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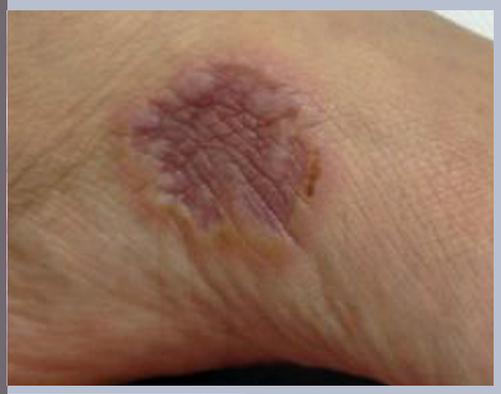

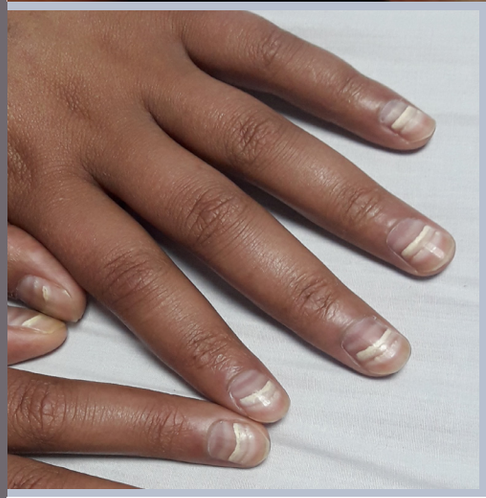

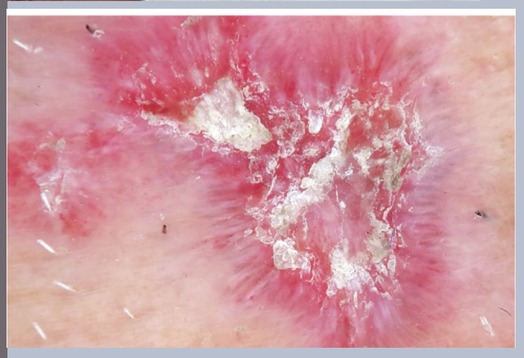

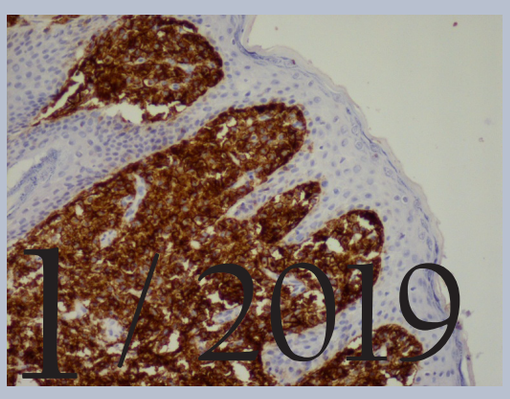

Editorial Pages

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Editorial Board

Contents

Contents

Contents

Our Dermatology Online

Original Article

Erythroderma: A clinical and etiological study of 92 patients

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ABSTRACT

Background: Ery throderma is a rare and severe dermatological manifestation of a variety of diseases. It is usually difficult to find the underlying cause. This is a dermatological emergency that can be life-threatening. **Objective:** study aimed to examine the causes, as well as the clinical and laboratory profiles of patients with erythroderma in our department. **Methods:** This retrospective study was carried out in the Department of Dermatology of CHU Hassan II of Fez. Between January 2013 and May 2017. We included all patients hospitalized for acute or chronic erythroderma **Results:** During the study period, 1700 patients were hospitalized in the Department of Dermatology. Erythroderma was diagnosed in 92 patients (5.41%), corresponding to an incidence of 20 cases/year. The average age was 49.6 years with a male to female ratio of 0.70. The average duration of evolution was 259 days. Erythroderma was dry in 82.6% of cases. Inflammatory anemia was found in 10.9% of patients. Eosinophilia in 44.6% of patients. Hepatic cytolysis and hyponatremia in 12.9%. Kidney failure in 18.3%. The most common cause of erythroderma was exacerbation of pre-existing dermatoses (43.5%), including psoriasis (27.2%), eczema (7.6%), pemphigus foliaceous (6.5%) and pityriasis rubra pilaris (2.2%). Followed by drug reactions (38%) dominated by the drug reaction with eosinophilia and systemic symptoms syndrome (30.4%). Cutaneous T-cell lymphomas constituted 15.2% of the causes of erythroderma. The evolution was good in 87% of the patients, whose methods of management were based on symptomatic measures in association with the etiological treatment. **Conclusion:**Erythroderma is a severe dermatological syndrome of various etiologies. Drug inducederythroderma is a very important entity to know. Indeed, its early diagnosis allows a fast and adequate care thus improving the prognosis of this entity.

Key words: Erythroderma; Etiologies; Department of dermatology

INTRODUCTION

Erythroderma or exfoliative dermatitis is a rare skin disorder that may be the result of many different causes. It represents an extreme state of skin irritation involving the whole or most of the skin surface. Because most patients are elderly and skin involvement is widespread, the disease implies an important risk to the life of the patient. The causative factors can be grouped as previous dermatoses, drug reactions, malignancies systemic diseases, infections and idiopathic disorders The four more common causes of idiopathic protracted erythroderma are probably atopic dermatitis of the elderly, intake of drugs overlooked by the patient, pre lymphomatous eruptions and occult malignancies [1,2]. Histopathology can help identify the cause of erythroderma in up to 50% of cases, particularly by multiple skin biopsies [3]. Many chronic dermatoses may be histologically indistinguishable in erythrodermic patients [4].

To date there are no published studies on the frequency of underlying causes of erythroderma from our country. In order to delineate the salient features of erythroderma in our region, we have reviewed the cases of erythroderma examined and treated in our department between 2013 and 2017 (four-and-a-half-year period). Our study

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aimed to examine the causes, as well as the clinical and laboratory profiles of patients with erythroderma in our department. Finally, our data are discussed and compared with previously published series.

MATERIALS AND METHODS

We conducted a retrospective analysis of the clinical and laboratory profile of all patients with erythroderma admitted to the Dermatology and Venereology Department of Hassan II University Hospital Center of Fez Morocco, between January 2013 and May 2017. Our department is one of the largest dermatology teaching units in Morocco, integrated in a tertiary referral hospital in the central region of the country. All the patients diagnosed with erythroderma had developed erythema involving more than 90% of the body surface area. Due to the risks that erythroderma implies for the patient's life and to study the cause in each case, they are always treated as inpatients. The records of these patients were carefully reviewed and the data recorded was personal data, past medical history (including history of skin diseases), drug consumption history, previous episodes of erythroderma onset and clinical data during the episode (pruritus, cutaneous signs, nail involvement, temperature, presence of edema, lymphadenopathy and visceral enlargement). Laboratory investigations including complete hematological parameters, serum electrolytes, liver and kidney function tests. electrocardiography, and chest radiography were performed for all the patients as a routine in the dermatology ward of our hospital. Whenever indicated, further investigations such as skin biopsies, lymph node biopsies, bone marrow investigation, flow-cytometry, immunophenotyping, patch tests and CT-scans were performed. We also examined follow-up data concerning the management, outcome and relapses when such information was available. For statistical analysis, data was compiled electronically into Excel programme and analysed using SPSS® (Statistical Package for the Social Sciences, v. 20). Clinical and laboratory data were analysed by chi-square (v2) and Fischer tests looking for a possible relationship between clinical data, laboratory tests, and corresponding etiologies. Statistical significance wa defined as $P<0.05$.

Ethics Statement

Patientshave given their informed consent and thatthe study protocol has been approved by the institute's committee onhuman research.

RESULTS

During the study period, 1700 patients were hospitalized in the Department of Dermatology. Erythroderma was diagnosed in 92 patients (5.41%), corresponding to an incidence of 20 cases/year. The average age was 49.6 years (range 3–96) with a male to female ratio of 0.70. Patients were seen in our department at a median of 259 days (range 2 to 3680 days) after the onset of erythroderma. A shorter duration was observed in patients with drug-induced erythroderma and a longer one with erythroderma caused by malignacies. In 48 (52.2%) of the 92 patients, erythroderma began acutely. As shown in table 1, erythroderma was dry in 82.6% of cases, pruritus was the most common complaint, was recorded in 82 (89.1%) patients; 57 of the 92 patients (62%) had a fever during the episode, 55 patients (59.8%) had pitting edema, palmoplantar keratoderma were found in 33 patients (35.9%) mostly in the group of psoriasis patients. Nail changes (including Beau's lines, onychodystrophy, pits, discoloration, subungual hyperkeratosis, onycholysis and paronychia) were observed in 30 (32.6%) patients. Nail changes were mostly found in malignancies and psoriasis patients. Other main symptoms included liver and/or spleen enlargement in 35 (38%) patients and lymph node enlargement in 50 (54.3%) patients.

Eosinophilia in 44.6% of patients. Kidney failure in 18.3%. Hepatic cytolysis and hyponatremia in 12.9%. Inflammatory anemia was found in 10.9% of patients.

Laboratory abnormalities are also summarized in table 1, including inflammatory anemia, eosinophilia, hyponatremia, hepatic cytolysis and renal failure. Cutaneous biopsy was performed in 55 (59.78%) patients. A skin biopsy was not performed in the rest of the patients because the cause of erythroderma was clear from the start as previous dermatosis. The biopsy was usually performed during the first three days after the patient was admitted to the hospital. The most common cause of erythroderma was exacerbation of pre-existing dermatoses (43.5%), including psoriasis (27.2%), eczema (7.6%), pemphigus foliaceous (6.5%) and pityriasis rubra pilaris (2.2%). Followed by drug reactions (38%) dominated by the DRESS syndrome (30.4%). Cutaneous T-cell lymphomas constituted 15.2% of the causes of erythroderma.

Etiologically, the 92 patients were categorized into four groups (Fig. 1): (1) Pre-existing dermatoses (49 patients, 43.5%): psoriasis 25 (27.2%), eczema

Table 1: Epidemiological, clinical and laboratory features of the 92 patients with erythroderma according to etiology

Etiology	Psoriasis n=25, 27.2%		Drug n=35, 38%		Eczema n=7, 7.7%		Malignancies n=16, 17.4%		Preexisting Dermatoses* n=40, 43,5%		Others** n=1, 1.1%		Total n=92, 100%	
Age at admission (years)														
Mean	41.9		48.8		58.3		55.6		48		47		49.6	
Range	3-87		13-96		35-77		17-81		3-87				3-96	
Ratio Male-Female	0.92		0.67		1.33		0.45		0.90		0 (1 female)		0.70	
Erythroderma's duration before admission (days)														
Mean	437		25		119		588		338		60		259	
Range	3-2880		2-90		15-368		30-3680		3-2880				2-3680	
Clinical findings	**n**	**%**	**n**	**%**	**n**	**%**	**n**	**%**	**n**	**%**	**n**	**%**	**n**	**%**
Erythroderma														
Dry	23	92.0	29	82.9	6	85.7	16	100.0	31	77.5	0	0.0	76	82.6
Wet	2	8.0	6	17.1	1	14.3	0	0.0	9	22.5	1	100.0	16	17.4
Pruritus	25	100.0	30	85.7	7	100.0	16	100.0	36	90.0	0	0.0	82	89.1
Peripheral edema	13	52.0	25	71.4	3	42.9	12	75.0	18	45.0	0	0.0	55	59.8
Fever	7	28.0	32	91.4	3	42.9	8	50.0	16	40.0	1	100.0	57	62.0
Palmoplantar keratoderma	19	76.0	2	5.7	4	57.1	6	37.5	25	62.5	0	0.0	33	35.9
Nail changes	14	56.0	2	5.7	2	28.6	10	62.5	18	45.0	0	0.0	30	32.6
Liver and/or spleen enlargement	6	24.0	14	40.0	3	42.9	12	75.0	9	22.5	0	0.0	35	38.0
Lymphadenopathy	7	28.0	20	57.1	2	28.6	16	100.0	14	35.0	0	0.0	50	54.3
Laboratory data	**n**	**%**	**n**	**%**	**n**	**%**	**n**	**%**	**n**	**%**	**n**	**%**	**n**	**%**
Anemia	3	12.0	5	14.3	0	0.0	0	0.0	4	10.0	1	100.0	10	10.9
Eosinophilia	4	16.0	22	62.9	4	57.1	8	50.0	11	27.5	0	0.0	41	44.6
Hyponatremia	2	8.0	12	34.3	0	0.0	2	12.5	2	5.0	0	0.0	12	13.0
Hepatic cytolysis	0	0.0	12	34.3	0	0.0	0	0.0	0	0.0	0	0.0	12	13.0
Renal failure	1	4.0	14	40.0	0	0.0	2	12.5	1	2.5	0	0.0	17	18.5

*Preexisting dermatoses: psoriasis, eczema, pemphigus foliaceus and pityriasis rubra pilaris.

**Others: subacute cutaneous lupus.

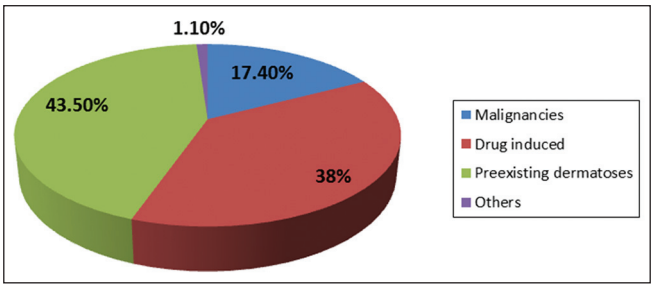

Figure 1: Groups of etiologies of erythroderma.

7 (7.7%), pemphigus foliaceus 6 (6.5%), pityriasis rubra pilaris 2 (2,2%). (2) Drug reactions (35 patients, 38%). (3) Malignancies (16 patients, 17.4%): mycosis fungoides and sezarysyndrom (14), lung cancer (1), cavum cancer (1). (4) Others (1 patient, 1.1% subacute cutaneous lupus).

Some etiologies were significantly associated with certain clinical signs and biological tests:

1 Acute onset and drug-induced erythroderma ($p = 0.000$).
2 Chronic onset and pre-existing dermatoses and malignancies ($p=0.000$).
3 Dry erythroderma and malignancies ($p=0.031$).
4 Fever and drug-induced erythroderma ($p = 0.000$).
5 Peripheral edema and drug-induced erythroderma and malignancies ($p=0.035$).
6 Palmoplantar keratoderma and pre-existing dermatoses (Psoriasis and Pityriasis rubra pilaris) ($p = 0.000$).
7 Nail changes and psoriasis and mycosis fungoïde ($p=0.000$).
8 Lymphadenopathy as well as liver/spleen enlargement were clinical features significantly associated with malignancy- related erythroderma cases ($p=0.000$ and $p=0.003$, respectively).
9 Lymphadenopathy and drug-induced erythroderma ($p = 0.000$).
10 Hypereosinophilia with drug-induced erythroderma ($p = 0.015$).
11 Hepatic cytolysis was more frequently objectified in the drug reactions ($p = 0.004$).

