *F. pedrosoi*), with the severity of the illness (edema and skin fibrosis can reduce the reach of antifungals to the skins), and with the election of the antifungal drug. So far, there is no consensus nor standardization to establish healing criteria for this mycosis [8,9]. Head and neck lesions do not respond with the same efficacy as other regions at distal level [1].

The preventive measure, aside of avoiding traumatic inoculation of fungi is the use of closed shoes.

Prognosis will depend on the stage of the lesions and their evolution.

In our environment, chromoblastomycosis is the third most common subcutaneous mycosis. It predominates in the lower limbs in warty form and *F. pedrosoi* is the most frequent etiological agent.

Consent

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

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

Our Dermatology Online

Case Report

Erythema gyratum repense like eruption in bullous pemphigoid: A case report

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ABSTRACT

Bullous pemphigoid (BP) is an autoimmune disease characterized by presence of bullous eruption on the trunk and extremities especially flexural aspects of the limbs. This disease usually occurs in the elderly. The initial presentation of BP is variable. An urticarial or erythematous rash may precede the appearance of the blister formation and can be associated with itch or pruritus. We presented 87 year old bedridden man diagnosed with BP who initially presented with erythema gyratum repens like eruption before blister formation. This case report discusses the presentation of figurate erythema in non-bullous phase of BP for clinicians.

Key words: Erythema; Pemphigoid; Bullous

INTRODUCTION

Bullous pemphigoid (BP) is an autoimmune bullous disease that is characterized by sub epidermal blisters on trunk and extremities and oral lesion is unusual. This disease may be signaled by urticarial or eczematous rash. Skin biopsy is the most reliable test for diagnosis. A direct immunofluorescence (DIF) shows deposition of Auto Antibodies (IgG) and complement (C3) at the dermo- epidermal junction [1,2].

The goal of this reporting is to explain variable features of non-bullous phase of BP and the association between BP and erythema gyratum repens like eruption in a 87 years old man.

CASE REPORT

A 87 years old bed ridden male admitted to hospital with chief complain of several blisters formation and pruritus. He started to have polycyclic erythema eruption at the back of the trunk since 20 days ago which was configured as Erythema gyratum repens like eruption.

The lesions were troublesome because of pruritus. Five days later, tense blisters were formed at the dorsal aspect of the right hand and were rapidly outspreaded through upper and lower extremities. He gave no history of weight loss or constitutional symptoms. His past medical history revealed cerebrovascular attack (CVA), femur fracture, dementia, BPH and nephrectomy. He was also incontinence since 3 years ago and was relatively bedridden before admission. The only drug he used was Finasteride 5 mg/day. His personal history was remarkable for cigarette smoking 60P/Y. He had no family history of cancer and skin disease.

Detailed cutaneous and mucosal examination showed erythematous plaques at the forehead and excoriated crusts on face and neck. Erythema gyratum repens like eruption was seen at the back of the trunk with typically "wood grain" appearance (Fig. 1). At the front aspect of the trunk there were multiple annular erythematous formations with crust. Multiple tense blisters and vesicles were detected all over the arms and legs (Figs. 2 and 3).

How to cite this article: Eftekhari H, Darjani A, Nazari M, Shafaei S, Yousefkhani L,Nickhah N. Erythema gyratum repense like eruption in bullous pemphigoid: A case report. Our Dermatol Online. 2016;7(3):309-311.

Submission: 27.11.2015; **Acceptance:** 25.01.2016

DOI: 10.7241/ourd.20163.83

The blisters were filled with clear fluid which aroused from erythematous skin. Nikolsky sign was negative and the rest of the examination of oral cavity and genitalia did not show abnormality. In general examination lymphadenopathy was not detected. Two plus edema of right foot was seen. He had wheeze all over the lung. His left hand was spastic and in an internal rotation position due to the past history of CVA.

Two biopsies were taken from the bullae formation of the hand and the figurated erythema at the back.

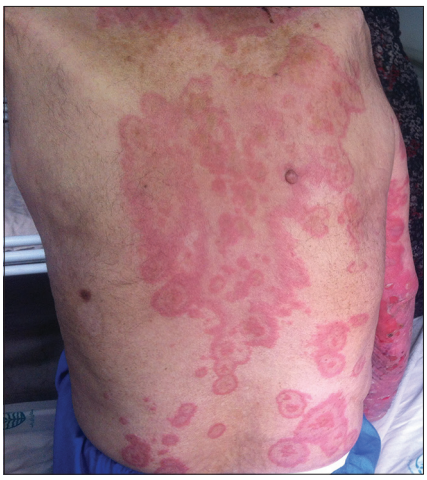

Figure 1: Erythema gyratum repense like eruption at the back.

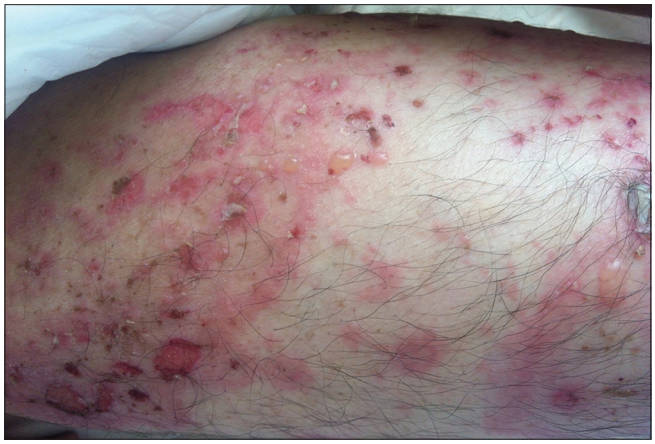

Figure 2: Tense bullae with erosion and crust.

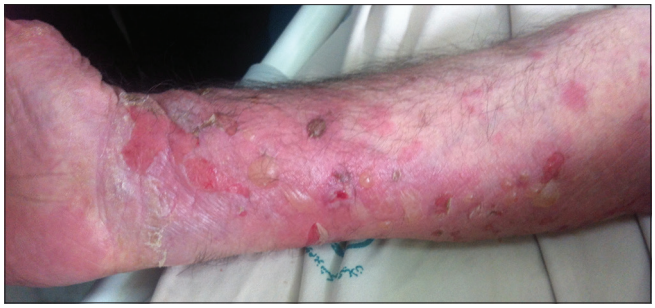

Figure 3: Tense bullae arising from erythematous skin.

The biopsy of the hand showed subepidermal blister containing some eosinophils (Figs. 4 and 5) with linear deposition of IgG and C3 along dermoepidermal junction and the biopsy from back lesion showed spongiotic dermatitis with eosinophils suggestive for early lesion of BP.

At the lab data he had white blood cell, 18700 cell/m^3 with marked eosinophilia (23%), hemoglobin 12.4 g/dl, MCV104 fl and albumin 3 g/dl with total protein 5.6 g/dl. Other lab tests showed CRP 1+ and ESR 20. CXR was normal.

The patient was started on 30 mg Prednisolone and Clobetasol ointment for topical use. The erythema gyratum repens like eruption disappeared with the start of treatment in a few days and new blisters did not appear.

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

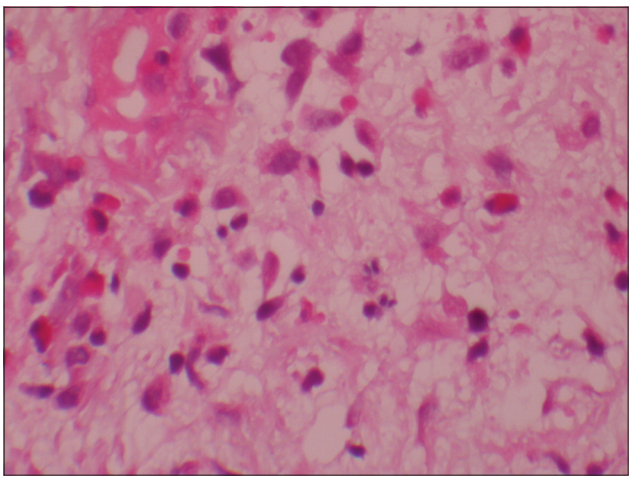

Figure 4: Sub epidermal bullae with eosinophils in the dermis.

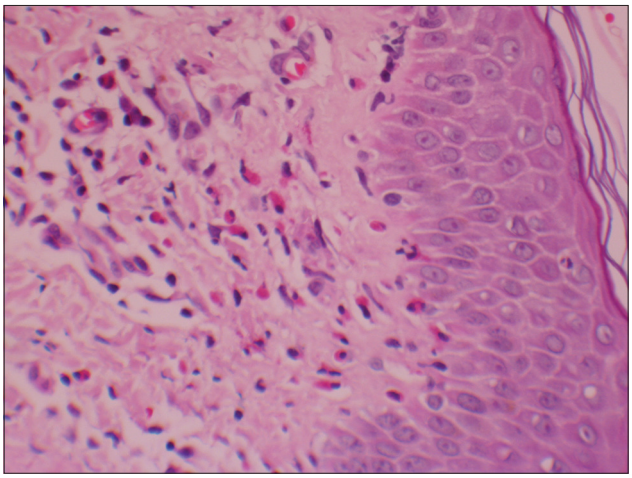

Figure 5: Eosinophilic infilteration in the dermis.

DISCUSSION

Bullous Pemphigoid is the most common autoimmune subepidermal blister disease that typically present in elderly. Before blister formation cutaneous manifestation of BP can be extremely polymorphic and nonspecific, from mild to severe pruritus alone to eczematous, papular or urticarial lesion. This signs and symptoms can be persistent for several weeks or months alone, and may remain as the only signs of BP.

The BP has association with some situation such as internal malignancy (GI, urinary bladder, lung), lymphoprolifrative disorder, autoimmune disease (such as IBD, SLE, and Dermatomyositis), trauma and neurologic disorders (such as MS, Stroke, dementia, and psychiatric disease). Patients with Erythema gyratum repens should be considered as having malignancy and should be mandatorily evaluated.

Several studies showed the association between figurate erythema and BP. In this case erythema gyratum repens like eruption was occurred before blister formation and there were no signs and symptoms of malignancy during six month follow-up.

Gilmour et al, reported a patient with urticarial arcuate lesion in a BP patient. Further assessment showed an underlying colon cancer [3]. In contrast Reynoso et al reported a patient with BP and figurate erythema without underlying malignancy [1].

Hadi et al studied 9 case of BP with figurate erythema that three of them had underlying malignancy (bronchogenic carcinoma and colon cancer) [4].

Urano-Saehisa et al, reported a patient with BP, bizarre figurate erythematous eruption and generalized pigmentation with underlying adenocarcinoma of stomach [5].

Geraham et al, report a case with figurate erythema resembling erythema gyratum repens in a patient with BP that have bronchial carcinoma [6]. Clinical Importance: Erythema gyratum repens can be a feature of bullous pemphigoid even absence of malignancy.

ACKNOWLEDGMENT

The authors thank the staff of Razi Hospital and the patient. There is no financial support as well as no conflict of interest in this study.

Consent

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

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

Our Dermatology Online

Case Report

Clinical diagnosis and a short-term treatment of bullous pemphigoid in an adult Yemeni female: A case report

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ABSTRACT

Bullous pemphigoid (BP) is an acquired, inflammatory, sub-epidermal, immunobullous disease. It usually occurs in the elderly population and reported cases of BP in adults are very rare. This report describes an adult Yemeni female who was clinically diagnosed to have BP, and successfully treated with a steroid containing short term regimen. It concludes the possibility of reaching a clinical diagnosis of BP in disadvantaged areas based on the clinical criteria, and the importance of considering a diagnosis of BP in young adults and even in children who present with itchy tense blisters of forearms, hands and feet. When accessible, further immunofluorescence studies are important to confirm the accuracy of the clinical diagnosis of BP.

Key words: Bullous; Pemphigoid; Adult; Clinical; Diagnosis; Treatment; Yemen

Abbreviations: BP, bullous pemphigoid; PV, bullous vulgaris; SJS, Stevens-Johnson syndrome; TEN, toxic epidermal necrolysis; SSSS, staphylococcal scalded skin syndrome; DH, dermatitis herpetiformis; EBA, epidermolysis bullosa acquisita; LABD, linear IgA bulous dermatosis; PCT, porphyria cutanea tarda; EM, erythema multiforme; NSAIDs, non-steroidal anti-inflammatory agents

INTRODUCTION

Bullous pemphigoid (BP) that was first described by Leverin 1953 is an acquired, acute or chronic, inflammatory, sub epidermal, immunobullous disease. Caused by autoantibody-mediated disruption of adhesion between basal keratocytes and the basement membrane [1,2].

Clinically, the earliest lesions may appear urticarial (like hives). Characteristic tense, usually symmetrical, ungrouped, with serous and/or hemorrhagic contents blisters of variable size eventually erupt either over normal or erythematous bases, most commonly at flexural areas of the limbs, trunk and abdomen. Any part of the skin surface can be involved; oral lesions have been reported, but other mucosal surfaces are usually not involved [2,3]. Clear diagnostic criteria can be lacking for definitive diagnosis in less than clear cut cases. If untreated, BP can persist for months or years, with periods of spontaneous remissions and exacerbations [4,5].

In Yemen, autoimmune disorders are among skin diseases groups with the lowest incidence rate. In general, BP is a rare, but the most commonly seen autoimmune blistering disease; most commonly occurs in elderly persons, but occasionally is seen in young adults and even in children [6,7].

This report describes an adult Yemeni female patient seen at Al Helal Hospital. Radaa district, of Al Baydaa governorate, in central Yemen, who was diagnosed to have BP based on history taking and physical examination, and successfully treated with a steroid-containing short-term regimen.

How to cite this article: Al-Kamel MA. Clinical diagnosis and a short-term treatment of bullous pemphigoid in an adult Yemeni female: A case report. Our Dermatol Online. 2016;7(3):312-315.

Submission: 09.12.2015; **Acceptance:** 15.02.2016

DOI: 10.7241/ourd.20163.84

CASE REPORT

A 29-years-old Yemeni female was referred to my service in Nov. 27, 2015, with numerous itchy, tense vesicles and bullae on erythematous bases, located predominantly on her palms and soles, dorsa of hands and feet and ventral forearms (Fig. 1), with two crusted erosions on the lower lip and soft palate, of three weeks duration, insidious onset and progressive course. No ocular or genital lesions, and the trunk was spared.

The blisters were sub-epidermal, filled either with clear or purulent fluid and concurrently occurred with diffuse pruritic urticarial patches which have started appearing few weeks prior to the visible blisters. The patient said that she evacuated some blisters by her hand.

Elicitation of clinical dermatological signs revealed positive Asboe-Hansen sign (Fig. 2), negative Nikolsky sign, and most of the solitary bullae showed regular rounded borders.

The patient reported a medical history with a frequent use of over-the-counter non-steroidal anti-inflammatory agents (NSAIDs), particularly ibuprofen to control a chronic daily headache. Prolonged sun-exposure was reported; past traumatic and family histories were unremarkable.

Review of systems reveals no other system complaints (fever, tachycardia, hypotension, altered level of consciousness, sore throat, chills, arthralgia, cough, dysuria, nor conjunctivitis).

Complete blood count, liver function and renal function tests were unremarkable, and the patients' cardinal signs were unaffected. Biopsy for histology and immunofluorescence testing was unavailable. Thus, a provisional diagnosis of bullous pemphigoid (BP) of this patient was made based on the history and clinical examination.

She was hospitalized, and the treatment regimen included oral 60 mg/day prednisolone as a starting dose (gradually tapered over the next17 days), oral azithromycin 500 mg/day for 6 days, betamethasone-clioquinol containing topical cream (Betnovate-C™) thrice a day and pheniramine maleate injections (first-generation antihistamines) twice daily. Intravenous fluids, Vitamins A, E and C tablets and potassium permanganate as soaks (diluted as 1:1000) were prescribed as an adjuvant therapy.

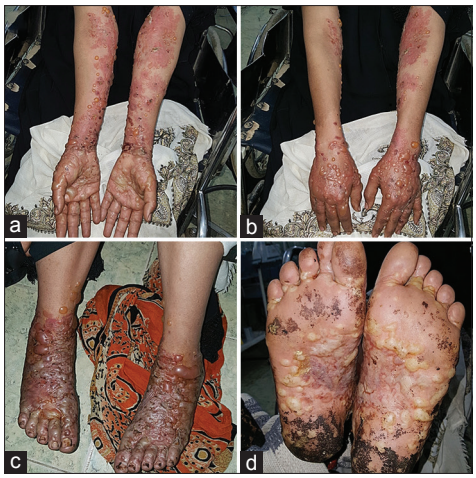

Figure 1: Clinical finding at the 1st presentation. Rounded, tense, sub-epidermal blisters on erythematous bases are noted on (a) palms, skin flexures of the elbows and wrists, (b) dorsa of the hands, (c) dorsa of the feet, (d) and soles.

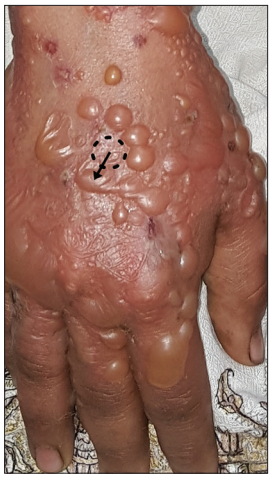

Figure 2: Dotted circle and the black arrow demonstrate the positive Asboe-Hansen sign.

The patient left the hospital on the 2nd day upon her request. On the 7th day, she was presented for a follow-up with complete remission of the blisters, but with post-inflammatory erythematous patches (Fig. 3). The patient was advised for regular future follow-up visits.

DISCUSSION

The diagnosis of bullous pemphigoid (BP) is generally made based on the clinical features which may be seen in patients with all forms of BP which include significant pruritus which is frequently present weeks or months before the appearance of any visible skin lesion and may be the only manifestation of the disease and tense fluid filled sub-epidermal blisters that do not rupture easily, supplemented by histologic features.

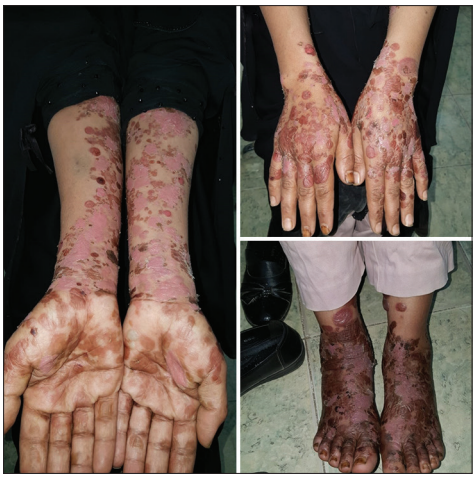

Figure 3: Clinical findings at the 7th day follow-up. All lesions were in complete remission.

Palmoplantar involvement with vesiculobullous lesions may also represent the only feature of the disease (dyshidrosiform pemphigoid), or occur in association with other more typical widespread lesions. Localized forms of BP account for up to 30 percent of cases. Blisters involves the mucosa in up to a quarter of patients [8-11].

Other clinical dermatological signs such as Asboe-Hansen sign (+ve in BP) and Nikolsky sign (-ve in BP) gave a clue to the probable possible diagnosis of BP [12].

In the current case, I relied on these cardinal symptoms and clinical signs to reach a clinical diagnosis of BP at such a suburban area lacking the advanced diagnostic.

Several triggers have been anecdotally implicated in the development of BP, such as ultraviolet irradiation, X-ray therapy, and exposure to some drugs. In the current case, non-steroidal anti-inflammatory agents (NSAIAs) and prolonged sun exposure are suspected trigger factors.

This patient had some clinical events, suggesting other blistering disorders which were considered and then ruled out on the basis of at least one characteristic clinical feature that distinguish each of them from BP, as the following:

Positive Asboe-Hansen sign and negative Nikolsky's helped in the exclusion of Stevens-Johnson syndrome (SJS), Hailey-Hailey disease, staphylococcal scalded skin syndrome (SSSS), and pemphigus vulgaris (PV); SJS and toxic epidermal necrolysis (TEN) are characterized by a prodrome of malaise and fever, and mucosal membranes are affected in 92 to 100 percent of patients, usually at two or more distinct sites (ocular, oral, and genital); TEN involves sloughing of greater than 30 percent of the body surface area; Skin lesions in PV are usually erosive, painful (but very rarely pruritic), superficial and do not form tense bullae; Blisters in dermatitis herpetiformis (DH) are the result of gluten sensitivity, often associated with other autoimmune disorders, and the lesions usually do not appear in the mouth; In epidermolysis bullosa acquisita (EBA), blisters tend to appear both spontaneously and as a result of trauma, predominantly on trauma-exposed body surfaces, and lesions heal with significant scarring; In linear IgA bulous dermatosis (LABD), blisters arise on normal, erythematous, or urticarial skin with a characteristic 'string of pearls' appearance; In porphyria cutanea tarda (PCT), patients present with blisters on sun-exposed areas of the body such as face, neck and hands, and the lesions are often painful, heal slowly with atrophic scars, milia, and post-inflammatory hyperpigmentation; In erythema multiforme (EM), the hallmark of which is the iris or target lesion [13-16].