No other cause of erythroderma was significantly associated with any of the remaining laboratory abnormalities analyzed (including anemia, hyponatremia or renal failure).

The evolution was good in 87% of the patients, whose methods of management were based on symptomatic measures in association with the etiological treatment. 10 patients died after transfer to an intensive care unit, either because of the etiology of erythroderma, the decompensation of an underlying pathology, or of infectious shock.

DISCUSSION

We collected 92 cases of erythroderma in an four-and-a-half-year period, corresponding to an incidence of 20 cases/year. A survey of erythroderma cases conducted in the Portugal identified an annual incidence of 9.4 cases/year [1], and another Tunisian series reported a hospital incidence of 6.3 cases/year [5], an increased incidence as we found, has never been noted in the literature, this could be explained by the delay of the consultation of patients. Erythroderma usually occurs in the fifth decade with a male predominance in the majority of reported studies [6-14].Our series is in accordance with the literature, concerning the age of patients, but with a female predominance. Typically, the onset of erythroderma is gradual and insidious [5,9-11], except in the drug-induced cases, where it tends to be sudden [5,12] In our series we identified similar results, given that drug-induced cases were associated with a shorter duration of erythroderma before admission. The approach of patients with erythroderma depends on their previous dermatological background. Patients with a history of dermatological disorder may develop erythroderma during a flareup. In such cases, the aetiological diagnosis is easy to reach. Otherwise, erythroderma remains a diagnostic challenge. In one series reported by Eugster et al., five patients among seven with malignancy-related erythroderma had a history of preexisting psoriasis.[12] Furthermore, drugs can also precipitate erythroderma in a well known psoriatic patient. So, it is important to consider other possible aetiologies even in patients who may have a clear history of pre-existing dermatosis [15,16].

The final diagnosis is a result of the evaluation of the clinical, biochemical, histological findings and the evolution of the erythroderma in each individual patient. Like many other series [4, 8-14], the majority of clinical features were non-specific. As in other studies [11,12,14], pruritus was the most common complaint of our patients and could not be traced to any specific cause of erythroderma. In this respect, only one retrospective study on the subject has found a significant association between pruritus and psoriasis [8]. As was the case of our series, other reports have found that palmoplantar keratoderma [8,9] as well as nail changes [8,9,12] are predictive clinical signs of psoriasis. Thus, in the absence of history of psoriasis, these cutaneous modifications may direct clinicians to psoriasis.

Edema was reported to be to drug reactions [17,18]. In our study, half of patients presented edema as it was seen in the Portugal survey [1]. We also found a significant relation between edema, drug consumption and malignancies. More than half of our patients had fever, comparable to the incidence reported in other studies [8,11-14]. We also noted that fever was more frequently associated with drug reactions, an association previously identified in three other recent series [1,8,9]. We found a higher percentage of lymphadenopathy and hepatic/splenic enlargement in comparison with previous series [8-14].

In previous publications, lymphadenopathy has been associated with cutaneous T-cell lymphomas, drug reactions and psoriasis [10] Hepatic or splenic enlargement have been reported in association with erythroderma caused by drug consumption [2]. In our study however, we only found significant associations between both lymphadenopathy and hepatic/splenic enlargement, and the group of patients with cutaneous T-cell lymphomas. Also between lymphadenopathy and drug consumption. In our series, drug-induced erythroderma was significantly associated with hypereosinophilia. The latter is, however, not a specific finding limited to drug reactions, as it has been previously reported in other studies to be also significantly associated with psoriasis, eczema and cutaneous T-cell lymphoma [1,5,8,9].

Comparison of the etiological groups among recent series of erythroderma and our own is given in Table 2. This table reveals a variation of the relative incidence of the different etiological groups of erythroderma. This may be partly related to genetic, geographical and social disparities. Our findings support the perception that non-malignant erythroderma, despite often bringing distress to patients, most often does not pose

Table 2: Comparison of earlier studies with present study for etiology of erythroderma.

Author,year, country	Number of patients	Preexisting dermatoses (%)	Drug Reaction (%)	Malignancies (%)	Others (%)	Idiopathic (%)
Present study	92	43.5	38	17.4	1.1	
Artur Cesar et al., 2016, Portugal (5)	103	65	18.4	12.6		3.9
Hulmani M et al., 2014, India (13)	30	63.3	16.6	3.3	1.2	16.6
Li J et al., 2012, China (12)	260	69.6	12.7	2.3	3.7	14.2
Yuan XY et al., 2010, China (11)	82	68.3	17	4.9	3.7	6.1
Khaled *et al.*, 2010, Tunisia (6)	82	43.9	21.9	4.9		25.6
Fernandes *et al.*, 2008, Brasil (10)	170	58.2	21.8	10.6		9.4
Akhyani M *et al.*, 2005, Iran (14)	97	59.7	21.6	11.3		7.2
Rym BM *et al.*, 2005, Tunisia (9)	80	72.5	11.3	8.8		7.5

a significant risk to the patient's life [19]. Despite its limitations as a retrospective design, this study provides us with interesting information related to clinical features of erythroderma. However, long-term prognostic assessment, close follow-up and better study design are needed in future studies.

CONCLUSION

Erythroderma is a dermatological clinical syndrome of generalized erythema and scaling, due to various etiologies. Although the causes may be diverse, most cases of erythroderma have a preexisting skin disease. Patients with previously normal skin often have drug-induced erythroderma or malignancy. Indeed, its early diagnosis allows a fast and adequate care thus improving the prognosis of these entities. In the uncommon cases were no underlying cause is found, close follow-up is recommended. Early diagnosis for skin diseases in our country avoids consultation at advanced stages as erythroderma, and decrease the frequency of erythroderma and its complications in our context.

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.

REFERENCES

1 Artur Cesar, Maria Cruz, Alberto Mota, Filomena Azevedo. Erythroderma. A clinical and etiological study of 103 patients. J Dermatol Case Rep. 2016;10:1–9.

2 Ndiaye M, Ly F, Dioussé P, Diallo M, Diop A, Diatta BA, et al. [The characteristics of severe forms of psoriasis on pigmented skins: A retrospective study of 102 cases in Dakar, Senagal]. Our Dermatol Online. 2017;8:138-42.

3 Caroline Ram-Wolff, Nadine Martin-Garcia,Armand Bensussan, Martine Bagotand Nicolas Ortonne. Histopathologic diagnosis of lymphomatous versus inflammatory erythroderma: a morphologic and phenotypic study on 47 skin biopsies.Am J Dermatopathol. 2010;32:755–63.

4 Nupur P, Singh R. Erythroderma: a clinico – etiological study in a tertiary hospital. Int J Scien Res. 2018;7:8179.

5 Khaled A, Sellami A, Fazaa B, Kharfi M, Zeglaoui F, Kamoun MR. Acquired erythroderma in adults: a clinical and prognostic study. J Eur Acad Dermatol Venereol. 2010;24:781-8.

6 Diallo M, Diadie S, Diatta BA, Diop A, Ndiaye MT, Ndiaye M, et al. Acquired erythroderma in adults in senegal: epidemiological and etiological aspects. Dermatol Case Rep. 2017;2:128.

7 Tan GFL, Kong YL, Tan ASL,Tey HL. Causes and Features of Erythroderma. Ann Acad Med Singapore. 2014;43:391-4.

8 Sehgal VN, Srivastava G, Sardana K, MNAMS. Erythroderma /exfoliative dermatitis: a synopsis. Int JDermatol. 2004;43:39–47.

9 Rym BM, Mourad M, Bechir Z, Dalenda E, Faika C, Iadh AM, et al. Erythroderma in adults: a report of 80 cases. Int J Dermatol. 2005;44:731-5.

10 Fernandes NC, Pereira FSM, Maceira JP, Cuzzi T, Dresch TFLR, Araújo PP. Eritrodermia: estudoclínico-laboratorial e histopatológico de 170 casos. An Bras Dermatol. 2008;83:532-36.

11 Yuan XY, Guo JY, Dang YP, Qiao L, Liu W. Erythroderma: A clinical-etiological study of 82 cases. Eur J Dermatol. 2010;20:373-7.

12 Li J, Zheng HY. Erythroderma: a clinical and prognostic study. Dermatology. 2012;225:154-62.

13 Hulmani M, Nandakishore B, Bhat MR, Sukumar D, Martis J, Kamath G, et al. Clinico-etiological study of 30 erythroderma cases from tertiary center in South India. Indian Dermatol Online J. 2014;5:25-9.

14 Akhyani M, Ghodsi ZS, Toosi S, Dabbaghian H. Erythroderma: a clinical study of 97 cases. BMC Dermatol. 2005;5:5.

15 Rothe MJ, Bernstein ML, Grant-Kels JM Life-threatening erythroderma: diagnosing and treating the "red man". Clin Dermatol. 2005;23:206-17.

16 Eugster R, Kisling S, Brand CU. Clinical aspects and aetiology of erythroderma: an analysis of 64 cases. ScweizRundscMed Prax. 2001;90:1449–54.
17 Mistry N, Gupta A, Alavi A, Sibbald RG. A review of the diagnosis nd management of erythroderma (generalized red skin). Adv Skin Wound Care. 2015;28:228-36; quiz 237-8.
18 Maldonado-García CA, Orozco-Anahuatib AP. Abordajediagnóstico de la eritrodermia en el adulto. Rev Med Inst Mex Seguro Soc. 2017;55:353-60.
19 Shirazi N, Jindal R, Jain A, Yadav K, Ahmad S. Erythroderma: A clinico-etiological study of 58 cases in a tertiary hospital of North India. Asian J Med Scien. 2015;6:20-4.

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Our Dermatology Online

Original Article

Neutrophil-lymphocyte ratio and platelet-lymphocyte ratio as an alternative to C-reactive protein in diagnostics of inflammatory state in patients with psoriasis

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ABSTRACT

Background: The ratio of neutrophils to lymphocytes (NLR), the ratio of platelets to lymphocytes (PLR), the ratio of monocytes to lymphocytes (MLR), and mean platelet volume (MPV) are considered novel inflammatory markers. We stated a hypothesis that apart from C-reactive protein (CRP) level, also NLR, PLR, MLR, and MPV are higher in patients with psoriatic arthritis (PsA, where both skin and joints are involved) than in patients with psoriasis vulgaris (PsV). **Material and Methods**: Our study is based on a retrospective analysis. We collected laboratory data, namely CRP levels as well as a total blood count, and calculated the following additional parameters: NLR, PLR, MLR, and MPVLR (mean platelet volume/lymphocyte ratio). For all data, two groups of patients were compared: PsV with N=80 and PsA with N=80. **Results**: CRP levels were significantly higher in PsA patients compared with those in PsV patients (median PsV – 19.43 nmol/L vs median PsA – 37.90 nmol/L, p=0.001). Similarly, both NLR and PLR were higher in PsA patients than in PsV patients (NLR, median PsV – 2.0 vs median PsA – 2.28, p=0.030; PLR, median PsV – 111.61 vs median PsA – 121.85, p=0.027). Moreover, for patients with psoriatic arthritis a weak to moderate positive correlation between C-reactive protein levels and WBC, neutrophils count, monocytes count, platelets count, NLR, PLR, and MLR was observed. **Conclusions:** CRP levels are higher in patients with PsA than in patients with PsV, which can be helpful in predicting arthritis in patients with psoriasis. Furthermore, similar information can be obtained from a blood count. In particular, NLR and PLR are simple predictors which can indicate ongoing joint inflammation in patients with psoriasis, hence they can be used as an alternative to the CRP level.

Key words: Psoriasis; Psoriatic arthritis; Inflammation; Neutrophil-lymphocyte ratio; Platelet-lymphocyte ratio; Low-cost test

INTRODUCTION

Psoriasis is a chronic immune-mediated inflammatory disease which affects around 2–4% of the population [1]. Occurrence of psoriasis is believed to be associated with both environmental and genetic factors [2,3]. Psoriasis vulgaris (PsV), or chronic plaque psoriasis, is the most common type of psoriasis and affects approximately 90% of patients with psoriasis. In psoriatic arthritis (PsA), there is an inflammation of joints, usually coexisting with a cutaneous manifestation of psoriasis. PsA occurs in up to 30% of patients with psoriasis [4]. Psoriasis is frequently associated with significant comorbidities including diabetes mellitus, obesity, dyslipidemia, inflammatory bowel disease, psychiatric disorders, osteoporosis, and obstructive sleep apnea, as well as cardiovascular diseases, e.g. hypertension, myocardial infarction, and stroke [5-7].

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Multiple investigations were conducted with the goal of finding useful biomarkers within peripheral blood samples of patients with psoriasis and psoriatic arthritis. Unfortunately, they have not found a single, simple and clinically useful biomarker which could be specific for psoriasis [8]. On the other hand, C-reactive protein (CRP) level, which is a known inflammatory blood test marker of inflammation in the body, is elevated among psoriatic patients and can be used to assess disease severity and progression [9]. Recently, the ratio of neutrophils to lymphocytes (NLR), the ratio of platelets to lymphocytes (PLR), and the ratio of monocytes to lymphocytes (MLR) have been recognized as inflammatory markers [10-12]. NLR, PLR, and MLR are assessed as a prognostic of poor neoplastic disease outcome [13-17], whereas mean platelet volume (MPV) has been studied as an inflammatory marker in several diseases [18-20]. There are also a few studies which investigate simple markers, viz. NLR, PLR, and MPV in patients with psoriasis [21-24].

In this study, we analyzed, i.a., CRP level, NLR, PLR, MLR, and MPV in Polish patients with psoriasis. In particular, we put forward a hypothesis that patients with psoriatic arthritis have higher inflammatory biomarker levels than patients with psoriasis vulgaris.

MATERIALS AND METHODS

The study was carried out at the Department of Dermatology, Pediatric Dermatology and Oncology Clinic, Medical University of Lodz, Poland. It was a retrospective analysis, conducted for Polish patients with psoriasis vulgaris and psoriatic arthritis, hospitalized from January to December 2017. Inclusion criteria covered patients who were from 18 up to 90 years of age and diagnosed with psoriasis at least 1 year before the test was recorded. Exclusion criteria comprised a current or recent treatment with biological therapies, known inflammatory processes in the body (except psoriasis), and present history of a neoplastic disease. The study group consisted of 80 patients with psoriasis vulgaris (group 1) and 80 patients with psoriatic arthritis (group 2). All patients included in group 1 had present skin changes, and all patients included in group 2 had both skin and joints manifestations of the disease.