As per this case, a short-term treatment regimen containing high dose of oral steroids, oral azithromycin, and potent topical steroids is an effective option in the treatment of BP.

CONCLUSION

This report concludes the possibility of reaching a clinical diagnosis of BP in disadvantaged areas through careful clinical examination, and the importance of considering a diagnosis of BP in young adults and even in children who present with itchy tense blisters of forearms, hands and feet. When accessible, further immunofluorescence studies are important to confirm the accuracy of the clinical diagnosis of BP.

Consent

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

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

Our Dermatology Online

Case Report

Autosomal recessive anhidrotic ectodermal dysplasia (Christ - Seimens - Touraine syndrome) in siblings

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ABSTRACT

Anhidrotic Ectodermal Dysplasia (Christ - Seimens - Touraine Syndrome)is a rare genodermatoses comprising triad of hypohidrosis (anhidrosis), hypotrichosis and anodontia. It is usually inherited as X-linked recessive. Autosomal dominant and recessive inheritance is very rare, here we report a case of autosomal recessive anhidrotic ectodermal dysplasia.

Key words: Anodontia; Genodermatoses; Hypotrichosis; Autosomal recessive; Ectodermal dysplasia; Christ-Siemens-Touraine syndrome

INTRODUCTION

Ectodermal dysplasias (EDs)are a heterogenous group of hereditary disorders characterized by certain shared structural and functional abnormalities in tissues derived from the ectoderm [1]. They are characterized by deficient function of at least 2 ectodermal derivatives such as skin, hair, teeth and sweat glands. Although more than 170 different subtypes of ectodermal dysplasia have been identified, these disorders are considered to be relatively rare with an estimated incidence of 1 case per 100,000 [2].

Ectodermal dysplasia is divided into two types based on the number and function of sweat glands: hidrotic ectodermal dysplasia (Clouston syndrome)and hypohidrotic (anhidrotic)ectodermal dysplasia (HED) (Christ-Siemens-Touraine syndrome) [3].

Hypohidrotic ED (Christ-Siemens-Touraine syndrome) is the most frequent form of ectodermal dysplasia, and genetic defects in ectodysplasin signal transduction pathways are the basis of this syndrome [4]. It is characterized by sparse hair, heat intolerance, and excessively dry skin due to the absence of sweat glands and abnormal spiky or absent teeth [5].

We report typical features of the CST syndrome in siblings, inherited in autosomal recessive pattern, which is a rare pattern of inheritance in this syndrome.

CASE REPORT

A 7-year old boy presented with the chief complaint of child being uncomfortable in warm weather and recurrent episodes of fever in summer. He also complained of necessity to take 6 to 8 baths per day with cold water in summer. There was similar history in the younger sibling, who was a 4 year old girl. The mother stated that her children had recurrent episodes of unexplained hyperpyrexia and thirst; and were not able to sweat, and she had to apply some precautions to protect them from overheating during physical exertion or warm weather. There was no similar history in the parents or any other family members and the children were a product of non-consanguineous marriage. On general examination both the siblings had sparse, thin, light, blond hair over the scalp, scanty eyebrow and eyelashes, depressed nasal bridge (Figs. 1a and 1b), frontal bossing (Fig. 2), and prominent supraorbital ridges. Lips were dry, everted, and prominent. The skin was dry and wrinkled. Hyper pigmentation was

How to cite this article: Chowdhry S, Shukla A, Gupta S. Autosomal recessive anhidrotic ectodermal dysplasia (Christ - Seimens - Touraine syndrome) in siblings. Our Dermatol Online. 2016;7(3):316-318.

Submission: 30.11.2015; **Acceptance:** 24.02.2016

DOI:10.7241/ourd.20163.85

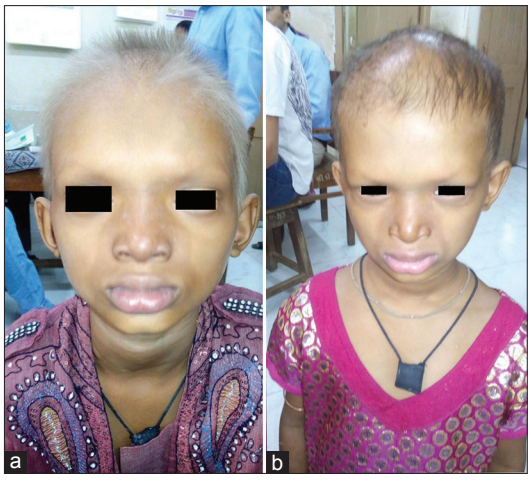

Figure 1: (a) Older Sibling-Sparse and light colored scalp hair with sparse eyebrows and eyelashes, Perioral and periorbital hyperpigmentation, Depressing nasal bridge and frontal bossing. (b) Younger sibling-with striking similar face as older sibling and similar features.

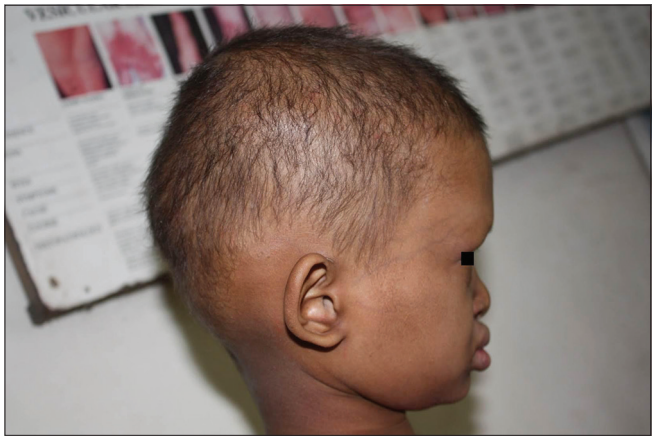

Figure 2: Sparse hair and frontal bossing.

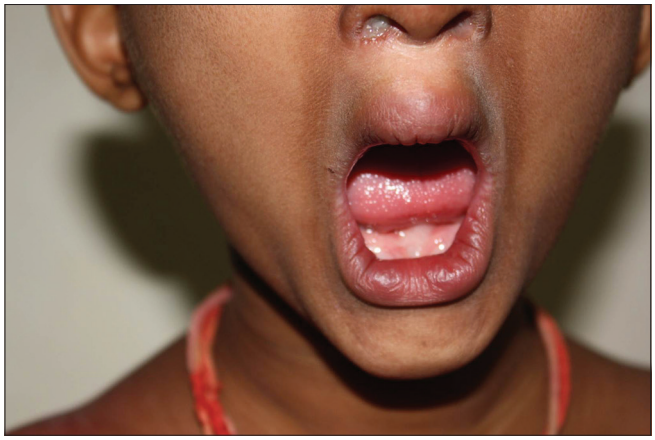

Figure 3: Complete anodontia.

evident around the eyes (Figs. 1a and 1b). There was a striking similarity in the facial features of both the siblings. Oral examination revealed complete anodontia. (Fig. 3)An ophthalmological examination revealed absence of lacrimal puncta. They had no nail dystrophy, and their intelligence was normal. With features of anhydrosis, anodontia, hypotrichosis and typical facial features both siblings were diagnosed as a case of Christ-Siemens-Touraine syndrome. Since there was no history of similar features in parents and both the siblings were involved and since they were of different sex, inheritance pattern was autosomal recessive. Parents were counselled about the inherited nature of the disease; however, they refused to undergo any genetic testing. Parents were counselled about maintenance of ambient cool temperature, application of sunscreen lotion, maintenance of hydration and prevention of children from exposure to excessive heat. We considered placement of removable partial denture as the best treatment option for anodontia for both children and referred them to a prosthodontist to receive professional prosthodontic procedures.

DISCUSSION

Ectodermal dysplasia is a rare disorder with defects in two or more of the following structures: the teeth and the skin and its appendages including hair, nails, eccrine, and sebaceous glands [4]. Anhidrotic ectodermal dysplasia (AED)is a rare disorder also known as Christ-Siemens- Touraine syndrome, Charles Darwin was one of the earliest observers to describe this condition. Subsequently, a full description of this disorder was given by Christ, Siemens and Touraine. It is commonly transmitted as an X linked recessive disorder. However, rarely autosomal recessive and autosomal dominant inheritance has also been seen [6]. Hypohidrotic ectodermal dysplasia manifests as a triad of defects, partial or complete absence of sweat glands, anomalous dentition and hypotrichosis. Affected children are unable to sweat, they may experience episodes of high fever in warm environment and may be mistakenly considered to have fever of unknown origin. The typical facies is characterized by frontal bossing, malar hypoplasia, flattened nasal bridge, recessed columella, thick and everted lips, wrinkled hyper pigmented periorbital skin and prominent low set ears. The skin over the entire body is dry, finely wrinkled and hypo pigmented. Anodontia or hypodontia with widely spaced conical teeth are consistent features. Poor development of mucous gland in the respiratory and gastro-intestinal tract may result in increased susceptibility to respiratory infections, purulent rhinitis, dysphonia and diarrhoea.

This disorder is non-progressive and life expectancy is normal or just below average. Mortality in early childhood is due to hyperthermia, failure to thrive and respiratory infections. Maintenance of cool, ambient temperature and psychological support is needed for the patient.

There is no specific treatment for this condition. Genetic counselling of involved families plays an important role.

Consent

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

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

Our Dermatology Online

Case Report

Cutaneous Rosai–Dorfmans nodules on cheek

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ABSTRACT

Sinus histiocytosis with massive lymphadenopathy (SHML) or Rosai–Dorfman disease (RDD) is a benign idiopathic histiocytic proliferative disorder that commonly involves the lymph nodes but secondarily involves the skin. However, purely cutaneous disease without lymphadenopathy or internal organ involvement may rarely occur. We report a patient who presented with asymptomatic non-specific enlarging skin nodules without evidence of lymphadenopathy or internal disease. Histopathological examination of skin lesions in this patient showed proliferation of large histiocytes with phagocytosed inflammatory cells characteristic of Rosai–Dorfman disease. The diagnosis of purely cutaneous Rosai – Dorfman disease may be complicated by its rarity, nonspecific clinical appearance of skin lesions and broad histopathological differential diagnosis of this disorder. The prognosis is excellent in most cases. Complete spontaneous regression is known to occur. Both physicians and pathologists need to be aware of rare conditions which masquerade as lymphoproliferative disorders. Here we describe one such clinical condition – "Cutaneous Rosai Dorfman disease" and an approach to such patients.

Key words: Rosai – Dorfman disease; Sinus histiocytosis; Massive lymphadenopathy; Emperipolesis

INTRODUCTION

Most cases of Rosai –Dorfman disease present with painless cervical lymphadenopathy. The disease can involve both nodal and extranodal sites, including the skin. It is generally considered a benign, self limited proliferation of histiocytes. Although cutaneous involvement in RDD is common, purely cutaneous disease is rare. It may present at any age and is common in the second decade. Afro – Caribbeans are frequently affected.

Two etiological hypothesis have been proposed. One favours disturbance in cell mediated immunity and the other proposes the role of infection due to Epstein – Barr virus, Klebsiella, Brucella or human herpesvirus 6 as the causative agent [1].

CASE REPORT

An otherwise healthy 55 year old man presented with a 2 years history of nonpruritic lesions, slowly growing as multinodular skin coloured non inflammatory nodules on the left cheek (Fig. 1). The lesion was excised by a surgeon (Fig. 2). After 6 months of postoperative period, he started developing new nodules over the site of excision. It was totally asymptomatic. The patient had no lymphadenopathy or hepatospleenomegaly. Biopsy specimens from lesions showed nodular dermal aggregates of foamy histiocytes among neutrophils, lymphocytes and plasma cells. The histiocytic nuclei were uniform, round and vesicular. Phagocytosed plasma cells and lymphocytes were seen with abundant eosinophils. The histopathology report confirmed Rosai- Dorfman disease. Histopathology was very characteristic showing dermal infiltrate composed predominantly of histiocytes with large vesicular nuclei, abundant pale cytoplasm and emperipolesis of histiocytes (Fig. 3). Typically the characteristic histiocytes of RDD were positive for S -100, negative for CD1a and variably positive for CD68. Total excision was done by a plastic surgeon with total reconstruction of the excision site. Postoperative period was uneventful. After one year, patient showed no signs of recurrence as in our picture below.

How to cite this article: Srinivasan SM, Parthasarathy S. Cutaneous Rosai–Dorfmans nodules on cheek. Our Dermatol Online. 2016;7(3):319-322.
Submission: 13.01.2016; **Acceptance:** 09.03.2016
DOI:10.7241/ourd.20163.86

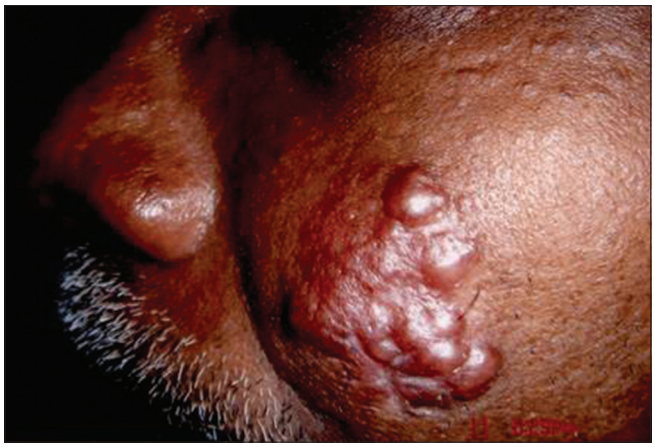

Figure 1: Multinodular skin coloured nodules over the left cheek.

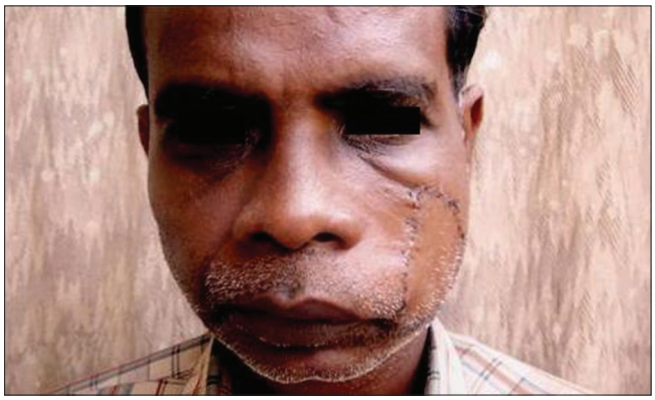

Figure 2: Postoperative clinical picture.

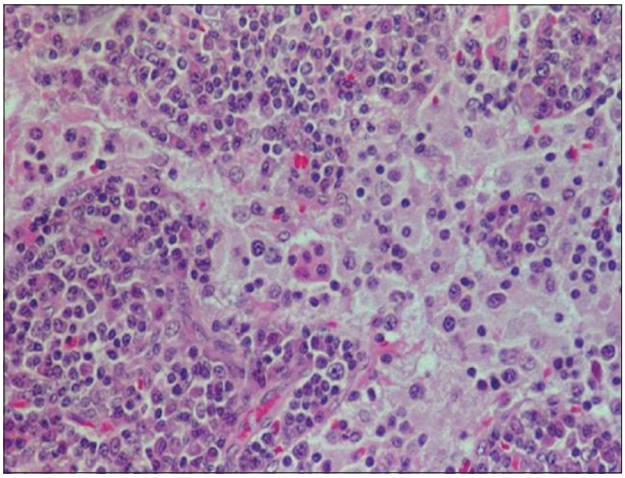

Figure 3: Biopsy of the nodule showing "Emperipolesis".

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

DISCUSSION

The term Cutaneous Rosai – Dorfman disease is used mostly for the forms of the disease in which involvement is restricted only to the skin, in order to differentiate it from sinus histiocytosis with massive lymphadenopathy in which there is systemic involvement of multiple sites including the skin. Extranodal forms occurs in about 43% of cases, out of which skin is the most common site [2]. The etiology of the disease still remains unclear, pointing towards a reactive process against infectious agents.

The purely cutaneous forms of the disease is more common in the older age groups when compared to the systemic forms. The most common site of lesions in CRDD is the face, followed by the back, chest, thigh, flank and shoulder. The lesions may resemble psoriasis or acne [3]. CRDD shows a marked female predominance (2:1). The majority of RDD patients are of African descent and the disease is rarely reported in Asian patients. Systemic RDD is more prevalent, while the pure cutaneous form accounts for only 3% of RDD [4] and involves only the skin and adjacent soft tissues without associated lymphadenopathy. Purely CRDD was reported for the first time by Thawerani et al in 1978 in a 48 year old male patient who presented with a solitary nodule on the shoulder [5]. Patients with CRDD are generally of normal health without fever, malaise, night sweat or other associated immune deficiency symptoms. Skin lesions are divided into three main types as papulonodular, indurated plaque and tumour. Based on literature, approximately 10% of patients have had cutaneous infiltrates, but very few of them have had lesions limited to the skin [6]. Cutaneous manifestations develop in about 10% of patients, characterized by asymptomatic xanthoma like, yellowish or reddish brown papules, nodules, and plaques which may ulcerate [7].

The retrospective review of the medical literature shows the following statistics in Tables 1 and 2.

Histopathology is very characteristic showing dermal infiltrate composed predominantly of histiocytes with large vesicular nuclei, abundant pale cytoplasm and emperipolesis of histiocytes. Emperipolesis means lymphocytophagocytosis (intact lymphocytes within the cells) which differs from phagocytosis in that lymphocytes taken up are not attacked by enzymes and appear intact within the histiocytes. Emperipolesis, a consistent finding in nodal disease, is often less apparent in extranodal sites and could be confidently identified in only 7 of 11 cases by hematoxylin and eosin staining morphology. Cutaneous sinus histiocytosis can be specifically diagnosed by skin biopsy [8].

Table 1: Differences between Nodal Rosai – Dorfman and purely cutaneous Rosai – Dorfman disease

Characteristics	Frequency (%)	Age (years)	Symptoms	Site	Emperipolesis
Nodal Rosai-Dorfman	60-70	20	Painless lymphadenopathy	Cervical region	Very comon
Purely cutaneous Rosai-Dorfman	3	40	Cutaneous lesion	Face, ears, trunk, extremities	Not very common

Table 2: Treatment modalities for Nodal Rosai – Dorfman and purely cutaneous Rosai – Dorfman disease

Nodal Rosai-Dorfman	Steroids	Chemotherapy	Methotrexate	Interferon α
Purely cutaneous Rosai-Dorfman	Steroids	Radiotherapy	Cryotherapy	Thalidomide

The distinctive histiocytes of RDD exhibit emperipolesis, association with numerous plasma cells and distention of lymph node sinuses or lymphatics in extra nodal sites. This helps in differentiating RDD from a variety of benign and malignant disorders such as melanoma and cancer metastasis to lymph nodes, in which phagocytosis of cells may be a prominent feature. Given the wide range of clinical presentations and the broad pathological differential diagnosis, the clinical hallmark of massive lymphadenopathy is often crucial for diagnosis of SHML. Immunophenotyping studies suggest that RDD can affect the antigen presenting activity of the skin dermal dendrocyte [9]. Although pathological evaluation is a key to definitive diagnosis, the variable presence of fibrosis, vascular proliferation, neutrophil microabscesses, lymphoid aggregates with germinal centers and background histiocytic proliferations of foam cells, multinucleated giant cell/or Touton cells may result in confusion of this disorder with a neoplastic, xanthomatous, infectious or other histiocytic process [10].

Clinical course of RDD can vary with some lesions healing spontaneously and others persisting for years or recurring after excision. In cases where it progresses to affect other organs, autoimmune hemolytic anemia, lymphocytopenia and neutropenia are common findings.

The differential diagnosis of RDD includes lymphoma, malignant histiocytosis, disseminated tuberculosis, and Langerhans cell histiocytosis (LCH). The phenomenon of emperipolesis is central in differentiating RDD as the rest of these diseases fail to exhibit lymphophagocytosis. Presence of weight loss, night sweats, hepatospleenomegaly and malignant cells staining positive for CD45 favours the diagnosis of lymphoma. Malignant histiocytosis differs from RDD clinically by its rapid downhill course and pathologically by the presence of malignant histiocytes having bizarre, pleomorphic nuclei. The histiocytes in LCH have a characteristic folded and grooved nucleus and exhibit CD1a positivity. Disseminated tuberculosis can be ruled out on the basis of absence of granulomas and negative staining for acid fast bacilli by Ziehl-Neelsen stain. The presence of characteristic histiocyte derived from circulating mononuclear cells, long history and an increased incidence of serum autoreactive antibodies during active disease suggest a possible pathogenic correlations with a dysregulatory process.