The following information was collected: C-reactive protein (CRP) level, red blood cells count (RBC), haemoglobin (Hb), white blood cells count (WBC), lymphocytes, neutrophils, monocytes and platelets counts, and mean platelet volume (MPV). These blood morphology results (UniCel DxH 800 Coulter Cellular Analysis System, Beckman Coulter Inc, USA) and CRP concentration (particle-enhanced turbidimetric method, Cobas 6000 Analyzer, Roche, Germany) were obtained from the hospital laboratory. On the basis of blood morphology data, the following markers (indexes) were calculated: PLR (platelet/lymphocyte ratio), NLR (neutrophil/lymphocyte ratio), MLR (monocyte/lymphocyte ratio), and MPVLR (mean platelet volume/lymphocyte ratio).

Statistical analysis was performed with the SciPy (v. 1.1.0) suite for the Python programming language. The Shapiro-Wilk test was used to evaluate the normality of data distribution, and Brown-Forsythe test was employed to check for variance homogeneity. Continuous parameters are described using medians and the interquartile range, with the Mann-Whitney U test used for significance testing. The calculated sample size for a power of at least 0.8 and significance of at least 0.05 was equal to 80 patients in a group. The correlation between parameters was determined with the use of the Spearman coefficient.

Ethics Statement

The authors state no conflict of interest. Data used in this study was collected retrospectively and it was fully anonymous.

RESULTS

A total of 160 patients diagnosed with psoriasis – 80 patients with PsV (45 males, 35 females; mean age 48.48±16.08 years; range 19–83 years) and 80 patients with PsA (46 males, 34 females; mean age 49.45±13.74 years; range 24–88 years) were included in the study. Complete blood count data, C-reactive protein level, NLR, PLR, MLR, and MPVLR results are presented in Table 1. One can observe that CRP levels were significantly higher in PsA patients when compared with those in PsV patients, and it was indeed the strongest predictor for the presence of arthritis in psoriasis patients. Moreover, both NLR and PLR were higher in PsA patients than in PsV patients. A statistically significant difference was also observed in terms of white blood cells, neutrophils and platelets counts, as well haemoglobin level between patients with psoriasis vulgaris and psoriatic arthritis. Let us

Table 1: Marker comparison between patients with psoriasis vulgaris (PsV) and psoriatic arthritis (PsA)

Parameter	PsV (N=80) Median (IQR)	PsA (N=80) Median (IQR)	p-value
C-reactive protein [nmol/L]	19.43 (9.52–36.76)	37.90 (16.76–60.00)	**0.001**
Red blood cells [x 10^{12}/L]	4.67 (4.31–5.00)	4.71 (4.35–5.12)	0.231
Haemoglobin [g/L]	147.5 (139.0–155.1)	143.5 (133.0–154.0)	**0.040**
White blood cells [x 10^9/L]	6.7 (5.5–8.0)	7.5 (6.0–9.0)	**0.017**
Lymphocytes [x 10^9/L]	1.9 (1.7–2.3)	2.0 (1.5–2.3)	0.447
Neutrophils [x 10^9/L]	3.85 (2.79–4.58)	4.35 (3.20–5.75)	**0.016**
Monocytes [x 10^9/L]	0.6 (0.5–0.7)	0.6 (0.5–0.8)	0.084
Platelets [x 10^9/L]	212.5 (192.4–259.5)	233.5 (197.4–298.0)	**0.034**
Mean platelet volume [fL]	9.25 (8.54–9.86)	9.30 (8.54–10.10)	0.242
Neutrophil to lymphocyte ratio	2.00 (1.43–2.49)	2.28 (1.61–3.23)	**0.030**
Platelet to lymphocyte ratio	111.61 (91.45–138.45)	121.85 (93.87–164.38)	**0.027**
Mean platelet volume to lymphocyte ratio	4.87 (3.89–5.66)	4.81 (3.97–5.81)	0.418
Monocyte to lymphocyte ratio	0.32 (0.24–0.39)	0.33 (0.24–0.47)	0.120

Continuous data are presented as a median with the interquartile range (IQR, lower quartile–upper quartile) in parentheses. Data was not normally distributed and significance was analyzed using the Mann-Whitney U test. P-value <0.05 was considered statistically significant (emphasized with the bold font).

also note that these differences between the two groups were in general less statistically significant for older groups of patients (e.g., born before 1970).

Other parameters, including RBC, lymphocytes, monocytes, MPV, MPVLR, and MLR were either similar or the difference between the two groups was not statistically significant. We also analyzed the Spearman correlation between the C-reactive protein level and other parameters, with results presented in Table 2. For PsA patients we observed a weak to moderate positive correlation with monocytes count (r_s=0.238, p=0.033), WBC (r_s=0.3, p=0.007), MLR (r_s=0.314, p=0.005), platelets count (r_s=0.372, p=0.001), neutrophils count (r_s=0.395, p<0.001), PLR (r_s=0.395, p<0.001), and NLR (r_s=0.439, p<0.001). Hence, the highest correlation coefficient for patients with psoriatic arthritis was discovered for neutrophils count, PLR, and NLR. Furthermore, we received negative statistically significant correlation between CRP and haemoglobin levels, lymphocytes counts, and MPV for PsA patients. In patients with psoriasis vulgaris the correlation between the aforementioned CRP level and other parameters was generally not statistically significant, with the sole exception of platelets count.

DISCUSSION

We searched for promising markers which might be useful for the prediction of arthritis in psoriasis patients. Let us reiterate that serum C-reactive protein level is a popular inflammatory biomarker, which might be employed for this task. Besides CRP, there exist other markers in psoriatic patients such as IL-6, TNF-α, E-selectin, or ICAM-1, however, they are costly to obtain and hence not commonly tested [25]. On the other hand, in this study we concentrated on parameters from routine blood tests and found that WBC, neutrophils, monocytes and platelets counts, NLR, as well as PLR were indeed higher for patients with PsA when compared to patients with PsV.

According to Solak et al. [21], both CRP level and NLR are promising predictors of inflammation in psoriasis (compared with the control group). In their study, PLR was not assessed and psoriatic arthritis was not considered. Polat et al. [22] evaluated CRP, NLR, and PLR in Turkish patients, which turned out to be significantly higher in people with PsV than in the control group. Kim et al. [23] compared parameters from routine blood tests in Korean patients with psoriasis vulgaris and psoriatic arthritis. Our results are in general consistent with their study, since they observed statistically significant differences in terms of CRP, WBC, neutrophils, platelets counts, NLR and PLR. Similarly to this work, both NLR and PLR were strong predictors for the presence of arthritis among psoriasis patients. On the other hand, Kim et al. reported monocytes counts to be significantly higher in PsA patients, a scenario which we did not observe. Asahina et al. [24] described a correlation between CRP and parameters including WBC, neutrophils, lymphocytes, platelets, MPV, NLR, and PLR in Japanese patients with psoriasis. Interestingly, they described significant correlations not only for PsA patients as in our study, but also for PsV patients. Canpolat et al. [26] investigated MPV in patients with psoriasis vulgaris, psoriatic arthritis, and healthy subjects. In their study, MPV was higher in patients with PsA than in patients with PsV, however, in our study MPV did not differ between the groups. In another note, mean platelet volume was regarded as

Table 2: Spearman correlation coefficient between C-reactive protein level and other parameters for patients with psoriasis vulgaris (PsV) and psoriatic arthritis (PsA)

Parameter	rs PsV, N=80	95% CI PsV	p-value PsV	rs PsA, N=80	95% CI PsA	p-value PsA
White blood cells	0.184	[-0.038, 0.388]	0.103	0.300	[0.086, 0.487]	**0.007**
Lymphocytes	0.025	[–0.196, 0.243]	0.826	–0.241	[–0.437, –0.022]	**0.031**
Neutrophils	0.177	[–0.044, 0.382]	0.116	0.395	[0.192, 0.566]	**<0.001**
Monocytes	0.104	[–0.119, -0.316]	0.359	0.238	[0.019, 0.435]	**0.033**
Platelets	0.230	[0.010, 0.428]	0.041	0.372	[0.166, 0.547]	**0.001**
Mean platelet volume	–0.169	[-0.375, 0.053]	0.134	–0.272	[–0.464, –0.055]	**0.015**
Neutrophil to lymphocyte ratio	0.103	[–0.119, 0.316]	0.363	0.439	[0.242, 0.600]	**<0.001**
Platelet to lymphocyte ratio	0.211	[-0.009, 0.412]	0.060	0.395	[0.192, 0.566]	**<0.001**
Mean platelet volume to lymphocyte ratio	–0.110	[–0.322, 0.113]	0.332	0.129	[–0.094, 0.339]	0.255
Monocyte to lymphocyte ratio	0.135	[–0.087, 0.345]	0.232	0.314	[0.101, 0.499]	**0.005**

Spearman correlation coefficient rs with a two-sided 95% confidence interval (CI). P-value <0.05 was considered statistically significant (emphasized with the bold font).

an inappropriate indicator of inflammation, which is again consistent with our results [27].

Let us also address the limitations of this study. The analysis was conducted retrospectively in a single university hospital. Even though we excluded patients who had a history of one or more significant inflammatory processes (except psoriasis), information regarding comorbidities might be incomplete, with a possible external influence on inflammatory biomarkers. Moreover, described parameters might be altered by many conditions which are difficult to control, such as dehydration or overhydration. We also did not consider skin lesions severity (which can be assessed, e.g. using Psoriasis Area and Severity Index).

CONCLUSIONS

Based on the obtained results, we regard CRP, NLR, and PLR as promising candidates for predictors of arthritis among patients with psoriasis. In particular, they were observed to be higher in patients with PsA than in patients with PsV. Hence, for the assessment of inflammation, the blood count might be sufficient, even without the need for a more complex CRP test. Moreover, higher white blood cell, neutrophils, or platelet counts might suggest a developing arthritis in people with psoriasis. These biomarkers are simple, easily accessible, and relatively inexpensive to obtain.

For achieving better results when it comes to psoriatic arthritis treatment, it is significant to begin the treatment early. Thanks to these aforementioned, easily obtainable biomarkers, chances to predict which patients with psoriasis are developing arthritis might be boosted. Furthermore, increased CRP level, NLR, and PLR indicate a greater inflammatory burden in patients with psoriatic arthritis compared to patients with psoriasis vulgaris. The elevation of inflammatory markers in patients with psoriasis could be associated with, for instance, increased cardiovascular risk. For this reason, patients with psoriasis who have high CRP level, NLR, or PLR, should be followed by a dermatologist more carefully, although more studies are required to show how these values could be used in clinical practice.

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.

REFERENCES

1. Parisi R, Symmons DP, Griffiths CE, Ashcroft DM. Global epidemiology of psoriasis: a systematic review of incidence and prevalence. J Invest Dermatol. 2013;133:377-85.
2. Herron MD, Hinckley M, Hoffman MS, Papenfuss J, Hansen CB, Callis KP, et al. Impact of obesity and smoking on psoriasis presentation and management. Arch Dermatol. 2005;141:1527-34.
3. Gruchala A, Gawrońska M, Galica K, Kaszuba A. Genetyczne aspekty łuszczycy i łuszczycowego zapalenia stawów [in Polish; Genetic aspects of psoriasis and psoriatic arthritis]. Dermatol Prakt. 2018;2:35-40.
4. Zachariae H. Prevalence of joint disease in patients with psoriasis: implications for therapy. Am J Clin Dermatol. 2003;4:441-7.
5. Sanchez-Carazo JL, López-Estebaranz JL, Guisado C. Comorbidities and health-related quality of life in Spanish patients with moderate to severe psoriasis: a cross-sectional study (Arizona study). J Dermatol. 2014;41:673-8.
6. Xu T, Zhang YH. Association of psoriasis with stroke and myocardial infarction: meta-analysis of cohort studies. Br J Dermatol. 2012;167:1345-50.
7. Machado-Pinto J, Diniz Mdos S, Bavoso NC. Psoriasis: new comorbidities. An Bras Dermatol. 2016;91:8-14.
8. Villanova F, Di Meglio P, Nestle FO. Biomarkers in psoriasis and

psoriatic arthritis. Ann Rheum Dis. 2013;72 Suppl 2:ii104-10.
9. Beygi S, Lajevardi V, Abedini R. C-reactive protein in psoriasis: a review of the literature. J Eur Acad Dermatol Venereol. 2014;28:700-11.
10. Balta S, Celik T, Mikhailidis DP, Ozturk C, Demirkol S, Aparci M, et al. The relation between atherosclerosis and the neutrophil-lymphocyte ratio. Clin Appl Thromb Hemost. 2016;22:405-11.
11. Özer S, Yılmaz R, Sönmezgöz E, Karaaslan E, Taşkın S, Bütün İ, et al. Simple markers for subclinical inflammation in patients with Familial Mediterranean Fever. Med Sci Monit. 2015;21:298-303.
12. Ji H, Li Y, Fan Z, Zuo B, Jian X, Li L, et al. Monocyte/lymphocyte ratio predicts the severity of coronary artery disease: a syntax score assessment. BMC Cardiovasc Disord. 2017;17:90.
13. Janik S, Raunegger T, Hacker P, Ghanim B, Einwallner E, Müllauer L, et al. Prognostic and diagnostic impact of fibrinogen, neutrophil-to-lymphocyte ratio, and platelet-to-lymphocyte ratio on thymic epithelial tumors outcome. Oncotarget. 2018;9:21861-75.
14. Templeton AJ, McNamara MG, Šeruga B, Vera-Badillo FE, Aneja P, Ocaña A, et al. Prognostic role of neutrophil-to-lymphocyte ratio in solid tumors: a systematic review and meta-analysis. J Natl Cancer Inst. 2014;106:dju124.
15. Fan W, Zhang Y, Wang Y, Yao X, Yang J, Li J. Neutrophil-to-lymphocyte and platelet-to-lymphocyte ratios as predictors of survival and metastasis for recurrent hepatocellular carcinoma after transarterial chemoembolization. PLoS One. 2015;10:e0119312.
16. Zhou WJ, Wu J, Li XD, Wang Q, Ni XF, Jiang JT, et al. Effect of preoperative monocyte-lymphocyte ratio on prognosis of patients with resectable esophagogastric junction cancer. Zhonghua Zhong Liu Za Zhi. 2017;39:178-83.
17. Tanrikulu AC, Abakay A, Komek H, Abakay O. Prognostic value of the lymphocyte-to-monocyte ratio and other inflammatory markers in malignant pleural mesothelioma. Environ Health Prev Med. 2016;21:304-11.
18. Gasparyan AY, Ayvazyan L, Mikhailidis DP, Kitas GD. Mean platelet volume: a link between thrombosis and inflammation? Curr Pharm Des. 2011;17:47-58.
19. Şenel E, Acar B, Demir E. Mean Platelet Volume: A reliable marker of inflammation in recurrent apthous stomatitis and Behçet disease? Indian Dermatol Online J. 2017;8:468-70.
20. Safak S, Uslu AU, Serdal K, Turker T, Soner S, Lutfi A. Association between mean platelet volume levels and inflammation in SLE patients presented with arthritis. African Health Scien. 2014;14:919 24.
21. Solak B, Dikicier BS, Erdem T. Impact of elevated serum uric acid levels on systemic inflammation in patients with psoriasis. Angiology. 2017;68:266-70.
22. Polat M, Bugdayci G, Kaya H, Oğuzman H. Evaluation of neutrophil-to-lymphocyte ratio and platelet-to-lymphocyte ratio in Turkish patients with chronic plaque psoriasis. Acta Dermatovenerol Alp Pannonica Adriat. 2017;26:97-100.
23. Kim DS, Shin D, Lee MS, Kim HJ, Kim DY, Kim SM, et al. Assessments of neutrophil to lymphocyte ratio and platelet to lymphocyte ratio in Korean patients with psoriasis vulgaris and psoriatic arthritis. J Dermatol. 2016;43:305-10.
24. Asahina A, Kubo N, Umezawa Y, Honda H, Yanaba K, Nakagawa H. Neutrophil-lymphocyte ratio, platelet-lymphocyte ratio and mean platelet volume in Japanese patients with psoriasis and psoriatic arthritis: response to therapy with biologics. J Dermatol. 2017;44:1112-21.
25. Dowlatshahi EA, van der Voort EA, Arends LR, Nijsten T. Markers of systemic inflammation in psoriasis: a systematic review and meta-analysis. Br J Dermatol. 2013;169:266-82.
26. Canpolat F, Akpinar H, Eskioğlu F. Mean platelet volume in psoriasis and psoriatic arthritis. Clin Rheumatol. 2010;29:325-8.
27. Leader A, Pereg D, Lishner M. Are platelet volume indices of clinical use? A multidisciplinary review. Ann Med. 2012;44:805-16.

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Our Dermatology Online

Original Article

Omega-3 fatty acids and quality of life in psoriasis: An open, randomised controlled study

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ABSTRACT

Background: Omega-3 fatty acids have been reported to reduce disease severity in psoriasis in various studies. This study aims to study the effects of omega-3 fatty acid supplementation on the quality of life in psoriasis.**Methods:** This open, non blinded interventional study divided patients of psoriasis with affected body surface area of less than <10% into two groups of 100 patients each. Group A was given topical paraffin with 1.8 grams of omega-3 fatty acids daily in three divided doses. Group B was advised topical paraffin application once a day. Oral antihistaminics were added as per need. Baseline patient characteristics like age, sex and severity of psoriasis by Psoriasis Area Severity Index and Dermatology Life Quality Index was assessed. The patients were followed up for 12 weeks and change in DLQI was noticed.**Results:** Baseline DLQI in group A changed by 2.96 points from 11.47 ± 3.90 to 8.51 ± 3.36. (p <0.00001) and by 1.27 points from 11.69 ± 3.75 to 10.42 ± 3.61 in group B (p<0.00001). The change in the DLQI between the two groups was statistically significant (p=0.0002).**Conclusion:** Omega-3 fatty acids appear to produce a statistically significant improvement in the quality of life in psoriasis patients.

Key words: Fish oils; Omega-3 fatty acids; Psoriasis

INTRODUCTION

Psoriasis is a chronic, immune mediated inflammatory disease characterised by well defined red scaly plaques over the extensors of body and the scalp [1]. The disease is chronic and marked by remissions and exacerbations. The clinical disease occurs due to activation of the Th1 immune response with a complex interaction of various inflammatory cells like lymphocytes, neutrophils, macrophages and dendritic cells with epidermal keratinocytes, leading to the production of various signalling molecules [2]. Psoriasis adversely impacts psychosocial well being and quality of life and may lead to self consciousness, embarrassment, social isolation and stigmatisation, leading to depression and decreased work productivity [3].

The role of diet and nutrition in the pathogenesis and management of the disease is now well established [4]. The greatest evidence in this respect has come with the use of omega-3 fatty acid supplementation [5]. Eicosapentaenoic acid (EPA) and Docosahexaenoic acid (DHA) are omega-3 fatty acids chiefly obtained from algae and marine fish. Omega-3 fatty acids lower the production of arachidonic acid derived pro inflammatory cytokines like LTB4, C4 etc. by metabolising through the same pathway as arachidonic acid and itself produces LTB5, a less potent keratinocyte stimulator and ten fold less strong chemotactic metabolite for neutrophils than LTB4 [6]. They also lead to an increased production of nitric oxide and decreased levels of Tumour necrosis factor alpha, IL-1 and other cytokines in the endothelium

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preventing vascular changes seen in psoriasis [7]. Altered intracellular signalling pathway, regulation of transcription factor activity and antioxidant effects have also been attributed to omega-3 fatty acids for their effect in psoriasis [8].

This study aims to find the beneficial effect of oral omega-3 fatty acids in the quality of life in patients with psoriasis.

METHODS

This prospective, open study was conducted in the department of Dermatology for a period of 2 years from March 2013 to March 2015. Ethical clearance from the institutional review board was obtained prior to the study. 100 clinically diagnosed cases of mild to moderate chronic plaque psoriasis attending the Out patient department of dermatology of the hospital (with involvement of <10% body surface area) [9] were selected and formed the study group (group A). All patients with age of 15 years to 50 years were included in the study. Patients with erythrodermic psoriasis, pustular psoriasis, palmoplantar psoriasis, pregnant and lactating women, patients with bleeding disorders, diabetes mellitus or a history of stroke or those having secondarily infected lesions were excluded from the study. Patients who were receiving any systemic therapy or phototherapy were included only after 4 weeks of washout period was passed. The same number of age and sex matched patients of chronic plaque psoriasis with <10% body surface involvement were taken as control (group B). Informed consent was given by all patients recruited for the study. Data was collected regarding the age, sex, duration of disease, age at onset and prior treatment taken. Group A was advised daily intake of 6 capsules of fish oils in three divided doses, with each capsule having 300mg of EPA and DHA (a total of 1.8 gram of omega-3 fatty acids a day). This treatment was continued for 3 months. Daily application of paraffin was advised. Antihistamines were prescribed as per need. The group B was advised daily topical paraffin therapy and antihistamines for 3 months.

Outcome of the study was measured by the change in Dermatology Life Quality Index (DLQI) score at the end of study at 12 weeks as compared to the baseline value. DLQI is a dermatology specific instrument for measuring health related quality of life index. It is a simple, 10 question validated questionnaire intended for use in adults (over 16 years of age). It is self explanatory and requires less than two minutes for completion. DLQI is calculated as the summation of the score of each question with a minimum marks of 0 and a maximum of 30 [10]. DLQI has been found to be reliable, valid, simple and easy to use [11]. Mattei et al evaluated 155 sources of randomised control trials and concluded that Psoriasis Area Severity Score (PASI) and DLQI scores are predictably correlated in patients of moderate to severe psoriasis undergoing treatment with biologicals [12].

Statistical analysis of the results regarding mean, standard deviation, percentage and tests of significance (paired and unpaired t tests and chi square tests) was done with the help of Statistical Package for Social Sciences version 16.0.

RESULTS

Both the groups in the trial consisted of 100 patients each. There were 61 males and 39 females in group A and 65 males and 35 females in group B. The mean age of patients in group A was 35.99 years with a standard deviation of 9.70 years. The group B had a mean age of 36.81 years and a standard deviation of 9.26 years. The two groups were comparable in statistical terms as far as the sex and age of the patients was concerned. The mean Psoriasis Area Severity Index (PASI) score of the two groups at baseline was also statistically not significant. The PASI score at baseline was 7.44 with a standard deviation of 2.84 for group A and 7.24 ± 2.99 for group B. After a drop out of 5 patients in group A and 7 patients in group B, 95 patients and 93 patients completed the study from the two groups respectively (Table 1). These findings have already been published in a paper by our group [13].

The Dermatology Life Quality Index for group A was 11.47 with a standard deviation of 3.90 points. This was statistically similar to the DQLI score for group B, which was 11.69 ± 3.75. The DQLI score at the end of the therapy decreased to 8.51 and with a standard deviation of 3.36 for group A and 10.42 ± 3.61 for

Table 1: Epidemiological characteristics of the patients

	Group A	Group B	p value
Male	65	61	0.5580
Female	35	39	
Mean age (in years)	35.99±9.70	36.81±9.26	0.5416
Group outs	5	7	0.5515
Mean baseline PASI score	7.44±2.84	7.24±2.99	0.6387

group B. There is a significant difference between the DLQI scores of the two groups at the end of therapy at 12 weeks. The decrease in DLQI in group A was 2.96 points or 25.81%. Group B showed a decease in DLQI by 1.27 points or 10.86% (Table 2). Both the groups showed statistically significant improvements in the quality of life at the end of therapy when compared to baseline. 15 patients achieved a 75% or more reduction in PASI (PASI 75) in group A and 2 patients reached PASI 75 in group B. None of the patients experienced any adverse effect of the treatment except for fishy odor in eructation in 5 patients and slight abdominal discomfort after taking capsules in one patient. None of the patients discontinued treatment due to side effects.

DISCUSSION

Both the groups in our study were statistically comparable in terms of age, sex and severity of psoriasis assessed objectively by the Psoriasis Area Severity Index carried out by the same assessor in a patient. There was a strong male predominance in our study, with males constituting more than 60% of both the groups. Studies from the western countries show an equal predilection for both sexes [14]. This is in contrast to previous studies from Asia where a male predisposition for the disease is seen [15]. Mabuchi et al postulated that this difference might at least partly be explained by the environmental factors such as diet, smoking and alcohol, which appear to be more common in males [16]. We observed that the mean age of patients in both the groups was around 35 years of age. Our findings are in concordance with other studies showing the mean age to be in the thirties [17]. The mean PASI scores of the two groups were 7.44 and 7.24 in the two groups. It is noteworthy that patients with mild to moderate severe psoriasis (body surface area of less than 10%) [9] were included in the study. Other studies also show similar PASI scores in patients of mild to moderate psoriasis [18,19].

The DLQI in our patients was more than 11 in both the groups, signifying severe impairment of quality of life. Psoriasis patients have been shown to have a large impact on the quality of life, comparable to those with diabetes mellitus or ischemic heart disease [20]. As much as a third of all psoriasis patients suffer from pathological worry and anxiety. Around 5% have been reported to develop suicidal ideation [21]. A reliable correlation between the severity of psoriasis measured by the physician via PASI and subjectively by the patient via DLQI only exists when a large reduction of PASI score, usually by more than 75% occurs [22], problems in public and sexual interactions do not correlate with the severity of psoriasis [23]. The feeling of stigmatisation, associated co-morbidities, social withdrawal exerts an adverse effect on the educational and professional life of patients, leading to decreased quality of life [24].

Table 2: Change in Dermatological Life Quality Index (DLQI)

DLQI Score	Group A	Group B	p value
Baseline	11.47±3.90	11.69±3.75	0.6939
At 12 weeks	8.51±3.36	10.42±3.61	0.0002
p value	<0.00001	<0.00001	

The DLQI decreased significantly in both the groups. Group A showed a statistically significant greater decline in DLQI than the group B. It improved by around 25% in group A, but the decrease was a little less than 11% in group B. Several trials have evaluated the beneficial role of omega-3 fatty acids in psoriasis taking objective measures as markers of improvement, the measures being PASI, erythema, scaling, area involved etc [25]. We could find just one trial evaluating the effect of omega 3 fatty acids in psoriasis patients' quality of life. This trial was conducted by Balbas et al, where 15 patients were advised topical tacalcitol with 640mg of omega-3 fatty acid capsules daily for 2 months. The control group was prescribed only tacalcitol for topical application. Statistically significant improvement was seen in DLQI with the test group showing an improvement of more than 6 points (78.19%) and the control group showing a 3 point improvement (53.44%) [26]. The results of our study are not as encouraging as those of Balbas et al despite using a higher dose of omega 3 fatty acids. The difference may be due to the topical agents used in the two studies. Our patients used paraffin and Balbas et al used tacalcitol, a topical vitamin D analogue.

Basra et al proposed that there should be a change in DLQI by at least 4 points to interpret it as a clinically significant change and produce a meaningful change in quality of life for the patient [27]. This means that omega 3 fatty acid supplementations failed to produce a clinically significant improvement in psoriasis in our trial, as did the group with topical emollients. The reason for this might be the low number of patients achieving PASI 75 at the end of study, indicating an incomplete clearance of the disease in most patients. Only 15 patients in group A and 2 patients in group B had achieved PASI 75 after 12 weeks. Reduction in

PASI by 75% is accepted as a reliable indicator of clinical response and has been extensively used in clinical trials to gauze the efficacy of newer drugs [28]. Several authors have argued that greater reductions in PASI lead to improved quality of life as reported by the patients, with greatest reductions in quality of life parameters reported with almost complete clearance of lesions of psoriasis [29,30].

The limitations of our study included its design which was an open, non blinded trial. This predisposed to bias by both the patients as well as the assessors. The study recruited patients only with less than 10% body surface area involved. Patients of more severe psoriasis were excluded as were patients with palmoplantar psoriasis who have a small area involved but have a much greater impairment of quality of life. This study used DLQI as an assessment tool for measuring quality of life. Several newer assessment tools are now available such as Skindex, Salford Psoriasis Index and SF-36.

CONCLUSION

This study shows that omega-3 fatty acids and emollients produce a statistically significant improvement in patient reported quality of life measures in psoriasis. This improvement is only modest and may not be clinically apparent. Nevertheless, omega 3 fatty acids have a beneficial effect on other chronic diseases including ischemic heart disease, which is associated with psoriasis. These are largely safe and free of side effects. Thus, omega 3 fatty acids may be used in conjunction to other treatments in patients of psoriasis. However, more studies are needed to accurately assess the role of omega-3 fatty acids in psoriasis in improving quality of life as well as to arrive to an appropriate dosage for supplementation.

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.