It is conceivable that some patients with purely cutaneous symptoms may have clinically undetectable systemic lesions. Lesions can mimic panniculitis [11]. However there have been no reported cases of cutaneous disease developing into systemic disease. The clinical and histopathological course is variable with some lesions resolving spontaneously over weeks to months. Histopathology is very characteristic showing emperipolesis of histiocytes [12]. Few lesions can also mimic vasculitis [13,14]. Pustular lesions may appear as acneiform eruptions and others persisting for years [15] or even recurring after excision as in this reported case. Few patients can also present as granuloma annulare, which is not associated with adenopathy. Treatment is generally not necessary for cutaneous RDD, but may be desired for cosmetic purposes or symptomatic relief. Cutaneous lesions have been reported to respond to radiotherapy, cryotherapy, excision, oral and topical corticosteroids and high dose thalidomide. Dapsone and thalidomide have been effective in cases refractory to other treatments. Steroid responsive CRDD can be associated with bilateral anterior uveitis and hypothyroidism. Many cases have reported improvement with combined application of Compound betamethasone and lidocaine, and intramuscular interferon and oral acitretin. But this mechanism requires further studies in the future. Utikal et al described a patient with complete remission of CRDD after receiving imatinib therapy, however a different study reported a patient with CRDD who was completely resistant to this treatment. Investigation of the true efficacy of any of these therapeutic modalities is complicated by the rarity of this disorder.

Scrutiny of the dermatological literature yields few cases of malignant histiocytosis presenting in the skin and breast using restrictive criteria as defined by Pileri et al (a malignant neoplasm of histiocytes, positive for one

or more histiocytic markers but negative for accessory/ dendritic cell markers. Several cases in the past have also been diagnosed as such, but do not adhere to the latest criteria/or lack the appropriate immunostaining techniques. To the best of our knowledge, this is the first case reported that adheres to Pileri 's criteria, yet does not show any evidence of systemic involvement.

Marshall in 1981 reviewed 320 cases based on literature, of which 6.9% presented chiefly with cutaneous lesions. These have been described on the face, scalp, chest, back and abdomen and more frequently on the extremities, especially the legs. Men are more often affected than women (2:2:1). Laboratory investigations may reveal pancytopenia, a raised ESR, eosinophilia, leukocytosis, altered liver function tests particularly increased aspartate aminotransferase, prolonged prothrombin time and cholestasis(in the absence of drug abuse or alcohol history).

CONCLUSION

Cutaneous Rosai Dorfman disease is a very rare cutaneous histiocytosis. When associated with lymphadenopathy and hepato-spleenomegaly, it can be fitted with sinus histiocytosis and the diagnosis is easier. Though we read about C-RRD, practical application of our knowledge is very meagre. This case report and literature survey emphatically emphasizes that histopathology is the concluding material to all dermatologists.

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

Our Dermatology Online

Case Report

Dermoscopic patterns in confluent and reticulated papillomatosis: a case report

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ABSTRACT

Confluent and reticulated papillomatosis (CRP) is an uncommon, progressive, distinctive dermatosis. It is characterized by persistent brown, scaly macules, papules, patches and plaques. It appears as tinea versicolor or acanthosis nigricans resulting in delay in the diagnosis. Hence a diagnostic tool which could help to diagnose this condition is dermoscopy. We reported a 24 year old male with skin lesions on upper back, neck and shoulders. Dermoscopic examination demonstrated fissures and ridges. Whitish structures were present on the ridges which are surrounded by grayish globules. These globules were lined by brown lines at the periphery. Fissures were brown in color. Global appearance was 'crocodile skin-like' pattern. Ridges and furrows in dermoscopy of CRP correspond to rete ridges in the histology. Whitish structures were there around follicular ostia which represent hyperkeratosis and acanthosis of follicular epidermis. Authors also observed sparse hairs in the involved area and V-sign under dermoscopy. Hence, pathogenesis of CRP probably revolves around hair follicles. CRP demonstrates specific dermoscopic patterns which correlate well with histopathologic changes. Therefore, authors propose that these patterns would aide in the diagnosis of CRP.

Key words: Acanthosis; Confluent reticulated papillomatosis; Dermoscopy; hyperkeratosis; Pathogenesis; Pattern; Whitish structures

INTRODUCTION

Confluent and reticulated papillomatosis (CRP) is an uncommon, progressive, distinctive dermatosis characterized by persistent brown, scaly macules, papules, patches and plaques [1].

CRP was first described in 1927 by Gougerot and Carteaud and it occurs predominantly in young adults and teenagers, affecting the neck, upper trunk and axillae with cosmetically displeasing appearance. Recognition of CRP is usually difficult by clinicians, including dermatologist as it frequently appears as pityriasis versicolor or acanthosis nigricans resulting in delay in the diagnosis [2]. Hence a diagnostic tool which could help to diagnose this condition is required to overcome this hurdle.

Dermoscopy is a noninvasive and quick auxiliary method that allows visualization of details of skin lesion which are subtle on examination by unaided eyes. And patterns are correlated with histopathological examination of the lesions. Thus, it is being employed commonly in clinical practice [3,4]. Therefore; dermoscopy can be utilized in the diagnosis of CRP. An attempt was made to evaluate the dermoscopic patterns in CRP. Authors believe that patterns are specific to CRP and they correlate to histopathological changes.

CASE REPORT

A 24 year old male presented with skin lesions on upper back, neck and shoulders since 10 months. Cutaneous examination revealed hyperpigmented, hyperkeratotic and scaly papules that coalesced into plaques in a reticulated pattern (Fig. 1). Lesions were asymptomatic. Tinea versicolor was suspected nevertheless potassium hydroxide preparation and culture for fungus was negative. Systemic examination

How to cite this article: Ankad BS, Dombale V, Sujana L. Dermoscopic patterns in confluent and reticulated papillomatosis: a case report. Our Dermatol Online. 2016;7(3):323-326.

Submission: 15.10.2015; **Acceptance:** 03.02.2016

DOI:10.7241/ourd.20163.87

was normal. Biochemical and hematological analyses were unremarkable. Dermoscopic examination was done using polarized dermoscopy. It demonstrated fissures and ridges. Whitish structures were present on the ridges which are surrounded by grayish globules. These globules were lined by brown lines at the periphery. Fissures were brown in color (Fig. 2). Global appearance was 'crocodile skin-like' pattern. A 4 mm punch biopsy was done and histopathology showed orthokeratosis, hyperkeratosis, mild acanthosis, follicular plugging and elongation of rete ridges in epidermis. There was inflammatory infiltrate surrounding hair follicle and sebaceous glands (Fig. 3). The patient was given minocycline 100 mg once daily for 8 weeks resulting in complete remission of lesions with hypopigmentation (Fig. 4). There was no relapse even after 15 months of follow up.

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

DISCUSSION

CRP is an uncommon dermatological condition. The etiology is unknown, although an abnormal keratinization has been implicated and sometimes has been associated with *Malassezia* species. It has a chronic course with remissions and exacerbations [5]. Accepted theories suggest that this condition might be the result of an exaggerated response to fungi [6].

Differential diagnosis of CRP include benign acanthosis nigricans, pseudoacanthosis nigricans, tinea versicolor, amyloidosis and importantly all these conditions present

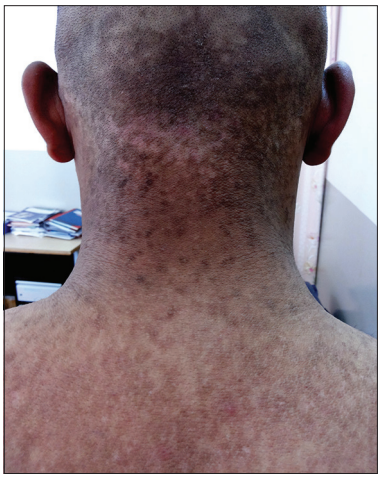

Figure 1: Clinical image of confluent and reticulated papillomatosis with hyperpigmented, hyperkeratotic and scaly papules in a reticulated pattern.

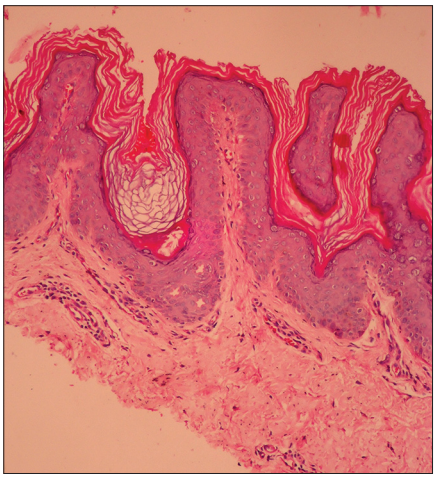

Figure 3: Histopathology showing orthokeratosis, hyperkeratosis, mild acanthosis, follicular plugging and elongation of rete ridges in epidermis (H and E, 10x).

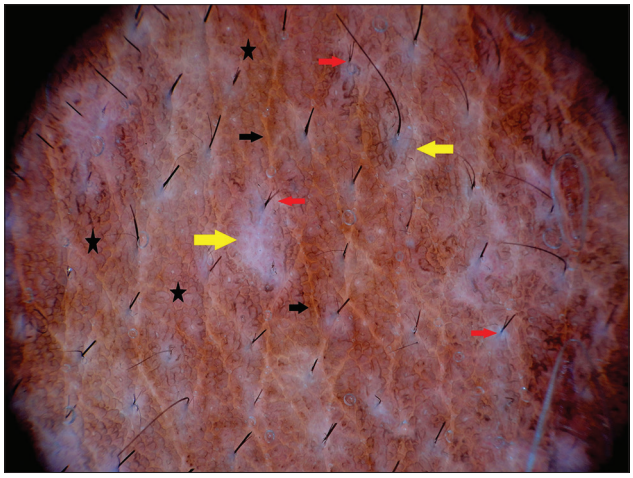

Figure 2: Dermoscopy showing ridges and fissures (black arrows), whitish structures (yellow arrows), grayish globules with brown lines (black stars) and V-hairs (red arrows).

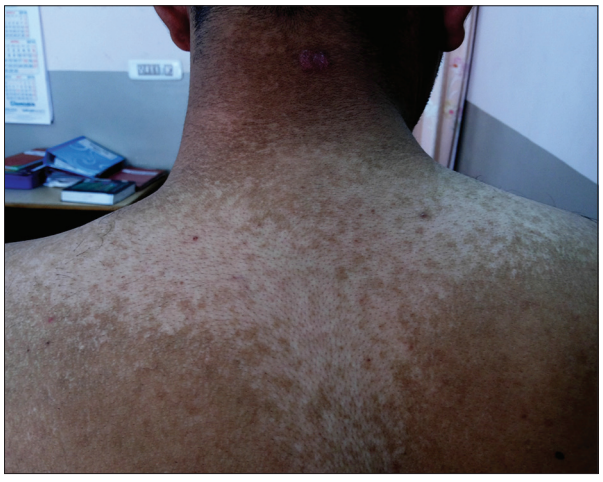

Figure 4: Clinical image showing improvement after treatment leaving behind hypopigmentation.

clinically as hyperpigmented patches and plaques that spread to become confluent and reticulated [7].

Dermoscopy is a new and valuable tool providing surface and subsurface microscopic views of skin and hair. This method fills the lacunae encountered in clinical diagnosis thus increasing accuracy of diagnosis [8].

Recently, dermoscopy is gaining importance in clinical practice as it is being used widely in the field of dermatology. Inflammatory conditions such as psoriasis, lichen planus, lichen sclerosis et atrophicus demonstrate specific patterns under dermoscopy [9].

Dermoscopy of CRP is very rare in the literature. In one report, authors observed sulci and gyri under dermoscopy in one patient with CRP [4]. However; patterns were not correlated with histological changes. In this study, dermoscopy of CRP demonstrated ridges and furrows which were arranged in 'crocodile skin-like' pattern in global view. Ridges and furrows were due to the confluent and reticulated nature of plaques and papules. Furrows were brown in color and were not well defined at few places. They were arranged in rhomboid pattern which correspond to rete ridges in the histology. Ridges and furrows are observed in seborrheic keratosis and they are in 'fat fingers or cerebriform' pattern [10,11].

Whitish structures were observed in the ridges and were centered on follicular ostia. These represent hyperkeratosis and acanthosis of follicular epidermis. Whitish structures are demonstrated in hypertrophic lichen planus and correspond to compact orthokeratosis, hyperkeratosis and acanthosis [12]. In lichen sclerosus, they appear as white chrysalis strands representing homogenization of collagen in the dermis, not the hyperkeratosis and acanthosis of epidermis [9]. In pyogenic granuloma, whitish structures are in 'white rail line' fashion revealing fibrous septa [13]. Therefore, whitish structures under dermoscopy indicate possible pathology in integument. Furthermore, these patterns should be considered in accordance with clinical features due to their varied pathological representation as mentioned above.

Brownish lines correspond to increased melanin in the epidermis and their pattern is different in each condition [10]. In solar lentigo, they are in 'uniform reticular' pattern and in acral nevus; they take 'parallel' pattern [11]. Here in CRP, brownish lines were at the periphery of grayish globules giving a wavy or curvilinear appearance. Thus pattern of pigmentary lines in dermoscopy gives clue to the diagnosis.

Authors observed interesting hair changes in CRP. Hairs were sparse in the involved area. Two hairs were emerging out of single follicular ostium at few points which is referred to as V-sign or V-hair. This trichoscopic finding is characteristic of trichotillomania and indicates trauma and damage of hair shafts [14]. The yeast, *Malassezia furfur*, which is sebaceo-folliculotrophic in nature, is implicated as etiologic organism in most of the cases of CRP [5, 6]. Furthermore, there is evidence of follicular plugging in histology which clearly states that follicles are involved in the pathogenesis [1]. Based on these observations along with hair changes and whitish structures around follicular ostia observed under dermoscopy in CRP, authors are of opinion that pathogenesis of CRP probably cornered around hair follicles. Nevertheless, this is a primitive observation, further evaluation is warranted.

Histopathology of CRP shows hyperkeratosis, acanthosis, elongation of rete ridges and undulated epidermis and follicular plugging with inflammatory infiltrate in the dermis [15]. In this study, hyperkeratosis, follicular plugging, mild acanthosis, elongation of rete ridges, papillomatosis and inflammatory infiltrate were noted in histopathology. However, authors could not observe the yeast.

CONCLUSION

Dermoscopy is gradually acquiring its importance as a diagnostic aide in clinical diagnosis in recent past. CRP demonstrates specific dermoscopic patterns which correlate well with histopathologic changes. Hence, authors propose that these patterns would aide in the diagnosis of CRP and recommend usage of dermoscopy in every day practice. Since, these observation were based on single case report, further studies of CRP involving large sample size are suggested.

Consent

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

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

Our Dermatology Online

Case Report

Colocalized nevus depigmentosus and lentigines

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ABSTRACT

Nevus depigmentosus (ND)is classically defined as a congenital nonprogressive hypopigmented macule, stable in size and distribution. A 17 year girl presented with hypopigmented patch with indented borders, present on the right side of face and neck since 3 years of age. Later on at the age of 5, numerous hyperpigmented punctiform spots appeared exclusively on the hyperpigmented area. On sun exposure, the hypopigmented area neither reddened nor burnt. On diascopy the margin of the hypopigmented lesion remained delineated. The dermoscopic examination showed 1-4 millimeters sized hyperpigmented lesions with a barely visible pseudonet, leading to the final diagnosis of colocalized nevus depigmentosus and lentigines.

Key words: Nevus; Hypopigmentation; Reverse; Mutation; Pigmentation

INTRODUCTION

Nevus depigmentosus (ND)is a rare, congenital, stable hypomelanosis first described by Lesser in 1884 [1]. The lesions usually present as dermatomal or quasidermatomal macules commonly on the trunk, lower abdomen, or proximal extremities. They are off-white in colour and have irregular, serrated, feathered, or geographic margins. The face, when involved, is a cause of social embarrassment for the patient. Unfortunately, there is no effective treatment for this condition. The case is being reported to highlight the phenotypic manifestation of reverse mutation.

CASE REPORT

A 17 year girl presented with hypopigmented patch with indented borders on the right side of face and neck which was first noticed by the mother of the child when she was 3 years old. Later on at the age of 5, numerous hyperpigmented punctiform spots appeared exclusively on the hyperpigmented area. With sun exposure the hypopigmented areas neither turned red nor did the skin burn at the site. On local examination, a single ill defined hypopigmented macule of size of about 25 X 8 centimeters was present at the angle of mouth on right side, further extending to lateral side of right ear, right angle of jaw, lateral right side of neck to about 6 centimeters below the clavicle. It was irregular in shape with serrated irregular margins. The surface was smooth and had multiple oval dark brown coloured macules of 1 to 4 mm in size (Fig. 1). On diascopy the margin of the hypopigmented lesion remained delineated. Adjacent oral mucosa was normal.

The dermoscopic examination (Fig. 2) showed 1-4 millimeter sized hyperpigmented lesions, with a barely visible pseudonet, leading to the final diagnosis of colocalized nevus depigmentosus and lentigines

DISCUSSION

The commonly used clinical diagnostic criteria for nevus depigmentosus (ND)are as follows [2-4]:

1. Leukoderma present at birth or of an early onset
2. No alteration in distribution of leukoderma throughout life
3. No alteration in texture or change of sensation in the affected area
4. Absence of hyperpigmented border

How to cite this article: Jaiswal P, Chowdhry S, D' Souza P. Colocalized nevus depigmentosus and lentigines. Our Dermatol Online. 2016;7(3):327-328.
Submission: 10.12.2015; **Acceptance:** 26.04.2016
DOI:10.7241/ourd.20163.88

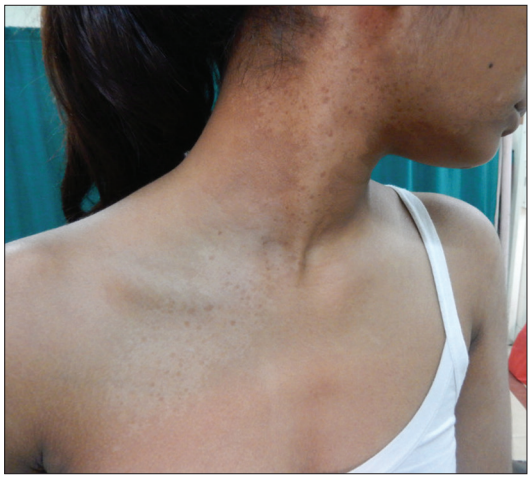

Figure 1: Single ill defined hypopigmented macule with lentigenes.

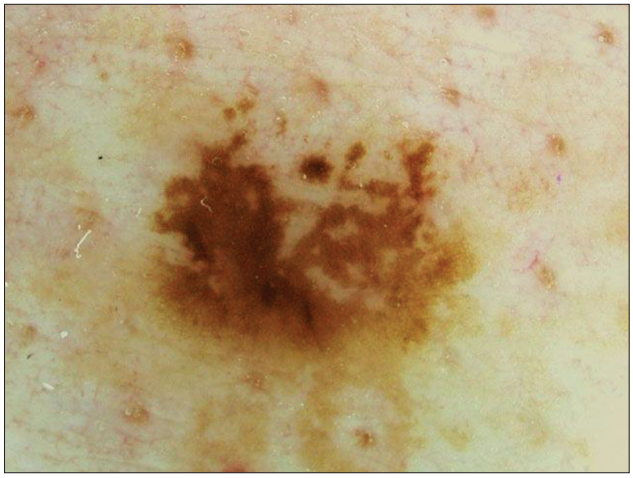

Figure 2: On dermoscopy dark network across the whole lesion.

It may be localized, segmental, or systematized [5]. Wood's lamp examination shows an off-white accentuation in ND (compared to chalky-white in vitiligo). Histopathologically, the numbers of melanocytes are normal or decreased [2,6] but DOPA reactivity is consistently reduced [2]. Melanosomes are usually normal in size, shape, and internal structure [1], but can be decreased in number, heteromorphic, aggregated in melanocytes, or located in membrane bound aggregates [5].

Until now 8 cases of colocalized nevus depigmentosus and lentigines have been reported [2-4,6,7,10]. The theory of twin spots does not apply to colocalized nevus. Two hypotheses were suggested for the colocalized nevus [8]. The first one, which applies to larger lesions such as syndrome of Ito, hypothesizes that a mutational event occurring in the first 4 weeks of life when the embryo is a single developmental field, leads to a polytopic malformation, namely more associated malformations. In our case, which is characterized by small nevi, the mutation affecting the same melanocytic function, is the more probable hypothesis [8] which is responsible for nevus depigmentosus. This mutation is followed by a reverse mutation of the gene involved in the pigmentation. The latter could restore the pigmentation incompletely. Thus, colocalization of lentigines can be regarded as a different form of repigmentation resulting from reverse mutation in one of the genes involved in pigmentation [2].

Consent

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

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

Our Dermatology Online

Case Report

Circumscribed palmar hypokeratosis: Report of 2 new Ecuadorian cases, dermatoscopic description and brief literature review

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ABSTRACT

The circumscribed palmoplantar hypokeratosis (CPH) is a dermatosis that is unfrequently reported around the world. It predominantly affects middle to advanced aged women, involving acral skin, asymptomatic lesions, and of unknown cause. However, its´ clinical, histopathologic and dermoscopic characteristics are distinctive. We report two cases of female patients, who come for a medical consultation presenting a long-term lesion of several years, located on the palm, asymptomatic and with no apparent cause.

Key words: Circumscribed hypokeratosis; Dermatoscopy; Cases report

INTRODUCTION

Circumscribed palmar or plantar Hypokeratosis was first described by Perez et al., in June of the 2002 [1], it refers to a rare dermatosis, characterized by an erythematous lesion, which is depressed, usually solitary, of a long-term history, asymptomatic, involving the hypothenar or thenar area of the palm and rarely, the sole [2,3] or dorsal area of the hand [4]. It mostly affects middle or advanced aged women, and its pathogenesis is not clear. Its´ histopathological characteristics consist of a well-defined depression of the epidermis. The epidermis that covers the depression shows a markedly thin stratum corneum and a thin granular layer [5], although thicker than the latter. More than 60 cases from Germany, Spain, Austria, Chile [1], Peru [5], United States, France [3], Italy, Korea, Japan, Malta, New Zeland [6] and Ecuador [7] have been reported so far.

CASE REPORT

Case 1

A 79-year old female, with no significant medical personal history, came for a dermatologist consultation, presenting an erythematous plaque, with superficial desquamation compatible with Actinic Keratosis localized on her right temporal region. Although on her physical examination, an oval lesion was noted, with erythematous base, loss of epidermis and sharp-cut borders, with a 2cm-diameter approximately, located on the external and palmar region of her left thumb. After inquiring the patient about this lesion, she referred a 10-year history, asymptomatic, with no previous trauma that she could recall of (Fig. 1).

A dermatoscopy study of the lesion was realized, noticing an erythematous base with irregular borders (Fig. 2). Amplifying the dermoscopic image, showed a stair-like edge and erythematous center, speckled with white macules and red dots (Fig. 3). A histopathological study was requested due to clinical suspicion of circumscribed palmar hypokeratosis. It reported acral skin with thick corneal layer which is characteristic of the anatomical area, presence of an area with abrupt well-defined loss of the corneal layer and the underlying dermis with slight superficial perivascular infiltrate, compatible with the diagnosis of circumscribed palmar hypokeratosis (Fig. 4).

How to cite this article: Uraga E, Morán A, Garcés JC. Circumscribed palmar hypokeratosis: Report of 2 new Ecuadorian cases, dermatoscopic description and brief literature review. Our Dermatol Online. 2016;7(3):329-333.

Submission: 03.02.2016; **Acceptance:** 10.05.2016

DOI: 10.7241/ourd.20163.89

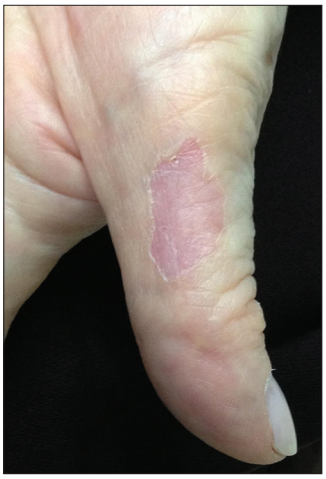

Figure 1: A depressed erythematous lesion with sharp cut borders can be observed.

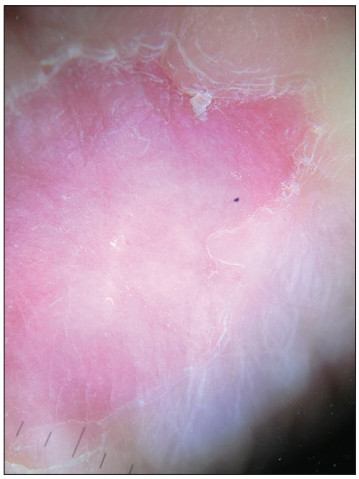

Figure 2: Dermoscopic image that shows erosion with erythematous base and irregular borders.

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

Case 2

A 53-year old female, without any relevant medical history, came for a dermatologist consultation for presenting a depressed lesion, with a slightly erythematous base, irregular well-defined border, of 1 cm diameter approximately, localized on left palm, asymptomatic, with a four year history. It had not presented any particular physical change during this time and the patient does not recall any local trauma (Fig. 4). The dermatoscopy image of the lesion, showed a depressed lesion with erythematous base and keratotic borders (Figs. 5-7). There was a clinical suspicion of circumscribed palmar hypokeratosis,

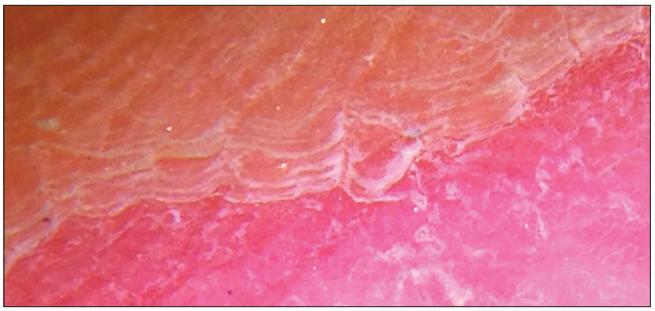

Figure 3: Stair-like desquamation on the border of the lesion. Erythematous base showing white macules and red dots.

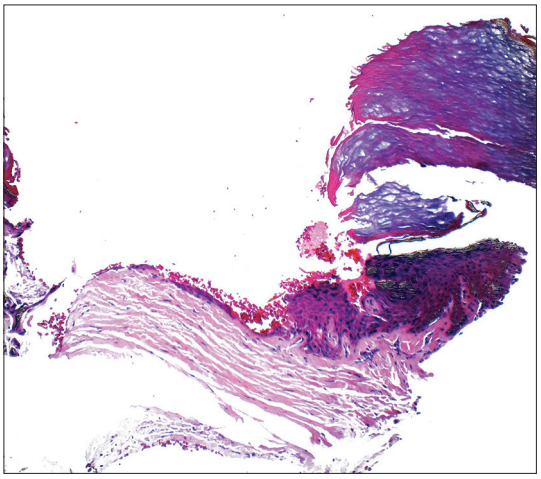

Figure 4: Histopathology that shows the abrupt well-defined loss of the corneal layer. The underlying dermis with slight superficial perivascular infiltrates.

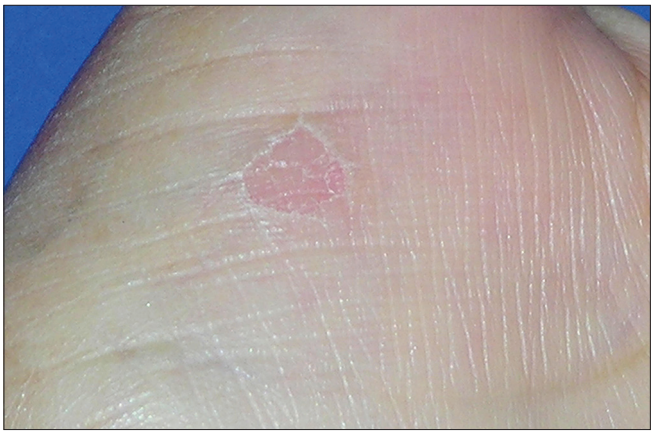

Figure 5: It can be observed a depressed lesion on left thenar area, with defined borders and erythematous base.

therefore, the histopathology was required; which confirmed the diagnosis.

DISCUSSION

Circumscribed palmoplantar hypokeratosis (CPH) was first described on 2002 by Perez et al., as an epidermic

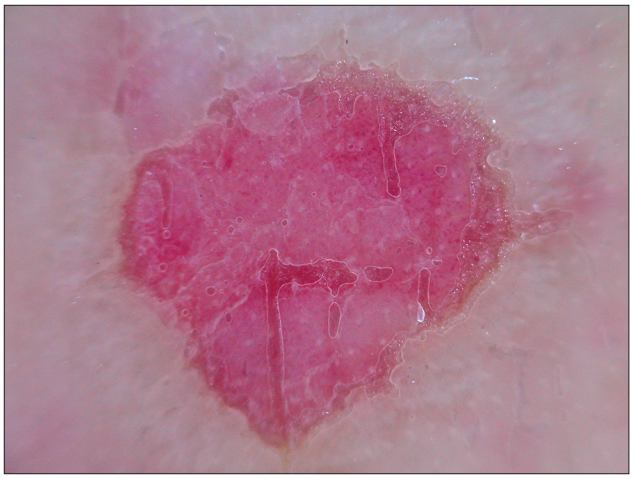

Figure 6: Dermastocopy shows image of depressed lesion with erythematous base.

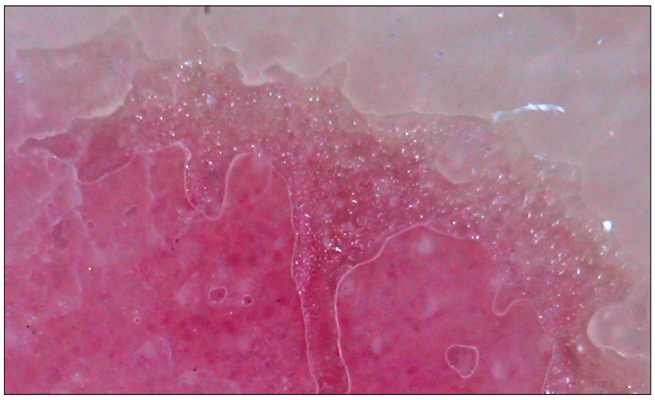

Figure 7: Dermatoscopy was amplified, showing well-defined keratotic borders and a moderate stair-like image, as well as, white macules and red dots spread on the erythematous base.

genetic disorder involving the acral regions, reporting a series of 10 patients who presented lesions with distinctive clinical and histopathological characteristics; 9 of them on the palm and 1 on the sole. Of these patients with palmar affectation, 6 of them had the lesion located on the thenar eminence, 2 of them on the hypothenar eminence and 1 of them on the palmar area of the left thumb [5]. It constitutes an independent well-characterized disorder, involving most frequently adult women, with a woman to man ratio of 4:1 [8], age may vary between 35 to 84 years old, with an average of 64.6 years old [9], and the lesions can last from 2 to 30 years [10], with an average of 12.8 years [9]. For this reason it has been categorized as an acquired disorder; although one congenital case on the United States was reported affecting an afroamerican 10-year old patient, who presented it since birth [11].

The pathophysiologic mechanism that develops CPH is not clear, Perez et al. proposed that it constitutes an epidermal benign clonal malformation, although, the late onset of the disease and the tendency to progression, suggest a proliferative dynamic disorder, without propensity to an oncogenic transformation [12]. CPH usually affects the dominant hand, which points towards a ¨forme fruste¨ of epidermolysis bullosa, with secondary lesions resulting from repetitive local trauma [13], however, just a small number of patients refer local trauma history or previous burns on the affected area [14-16].

On the other hand, Böer et al., mentioned the finding of human papilloma virus (HPV) type 4 in 2 reported cases, to which a polymerase chain reaction (PCR) was realized that suggest that CPH could be a rare morphological manifestation of common warts. Berk reported the presence of HPV type 6 in 1 of his 3 reported cases [17]. Nevertheless, the study for this virus has been done on other reported cases and the results has come up negative [10,16]. On 2007 Ishiko et al. realized immunohistochemical studies to the skin sample, from which it was found an overexpression of AE1/AE3 and diminished K9 and K2e. The expression of K16 and ki 67 did not show distinctive difference from normal nor perilesional skin, which suggests it could be associated with a keratinization disorder [10,12,18]. On 2009, Tanioka et al. proposed the existence of two subtypes of the disease. The most common one characterized by the erythematous palmar lesion, associated with diminished K6 and K16 and normal K9 immunohistochemically reported. The other subtype usually affecting the plantar region, without or hardly any erythema, associated with diminished k26 and connexin 9 expression [2]. Yamamoto et al. on 2011 advocated that it is a cell fragility disorder, due to a precipitated decomposition of desmosomes on the superior granular layer and of corneodesmosomes on the lowest corneal layer, which could be consequence of a high proteolytic activity associated with abundant Odland bodies present on this disease. These findings suggest the existence of two different pathogenic subtypes of CPH, the first one induced by an elevated corneocyte fragility and the second one caused by an abnormality on adhesion structures [14,19]. For Urbina and col., it is about a keratinization disorder expressed morphologically on the granular and corneal layers only. It consists on a reduction of keratin filaments and keratohyaline granules combined with an increased number of lipids on the corneal layer [20]. The transmission electron microscopy (TEM) does not identify abnormal mitosis or atypical cells, which supports the theory that subclinical local trauma

could generate a defect during the development of the keratin [21].

Other authors noted that the hypokeratotic epidermis found on CPH could be consequence of a clonal expansion of the keratinocytes which lose the ability to differentiate towards hyperkeratotic palmoplantar keratinocytes. The exact mechanism of this referred abnormal differentiation is unknown, even so, the existence of patients suffering of multiple CPH lesions suggest some genetic predisposition to this defect [21].

CPH is also known as circumscribed acral hypokeratosis [22] and it is a chronic dermatosis, of benign progression, characterized by a lesion that shows a slow evolution or it could even remain stable throughout many years. Only 1 case of a malignant transformation of these lesions has been reported, so still a close clinical follow up is recommended [21].

Clinically, CPH typically presents a solitary skin lesion, circular, erythematous, depressed, well-defined, of 1-4cm of diameter, asymptomatic, localized on the thenar or hypothenar eminence of the hand or in the center of the sole [22,23], nonetheless a few reports exist of patients presenting multiple lesions [24], or lesions localized on the dorsal region of the fingers of the hands [4,22] or in a no acral area, such as the case of a 63 year old male presenting a lesion on his anterior thorax [7].

The diagnosis of CPH is based on the clinical features, the anatomical location, and the histopathologic characteristics of the lesion [7]. The dermatoscopy currently depicts distinctive findings. The histology describes a depression of the epidermis with a stair-like border between affected and normal skin. The epidermis that covers the depressed area, shows a thinner granular and corneal layer, if compared to perilesional skin.

The corneal layer that covers the depression is ortokeratotic with a basket basophilic pattern [1], while the adjacent area to the hypokeratosis shows a frayed pattern [12].

Exists a morphologic change of the corneocytes on the edge of the hypokeratotic area. Specifically, the superficial corneocytes show a vacuolar appearance and are basophilic; on the other hand the deeper corneocytes appear hypereosinophilia. A slight lymphocytic infiltrate surrounding the capillaries on the superficial dermis was present [12]. Groysman et al reported la existence of granular parakeratosis, local subepidermal fibin deposition and ulcers which can be related with a post traumatic skin lesion [7].

The differential diagnosis most commonly considered are Bowen´s disease and Porokeratosis of Mibelli, however the absence of atopy and cornoid lamillation void these diagnosis [24].

On dermatoscopy and microscopy with oil immersion, Nishimura et al. described the presence of ¨stair-like¨ desquamation on the borders of the lesion and well-defined erythema with scattered white macules and small red dots. The findings on the edge of the lesion correspond histologically to the thinning of the corneal layer, the erythema and red dots are consequence of the dilated capillaries on the papillary and reticular layers of the dermis, and the white macules could represent the sweat gland ducts which become accentuated because of the thinner corneal layer [25-27].

Currently there is no established treatment for this condition. Many local topical therapies have been tested; neither single nor combined therapies have been successful, getting no improvement or only partial remission. The use of corticosteroids, hydrocolloids or topical retinoid appears ineffective [26]. Urbina et al. reported healing almost completely a lesion using calcipotriol, after a 4-5 year continued treatment [20]. Dynamic Phototherapy resulted on a partial improvement after two cicles of treatment [28]. The superficial excision of the lesion is rarely done, but offers a complete cure after a 6-month follow up. Exists one reported case of a lesion being cured with the use of cryotherapy [26]. Wilk et al. informed a partial improvement on a patient being treated with topical 5-fluoruoracil for 18 months, but, the lesion reappears 3 months after discontinuing the treatment. Finally, a case of a patient with bilateral lesions was spontaneously healed [29]. On our two cases reported, we initiated treatment with topical calcipotriol, getting no improvement for three months, so both patients decided to discontinue the treatment.

CONCLUSION

To conclude, we agree with all the other authors with the fact that it is an under-diagnosed dermatosis [30]. Due to the fact that the lesion is asymptomatic, it is usually a random finding during a physical medical

examination, so it is required to be attentive during the examinations as more and more cases are being reported. The distinctive dermatoscopy referred, such as the erythematous base, ¨stair-like¨ edge, white macules and red dots, become strong decisive findings for the diagnosis of this disorder. On our country, Ecuador, our cases combined with Pinos et al [7]. add up to 5 cases reported of this unusual disease.

Consent

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

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

Our Dermatology Online

Case Report

Rare splenic calcifications in a child with cutaneous systemic lupus and a review of the literature

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ABSTRACT

Lupus erythematosus is a complex autoimmune multisystemic disease that can affect the skin and other organs including the spleen where Various anatomical changes can occur but the recognition of spleen calcifications in lupus patients is really a rare event which may be a marker of poor prognosis and renal involvement of lupus, our observation is the 13th described in the literature, and the to our knowledge the first one that concerns a child which is even more unusual.

Keys words: Lupus; Spleen calcifications; Renal involvement; Child

INTRODUCTION

Lupus erythematosus is a complex autoimmune multisystemic disease that can affect the skin and other organs including the spleen [1]. Various changes can occur in the spleen affected by the inflammatory process such as splenomegaly, hyposplenism, infarction, spontaneous rupture, functional asplenia, and periarterial thickening in an "onion skin" pattern [2].

The recognition of calcifications in systemic lupus have been unusually reported concerning other organs like the brain [3], the skin [4], the breast [5,6], the coronary system [7] and submandibular gland [8], in periarticular [9] and in Soft tissues, but their existence in the spleen of patients with systemic lupus is really a rare event since our observation is the 13th described in the literature, and the occurrence of this feature in a child as the case of our patient is even more unusual.

CASE REPORT

A 14-years-old child admitted in our department for scaly erythematous lesions in the face and the hands with acrocyanosis and photosensitivity that started 3 months before; the dermatological examination revealed a maculopapular erythematous patches of the nose and the malar area (butterfly mask),in the palmar surface and interarticular surfaces of the dorsum of the hands and feet with buccal mucosal ulcerations. Cutaneous lupus was confirmed histologically (Figs. 1 and 2).

The results of paraclinical examinations of blood and urine had revealed a systemic lupus with hematological (anemia, thrombocytopenia and leucopenia), immunological (positive antinuclear antibodies with diffuse pattern anti-dsDNA) and renal involvement (hematuria, proteinuria) confirmed histologically as focal lupus nephritis Class III.

The rest of laboratory investigations showed a high erythrocyte sedimentation rate and a hypocomplementemia CH50, C3, C4.

On a chest radiography taking the diaphragmatic domes, we had noticed calcifications in the splenic area with some calcifications in the chest (Figs. 3 and 4). The abdominal ultrasound revealed extensive calcifications over the entire height of the spleen.

How to cite this article: Kelati A, Gallouj S, Mernissi FZ. Rare splenic calcifications in a child with cutaneous systemic lupus and a review of the literature. Our Dermatol Online. 2016;7(3):334-337.

Submission: 29.01.2016; **Acceptance:** 07.03.2016

DOI:10.7241/ourd.20163.90

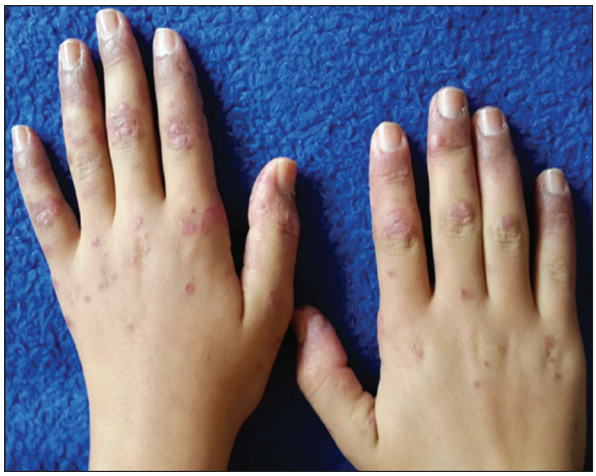

Figure 1: Clinical image showing patches of lupus on the dorsum of the hands.

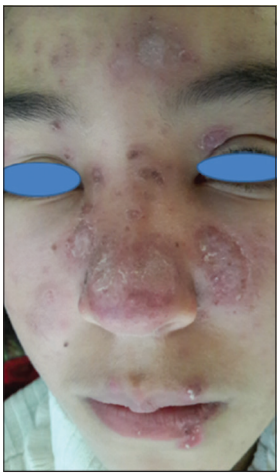

Figure 2: Clinical image showing erythematous patches of the nose and the malar area (butterfly mask).

During all the investigations, the serum calcium, phosphorus, and alkaline phosphatase levels were normal.

The patient was treated initially with bolus corticosteroids during 3 days for the renal lupus and then oral corticosteroids, in association with hydroxychloroquine 200 mg/d, and we transferred the child to the department of pediatrics for appropriate management of his systemic lupus.

DISCUSSION

Splenic calcifications have been reported in connective tissue disorders such as rheumatoid arthritis, systemic sclerosis, infections, sickle cell disease, splenic haemangiomas, and cysts, and in B-cell lymphoma [10].