REFERENCES

1. Griffiths C, Barker JN. Psoriasis. In: Burns T, Brethnach S, Cox N, Griffiths C, editors. Rook's textbook of dermatology. 8th ed. West Sussex: Blackwell Publishing; 2010. p.1-22.
2. Gudjonsson JE, Elder JT. Psoriasis. In: Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffel DJ, Wolff K, editors. Fitzpatrick's dermatology in general medicine. 8th ed. New York: McGraw Hill; 2012. p. 197-224.
3. Armstrong AW, Schupp C, Wu J, Bebo B. Quality of Life and Work Productivity Impairment among Psoriasis Patients: Findings from the National Psoriasis Foundation Survey Data 2003–2011. PLoS One. 2012;7:e52935.
4. Araujo MLD, Burgos MGAP, Moura ISCM. Nutritional influences in psoriasis. An Bras Dermatol. 2009;84:90-2.
5. Millsop JW, Bhatia BK, Debbaneh M, Koo J, Liao W. Diet and psoriasis, part III: role of nutritional supplements. J Am Acad Dermatol. 2014;71:561-9.
6. Ricketts JR, Rothe MJ, Grantkels JM. Nutrition and Psoriasis. Clin Dermatol. 2010;28:615-26.
7. Traub M, Marshall K. Psoriasis– pathophysiology, conventional and alternative approaches to treatment. Alt Med Rev. 2007;12:319-30.
8. Passi S, de Pita O, Cocchi M. Psoriasis and diet. Progress Nutrition. 2004;6:231-47.
9. Assessing a psoriasis patient. In: Voorhees AV, Feldman SR, Koo JYM, Lebwohl MG, Menter A, editors. Psoriasis and psoriatic arthritis pocket guide: treatment algorithms and management guide. 3rd ed. Portland: National Psoriasis Foundation; 2009. p 9-44.
10. Finley AY, Khan GK. Dermatology Life Quality Index (DLQI)--a simple practical measure for routine clinical use. Clin Exp Dermatol. 1994;19:210-6.
11. Bronsard V, Paul C, Prey S, Puzenat E, Gourraud P-A, Aractingi S, et al. What are the best outcome measures for assessing quality of life in plaque type psoriasis? A systematic review of the literature. J Eur Acad Dermatol Venereol. 2010;24:17-22.
12. Mattei PL, Corey KC, Kimball AB. Psoriasis Area Severity Index (PASI) and the Dermatology Life Quality Index (DLQI): the correlation between disease severity and psychological burden in patients treated with biological therapies. J Eur Acad Dermatol Venereol. 2014;28:333-7.
13. Adil M, Singh PK, Maheshwari K. Clinical evaluation of omega-3 fatty acids in psoriasis. Przegl Dermatol. 2017;104:314-23.
14. Kurd SK, Gelfand JM. The prevalence of previously diagnosed and undiagnosed psoriasis in US adults: results from NHANES 2003-2004. J Am Acad Dermatol. 2009;60:218-24.
15. Dogra S, Yadav S. Psoriasis in India: prevalence and pattern. Indian J Dermatol Venereol Leprol. 2010;76:595-601.
16. Mabuchi T, Ota T, Manabe Y, Ikoma N, Ozawa A, Terui T, et al. HLA-C*12:02 is a susceptibility factor in late-onset type of psoriasis in Japanese. J Dermatol. 2014;41:697-704.
17. Puri N, Mahajan BB, Sandhu SK. Clinical evaluation of different therapeutic modalities in psoriasis by PASI score. Our Dermatol Online. 2013;4:16-22.
18. Choonhakarn C, Busaracome P, Sripanidkulchai B, Sarakarn P. A prospective, randomized clinical trial comparing topical aloe vera with 0.1% triamcinolone acetonide in mild to moderate plaque psoriasis. J Ear Acad Dermatol Venereol. 2010;24:168-72.
19. Gisondi P, Tessari G, Conti A, Piaserico S, Schianchi S, Peserico A, et al. Prevalence of metabolic syndrome in patients with psoriasis: a hospital-based case–control study. Br J Dermatol. 2007;157:68-73.
20. Langley RGB, Krueger GG, Griffiths CEM. Psoriasis: epidemiology, clinical features, and quality of life. Ann Rheum Dis. 2005;64(Suppl II):ii18–23.
21. Gupta MA, Schork NJ, Gupta AK. Suicidal ideation in psoriasis. Int J Dermatol 1993;32:188-90.
22. Puig L, Thom H, Mollon P, Tian H, Ramakrishna GS. Clear or almost clear skin improves the quality of life in patients with

moderate-to-severe psoriasis: a systematic review and meta-analysis. J Eur Acad Dermatol Venereol. 2017;31:213-20.
23. Schmid-Ott G, Schallmayer S, Calliess IT. Quality of life in patients of psoriasis and psoriatic arthritis with a special focus on stigmatisation experience. Clin Dermatol. 2007;25:547-54.
24. Kawro T, Zalewska-Janowska A, Hawro M, Maurer M. Impact of psoriasis severity on family income and quality of life. J Eur Acad Dermatol Venereol. 2015;29:438-43.
25. Upala S, Yong WC, Theparee T, Sanguankeo A. Effect of omega-3 fatty acids on disease severity in patients with psoriasis: a systematic review. Int J Rheum Dis. 2017;20:442-50.
26. Balbás GM, Regana MS, Millet PU. Study on the use of omega-3 fatty acids as a therapeutic supplement in treatment of psoriasis. Clin Cosmet Investig Dermatol. 2011;4:73-7.
27. Basra MK, Salek MS, Camilleri L, Sturkey R, Finlay AY. Determining the Minimal Clinically Important Difference and Responsiveness of the Dermatology Life Quality Index (DLQI): Further Data. Dermatology. 2015;230:27-33.
28. Feldman SR, Kreuger GG. Psoriasis assessment tools in clinical trials. Ann Rheum Dis. 2005;64(Suppl II):ii65-8.
29. Takeshita J, Callis Duffin K, Shin DB, Krueger GG, Robertson AD, Troxel AB, et al. Patient-reported outcomes for psoriasis patients with clear versus almost clear skin in the clinical setting. J Am Acad Dermatol. 2014;71:633-41.
30. Viswanathan HN, Chau D, Milmont CE, Yang W, Erondu N, Revicki DA, et al. Total skin clearance results in improvements in health-related quality of life and reduced symptom severity among patients with moderate to severe psoriasis. J Dermatolog Treat. 2015;26:235-9.

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Our Dermatology Online

Original Article

Clinical findings and outcomes in patients with pyoderma gangrenosum: A single tertiary centre experience

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ABSTRACT

Background: Pyoderma gangrenosum (PG) is an uncommon neutrophilic dermatosis which has a great variety of clinical presentation and course. **Material and methods:** We reviewed the medical records of PG patients who were diagnosed and treated in our department between 2015-2018 years. **Results:** Our study included 16 patients (10 female, 10 male). Lower extremity was the most common location (81%). Of the 16 patients, 8 had multiple ulcerations at the time of diagnosis. Regarding associated comorbidities, haematological disorders (25%) and seronegative arthritis (25%) were the most frequent. Surgery and trauma were detected in two patients as a triggering factor. Pathergy positivity was documented in 6 patients. First-line immunosuppressive monotherapy was effective in 7 (43%) patients. Mean duration to complete remission was 4.6 months. **Conclusions:** In our study we present our experience with some unusual aspects. Our series represented male predominance and higher rates of associated haematological disorders. Initial treatment with systemic corticosteroids is likely to be associated with faster response.

Key words: Cutaneous ulcers; Neutrophilic disorders; Pyoderma gangrenosum

INTRODUCTION

Pyoderma gangrenosum (PG) is an uncommon chronic inflammatory disorder with neutrophilic inflammation. PG is still a challenging disease which is poorly characterized in terms of diagnostic criteria and treatment outcome. Patients with PG usually present with one or more very painful ulcerations with violaceous undermined borders. Major clinical types of PG are as follows: ulcerative, pustular, bullous, and vegetating/superficial granulomatous [1]. However, wide variety of clinical presentations and course can be seen. Exact pathogenetic mechanisms are not fully elucidated yet but association with other immune-mediated disorders and typically favorable treatment response to immunomodulatory drugs including corticosteroids, intravenous immunoglobulin, antitumor necrosis factor- α modalities, plasmapheresis support the immune mediated pathogenic pathways for PG [2].

PG is associated with underlying systemic comorbidites in 50-75% of the cases. Inflammatory bowel diseases, seronegative arthritis, heamatological disorders including malignancies and monoclonal gammopathy of undetermined significance (MGUS) and Behcet disease are common diseases related with PG. It has been reported that in pediatric population the association with inflammatory bowel disease is common[3] Also, surgical procedures may trigger the condition[4]. The disorder may arise as a component of syndromic forms which belong to a spectrum of autoinflammatory conditions characterized with over activated innate immune system [5]. These syndromes are PAPA (pyogenic arthritis, PG and acne), PASH (PG, acne and suppurative hidradenitis), PAPASH (pyogenic arthritis, acne, PG and suppurative hidradenitis) and PASS (pyoderma gangrenosum, acne conglobata, suppurative hidradenitis and seronegative spondiloarthropaty).

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Since PG is considered an orphan disease, prospective randomized studies are lacking in the literature. In addition, due to the low incidence, there is still currently no uniform therapeutic approach. However, PG is potentially life threatening disorder which may show extracutanoeus manifestations [6]. Presence of systemic manifestations including renal and pulmonary involvement and association with comorbidities, seriously affect the choice of treatment.

The main aim of our study was to describe the characteristics of patients with PG in terms of demographics, clinical characteristics, comorbidities, coincidental findings, treatment modalities, and treatment outcomes. Here we document our observations on remarkable clinical findings of PG patients in order to help better understanding of this peculiar disorder.

MATERIALS AND METHODS

A retrospective single-center study was performed to evaluate the patients who were diagnosed with PG in our dermatology department between July 2015 and July 2018. In our department we reviewed the data of patients with the ICD code (L88) on medical charts. History and demographical data of the patients, clinical characteristics, laboratory analyses, associated diseases, coincidental findings, therapeutic interventions and recurrence during follow-up period were recorded. The patients with inadequate data, inconsisted diagnosis of PG or miscoded diagnosis were excluded. Subjects without histopathologic evidence of PG were not included. Follow-up data of the patients were obtained from both inpatient and outpatient medical records. Complete remission was defined as 100% healing of ulcer size or improvement all the lesions, partial remission as a decrease from 50% to 100% ulcer size or lesions. Patients with a <50% decrease of ulcer size or improvement <50% of lesions are grouped as persistent disease.

Statistics

Statistical analyses were carried out using SPSS software (version 21.0 for Windows; SPSS Inc, Chicago, IL, USA). Parametric variables were presented as means and standart deviations and nonparametric variables were presented as medians and interquartile ranges. For categorical variables number of cases and percentages were used. Kolmogorov Smirnov and histogram analyses were used to determine whether continous variables were normally distributed.

Ethics

The methods were in accordance with ethical principles of Declaration of Helsinki and the approval letter of ethics committee was obtained.

RESULTS

Demographics and Clinical Characteristics

Demographical and clinical data of the PG patients were summarized in the Table 1.

Among 34 patients with the presumptive diagnosis of PG during this 3-year period, 16 patients were finally diagnosed with PG. All of the PG patients included in this study required hospitalization. There were 10 (62%) men and 6 (38%) were women. The mean (SD) age at diagnosis was 41.5 (12.3, 25-59) years. The mean duration of disease was 3.5 months and median follow up time was 13 (range 6-22) months.

Twelve (75%) of the recorded cases presented with classical(ulcerative) type. In 7 (43%) of the patients, these ulcerative lesions were exclusively located on the lower limbs (Figs. 1a and 1b). Fifty percent of the patients exhibited multiple ulcerations (Table 1). Cribriform scarring or wrinkled paper scars were observed at healed ulcer sites in 6 (50%) patients with ulcerative lesions (Fig. 2).

Pathergy positivity was noted in 37% of the cases. Trauma was precipitating factor in two patients. One of the male patients reported that he had received wet cupping therapy for chronic leg swelling and pain two weeks before the occurrence of PG lesions (Fig. 3). Pustular type was observed in two patients and the bullous in two. There was no vegetative variant in our series. Three (18%) patients had multiple attacks. Lower limbs were the most commonly affected site, mainly the pretibial areas.

Comorbidities and Coincidental Findings

In our series, 5(31%) of the patients had none of the comorbid diseases previously described in the literature (Table 1). Among these patients two patients had triggering factor: one with breast surgery and one with cupping treatment. Common comorbid diseases diagnosed before the occurrence of PG were listed in the Table 2. Type 2 diabetes mellitus(2), Hashimoto thyroiditis(1), deep venous thrombosis(1) were other medical conditions diagnosed before PG.

Table 1: Data of the patients diagnosed with PG in our department between July 2015 and July 2018

	Men(n=10)	Women (n=6)	Total (n=16)
Age at onset, mean±SD	37.4±11.1	48.5±11.7	41.5±12.3
Duration of the disease, mean±SD, months	4.7±7.1	1.6±1.3	3.5±5.8
Initial presentation			
Ulcer, n(%)	8(50%)	4(25%)	12(75%)
Pustule, n(%)	1(6%)	1(6%)	2(12%)
Blister, n(%)	1(6%)	1(6%)	2(12%)
Total number of ulcers	2.6±1.6	2.6±1.6	2.6±1.6
▪ Solitary ulcer, n (%)	2(12%)	2(12%)	4(25%)
▪ Multiple ulcers(≥2), n(%)	6(37%)	2(12%)	8(50%)
Ulcer size, min-max, cm	3-40	2-8	2-40
Attack number, min-max	1-4	1	1-4
Pathergy positivity, n(%)	4(25%)	2(12%)	6(37%)
Localization			
Lower extremity, n(%)	9(56%)	4(25%)	13(81%)
Upper extremity, n(%)	3(18%)	-	3(18%)
Breast , n(%)	-	3(18%) 2(12%)	3(18%)
Trunk, n(%)	-		2(12%)
Triggering factors			
Trauma (surgery), n(%)	1(6%)	1(6%)	2(12%)
Comorbidities			
▪ No comorbidites*	3(18%)	2(12%)	5(31%)
▪ One comorbidity	4(25%)	2(12%)	6(37%)
▪ Multiple comorbidity (≥2)	3(18%)	2(12%)	5(31%)

*Diabetes, autoimmune thyroid disorders and peripheral vascular disorders were not included.

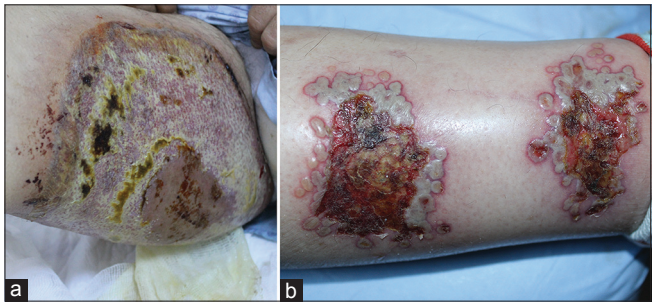

Figure 1: Initial presentation of a classical type PG in a 55-year old male patient without known triggering factor (1a). Multiple confluent vesicles and tiny vesico-pustules surrounding ulcerations in a female patient with coexistent multiple myeloma (1b).

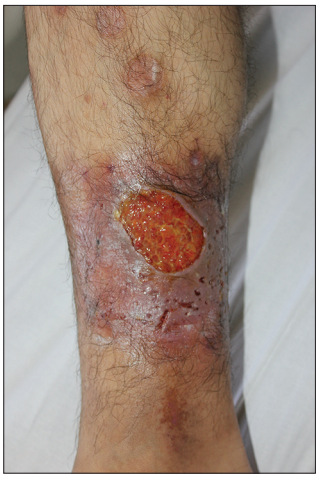

Figure 2: Characteristic appearance of healing period of an ulcerative PG demonstrating wrinkled-paper and cribriform scarring.