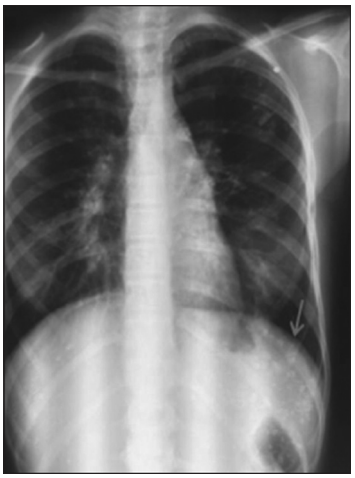

Figure 3: Radiologic image showing spleen calcification.

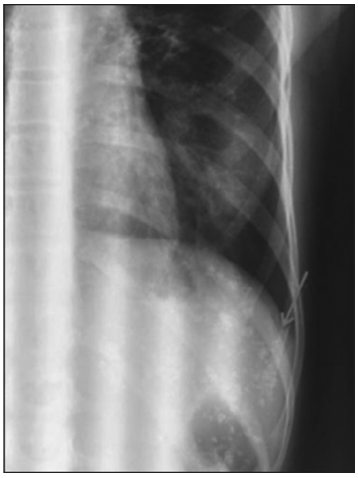

Figure 4: Radiologic image showing spleen calcification (close view).

Yet, these calcifications have barely been described among the anatomical and physiological changes in the spleen of patients having systemic lupus erythematous (SLE).

On the other hand, calcifications have been recognized in other organs in lupus patients, such as the brain[3], the skin [4], the breast [5,6], the coronary system [7] and submandibular gland [8], in periarticular [9] and in Soft tissues in general. The largest study carried out concerning the prevalence of soft tissue calcifications in patients with SLE had been done 15 years ago and had found that in 6 patients, the prevalence of ectopic calcification in SLE was 40% and that the incidence of lupus nephritis and nephrotic syndrome were significantly higher in the calcification positive group than the negative one. This renal involvement was also noticed in a woman having systemic lupus

with spleen calcifications [10] which is the same of our case, so, based on these results, we may deduce that these spleen calcifications could be a marker of poor prognosis and renal involvement especially in children where the lupus is known more dangerous [11].

Another important findings were reported in a patient having lupus erythematosus with diffuse soft tissue and vascular calcification in addition to a chronic kidney disease [12]. In this publication, the authors have explained these calcifications by the systemic disorder of mineral and bone metabolism which is related to chronic kidney disease.

As a result, if we combine all these facts, that the spleen calcifications are significantly related to the renal involvement of lupus and that a kidney disease could cause systemic disorder of mineral and bone metabolism, thus, it may cause calcium deposit in soft tissues like the spleen, but this theory does not explain why our patient whose renal disease was acute -instead of chronic- have developed these calcifications.

Moreover, three mechanisms have been proposed to explain the formation of calcifications as a whole in lupus patients, the first one was suggested by Neuman et al, that these calcifications could be enhanced by high alkaline phosphatase concentration since this alkaline phosphatase can remove the organic phosphate which inhibit the calcium deposit, another theory and the most logical one, has been demonstrated in necrotic tissue by Moss and Urist who proposed that the calcifications may be precipitated by chronic inflammation and tissue necrosis, injuries and ulcers, this explanation could explain calcifications in a spleen affected by the inflammatory process. Another Hypothesis for soft tissue calcifications is a pressure phenomenon producing ischaemia which was reported by Powell et al in 1974. While the exact significance of diffuse splenic calcification is still unknown, this unique radiologic finding may be a result of the disease process itself [13,14].

Furthermore, after a review of the literature from 1947 to 2015, 12 observations of these spleen calcifications were reported [1,13,14]. With a pathologic confirmation in an autopsy of the spleen in one patient [15], our observation is the 13th. The age of these patients ranged from 24 to 73 years old in contrary with the age of our patient (14 years old), this fact increase the originality of our case because as we already noticed, it may have a prognosis signification. Therefore, it may be important to choose invasive therapeutic options from the beginning in patients having these calcifications especially in children whose lupus is usually more severe [11].

CONCLUSION

The originality of our case is the occurrence of spleen calcifications in a child having cutaneous and systemic lupus with renal involvement which may confirm the theory that these calcifications have a prognosis signification.

Consent

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

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

Our Dermatology Online

Case Report

Terra firma-forme dermatosis – a dirty dermatosis: report of two cases

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ABSTRACT

Terra firma-forme dermatosis is characterized by 'dirty' brown-grey cutaneous patches and plaques that can simply be eradicated by forceful swabbing with alcohol pads. The pathogenesis has been attributed to abnormal and delayed keratinization. Although affected patients present with typical lesions, the disorder is not well-known by physicians. From a clinical point of view, we lay emphasis on its unique expression and diagnosis/treatment. From a histological perspective, we highlight its resemblance to dermatosis neglecta and speculate on the role of 'neglect' in a patient with seemingly adequate hygiene. The role of urea containing emollients in the development of this disorder remains to be determined.

Key words: Dermatitis neglecta; Dermatosis neglecta; Terra firma-forme dermatosis

INTRODUCTION

Terra firma-forme dermatosis (TFFD) is a bizarre, acquired and idiopathic dermatosis [1]. The disorder was initially reported as a distinct entity by Duncan *et al*., in 1987. The Latin phrase *terra-firma* mean'dry land' and denotes a cutaneous discoloration resembling 'dirt' (hence the alternative name Duncan's Dirty Dermatosis) [2].

CASE REPORTS

Case 1

A 26-year-old male presented with a 15 days history of persistent scaly pigmented patches on the nape of neck. Patient gives history of not taking bath for a period of 7 days.

Cutaneous examination revealed brown-grey patches on nape of neck (Fig. 1).

There were islands of normal skin within the patches. Initial tentative clinical diagnoses were terra firma-forme dermatosis, pomade crust, confluent and reticulated papillomatosis and acanthosis nigricans. A 'wipe test' with 70% ethyl alcohol accomplished clearance of lesions (Fig. 2).

Case 2

A 70-year-old male presented with a 25 to 30 days history of persistent scaly pigmented patches on the left leg. Patient gives history of admission in hospital for a systemic condition since 10 days.

Cutaneous examination revealed brown-grey patches on anterior surface of left leg (Fig. 3). There were islands of normal skin within the patches. A 'wipe test' with 70% ethyl alcohol accomplished clearance of lesions (Figs. 4 and 5).

DISCUSSION

TFFD is a recently described cutaneous pigmentation disorder. It is probably more prevalent than cited in the Literature. The disorder affects all age groups (age range: 3 months to 72 years) and both genders [1,3]. Clinically, TFFD is characterized by brown-grey, velvety, pigmented patches or plaques [2]. Involvement

How to cite this article: Lunge S, Supraja C. Terra firma-forme dermatosis – a dirty dermatosis: report of two cases. Our Dermatol Online. 2016;7(3):338-340.
Submission: 02.11.2015; **Acceptance:** 28.01.2016
DOI: 10.7241/ourd.20163.91

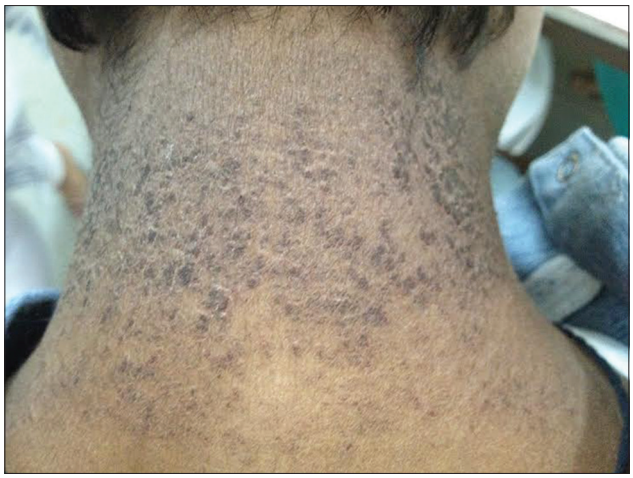

Figure 1: Multiple scaly pigmented patches on the nape of neck.

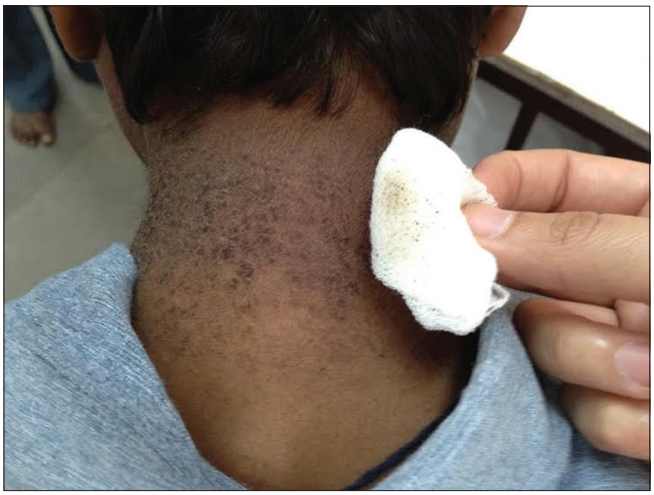

Figure 2: Wipe test.

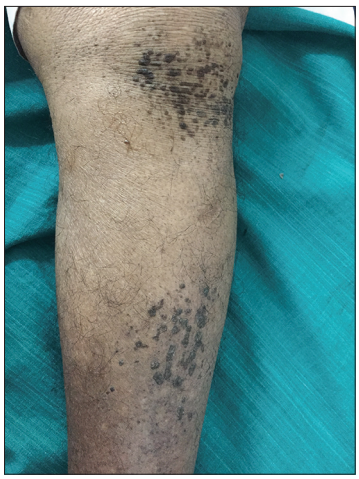

Figure 3: Multiple scaly pigmented patches.

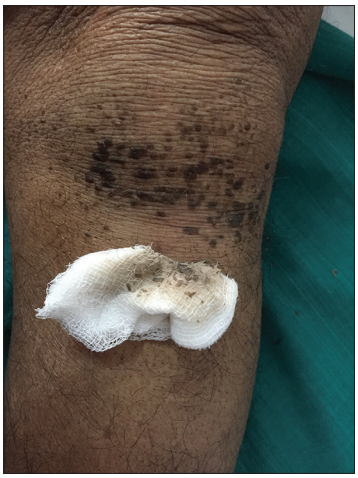

Figure 4: Wipe test.

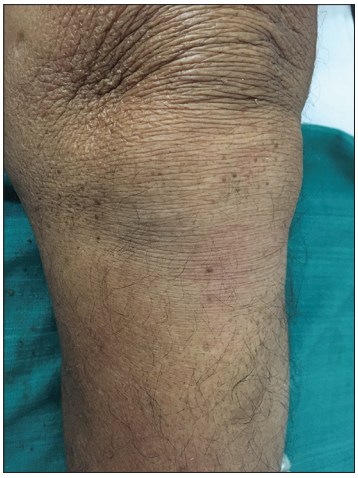

Figure 5: Clearance of the lesions.

of the face, neck, trunk or ankles is usual, although unusual sites such as scalp, lips, chest, axilla, back, umbilical area, pubis, arms and legs have been reported [3-5]. The distribution may be localized, generalized or symmetrical. Rarely palpable verrucous or papillomatous plaques, reticulated patches and focal slight scaling have been observed [4].

Symptomatic cases are rare; however, TFFD is a cause of cosmetic concern [1].

The histopathological description encompasses prominent lamellar hyperkeratosis with focal areas of compact whorled orthokeratosis [1], papillomatosis, mild acanthosis and deposition of keratotic material within the valleys between the papilla [5]. There is no parakeratosis. Dermal inflammation is trivial [2,5]. Other findings with special stains include increased melanin content in the compact hyperkeratotic and basal areas of the epidermis (Fontana-Masson) [1], scattered keratin globules throughout stratum corneum (Toluidine blue) [1,5] and occasional yeast cells indicative of Pityrosporum (Periodic Acid Schiff) [5,6].

The diagnosis of TFFD is confirmed by forceful rubbing with a gauze pad immersed into 70% isopropyl alcohol [1,2] or ethyl alcohol. This diagnostic test prevents unnecessary laboratory work-up or biopsy [3]. In addition, it offers a magical and prompt therapeutic cure for the disorder [1,2]. A pink normal skin underneath is exposed after the wiping procedure [5]. Reappearance isunusual once the skin is devoid of 'dirty' appearance. Prophylactic weekly application of alcohol has been recommended for resistant or recurrent cases [1]. Patients should be reassured about the benign nature of TFFD and educated about the cleaning procedure.

Differential diagnosis includes acanthosis nigricans, confluent and reticulated papillomatosis, pityriasis versicolor, epidermal nevi, dirty neck syndrome of atopic dermatitis and dermatosis neglecta (DN) [1,3,5].

Most of these disorders can be excluded by alcohol swab test [1,3]. DN results from 'neglect', i.e. poor hygiene or inadequate skin cleansing [6]. TFFD is distinguished from DN arbitrarily by presence of adequate hygiene, absence of cornflake-like brownish scales and successful eradication of pigmentation with isopropyl alcohol in the former and effective clearance of lesions with soap and water in the latter [1,2,4-6].

However, isopropyl alcohol is operative in both disorders [1,5].

Histologically, DN is similar to TFFD, except for the absence of whorled hyperkeratosis in DN [1,5]. Nevertheless, the distinction between TFFD and DN is blurred and there seems to be a considerable clinical and histological overlap between the two disorders.

CONCLUSION

Dermatitis neglecta is an underreported, asymptomatic, but aesthetically bothersome dermatosis. Physicians need to be aware of this condition that can be clinically diagnosed and effectively and inexpensively treated.

Consent

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

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

Our Dermatology Online

Clinical Image

Giant acrochordons arising from genitals in a postmenopausal non-obese woman

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A 65 year old post menopausal female presented to our dermatological out patient department with a chief complaint of mass hanging from the genital area for the last two and a half years. Initially the swelling was very small which has progressively increased to form a bigger mass to produce a feeling of discomfort due to the weight of the lesion during walking. The apprehension of increasing size was one of the concerns for visiting hospital. There was no history of pain, itching, fever, ulceration, bleeding or any discharge from the lesion. There was no history of any sudden increase, postural or diurnal change in the size of swelling. Patient didn't report any aggravating/relieving factors in relation to the size of the swelling.

On examination, she was non obese with a BMI of 20.77. Cutaneous examination revealed two major, skin coloured, pedunculated, soft fleshy masses measuring 15 × 5 cm and 8 × 5 cm with a thick stalk arising from the anterior surface of left labia majus and vulvar region (Fig. 1). The surface of the swellings was lobulated. There were also 3-4 smaller swellings arising from the upper aspect of one of the major swellings around the size of 2.5 × 1.5 cm. Skin overlying the swellings was normal with no redness, discharge or ulceration. There were no visible pulsations. The swelling was nonreducible with no palpable thrill. Cough impulse was absent. No bruit was audible on auscultation. There was no regional lymphadenopathy. Hernial orifices were intact. Systemic and genitourinary examination was unremarkable. Her Blood sugar and lipid profile were normal. The masses were diagnosed as acrochordons based on history and suggestive clinical findings. The lesions were surgically excised to confirm the diagnosis and to rule out any malignant change.

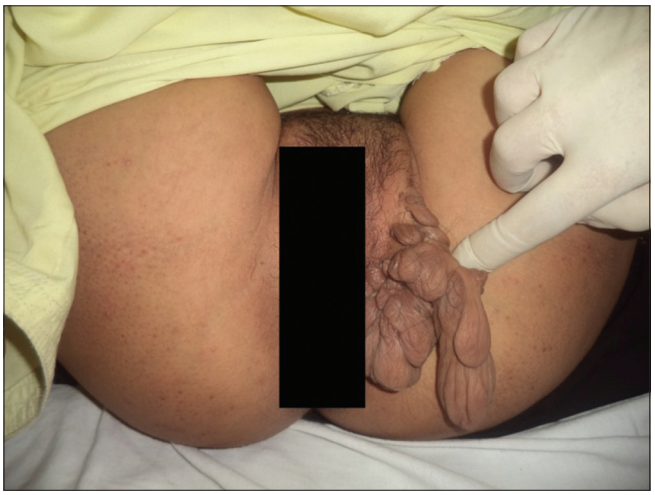

Figure 1: Giant acrochordons in a post menopausal non-obese woman.

The swellings in the present case were giant acrochordons. An Acrochordon (fibroepithelial polyp) is a common benign tumor mainly seen in obese individuals. In the general population, its incidence has been around 46% [1]. Most acrochordons vary in size from 2 to 5 mm in diameter [2]. The unusually large sized acrochordons have been rarely reported. In women, peak incidence is seen in 20-40 years of age. Regarding genital acrochordons in women, these are more commonly seen in vagina than vulva and cervix [2]. Postmenopausal cases are rarely seen. Various conditions have been associated with acrochordons which include obesity, dyslipidemias, pregnancy, insulin resistance, type 2 diabetes mellitus, genetic

How to cite this article: Arif T, Amin SS. Giant acrochordons arising from genitals in a postmenopausal non-obese woman. Our Dermatol Online. 2016;7(3):341-342.
Submission: 25.12.2015; **Acceptance:** 23.04.2016
DOI: 10.7241/ourd.20163.92

predisposition, human papilloma virus 6 and 11 and acromegaly [3].

The presence of unusually large acrochordons, in a postmenopausal non-obese woman arising from vulvar/labial site with no associated clinical conditions makes this case a rare one and hence reported.

Consent

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

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

Clinical Image

Dermatosis de Los pliegues periungueales [Dermatoses of the periungual nail folds]

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Paciente masculino de 27 años de edad que consulta por presentar "bolita que le sangra en ocasiones" en dedo medio derecho desde hace 6 semanas pero que se ha hecho más frecuente en la última semana por lo que consulta.

Al examen físico se encontró una dermatosis localizada al pliegue lateral del dedo medio derecho constituida por neoformación rojiza con costra sanguínea y pequeña ulceración central de más o menos 0.5 cm, la dermatoscopía acentúa estos mismos aspectos clínicos (Fig. 1). Resto del examen físico dentro de límites normales.

Inicia su padecimiento hace más o menos 6 semanas después de haberse arrancado un "pellejito" en ese dedo y poco a poco le empezó a crecer una bolita roja que le sangra y es molesto para el motivo por el cual consulta.

Antecedentes familiares y personales sin importancia.

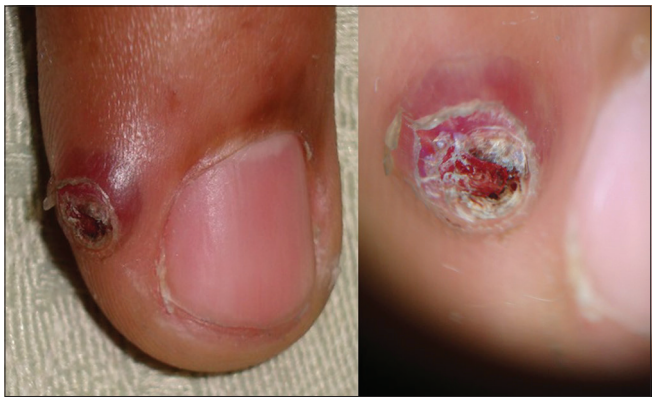

Figure 1: Aspecto clínico y dermatoscópico de la neoformación vascular ulcerada en pliegue lateral dedo medio derecho de un granuloma piógeno.

Con estos datos clínicos se hace el diagnóstico de granuloma piógeno del pliegue lateral externo del dedo medio derecho al cual se le realiza electrocirugía resolviendo su afección cutánea.

Los pliegues ungueales tanto el proximal como los laterales son parte importante del aparato ungueal, dando soporte y protección a la placa ungueal [1].

El proximal es una continuación de la parte dorsal de la piel de los dígitos [1]. La cutícula es el producto corneo final de este pliegue actuando como un sello protector para prevenir infecciones [2,3]. Los pliegues laterales cubren y protegen la mayor parte de la matriz y la superficie de la uña [1].

Diversas enfermedades como las dermatológicas, sistémicas, infecciosas, tumorales, reaccionales, reacciones medicamentosas, traumáticas, ocupacionales entre otras pueden afectar estos pliegues [3].

El trastorno más frecuente de los pliegues periungueales es la paroniquia [1,3] que es la inflamación del tejido alrededor de la lámina ungueal. Las vías de acceso de infección pueden ser traumatismos o mecanismos que interrumpan la continuidad de la piel. Se puede clasificar como aguda o crónica, la causa más común de paroniquia aguda es el traumatismo directo o indirecto a la cutícula o a los pliegues laterales. (Fig. 2) [2,4].

En todas las enfermedades del colágeno, el pliegue proximal es el sitio más afectado, siendo el eritema y las telangiectasias hallazgos frecuentes; las hemorragias y la necrosis focal también son observados [5].

How to cite this article: Chang P, Montejo Camposeco EY. Dermatosis de Los pliegues periungueales [Dermatoses of the periungual nail folds]. Our Dermatol Online. 2016;7(3):343-346.

Submission: 10.01.2016; **Acceptance:** 15.03.2016

DOI: 10.7241/ourd.20163.93

Los tumores de Koenen y el quiste mucoide son los tumores más frecuentes a este nivel [2].