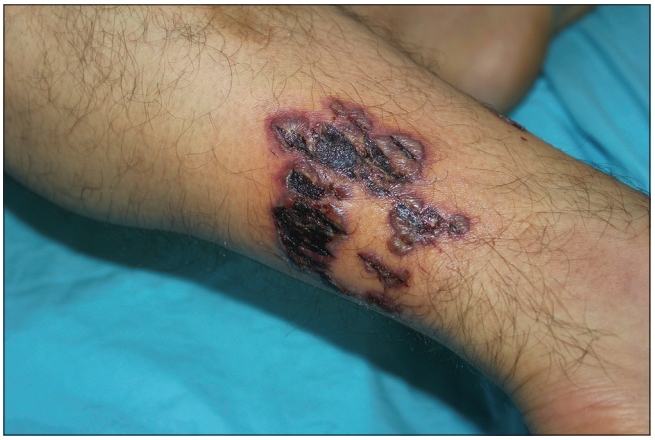

Figure 3: Hijama-induced multiple linear ulcerations with erythematous borders.

Extended laboratory investigations in our center revealed other concurrent diseases: sigmoid colonic polyp(1), monoclonal gammopathy of undetermined significance (MGUS)(1), cryoglobulinemia(1), hydatid cyst(1), multiple myeloma(1), factor V leiden mutation(1), benign thyroid nodules(2). One of the female patients with disseminated sterile pustules and tender nodules, developed multiple myeloma from preceding MGUS during follow up period (Fig. 4). Notably, one of the patients was under therapy for alcohol and cocaine addiction. Two of the patients who were suffered from ulcerative lesions with vegetating aspects were diagnosed with PASH (Fig. 5) and PASS

Table 2: Comorbidites, coincidental findings and associated syndromes in the patients with PG.

Comorbidities	n(%)
Ulcerative colitis	2(12)
Behcet disease	2(12)
MGUS	2(12)
Multiple myeloma∞	3(18)
Psoriasis	2(12)
Psoriatic arthritis	2(12)
Seronegative arthritis	2 (12)
Coincidental conditions	**n(%)**
Sigmoid colonic polyp	1(6)
Cryoglobulinemia,	1(6)
Hydatid cyst	1(6)
Factor V leiden mutation	1(6)
Benign thyroid nodules	2(12)
Peripheral vascular disease	1(6)
Type 2 diabetes mellitus	2(12)
Hashimoto thyroiditis	1(6)
Syndromic forms*	2(12)

∞One of the cases with MGUS progressed into MM during the follow-up period.

*Two male patients were diagnosed with PASS and PASH syndrome.

Table 3: Treatment regimes and outcomes in patients with PG.

	n(%)
Treatment modality	
Immunosuppressive monotherapy	10(10)
Immunosuppresive polytherapy	4(25)
Exclusively topical wound careα	2(12)
Systemic therapy	
Methylprednisolone*	4(25)
Prednisolone*	4(25)
Dapsone	1(6)
Methotrexate	1(6)
Pulse methylprednisoloneα + Methotrexate (SC)	1(6)
Methylprednisolone+Cyclosporine	1(6)
Methylprednisolone+Mycophenolate mofetil	1(6)
Prednisolone+Infiliximab	1(6)
Infliximab	1(6)
Tretment outcome	
Complete remission	11(75)
Partial remission	3(18)
Persistent disease	1(6)
Contact lost to patient	1(6)
Mean duration to complete remission, mean±SD, months	4.6±2.3
Recurrence	3(18)

*Glucocorticosteroid at 1-2 mg/kg/day with gradually tapering was used.

α1g/day 5 consecutive days

syndrome because of the presence other clinical manifestations.

Treatment Modalities

Most patients (87.5%) were treated with systemic therapy. Systemic corticosteroids were initial treatment regimen in 11 patients (68.7%) either alone in 7 patients or in combination with cyclosporine in one (6%), methotrexate in one (6%), mycophenolate mofetil in one (6%), infliximab in one (6%). Corticosteroid and infliximab combination was chosen as first line therapy

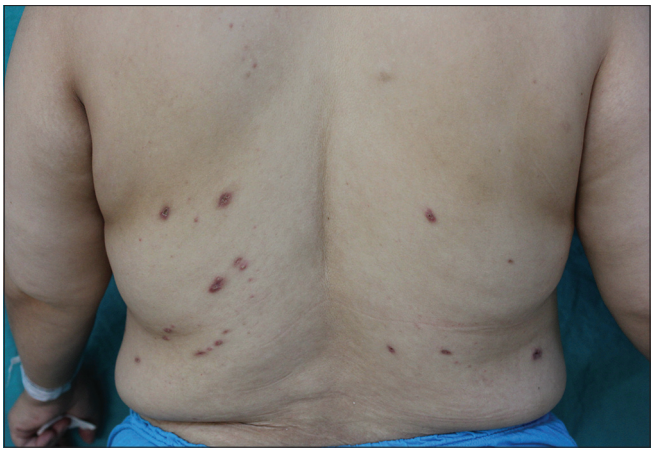

Figure 4: 44-year old female presenting with disseminated sterile pustules on the trunk in association with MGUS.

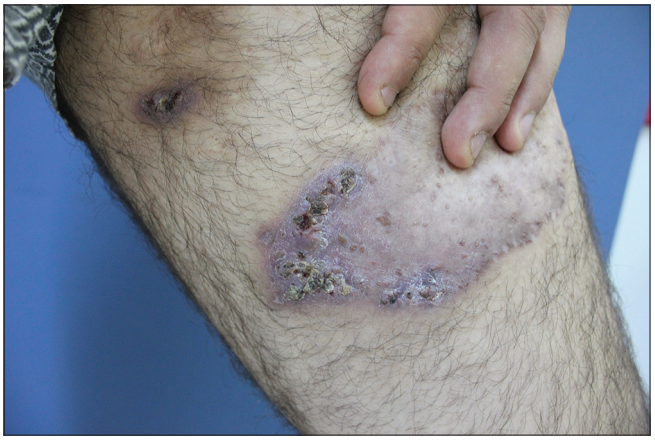

Figure 5: Vegetating appearance of an ulcerative lesion in a 28-year old male diagnosed with PASH syndrome.

for the patient with PASS syndrome. In three patients who had comorbidities that contraindicated the use of systemic steroids treatment regimens were as follows: dapsone(1), methotrexate(1) and cyclosporine(1). Topical wound care was carried out in all patients. Two patients treated with topical wound care and topical super-potent corticosteroids. Treatment regimens were included in Table 3.

Treatment Outcomes

Systemic therapy at first line was effective with complete healing in 9 (56%) patients on average 4.4±1.3 months. Patients (12%) who were exclusively treated with topical wound care and topical corticosteroids eventually achieved complete response. One of three patients with partial response infliximab was added to initial therapy because of steroid failure. In a patient with ulcerative colitis who was free from PG for 10 months recurrence was noted after one year. Systemic antibiotic treatment was promptly initiated in five patients who had clinical

findings such as erythema, swelling, fever and elevated C-reactive protein and positive culture from smears.

DISCUSSION

PG may affect any age group but those between 20-50 years are particularly at risk [1, 7]. Similarly, in our study mean age of onset of total study population was 41.5. Female patients were older than male patients at onset of disease however the difference did not reach statistical significance (Table 1). In contrast to many previous studies, there was men predominance (1.6:1) in our study group as it was reported by some authors [8].

Presentation with ulcerative lesions was the most common clinical type in our study as in other series [7,9]. Almost 81% of the PG lesions were located on lower extremity. Interestingly, one of these cases was triggered with wet cupping (or called 'hijama') therapy which is a form of alternative medicine (Fig. 3). In this technique before the cups are applied, tiny incisions are made in the skin for the elimination of toxic blood or fluids. We emphasize this patient because of distinct clinical picture with sterile bullae rapidly progressing to shallow ulceration. This patient with preceding hijama history, seem to be the first reported in the literature. He was successfully treated with corticosteroid monotherapy.

PG should be included in the differential diagnosis of breast ulcers. Among female patients (6) breast involvement was also common (50%) in our series. It is well known that postoperative pyoderma gangrenosum commonly affects breasts [10]. One of our patients with bilaterally breast involvement had had breast reduction immediately before the occurrence of PG lesion. Recently, Maverakis et al. reported the results of Delphi consensus on diagnostic criteria of ulcerative PG [11]. In their report presence of multiple ulcers at least one anterior lower leg was one of the newly introduced diagnostic criteria. As they have stated we observed involvement of the anterior leg involvement in 58% of patients. Also, as the panel has agreed that cribriform or wrinkled paper scarring is useful in the diagnosis of PG, we observed these scars during healing process in half of the patients with ulcerations.

In the literature, percentage of the association with underlying diseases varies between 50%-70% [7, 2]. PG is associated with systemic diseases in 68.7% of our patients. It has been suggested that comorbidities including diabetes and PVD may be contributing factors for the development of PG [7]. However we did not include newly reported associations including endocrinopathies [12]. Hematological disorders were the most common underlying diseases in our series. Of note coincidental findings which were detected after the diagnosis of PG were considerable (Table 2). Interestingly two of our patients had the diagnosis of PG associated genetic syndromes (PASS and PASH syndrome) in our department. Taken together these results may suggest that individuals in a tertiary care setting are more likely to have concurrent serious or complicated diseases.

Several mucocutaneous diseases have been associated with cocaine consumption. Cocaine induced pyoderma gangrenosum (CIPG) is one of the cutaneous manifestations associated with cocaine use[13]. Clinically, lesions are not indistinguishable from other forms. However they tend to be of greater size and multiple [13]. A male patient with a history of cocaine abuse presented with multiple ulcerative lesions which were located on the upper extremities. He was treated with pulse methylprednisolone and methotrexate (15 mg/weekly, sc). All lesions healed within 4 months but recurrence was noted during follow-up. This would support that CIPG is seem to be more refractory and recurrent.

To date, there are still no established gold-standard treatment strategy and uniformly effective therapy for PG. The main aim is to promote wound healing and to reduce the pain by reducing inflammation. Milder forms of disease may be treated with local wound care [14]. In our clinical practice we usually prefer topical modalities in combination with systemic treatments. In this series, we had two patients who were exclusively controlled with topical wound therapy. Because of small sample size our study can provide little guidance regarding the choice of systemic therapy. However we observed shorter periods of healing in patients who were initially treated with systemic corticosteroids (usually oral corticosteroids). In our series, four patients required immunosuppresive polytherapy. All these patients had severe form of PG and severe comorbidities requiring faster taper of corticosteroids. Anti-tumor necrosis factor alfa (anti-TNF-α) agents are another treatment option for resistant PG [15]. TNF- α is the main inflammatory mediator of PASS syndrome [5]. We preferred to use infliximab in two patients who had the diagnosis of Behcet disease and PASS syndromes.

In our retrospective study of PG patients healing was achieved approximately within 4 months in 75% of the cases. Forty three percent of the patients were successfully treated with first-line immunosuppressive monotherapy. Our results were comparable with previous reports [9, 16]. It has been reported that recurrence rates of PG is 17-61% [9]. In our series of 16 patients, only 3 (18.7%) patients who were suffering from co-morbid (psoriatic arthritis in one, ulcerative colitis in one and seronegative spondyloarthropathy in one) chronic diseases had recurrent attacks.

Our study has several limitations related to its retrospective design and evaluation of small number of patients. So we were unable to detect a statistical difference in many of parameters between subgroups. Because of rarity the disorder it is quite difficult to present large number of cases. Nevertheless it is not possible for us to draw strong conclusions based on the data reported here.

CONCLUSION

These results described here represent our tertiary centre experience to date. Our study demonstrated a series of 16 patients with PG. Underlying systemic diseases were observed in 68% of the patients. Haematological disorders (MGUS and multiple myeloma) and seronegative arthritis were the most common (25%) associated diseases. Development of new lesions after trauma and surgery was documented in two of the patients. Most of the patients (43%) were controlled with first line immunosuppressive monotherapy, mainly systemic corticosteroid therapy. Mean duration to complete remission was 4.6 months. Recurrence was noted in three (18%) individuals.

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.

REFERENCES

1. Binus AM, Qureshi AA, Li VW, Winterfield LS. Pyoderma gangrenosum: a retrospective review of patient characteristics, comorbidities and therapy in 103 patients. Br J Dermatol. 2011;165:1244-50.
2. Kridin K, Cohen AD, Amber KT. Underlying Systemic Diseases in Pyoderma Gangrenosum: A Systematic Review and Meta-Analysis. Am J Clin Dermatol. 2018;19:479-87.
3. Diatta BA TF, Diop A, Diadie S, Ndiaye M, Diallo M, Niang SO, et al. Pyoderma gangrenosum among children in Senegal: 6 cases. Our Dermatol Online. 2017;8:463-6.
4. Hiraiwa T FH, Yamamoto T. Pyoderma gangrenosum triggered by surgical procedures in patients with underlying systemic diseases. Our Dermatol Online. 2014;5:432-3.
5. Cugno M, Borghi A, Marzano AV. PAPA, PASH and PAPASH Syndromes: Pathophysiology, Presentation and Treatment. Am J Clin Dermatol. 2017;18:555-62.
6. Wollina U. Pyoderma gangrenosum--a systemic disease? Clin Dermatol. 2015;33:527-30.
7. Adisen E, Erduran F, Gurer MA. Pyoderma Gangrenosum: A Report of 27 Patients. Int J Low Extrem Wounds. 2016;15:148-54.
8. Bhat RM, Nandakishore B, Sequeira FF, Sukumar D, Kamath GH, Martis J, et al. Pyoderma gangrenosum: an Indian perspective. Clin Exp Dermatol. 2011;36:242-7.
9. Pereira N, Brites MM, Goncalo M, Tellechea O, Figueiredo A. Pyoderma gangrenosum--a review of 24 cases observed over 10 years. Int J Dermatol. 2013;52:938-45.
10. Tolkachjov SN, Fahy AS, Cerci FB, Wetter DA, Cha SS, Camilleri MJ. Postoperative Pyoderma Gangrenosum: A Clinical Review of Published Cases. Mayo Clin Proc. 2016;91:1267-79.
11. Maverakis E, Ma C, Shinkai K, Fiorentino D, Callen JP, Wollina U, et al. Diagnostic Criteria of Ulcerative Pyoderma Gangrenosum: A Delphi Consensus of International Experts. JAMA Dermatol. 2018;154:461-6.
12. Abdel-Mohsen MA, El-Braky AA, Ghazal AAE, Shamseya MM. Autophagy, apoptosis, vitamin D, and vitamin D receptor in hepatocellular carcinoma associated with hepatitis C virus. Medicine (Baltimore). 2018;97:e0172.
13. Moreno-Artero E, Querol-Cisneros E, Rodriguez-Garijo N, Tomas-Velazquez A, Antonanzas J, Secundino F, et al. Mucocutaneous manifestations of cocaine abuse: a review. J Eur Acad Dermatol Venereol. 2018;32:1420-6.
14. Feldman SR, Lacy FA, Huang WW. The safety of treatments used in pyoderma gangrenosum. Expert Opin Drug Saf. 2018;17:55-61.
15. Ahronowitz I, Harp J, Shinkai K. Etiology and management of pyoderma gangrenosum: a comprehensive review. Am J Clin Dermatol. 2012;13:191-211.
16. Duman N, Evans SE, Karaduman A, Elçin G, Özaygen GE, Atakan N. et al. Pyoderma Gangrenosum: A Retrospective Study of 25 Cases and Review of the Literature Findings. Turk Klin J Dermatol. 2013;23:77-83.