Otras alteraciones del pliegue proximal son los hematomas por el uso del oxímetro (Fig. 3), la onicofagia, dermatitis reaccionales (Figs. 4 and 5), reacciones medicamentosas como el eritema fijo pigmentado (Figs. 6 and 7), síndrome de Steven Johnson, Lyell y VIH,vasculitis (Fig. 8), peri onicofagia, padrastros [3,6] (Fig. 9).

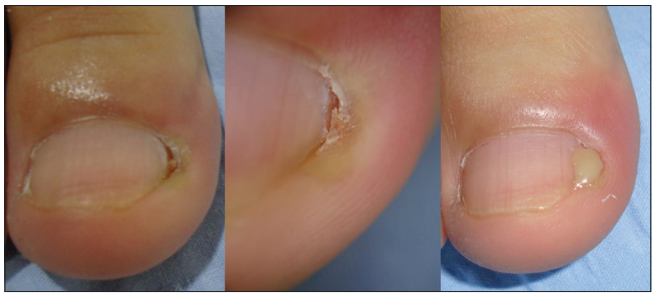

Figure 2: Paroniquia aguda.

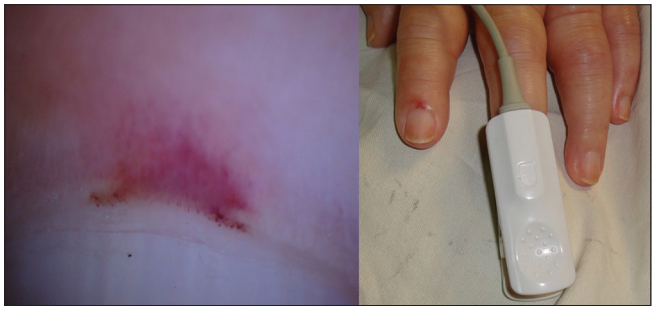

Figure 3: Hematomas del pliegue proximal por uso del oxímetro.

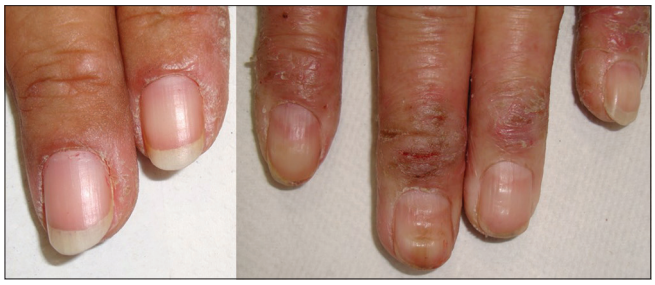

Figure 4: Lesiones eccematosas de los pliegues periungueales.

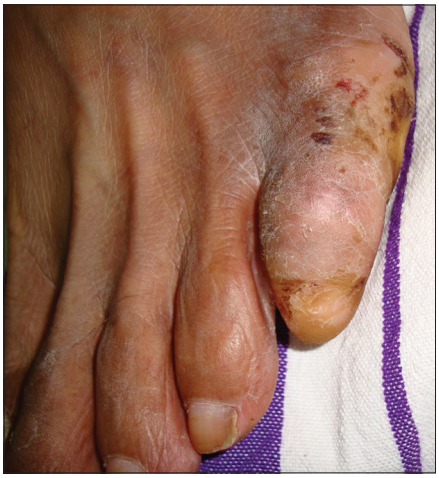

Figure 5: Liquen simple crónico 5to ortejo izquierdo.

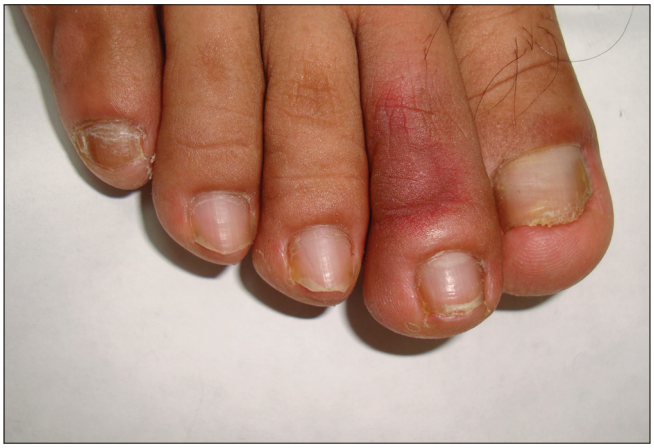

Figure 6: Erupción fija por alopurinol

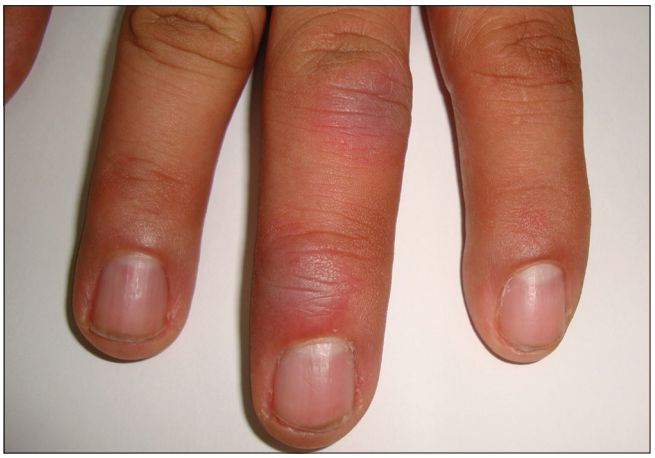

Figure 7: Erupción fija por sulfas.

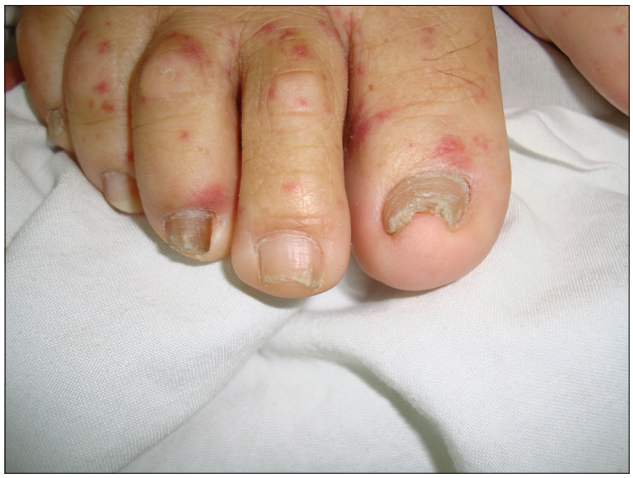

Figure 8: Vasculitis por azitromicina.

Las alteraciones más frecuentes de los pliegues laterales son la onicocriptosis y la hipertrofia congénita de los mismos [1].

En la onicocriptosis la superficie de la uña se incrusta en diversos grados en los tejidos periungueales [7]. Se clasifica en 3 estadios clínicos: Grado I: edema y dolor. Grado: II, infección con secreción purulenta. Grado III: signos y síntomas intensificados de los grados anteriores, con hipertrofia de pliegues laterales y tejido de granulación [5] (Fig. 10).

La hipertrofia congénita de los pliegues laterales de las uñas del primer ortejo describe una anomalía caracterizada por hipertrofia de los pliegues, que cubre parcialmente la superficie de la uña. (Fig. 11) [8].

Las verrugas periungueales son lesiones benignas localizadas alrededor de las uñas (Fig. 12). Algunas lesiones malignas como el carcinoma epidermoide, también pueden afectar estos pliegues, en este caso es de crecimiento lento, tiene bajo grado de malignidad y raramente metastatiza [2].

Los pliegues laterales también pueden ser afectados además de las causas antes descritas por traumatismos, xerosis, infecciones por el virus del herpes simple, vasculopatías asociadas a enfermedades como la diabetes mellitus (Fig. 13), lupus eritematoso sistémico (Fig. 14), artritis reumatoidea, dermatomiositis (Fig. 15), esclerosis sistémica (Fig. 16) entre otras [1,9,10].

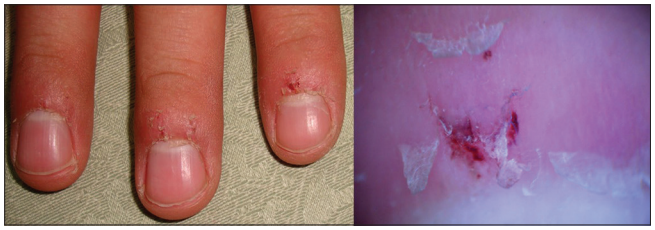

Figure 9: Padrastros periungueales.

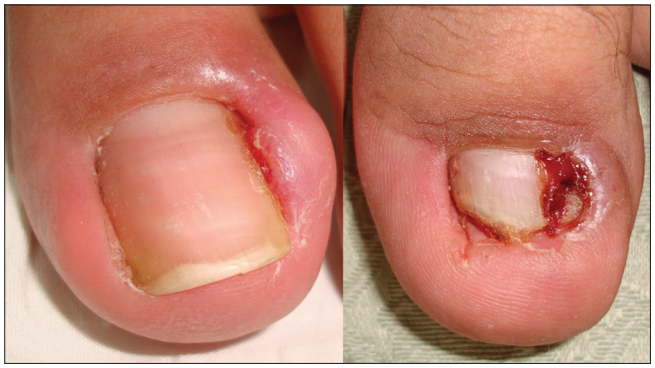

Figure 10: Onicocriptosis de diferentes grados.

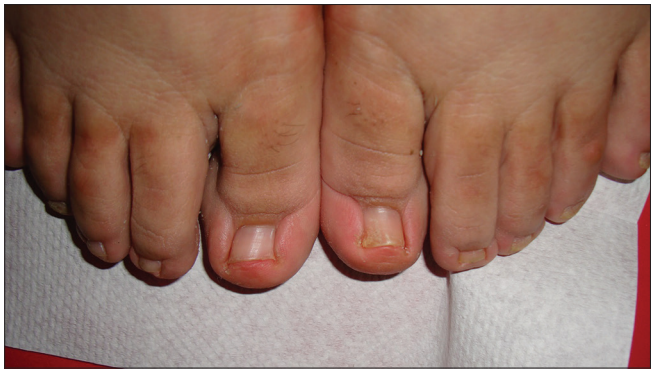

Figure 11: Hipertrofia de los pliegues laterales.

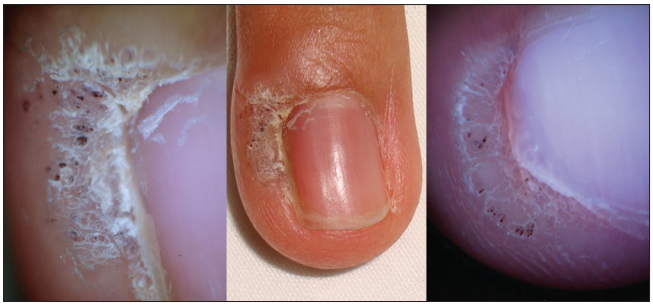

Figure 12: Verrugas periungueales.

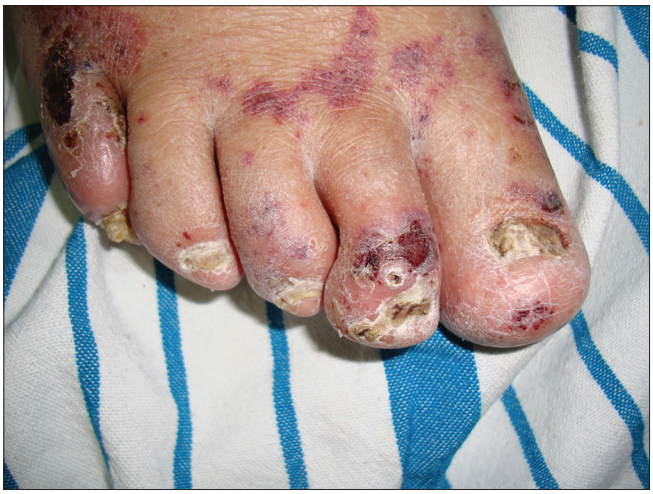

Figure 13: Alteraciones periungueales por diabetes mellitus.

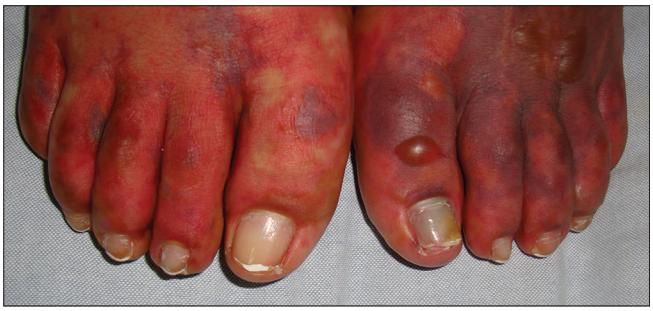

Figure 14: Manifestaciones subcutáneas por LES.

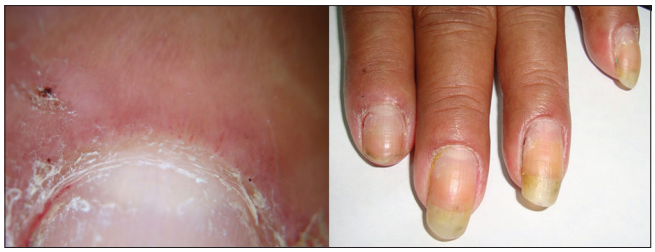

Figure 15: Telangiectasias del pliegue proximal por dermatomiositis.

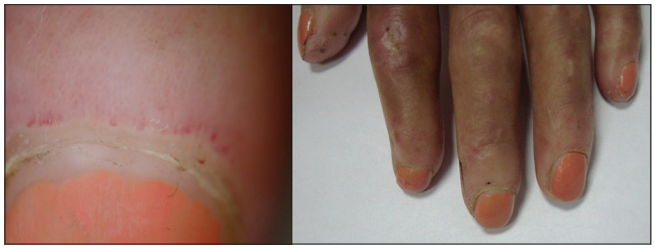

Figure 16: Telangiectasias del pliegue proximal por esclerosis sistémica.

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

Our Dermatology Online

Letter to the Editor

Benign nevus with nerve sheath differentiation

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Sir,

We report a 74 year-old woman who presented with lump on the right elbow. The decision was to remove the lump. The patient underwent a minor operation and the specimen was sent to the Histopathology department of Salah Azaiez institute. Grossly, an ellipse of skin was measuring 30 x 15mm with a central nodule measuring 10 x 8mm. It was transacted and 2 representative sections were put into blocks. Histological examination showed a well circumscribed dermal lesion made partly of nests of melanocytes (at the periphery of the lesion)and partly of spindle shaped cells with elongated and wavy nuclei (in the middle of the lesion), suggestive of a nerve sheath tumour (Fig. 1). There was no atypia and no mitoses (Fig. 2). The Immunohistochemical study showed that both the naevus cells and the spindled cells in the nerve sheat tumour-like area were positive for melaninA, S100 protein, EMA (Epithelial Membrane Antigen)and CD34. The desmin and SMA (smooth muscle actine) were negative. These features confirmed the diagnosis of benign nevus with nerve sheath differentiation. At 5 years of follow- up the patient was asymptomatic and there was no recurrence.

The patient's informed consent was obtained.

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

Neural differentiation by melanocytic nevi represents a well-recognized phenomenon, and melanocytic nevi with perineural differentiation have been reported [1]. Melanocytic nevi with neural differentiation are not rare. They retain some features of schwann cells and usually express S100 protein. Peripheral nerve sheath tumour and melanocytes are closely-related cells originating from the neural crest. It has been well-known that both benign and malignant melanocytic proliferation can show various type of neural differentiation [2,3].

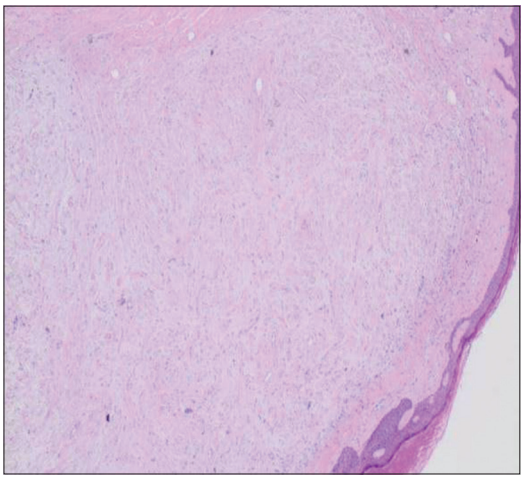

Figure 1: A well circumscribed dermal lesion made partly of nests of melanocytes and partly of spindle shaped cells (HEx4).

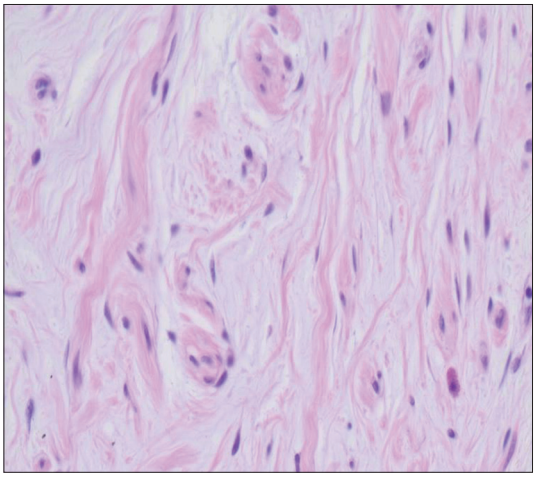

Figure 2: Spindle shaped cells with elongated and wavy nuclei without atypia and mitosis (HEx40).

How to cite this article: El Amine Elhadj O, Leila B, Goucha A, Gamoudi A, Elmay A. Benign nevus with nerve sheath differentiation. Our Dermatol Online. 2016;7(3):347-348.

Submission: 10.01.2016; **Acceptance:** 16.02.2016

DOI:10.7241/ourd.20153.94

Features of peripheral nerve sheath differentiation such as neuroid cords, nerve corpuscles, fascicle-like structures, and, exceptionally, palisading has been reported in melanocytic nevi [4]. Benign tumors of peripheral nerve sheath include mainly three subtypes: schwannoma, neurofibroma and perineurioma [5].

The main differential diagnoses for our case are benign naevus with nerve sheath differentiation or collision lesion tumour of naevus and nerve sheath tumour. The intimate mingling and merging of the naevus cells and the spindle-shaped cells suggest that this is a single lesion consisting of a benign intradermal naevus with nerve sheath differentiation. The immunohistochemistry results shows a mixture of S-100, EMA and CD34 positive cells.

Consent

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

Written informed consent was obtained from the patient for publication of this article.

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

Letter to the Editor

Blaschkoid lichen planus: A rare presentation of a common disease

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Sir,

Lichen planus (LP) is a common dermatological disease that classically presents with purple, polygonal, pruritic, flat topped papules on the skin, but can also involve mucosae and nails [1]. Many morphological variants of LP have been described. The linear variant following the lines of Blaschko is an extremely rare entity and must be differentiated from the commoner linear LP as a result of koebnerization due to trauma and zosteriform LP [2]. The rarity of this pattern of the LP inspired us to present this case.

A 17 year old healthy male presented with multiple purple colored lesions on the right side of his lower trunk which he had noticed one month back. These lesions were associated with itching. The patient did not give any history of trauma, topical applications of any medications or any recent medical or surgical illness. On examination, there were multiple, discrete, violaceous, closely set, flat topped, 2-5 mm papules, strictly unilaterally on the right side of the body in a distinct S- shaped pattern arising from the right side of suprapubic region anteriorly to the lower back (Figs. 1a and 1b) along the lines of Blaschko. The scalp, hair, mucosae and nails were normal. Routine Laboratory investigations were unremarkable. Serology for HIV, Hepatitis B and C was negative. A 5 mm punch biopsy from the lesion was taken and sent for histopathological examination which showed hyperkeratosis, hypergranulosis, vacuolar degeneration of basal layer with a band like infiltrate at the dermo-epidermal junction along with melanin incontinence. Civatte bodies were identified in the epidermis. Based on the clinical and histopathological findings, a diagnosis of Blaschkoid LP was made and the patient

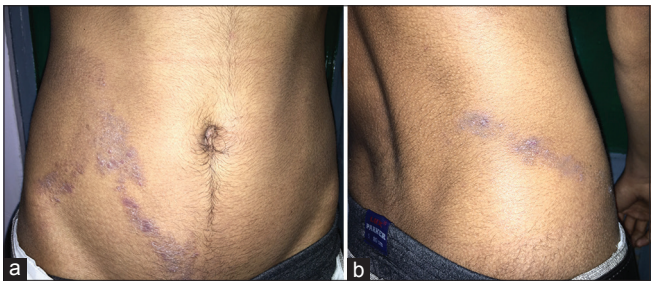

Figure 1: Multiple Violaceus papular lesions in a typical blashkoid pattern over abdomen (a) and back (b).

was treated with topical corticosteroids (mometasone) and oral antihistamines. The patient responded favorably to treatment with resolution of the lesions and decreased itching.

Lichen planus is a fairly common dermatosis but unilateral LP along the lines of Blaschko has been rarely reported [3]. Lines of Blaschko are lines that represent the boundaries between normal and mutant skin due to mosaicism, and do not correspond to any vascular, lymphatic or neural structures [4]. These are V-shaped over the upper spine, S-shaped over the abdomen, U-shaped on the chest and upper arm and perpendicular on the extremities. Some congenital nevi and inflammatory diseases like psoriasis, vitiligo and LP may present in a nevoid form [5]. The linear variety of LP can simply be a result of Koebner's phenomenon, segmental LP or zosteriform LP. It must also be differentiated with lichenoid epidermal nevus, lichen striatus, linear psoriasis or inflammatory linear verrucous epidermal nevus [6]. Happle (1996) was of the view that many cases of lichen planus along Blaschko lines are misdiagnosed as zosteriform lichen planus [7]. Blaschkoid LP is not associated with Hepatitis C or hepatocellular carcinomas while other varieties of

How to cite this article: Adil M, Amin SS, Arif T. Blaschkoid lichen planus: A rare presentation of a common disease. Our Dermatol Online. 2016;7(3):349-350.
Submission: 30.12.2015; **Acceptance:** 26.04.2016
DOI: 10.7241/ourd.20163.95

linear LP can show such an association [8,9]. Typical pattern of distribution of the lesions with suggestive histopathology can help to settle the diagnosis.

Lichen planus in a Blaschkoid distribution is rare with only some case reports published till date [1-6,9,10]. A correct diagnosis of Blaschkoid LP and its differentiation from other causes of linear LP is important as unlike the former, the latter is associated with systemic diseases. The recognition of this rare entity will prevent unnecessary investigations, cutting monetary costs, saving time and will also prevent undue anxiety in the patient.

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

Letter to the Editor

Erythema nodosum induced by kerion Celsi in a Tunisian child: A case report

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Sir,

Erythema nodosum (EN) is the most frequent form of acute nodular panniculitis [1]. The combination between EN and dermatophytosis of the scalp (kerion Celsi) is infrequently present in the literature [2]. We report another case of EN induced by kerion Celsi in Tunisian child.

A 7-years-old boy consulted us for a scaled plaque of the scalp starting 2 weeks ago. The child used to play with the rabbits according to the mother. Dermatologic examination showed a 3 x 6 cm suppurative and scaly plaque, spangled with pustules, and located on the temporal area of the scalp (Fig. 1). Over the 2 legs, we found a multiple and painful, erythematous subcutaneous nodules (Fig. 2). Mycologic direct examination of scalp lesion showed large-spore ectoendothrix invasion of the hair fragment and *Trichophyton Mentagrophites*. Histologic examination of skin biopsy of leg lesion was consistent with diagnosis of EN (Fig. 3). No other causes of EN were founded allowing us to retain the diagnosis of erythema nodosum complicating kerion celsi. The child was treated by griseofulvin 20 mg/kg/day associated to econazole nitrate cream for his kerion celsi and by mefenamic acid 500 mg/day for his EN. EN lesions regressed by the 7th day and the kerion lesions resolved without sequelae by the 6th week of antifungal treatment.

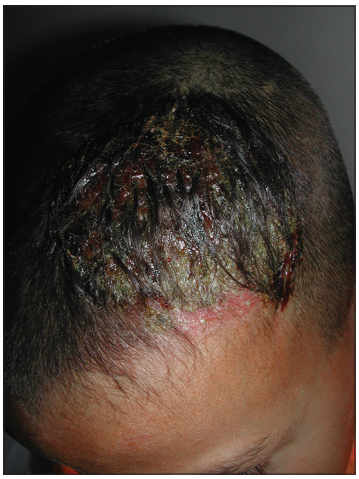

Figure 1: Kerion Celsi, 8 cm in diameter, localised in the frontal area of the scalp.

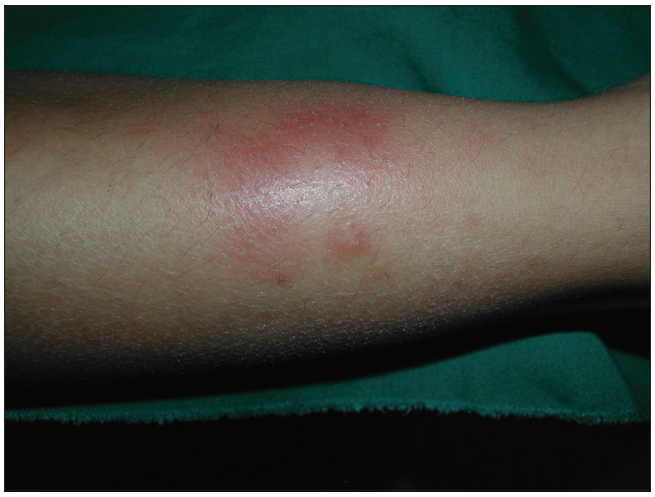

Figure 2: Erythematous nodules over the lower limbs.

Erythema nodosum is the most frequent form of acute panniculitis [1]. It can be associated to a wide variety of processes including dermatophytic infections like kerion Celsi [2]. EN complicating kerion celsi could be assimilated to a dermatophitide also called "id reaction": a distant cutaneous allergic reaction to a

How to cite this article: Mohamed M, Belkahla M, Hammedi F, Belhadjali H, Zili J. Erythema nodosum induced by kerion Celsi in a Tunisian child: a case report. Our Dermatol Online. 2016;7(3):351-352.

Submission: 05.02.2016; **Acceptance:** 01.04.2016

DOI: 10.7241/ourd.20163.96

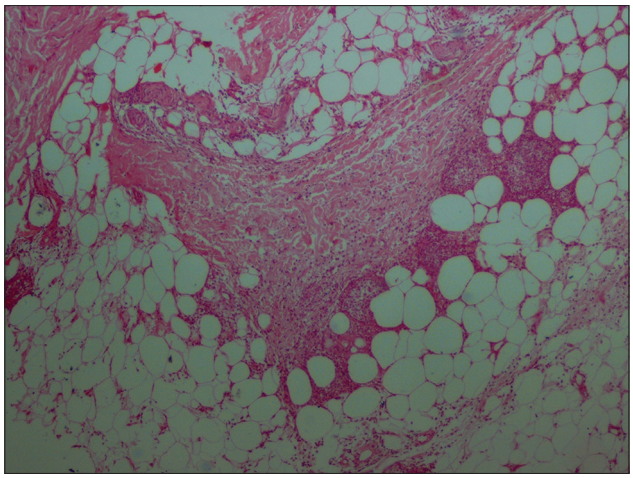

Figure 3: The histological examination of a lesion on the leg showed a thickened septa of the subcutaneous fat with abundant infiltration of neutrophils and multinucleated giant cells (HE stain x100).

fungal infection. The association between EN and kerion celsi was reported in only 15 cases (14 children and 1 woman) [2,3]. The pathogenic mechanism of this association has not been fully elucidated. Type IV hypersensibility seems to play a major role in this phenomenon. Llorente et al [4] founded increased TH1 cytokine expression in the skin lesions and peripheral blood of most of the patients with erythema nodosum compared to a control group. In the same time, it was been showed that the skin lesions produced by dermatophytic infection of zoophilic agents specifically produced by *Trichophyton Mentagrophites* are caused by TH1 response involved in the host defense against the dermatophytosis [5]. That could explain the predominance of *Trichophyton Mentagrophites* in the reported cases of EN associated to kerion Celsi, including our, (13/16 cases). On the other hand, the role of griseofulvin as causal agent of reported cases of EN associated with kerion celsi was discussed. In fact, EN occurred after the use of griseofulvin in 7 of the 15 reported cases. However, In seven other cases, as it is in our case, EN occurred before the use of griseofulvin [2]. Furthemore, improvement of EN is usually obtained using antifungal therapy.

We reported here another case of unusual association between EN and kerion Celsi, of which there are only 16 cases (including ours) in the international literature after the Second World War.

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

Our Dermatology Online

Letter to the Editor

Facial porokeratosis - Rare presentation of an uncommon disease

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Sir,

Porokeratoses are a group of hereditary or acquired disorders of keratinization. Several types have been described. Disseminated superficial actinic porokeratosis (DSAP) is the most common presentation of the disease. Facial lesions have been seen in 15% of the DSAP patients while majority of the reported cases involve extrafacial sites such as the extensor surfaces of the extremities. However, Exclusive facial presentation of DSAP has rarely been reported. In this article, the authors report a 20 year female with exclusive facial porokeratosis.

A 20 year old female visited our dermatology outpatient department (OPD) for multiple brownish lesions on the face for the last 3 months. There is no history of photosensitivity. There is no history of such lesion in her family members. There is no excessive sun exposure during these 3 months with respect to her routine work. There was no history of recurrent infections. Dermatological examination revealed multiple brownish hyperkeratotic macules of size 2-8 mm present over cheeks and infraorbital area on both sides with majority of lesions on the right side of face [Figure 1]. Most of the lesions had dense hyper pigmented (brownish) and hyperkeratotic borders with less pigmented atrophic centres. The examination of hair, mucosae and nails was normal. Systemic examination was unremarkable. Her laboratory investigations were within normal limits. Histopathology of skin biopsy revealed hyperkeratosis, columns of parakeratotic cells (coronoid lamellae) overlying an interrupted stratum granulosum and dyskeratotic keratinocytes which confirmed the diagnosis of porokeratosis. She was prescribed topical tazarotene and sunscreens. Though initially there was some benefit with respect to hyperpigmentation and scaling, but at 3 months follow up her lesions were persistent with minimal improvement.

Porokeratoses, a disorder of epidermal keratinization, includes a heterogeneous group of disorders that are mostly inherited in an autosomal dominant fashion [1]. Several clinical variants have been described which include classic porokeratosis of Mibelli, disseminated superficial porokeratosis (DSP), disseminated superficial actinic porokeratosis (DSAP), linear porokeratosis, porokeratosis palmaris et plantaris disseminata and porokeratosis punctata [2]. Other uncommon variants are giant porokeratosis, porokeratosis ptychotropica and porokeratoma, Porokeratotic Eccrine Ostial and Dermal Duct Nevus (PEODDN), punched-out, hypertrophic verrucous and reticulate porokeratosis [3,4]. Most of these variants are clinically characterized by a thin hyperkeratotic rim bordering a slightly atrophic macule or plaque with a tendency to extend in a centrifugal fashion [5]. DSAP is the most common clinical variant of the disease with multiple lesions predominantly in sun-exposed sites in middle-aged individuals [6]. Majority of the lesions occur on extrafacial sites including extremities with only 15% cases having lesions on face. Lesions of DSAP exclusive to face are rare and hence reported [7].

Various conditions have been associated with porokeratosis like Psoriasis and phototherapy (UVA, NB-UVB and BB-UVB). In addition, DSAP has also been linked with HIV infection, following administration of

How to cite this article: Arif T, Amin SS, Adil M. Facial porokeratosis - Rare presentation of an uncommon disease. Our Dermatol Online. 2016;7(3):353-354.
Submission: 19.01.2016; **Acceptance:** 24.04.2016
DOI: 10.7241/ourd.20163.97

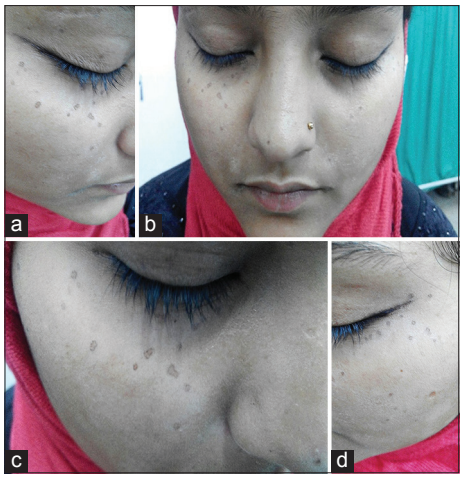

Figure 1: (a,b,c,d) Multiple 2-8 mm brownish macules with well marginated hyperkeratotic and hyperpigmented borders with less pigmentation at the centre.

immunomodulating drugs used to treat autoimmune diseases and after organ transplantation, diabetes mellitus, liver cirrhosis, acute pancreatitis, solid malignancy, Crohn's disease, etc. However our case had no systemic associations [8,9].

The histopathology is diagnostic in porokeratosis. The edge of the lesion shows characteristic microscopic findings. A hyperkeratotic stratum corneum, column of poorly staining parakeratotic keratinocytes termed as coronoid lamella with underlying interrupted granular layer and dyskeratotic cells are highly suggestive of porokeratosis. The central area of a lesion may be atrophic or normal or grossly hyperkeratotic [5,6].our case had similar findings on histopathology.

The treatment of porokeratosis is difficult.Various treatments have been tried which include cryotherapy, photodynamic therapy, erbium YAG and CO2 lasers, 3% diclofenac gel, 5- fluorouracil cream, Keratolytics, topical tacalcitol, imiquimod cream and oral etretinate [6,7]. However our case was prescribed topical tazarotene 0.05% gel to be applied at bed time on the lesions and topical sunscreens during the day. Initially the hyperkeratosis and the pigmentation improved. After a follow up of 3 months the lesions were persistent with minimal improvement in the lesions.

CONSENT

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

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

Our Dermatology Online

Historical Article

Eponyms in Tuberculosis

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Tuberculosis (TB) is an old disease and the most common cause of infection-related death worldwide. In 1993, the World Health Organization (WHO) declared TB to be a global public health emergency.

There are several eponyms related to TB. Some of these eponyms are rarely used in the present time. For example some medical dictionaries mention about "Lorenz sign", which is an

Table 1: Selected eponyms in tuberculosis

Selected eponyms in tuberculosis	Remarks
Erythema induratum of Bazin [2]	In 1861, Bazin gave the name erythema induratum to a nodular eruption that occurred on the lower legs of young women with tuberculosis. Erythema induratum/nodular vasculitis complex is classified into 2 variants: erythema induratum of Bazin type and nodular vasculitis or erythema induratum of Whitfield type. The Bazin type is related with tuberculous origin, but Whitfield type is not. Ernest Bazin (1894-1964), (Fig. 2), was a French physician
Ghon's complex [3,4]	It is the initial site of parenchymal involvement at the time of the first infection. It consists of a calcified focus of infection and an associated lymph node. These lesions are particularly common in children and can retain viable bacteria, so are sources of long-term infection and may be involved in reactivation of the disease in later life Anton Ghon (1866-1936), was an Austrian pathologist
Heaf test [5]	It is a diagnostic skin test, was long performed to determine whether or not children had been exposed to tuberculosis infection. It is named after, Professor Frederick Roland George Heaf (1894-1973), (Fig. 3), who was a British physician. The test was discontinued in 2005 because the manufacturer deemed its production to be financially unsustainable after manufacturers could not be found for tuberculin or Heaf guns. Until 2005, the test was used in the United Kingdom to determine if the BCG vaccine was needed
Mantoux test [6-8]	Also known as Mendel-Mantoux test, Mantoux screening test, tuberculin sensitivity test, Pirquet test, or purified protein derivative (PPD) test, is a screening tool for tuberculosis (TB). It is named after Charles Mantoux (1877-1947), (Fig. 4), how was a French physician. Mantoux built on the work of Robert Heinrich Herman Koch (1843-1910) and Clemens Peter Freiherr von Pirquet (1874-1929) to create his test in 1907. Koch (Fig. 5) was a celebrated German physicianand pioneering microbiologist. The founder of modern bacteriology, he is known for his role in identifying the specific causative agents of several pathogens including tuberculosis. His work with this disease won Koch the Nobel Prize in Physiology and Medicine in 1905. Pirquet (Fig. 6) was an Austrian scientist and pediatrician .He introduced the term "allergy" in 1906. He and his wife committed suicide with potassium cyanide in 1929. One more scientist to be mentioned in this regards is Florence Barbara Seibert (1897-1991), (Fig. 7), who was an American biochemist known for isolating a pure form of tuberculin used in the standard TB test
Pott disease [9-11]	It is tuberculosis of the spine. It is named after Percivall Pott (1714-1788), (Fig. 8), a British surgeon. He was one of the founders of orthopedy, and the first scientist to demonstrate that a cancer may be caused by an environmental carcinogen
Ranke's stages of tuberculosis [12]	This term is no longer used in the current literature. It was based on a hypothesis that lung tuberculosis develops in three stages. Named after, Karl Ernst Ranke (1870-1926), who was a German internist, pediatrician and pulmonologist known for his research of tuberculosis. His name was also associated with the so-called "Ranke complex", a combination of a Ghon focus and enlarged or calcified lymph nodes
Rasmussen's aneurysm [13-16]	It is a pulmonary artery aneurysm adjacent or within a tuberculous cavity. It is caused by erosion from an adjacent tuberculous cavity. It occurs in up to 5% of patients with such lesions. It may lead to rupture and hemorrhage. It is named after, a Danish physician, Fritz Valdemar Rasmussen (1837-1877), (Fig. 9)
Ziehl-Neelsen stain [17]	Also known as the acid-fast stain, which is used to identify acid-fast bacteria. In this stain, Mycobacteria will appear bright red. Dr. Franz Ziehl (1857-1926), (Fig. 10), was a German bacteriologist. He was a professor in Lübeck. Franz Ziehl introduced the carbolfuchsin stain for the tubercle bacillus in 1882. With a Friedrich Carl Adolf Neelsen (1854-1898), (Fig. 11), who was a German pathologist, he developed the Ziehl-Neelsen stain

How to cite this article: Al-Aboud NM. Eponyms in Tuberculosis. Our Dermatol Online. 2016;7(3):355-358.
Submission: 22.09.2015; **Acceptance:** 06.12.2015
DOI: 10.7241/ourd.20163.98

obsolete term for stiffness of the thoracic spine in early pulmonary tuberculosis. Named after, Adolf Lorenz (1854 - 1946) (Fig. 1), who was an Austrian surgeon [1].

One may find little information in the literature about the origin of some of the eponyms related to tuberculosis. An example of this is "Löwenstein-Jensen" media used for TB culture.

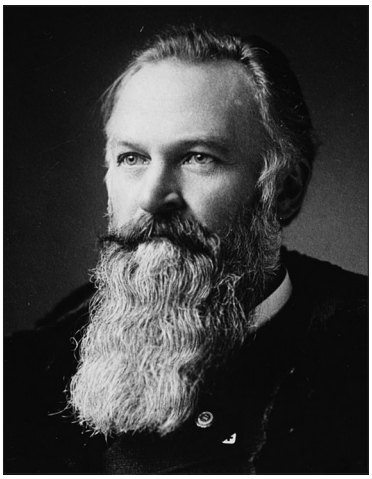

Figure 1: Adolf Lorenz (1854-1946).

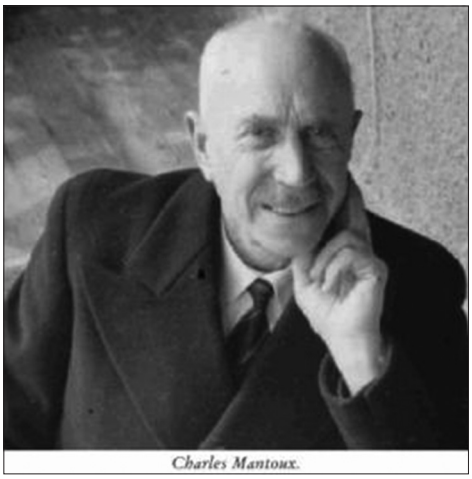

Figure 4: Charles Mantoux (1877-1947).

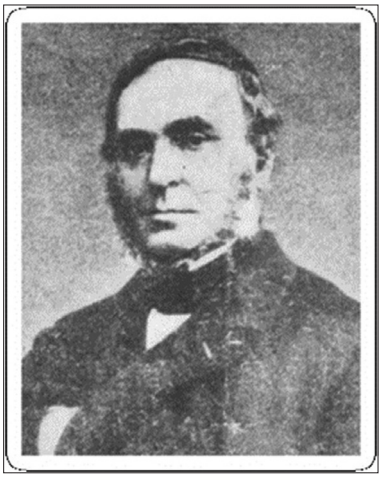

Figure 2: Ernest Bazin (1894-1964).

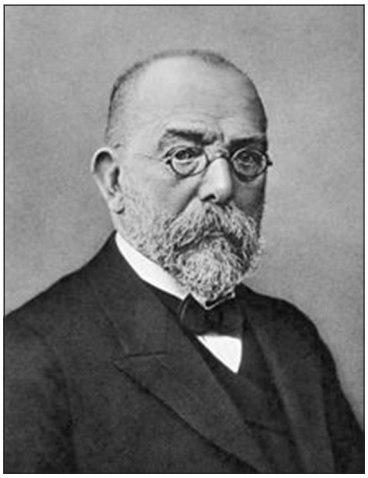

Figure 5: Robert Heinrich Herman Koch (1843-1910).

Figure 3: Frederick Roland George Heaf (1894-1973).

Figure 6: Clemens Peter Freiherr von Pirquet (1874-1929).

Figure 7: Florence Barbara Seibert (1897-1991).