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Our Dermatology Online

Original Article

Mélanome acral du pied: a propos de 9 cas et revue de la littérature [Acral melanoma of the foot: a study of 9 cases and guidelines update]

Sid'Ahmed Mohamed Limam[1], Cheikh Elmoustapaha Erebih[2], Abdelaziz Beyrouk[1], Khaled Isselmou Boye[3], El Hassen Didi[1], Seyidna Oumar Ely[4], Cheikh Ne[2]

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ABSTRACT

Background: Malignant Melanoma is a malignancy of pigmented-producing cells (melanocytes). Acral melanoma is located on non-hair bearing skin of palms and soles or under the nail bed. This histological subtype is often described in darker skin. Patients commonly display advanced stage of disease at presentation. This leads, inevitably, to poor disease prognosis. The functional requirement together with the wide excision in the foot makes treatment of melanomas a challenging for the surgeons. **Methods:** We retrospectively collected clinical, pathological, surgical and follow-up data of 12 acral melanoma cases in the foot. Three patients whose records contained insufficient information were excluded. **Results:** The mean age was 65 years (43-70years). 5 patients were female and 4 were male. The heel was the most commonly involved location with 50 % of the patients. Clark's Level showed advanced stages IV and V with more than 88, 88 % of the patients. Wide excision until the plantar fascia was performed in all patients (9 cases) and for metastatic sentinel nodes; complete lymphadenectomy was performed in 2cases. Lymphadenectomy of retroperitoneal nodes was made in 1 case; resection of crural metastasis was done in 2 cases.

In Postoperative coverage: primary closure was feasible (n=2), skin graft (n=2) and spontaneous wound healing (n=5). **Conclusion:** Acral melanoma is the commonest melanoma in our practice. Most of the cases presented with advanced stage disease.

Key words: Acral melanoma; Foot, Surgery; Mauritania

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RÉSUMÉ

Introduction: Le mélanome malin est un cancer cutané développé aux dépend des cellules productrices de pigments cutanés: les mélanocytes. Le mélanome acrolentigineux touche la peau palmo-plantaire et sous-unguéale. Il constitue un type histologique fréquent chez le sujet à peau pigmentée. Le diagnostic tardif de ces tumeurs conduit souvent à une lourde mortalité par métastases ganglionnaires et viscérales. L'atteinte du pied constitue un challenge thérapeutique pour le chirurgien reconstructeur; car l'exérèse doit concilier les soucis carcinologiques et fonctionnels. **Matériels et méthodes:** 12 cas de mélanome acral du pied ont été consécutivement réunis, mais 3 dossiers incomplets ont été exclus. **Résultats:** L'âge moyen était de 65 ans (43-70 ans); une légère prédominance féminine a été notée (5 femmes, 4 hommes). La topographie lésionnelle était dominée par le talon retrouvé dans 50% des cas. La classification de Clark-Mihm retrouvait les types IV et V chez 88,88% des patients. Le bilan d'extension a montré une atteinte ganglionnaire chez 4 malades. Le traitement chirurgical a consisté en une exérèse large des tumeurs chez les 9 malades; associé à un curage ganglionnaire inguinal chez 4 malades. Un curage retro-péritonéal a été réalisée dans un cas. Une métastasectomie locorégionale a été réalisée chez deux cas. Pour la couverture de la perte de substance cutanée; une cicatrisation dirigée avait été suivie chez 5 malades, une greffe de peau, dans 2cas et une suture primitive avec ou sans autoplastie cutanée chez 2 malades. **Conclusions:** le mélanome acral reste le mélanome le plus fréquent dans notre pratique; son diagnostic est tardif ce qui conduit à un traitement difficile et un pronostic sévère.

Mots clefs: Mélanome acral; Pied; Chirurgie; Mauritanie.

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INTRODUCTION

La protection contre les rayons ultra-violets (UV) est l'une des fonctions principales de la peau, elle est assurée par un pigment cutané appelée la mélanine.

Le mélanome malin (MM) se développe aux dépend des cellules productrices de pigments cutanés (les mélanocytes) [1-4].

La prévalence du MM en Afrique reste sous-estimée; Il reste globalement moins fréquent qu'en Europe, car la peau noire est plus riche en mélanine. Cependant ce pigment est absent au niveau des paumes des mains et des plantes des pieds ce qui explique la vulnérabilité de ses régions [2].

Le mélanome acral (MA) touche la peau non pigmentée palmo-plantaire et sous unguéale, ainsi que les muqueuses. Son extension en profondeur est à l' origine du retard diagnostique et du mauvais pronostic.

En France le MM représente 1% de tous les néoplasmes et 5% des cancers cutanés [2]. Le MM est responsable de 80% de la mortalité des cancers cutanés [3]. La mortalité du MM est due à sa tendance métastatique lymphatique et hématogène; son pronostic est lié à l'épaisseur de la tumeur évaluée par l'indice de Breslow et le niveau de Clark-Mihm [2].

MATÉRIELS ET MÉTHODES

Un travail continu a été mené au service de Traumatologie-Orthopédie de l'Hôpital Cheikh Zayed de Nouakchott-Mauritanie, en collaboration avec le Centre National d'Oncologie (CNO).

Nous avons ainsi recruté 12 malades, mais 3 ont été exclus car leurs dossiers n'étaient pas complets. Un dermato-cancérologue recrutait les patients, réalisait la biopsie, vérifiait le bilan d'extension et nous référait les patients pour réaliser l'acte chirurgical.

Le traitement chirurgical était réalisé par un chirurgien orthopédiste (auteur correspondant). Chez 4 patients nous avons eu recours à un chirurgien cardio-vasculaire pour le curage ganglionnaire inguinal et retro péritonéal.

Le bilan d'extension consistait à faire une radiographie pulmonaire, une échographie abdomino-pelvienne, et dans certains cas un scanner-TAP (thoraco-abdomino-pelvien) était demandé.

Le suivi de la plaie est assure par l'équipe chirurgicale jusqu'a cicatrisation. Puis le malade est confié au dermato-cancérologue pour suivi oncologique.

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.

RÉSULTATS

L'âge moyen de nos patients était de 65 ans (extrêmes de 43 et 70 ans), une légère prédominance féminine a été retrouvée (5 femmes contre 4 Hommes).

La topographie lésionnelle se distribuait comme suit: le talon 4 cas, l'avant-pied: 2 cas, les orteils: 2 cas et le tronc 1: cas (Figs. 1a et 1b.)

L'exérèse était carcinologiquement satisfaisante dans les berges latérales et profondes; chez les neuf malades (Fig. 2).

Dans les 4 cas ou un curage ganglionnaire était nécessaire, la lame ganglionnaire inguinale était reséquée en totalité avec 2 formations ganglionnaires atteintes respectivement (2N+/9N) et (2N+/2N) chez 2 malades (Fig. 3).

Un curage extensif de tout le réseau lymphatique de la région crurale a été réalisé chez deux malades, avec chez un parmi eux un curage retro-péritonéal sacrifiant

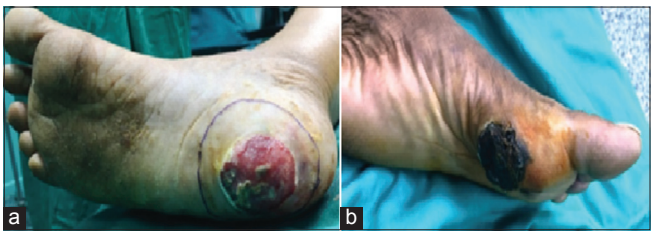

Figure 1: Topographie lésionnelle. a-Mélanome superficiel ulcéré du talon; b- Mélanome nodulaire de l'avant- pied.

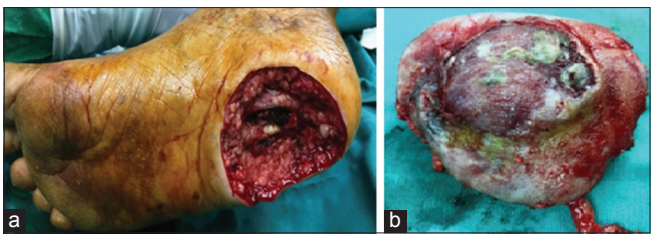

Figure 2: Exérèse d'un mélanome du talon ulcéré. a- Lit tumoral après exérèse; b- Pièce opératoire.

le pédicule iliaque externe et conduisant a un double pontage artériel et veineux iliaque externe (Figs. 4 et 5).

Deux cas d'amputation aux orteils ont été nécessaires dans notre série (Fig. 6).

La classification de Clark-Mihm retrouvait: un type III (n=1), un type IV (n=3) et un type V (n=5).L'indice pronostique de Breslow a été établi chez 3 malades avec un résultat de 3mm, 3,5 mm et 11.5 mm.

La classification histologique a retrouvé les types suivants: mélanome acrolentigineux ou acral lentiginous melanoma (ALM: 4 cas), mélanome nodulaire ou nodular melanoma (NM: 3 cas) et mélanome a extension superficielle ou superficial spreading melanoma (SSM: 2 cas).

L'infiltrat lymphocytaire tumoral (Tumour Infiltrating Lymphocytes: TIL) a été retrouvé avec un non-brisk chez 1 malade. Les marqueurs HMB-45 et S 100 ont été retrouvés chez un patient (Fig. 7).

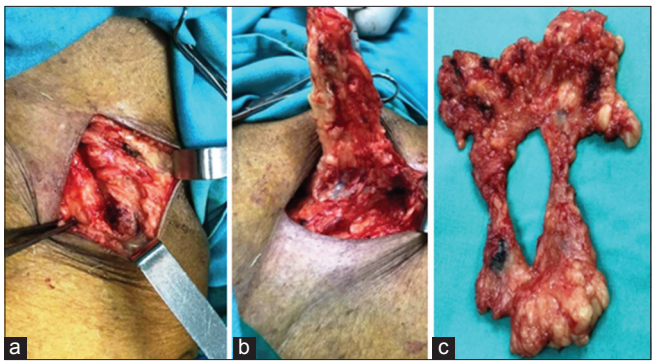

Figure 3: Ccurage ganglionnaire: (a) début de dissection de la lame inguinale; (b) fin de la dissection; (c) pièce opératoire contenant plusieurs ganglions manifestement malades.

DISCUSSIONS

Le MM constitue une tumeur maligne des cellules productrices de mélanine. Son épidémiologie est assez paradoxale; il représente seulement 4-5% des cancers cutanés; mais on lui attribue 80% de la mortalité liée à ces tumeurs [3,4].

Le MM se divise en 4 types histologiques (selon la classification de l'OMS 2006) rapportés ici par ordre de fréquence décroissante: le mélanome à extension superficielle (SSM), le mélanome nodulaire(NM), le mélanome lentigo malin(LMM) et le mélanome acrolentigineux(ALM) [5,6].

Le mélanome acrolentigineux (ALM) ou mélanome acral (MA) touche la peau palmo-plantaire et sous-unguéale; Reed fût le premier à le décrire en 1976 comme type de MM [6].

Le mélanome plantaire compte pour 3-5 % des MM; l'atteinte du pied constitue un challenge thérapeutique car le traitement adéquat doit concilier l'exérèse carcinologique de la tumeur aux soucis de préservation de la fonction du pied (la marche) [7].

Le MA est considéré comme le type le plus agressif parmi les mélanomes et par conséquent corrélé à un pronostic réservé. Des études récentes ont expliqué ce mauvais pronostic par le diagnostic tardif de ces tumeurs plus que par leur nature [8].

Il reste le type le plus décrit chez les sujets à peau pigmentée; en Afrique [1] et en Asie [8-13]. Chez le caucasien, le mélanome acral est rare [3,6].

Le MA reste une maladie du sexagénaire; l'âge moyen de nos malades était de 65 ans, Coulibaly au Mali avait retrouvée une moyenne de 60.37 ans, Jiaojie en

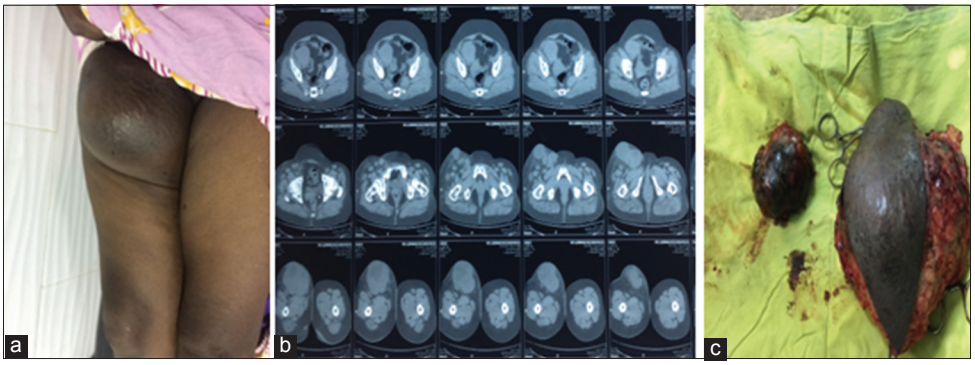

Figure 4: Métastasectomie crurale avec curage retro-peritonéal: (a) Grosse métastase crurale droite sur mélanome récidivant de l'avant-pied; (b) Scanner pelvien montrant la métastase crurale avec une autre métastase retro péritonéale; (vc) Pièces opératoires avec à droite la grosse métastase crurale et à gauche la métastase retro péritonéale.

Chine 62 ans, la même moyenne avait été retrouvé par Rashid aux USA [2,7,10].

Pour le sexe; nous avons retrouvé une prédominance féminine (5 femmes contre 3 hommes); ce qui a été retrouvé par Rashid et al. (31 F contre 15 H) et par Tseng et al. (77 F contre 39 H). Mais d'autres auteurs ont retrouvé le contraire: Jiaojie et al. (84 H contre 55 F), Zainal et al. (23 H contre 10 F). Dans d'autres séries les deux sexes se retrouvaient sensiblement répartis: Coulibaly (19H contre 15 F) [1,2,7,14,10].