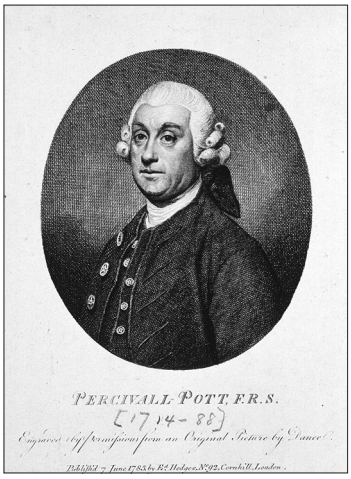

Figure 8: Sir Percivall Pott (1714-1788).

Figure 9: Fritz Valdemar Rasmussen (1837-1877).

In Table I, we tried to summarize the available literature for selected eponyms linked to TB.

Figure 10: Franz Ziehl (1857-1926).

Figure 11: Friedrich Carl Adolf Neelsen (1854-1898).

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

Our Dermatology Online

Historical Article

Dermatology Eponyms – sign –Lexicon (P). Part 2

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ABSTRACT

Eponyms are used almost daily in the clinical practice of dermatology. And yet, information about the person behind the eponyms is difficult to find. Indeed, who is? What is this person's nationality? Is this person alive or dead? How can one find the paper in which this person first described the disease? Eponyms are used to describe not only disease, but also clinical signs, surgical procedures, staining techniques, pharmacological formulations, and even pieces of equipment. In this article we present the symptoms starting with (P) and other. The symptoms and their synonyms, and those who have described this symptom or phenomenon.

Key words: Eponyms; Skin diseases; Sign; Phenomenon

Port-Light Nose sign

Rhinophyma as a progressive, disfiguring disease of the nose, is considered the final stage of acne rosacea [1]. Also called bottle nose (Fig. 1).

Possum Boil Sign

Leptospirosis infection [2]. Also known as Opossum and Skunk sign.

Pot Belly Sign

A classic presentation of pellagra (Figs 2a and 2b) [3-5].

Practolol Sign

Hyperkeratosis of the feet and palms. Associated with carcinoma of the oesophagus and psoriasis [6]. Even though it is a syndrome of low incidence, hyperkeratosis or tylosis palmoplantaris is widely related with the onset of squamous cell carcinoma of the esophagus. Such correlation receives the name Howel-Evans and has a family penetrance close to 100%.

Prayer Sign

1. It is described in relation to diabetic cheiroarthropathy, wherein the patient is requested to bring both the palmar surface of the hands together as at prayer. Prayer sign is said to be positive when patient is unable to bring both the palmar surface together completely and it indicates limited joint mobility. Limited joint mobility is secondary to nonenzymatic glycosylation of collagen and its deposition in the small joints of the hand [7].

2. Callosity on the forehead, from the Moslem style of praying several times a day, with the forehead touching the ground [8].

How to cite this article: Brzeziński P, Tanaka M, Husein-ElAhmed H, Castori M, Barro/Traoré F, Kashiram Punshi S, Chiriac A. **Eponyms – sign –Lexicon (P). Part 2.**. Our Dermatol Online. 2016;7(3):359-365.

Submission: 21.12.2014; **Acceptance:** 12.03.2016

DOI:10.7241/ourd.20163.99

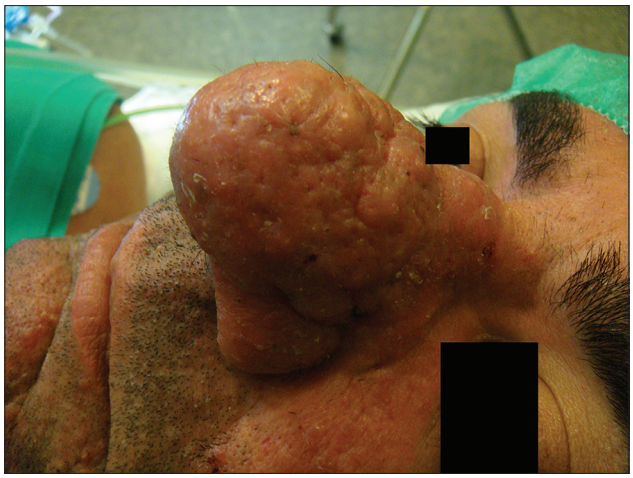

Figure 1: Port-Light Nose sign

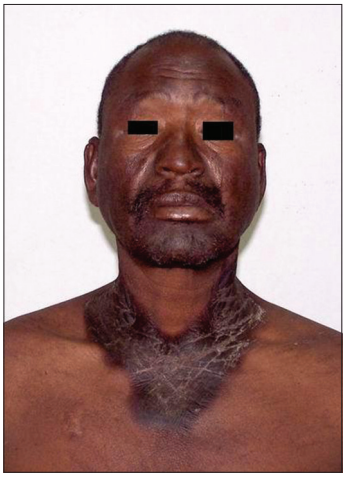

Figure 2: (a) Pot Belly sign

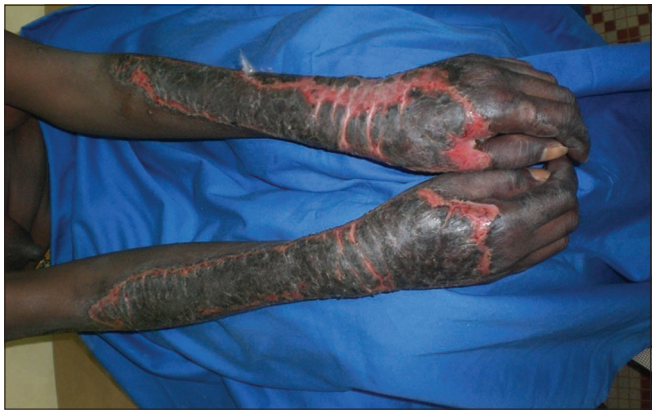

Figure 2: (b) Pot Belly sign

Preston's Sign

Keratolysis or deciduous skin, a condition in which the whole skin is cast off like a snake in huge contiguous pieces. Also called Serpent sign. Deciduous skin can be defined best as continuous, periodic or seasonal shedding or peeling of the epidermal layer of the skin of more or less generalized distribution [9].

Price's Sign

Press the thumb against the sternum and hold it there for a few seconds, then remove it quickly. If the blanched area does not turn pink in less than a second, this is an indication of early shock. Sign described by **Philip Barton Price,** American surgeon.

Primary Sign

Synonyme: chankroid. Induration, resulting in the site of entry of Treponema pallidum in syphilis [10].

Prosector's Rot Sign

Paronychia primarily from Mycobacteruim tuberculosis, contained within tissue sections [11].

Proud Flesh Sign

Exuberant granulation tissue around the opening of sinus track. A sign that signifies necrosis of bone is occurring in the depths of the track.

Prussic Sign

Giddiness, eyes staring, and teeth clenched. A sign of hydrocyanic acid poisoning [12].

Purple Foot Sign

Trench foot, mmersion foot was originally described in the military literature during World War I [13,14]. A characterized signs are pedal swelling, numbness, and pain.

In your work Atenstaedt described [15]:

"…. Trench foot appeared in the winter of 1914, characterized by pedal swelling, numbness, and pain. It was quickly recognized by military-medical authorities. There was little debate over whether it was frostbite or new condition, and it was quickly accepted as a specific disease. The major etiologies proposed were exposure, diet, and infection. The opinion emerged that it was caused by circulatory changes in the foot caused by cold, wet, and pressure. Predisposing factors included dietary inadequacy and fatigue. A number of labels were first given to the disease. However, the name "trench foot"

was eventually officially sanctioned. Trench foot became a serious problem for the Allies, leading to 75 000 casualties in the British and 2000 in the American forces. Therapy for trench foot involved a number of conventional, tried-and-tested, and conservative methods. Some more innovative techniques were used. Amputation was only used as a last resort. Prevention involved general measures to improve the trench environment; modification of the footwear worn by the men; and the provision of greases to protect them from moisture. The medical reaction to this condition seems to have been relatively effective. The causation was identified, and prophylactic measures were introduced to fit this model; these seem to have been successful in reducing the prevalence of the condition by 1917-18…"

Purple Hair Sign

Hair becomes purplish brown when the scalp is treated with chrysarobin and then washed with an alkali soap [16].

Pseudo-acanthosis Nigricans Sign

Pseudo-acanthosis is a type of acanthosis nigricans that is seen in obese persons and is reversible on weight reduction [17].

Pseudo-acne Sign

Acneiform red papules along transverse nasal crease in preadolescent individuals. They are basically keratin granulomas derived from ruptured and inflamed milia. Due to their clinical similarity to acne, they have been named pseudo-acne [18].

Pseudo-ainhum Sign

Ainhum is the autoamputation of a digit, due to development of constricting bands around bilateral fifth toes, seen in underdeveloped countries of Africa (Fig. 3). In the remainder of the world, constricting bands that mimic ainhum are termed pseudo-ainhum. Causes of pseudo-ainhum are amniotic bands, constrictions associated with keratotic disorders, infections, and trauma [19].

Pseudo-bubo Sign

Seen in Donovanosis. Sub-cutaneous swellings in inguinal areas, which may break down to form typical granulomas, are known as pseudo-bubos since there is no lymphadenitis on microscopic examination [20].

Pseudo-chancre Redux Sign

This term is used to describe a solitary gumma of the penis at the site of the original chancre [21].

Pseudo-cowpox Sign

Milker's nodule is otherwise known as pseudo-cowpox. It is a paravaccinia (parapox) virus infection seen in individuals in close contact with cattle, like milkers, veterinarians, and workers in meat industry. Cutaneous lesions can be one or several papulovesicles to bluish nodules, which may rupture to form ulcers with eschar. Cowpox is caused by the cowpox virus (orthopox) [22].

Pseudo-Darier Sign

When firmly stroked, fasciculation of the skin causes the appearance of goosebumps or cobblestones from the stimulation of aberrant arrector pili muscles - Smooth muscle hamartoma.

Pseudo-eccrine Chromhidrosis Sign

This refers to the development of colored sweat when surface compounds or molecules mix with sweat to produce pigment e.g. the formation of blue sweat in copper workers. Extrinsic dyes, paints, fungi, and chromogenic bacteria are other causes of pseudo-eccrine chromhidrosis [23].

Pseudo-elephantiasis Sign

Pseudo-elephantiasis is a condition, caused due to inflammation, edema or obstruction of lymphatics, triggered by non-filarial infections like donovanosis, lymphogranuloma venereum, syphilis, tuberculosis [24].

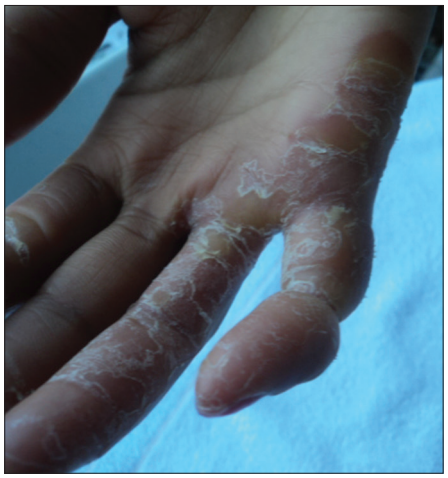

Figure 3: Pseudo-ainhum sign

Pseudo-folliculitis Sign

Inflammatory papules resulting from re-entry of sharp tips of cut hair into the skin. Pseudo-folliculitis usually affects the beard area, when it is known as pseudo-folliculitis barbae [25].

Pseudo gall Sign

Eliminate certain parasitic pruritus such as scabies, difficult to diagnose in patients with strict hygiene, in the absence of specific signs (pseudo-bladder and groove), possibly by a testing treatment (Vulvar and anal pruritus) [26].

Pseudo-granuloma Inguinale Sign

It is a clinical variant of chancroid resembling granuloma inguinale (Donovanosis). Selective culture media isolates Hemophilus ducreyi from the penile ulcer, which clinically looks like granuloma inguinale [20].

Pseudo-Hutchinson's Sign

Discoloration of the eponychium: Congenical nevus of the nail apparatus. (Fig. 4) [27].

Pseudo sign of Lesar Trelat

Inflammation of seborrheic keratosis caused by cytarabine: A pseudo sign of Leser-Trelat. A Leser-Trelat sign refers to multiple eruptive seborrheic keratosis, in the context of an internal malignancy [20].

Pseudo-monilethrix Sign

It is a hair shaft abnormality, where the hair shaft shows irregular, flattened, expanded areas that have an indented appearance, unlike monilethrix where the hair shafts have beaded appearance because of alternate zones of spindle-like thickening and thinning, placed at a regular intervals [28].

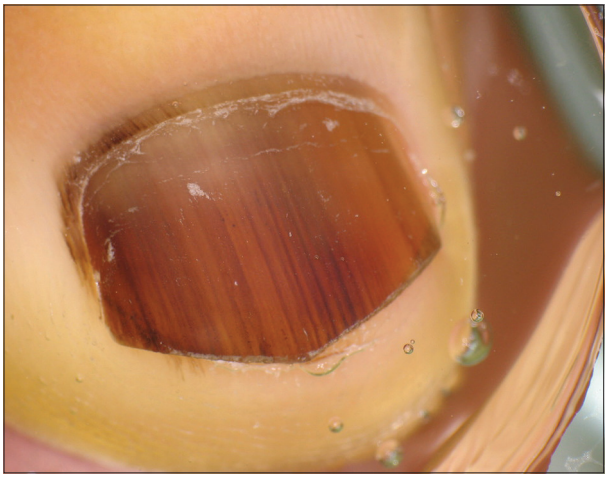

Figure 4: Pseudo-Hutchinson's sign

Pseudo-mycosis Sign

Botryomycosis is otherwise known as pseudo-mycosis. Cutaneous botryomycosis resembles a fungal infection but is actually a chronic, inflammatory response to bacterial infection. Staphylococcus aureus is the most commonly implicated bacteria, followed by Pseudomonas aeruginosa [29].

Pseudo-Nikolskiy Sign

The method and mechanism of elicitation is the same as for Nikolskiy sign but and can be elicited only on the involved or erythematous areas. The underlying mechanism here is the necrosis of epidermal cells and not acantholysis as in true Nikolskiy sign. This sign is positive in Stevens-Johnson syndrome, toxic epidermal necrolysis and in some cases of burns and bullous ichthyosiform erythroderma [20].

Pseudo-nits Sign

Peipilar keratin casts or hair casts or pseudo-nits are thin, elongated, firm, cylindrical concretions that encircle the hair shafts and can be easily dislodged, unlike true nits, which are oval, glistening, translucent, whitish lidded capsule attached to the side of the hair shaft by a firm chitinous sheath [20].

Pseudo-ochronosis Sign

Bluish-black macular pigmentation seen on the upper extremities of jewelry workers. Histopathologically, it is seen as localized argyria. The pigmentation simulates the typical pigmentation seen in endogenous ochronosis (alkaptonuria) [30].

Pseudo-paralysis of Parrot Sign

Originally described by Parrot, this pseudo-paralysis represents decreased movement of the extremity secondary to painful syphilitic periostitis [20].

Pseudo-pseudoxanthoma Elasticum

It is a penicillamine-induced degenerative dermatosis, histopathologically characterized by abnormal elastic fibers in the dermis. Clinically, the patient develops

multiple small yellowish papules on both sides of neck mimicking pseudo-xanthoma elasticum, after long term therapy with D-penicillamine [31].

Pseudo-rheumatoid Nodule Sign

Sub-cutaneous granuloma annulare is otherwise known as pseudo-rheumatoid nodule, because of their clinical resemblance to rheumatoid nodule. Lesions are nodular and predominantly occur on the scalp and pretibial regions [32].

Pseudo-scleroderma Sign

Pseudo-scleroderma refers to sclerosis of skin in conditions other than morphea or systemic sclerosis. This is seen in eosinophilic fasciitis, dermatomyositis, systemic lupus erythematosus, porphyria cutanea tarda, phenylketonuria or paraproteinemia [33].

Pseudo-syndactyly Sign

Pseudo-syndactyly is seen in dystrophic epidermolysis bullosa, autosomal-recessive forms, where repeated blistering with progressive scarring causes fusion of adjacent fingers and toes, thus giving rise to pseudo-syndactyly. True syndactyly is a congenital malformation, where there is failure of differentiation, in which the fingers fail to separate into individual appendages [34].

Pseudo-verrucous Papules Sign or Pseudo-verrucous Papules Nodules Sign

Pseudo-verrucous papules and nodules occur in the diaper and perineal areas in patients, of any age, with a predisposition to prolonged wetness. Children who wear diapers due to chronic urinary incontinence are prone to this type of dermatitis [35].

Pseudo-xanthoma Elasticum Sign

Pseudo-xanthoma elasticum is an inherited disorder characterized by generalized fragmentation and progressive mineralization of the elastic fibers in various tissues, involving the dermis (in flexural sites), eye (angioid streak and retinal defects), and cardiovascular system (hypertension and vascular disease). Cutaneous lesions are small, yellowish papules occur in a linear or reticular pattern, commonly involving areas like the sides of neck, axillae, groins, and perineum [36].

Punshi's Sign

In young women and girls having from vitiligo the original white color of vitiligo macules turns to red-pink during menstruation and after the menstruation, it turns to the original colour [37]. Sign described in 1970.

Satish Kashiram Punshi

Dermatologist from India (Fig. 5). He was born in the year 1939. He is a well known writer, a thinker, philosopher, a social worker and a dermatologist of international repute, all rolled up in one. He is one of the most famous author of medical books on the topic Leucoderma. He is one who is pioneer for introducing placental extract therapy in Vitiligo (Leucoderma) and winner of prestigious International Lions award for Leprosy eradication. Dr. S. K. Punshi's work on Leucoderma is quoted in various books and Journals of Dermatology. He completed his Medical Dgree M.B.B.S. from the Indore and went on to get D.D.V. from Mumbai, his academic qualification also includes F.I.M.S. , F.D.S. (London). Though being engaged with writing of Medical books and serving the patients of skin diseases, he being awoken soul always find time to spend for the social works and thinks about the development of deprived section of society. During his long illustrated social life at Amravati city Dr. Punshi is active with many social organizations like Amravati Junior Chamber [Ex. Vice President], Loins Club of Amravati Central [Ex. President] along with being the life member of Red Cross Society, Sangit Kala Upasak Mandal, Cancer relief society, Vivekananda Ashram Society and many more. He also attended many national and international conferences and at few he was the Guest Speaker. In the Sindhi literature, Dr.Punshi is the famous personality for being the Author of Biography of Famous sindhi Saint Bhagat Kanwar Ram and spiritual master Sant Satramdas Sahib Ji. His many articles are published in Hindvasi (Sindhi), Illustrated weekly of India, Hindustan, Times of India, Hitvada and Nagpur times etc. Divine knowledge (A scientific approach to the study of Geeta), Biography of Dr. A. V. Mudliyar, Sindh darshan and Saint Kashiram- Roohani Rahbar are the some of other books written by him.

Dr. Punshi with all his achievements remains simple, solumn, sober, without much pomp and show and fanfare. He believes in God says "All is God's will; man is just an instrument in the hands of God".

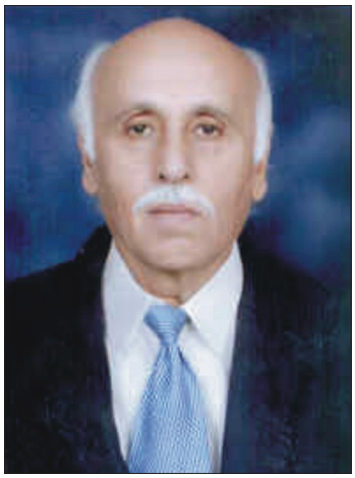

Figure 5: Satish Kashiram Punshi

He has more of Humanistic and Holistic approach to life". He is a humble servant of humanity. He has lived his life for the cause and service of others. Nothing did for the self accept suffering.

Author Medical Books: 1. Vitiligo – Diagnosis and treatment; 2. Vitiligo and placental extract; 3. Vitiligo – quarterly medical review; 4. A Hand books of Scabies; 5. A Hand books of Leprosy; 6. Dermatology for general practitioners; 7. Colour Atlas of Vitiligo (Jaypee Brothers); 8. Diagnosis & Management of Dermatology made easy. (Jaypee Brothers).

Awards: 1. All India Award by skin institute Delhi in 1977 for research on use of placental extract in Leucoderma; 2. ' Ambady Oration' award I 1986; 3. International Lions award for Leprosy eradication; 4. Lifetime achievement award I.A.D.V.L- Maharashtra state 2010; 5. Saint Kanwaram award; 6. Proffesor Hasanand memorial award.

"One who works in devotion, who is a pure soul, and who controls his mind and senses is dear to everyone, and everyone is dear to him. Though always working, such a man is never entangled."

Pup-tent Sign

It is seen in nail lichen planus, in which the nail splits and elevates longitudinally with downward angle of lateral nail edge [38,39].

Puumala Sign

Rapid fever, kidney failure, severe back pain, and bleeding rash which progresses to death in 15 percent of victims. Caused by a zoonotic hantaviral infectious process known as hemorrhagic fever with renal syndrome disease [40].

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

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3.2016 (05.July.2016)