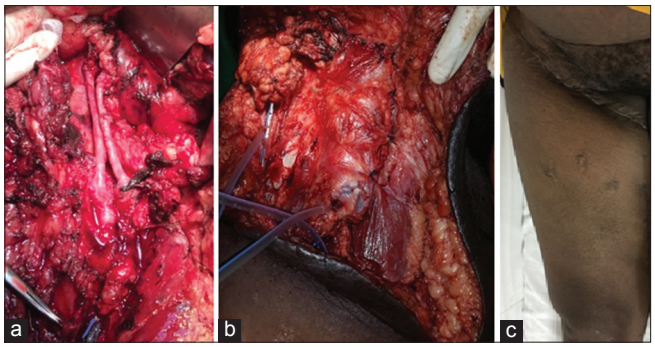

Figure 5: Double pontage du pédicule iliaque externe: (a) Déclampage de l'artère et la veine iliaque; (b) Protection du pontage par lambeau du muscle sartorius; (c) Cicatrisation et bon contrôle oncologique. (Même malade que la figure 4)

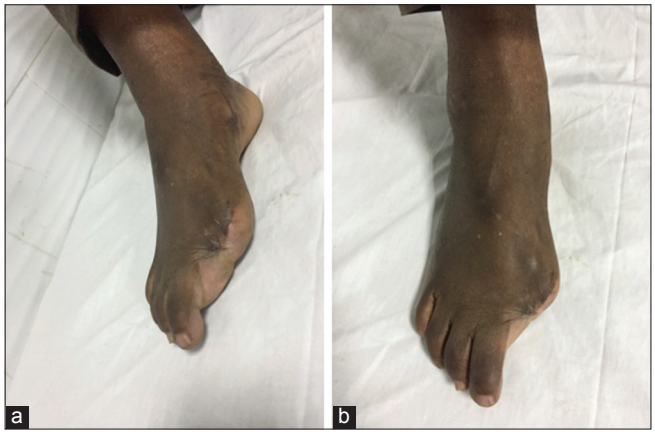

Figure 6: Désarticulation du gros orteil sur un mélanome nodulaire: (a) Vue de face; (b) Vue oblique.

Nous en avons conclu que le MM se répartit indifféremment sur les deux sexes.

En ce qui concerne la topographie lésionnelle la moitié de nos malades était localisée au niveau du talon (4 cas); suivi par l'avant-pied et orteils (4 cas) et en fin le tronc (1 cas).

L'atteinte plantaire reste largement rapportée: Coulibaly avait retrouvé sur 34 cas la répartition suivante: mélanomes plantaires (n=23), talon (n=10) et cuisse (n=1) [1].

Jiaojie retrouvait sur une série de 142 cas; dont 122 MA: plante du pied (59 cas), talon (38 cas), Dos du pied (8 cas) et orteils (10 cas) [10].

Zainal sur une sérié de 60 MM; dont 33 MA: talon (n=13), plante du pied (n=8) et orteils (n=11) [1].

Sur une série exclusivement consacrée au mélanome acrolentigineux (n=21) Liu et al ont retrouvé: plante (n=7), talon (n=4), dos du pied (n=3), cheville (n=5) et orteils (n=2) [4]. Ceci a été expliqué par les traumatises liés à la zone d'appui [4].

La classification de Clark-Mihm est considérée comme un critère pronostique; nous avons retrouvé dans 88.88 % des cas les types IV et V.

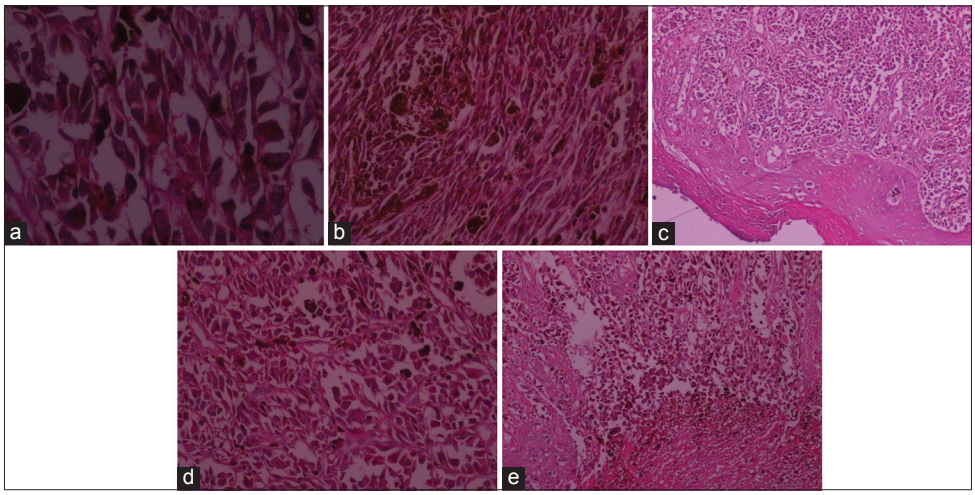

Figure 7: Aspects anatomopathologiques de l'un d nos cas cliniques: (a) Cellules tumorales atypiques et mitotiques; (b) Détails de la prolifération tumorale; (c) Migration pagétoide au niveau du revêtement épithélial; (d) Prolifération de cellules chargées en mélanine; (e) Ulcération du revêtement de surface.

Morton et al. Ont étudié la valeur pronostique de la classification de Clark-Mihm en réalisant une analyse multifactorielle des registres d'une vaste base de données impliquant 5575 patients dont 3323 dossiers exploitables; les résultats étaient comme suit: la survie à 5 ans pour le MM stade V était de 47%; pour le stade IV une survie de 68%; ce chiffre passe à 81% pour le type III et à 95% pour le type II de Clark-Mihm [15].

Zainal et al. Ont retrouvé 75% des types IV et V de Clark-Mihm. Jiaojie a retrouvé 90.14% des mêmes types IV et V [1,10].

Ces résultats suggèrent que le diagnostic du mélanome acrolentigineux survient souvent à un stade tardif de la maladie; à cause de son évolution silencieuse.

L'indice de Breslow est le meilleur facteur pronostique du MM d'après la méta-analyse de Morton et al [15]. Cependant l'AJCC (American Joint Comittee on Cancer) recommande de coupler son interprétation avec celle du niveau de Clark-Mihm.

Dans notre série cet indice a été réalisé chez trois malades et les valeurs étaient de 3mm; 3,5 et 11.5 mm. Jiaojie en 2016 a trouvé un indice de Breslow supérieur à 2mm chez 80% de ses patients [10]. Ce qui dénote du stade diagnostique tardif déjà; retrouvé avec la classification de Clark-Mihm. L'absence d'un micromètre dans notre service d'anatomie pathologique fait que les biopsies traitées à notre niveau sont dépourvue de l'indice de Breslow.

Balch et al. En 1977 sur 211 MM trouvèrent un indice de Breslow supérieur à 1.5 mm chez 57% des patients. En 1993 Morton et al. Trouvèrent 40% de patients pour la même tranche [15,17].

L'infiltrat tumoral lymphocytaire ou Tumour Infiltrating Lymphocytes (TIL) est un facteur de bon pronostic. Herbermann fût le premier à parler de son rôle inhibiteur de l'extension du MM en 1992. Le même constat a été réitéré par Thorn et al. en 1994 [16]. Zainal et al.ont décrit la réponse au TIL en trois catégories: brisk, non-brisk et absence. La réponse brisk se définit comme une bande continue des lymphocytes vue à la base du mélanome acral; elle est corrélée à un taux de survie élevé [1].

Dans notre série le TIL a été mis en évidence chez un seul patient; avec un non-brisk. On ne peut pas en déduire un rôle pronostique.

Sur le plan thérapeutique la chirurgie reste le meilleur traitement du MM. les marges adéquates restent un sujet de débat; cependant plusieurs auteurs s'accordent à considérer l'épaisseur tumorale mesurée par l'indice de Breslow comme déterminant des marges chirurgicales: (<1mm: 0,5cm de marges; <2mm: 1 cm de marges; >2mm: 2cm de marges) [2,3,6,11,12,14]. L'exérèse doit toucher le tissu cellulaire sous cutané; ainsi que le fascia superficiel pour être sûr d'emporter toutes les chaines lymphatiques. Si le fascia n'est pas touché par le MM; son exérèse n'améliore guère le taux de récidive [6].

Les larges pertes de substance cutanées sont traitées par des lambeaux cutanés locaux de rotation ou d'avancement ou par des greffes de peau [14,18]. Dans notre travail le retard du résultat de l'anatomie pathologique nous a contraints à suivre une cicatrisation dirigée chez 4 malades; chez deux malades une suture primitive a été possible; deux malades ont bénéficié d'une greffe de peau mince; une plastie cutanée était nécessaire dans un cas.

Les MM des doigts et orteils sont traités par amputation [4,6]. Ce qui été réalisé chez deux malades de notre série.

L'atteinte des nodules lymphatiques constitue un facteur de mauvis pronostic. Tous les ganglions malades doivent être reséqués; sauf ceux associés à des métastases non résécables [6]. La résection des ces ganglions à contenu micro-métastatique avant la dissémination vers des métastases à distance; améliore la survie sans événement tumoral et peut guérir les malades [6,14]. Dans notre série l'atteinte ganglionnaire a été objectivée chez quatre malades; l'exérèse de la lame ganglionnaire a été réalisée chez deux et un curage extensif ganglionnaire a été indiqué chez deux autres; associé chez un parmi ces derniers à un curage retro-péritonéal ayant conduit à emporter le pédicule iliaque externe remplacé par un double pontage artériel et veineux iliaque externe; ce genre d'extension n'a pas été rencontré dans la littérature. Parmi ces quatre malades deux sont décédé par AVC, sans qu'un antécédent ou facteur de risque cardio-vasculaire ne soit connu chez ces malades. La seule explication retenue c'est éventuellement une hémorragie due a une métastase cérébrale.

CONCLUSIONS

Le mélanome acral est le mélanome le plus fréquent dans notre pratique. Son diagnostic est souvent tardif;

ce qui conduit à un pronostic grave. L'indice de Breslow reste indispensable pour l'approche thérapeutique et pronostique de ces tumeurs. L'exérèse large reste le meilleur traitement associé au curage ganglionnaire inguinal en cas d'atteinte ganglionnaire.

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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.

BIBLIOGRAPHIE

1. Zainal AI, Zulkarnaen M, Norlida DK, Syed Alwi SA. Acral melanoma of the extremities: a study of 33 cases Sarawakian patients. Med J Malaysia. 2012;67:60-5.
2. Coulibaly DK. Thèse de Médecine, Faculté de Médecine, de Pharmacie et d'Odontostomatologie. Université de Bamako- Mali: 2007- 2008.
3. Sue GS, Hanlon A, Lazova R, Narayan D. Case report: use of imiquimod for residual acral melanoma. BMJ Case Rep. 2014;2014:pii: bcr2014203826.
4. Liu J-F, Zhao L-R, Lu L-J, Chen L, Liu S-G, Gong X, Liu B. Limb Salvage Surgery following resection of a melanoma: Foot and ankle reconstruction using cutaneous flap: Oncol Lett. 2014;8:1966–72.
5. Scolyer RA, Long GV, Thompson JF. Evolving concept in melanoma classification and their relevance to multidisciplinary melanoma patient care. Molec Oncol. 20111;5:124-36.
6. Kosmidis C, Efthiamidis C, Anthimidis G, Grigoriou M, Vasiliadou K, Ioannidouet G, al. Acral Lentiginous Melanoma: A Case Control Study and Guidlines Updates. Case Rep Med. 2011;2011:670581.
7. Rashid OM, Chaum JC, Wolf LG, Brinster NK, Neifeld JP. Prognostic variables and Surgical management on Foot Melanoma: Review of a 25- years Institutional Experience. ISRN Dermatol. 2011;2011:384729.
8. Oh TS, Bae E, Ro KW, Seo SH, Son SW, Kim IH. Acral lentiginous melanoma developping during long-standing atypical melanosis: usefullness of dermatoscopy for detection of early acral melanoma. Ann Dermatol. 2011;23:400–4.
9. Bae JM, Kim HO, Park YM. Progression from in situ to Invasive Acral Lentiginous Melanoma. Ann Dermatol. 2009;21:185–8.
10. Lv J, Dai B, Kong Y, Shen X, Kong J. Acral melanoma in Chinese: A clinicopathological and prognostic study of 142 cases. Scien Rep. 2016;6:31432.
11. Roh MR, Kim J, Chung KY. Treatment and outcomes in acral location in Korean patients: Yonsei Med J. 2010;51:562–8.
12. Uehara J, Ito Y, Takahashi I, Honma M, Yamamoto AI, Matsuo S, et al. Case Rep Dermatol. 2010;2:201-6.
13. Caldeira Xavier-Júnior JC, Munhoz T, Souza V, Pires de Campos EB, Stolf HO, Alencar Marques ME. Focal invasivness in complete histological analyses of large acral lentiginous melanoma. Diagn Pathol. 2015;10:73.
14. Tseng JF, Tanabe KK, Gadd MA, Cosimi AB, Malt RA, Haluska FG, et al. Surgical management of primary cutaneous melanoma of the hands and feet Ann Surg. 1977;225:544-53.
15. Morton DL, Davtyan DG, Wanek LA, Foshag LJ, Cochran AJ. Multivariate analysis of the relationship between survival and the microstage of primary melanoma by Clark level and Breslow thickness. Cancer. 1993;71:3737-43.
16. Thörn M, Pontén F, Bergström R, Sparén P, Adami HO. Trends in tumour characteristics and survival of malignant melanoma 1960-84: a population-based study in Sweden. Br J Cancer. 1994;70:743-8.
17. Balch CM, Murad TM, Soong SJ, Ingalls AL, Halpern NB, Maddox WA. A multifactorial analysis of melanoma: prognostic histopathological features comparing Clark's and Breslow's staging methods. Ann Surg. 1978;188:732-42.
18. Maker AV, Iteld L. Closure of melanoma defects on the sole of the foot using glabrous skin: the end of the flap. Ann Surg Oncol. 2015;22:4081-2.

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Our Dermatology Online

Brief Report

A cyanobacterium, priorly stressed by chemical way, could represent the occult Tantra for tanning phototypes I and albinoes

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ABSTRACT

Background: Since the recent discover of seven UV-absorbing pigments (UVP), isolated from various marine organisms and identified as a series of mycosporine-like amino acids (MAAs), that are reputed to absorb in wavelengths ranging from 310 to 365 nm, spanning both UV-B and UV-A (320-400nm) portions of the solar spectrum (mycosporine-glycine, shinorine, porphyra-334, palythine, asterina-330, palythinol, usujirene and palythene). We have made up our mind to select one cyanobacterium (Aphanothece sacrum), admitted as cosmetic ingredient, apt to increase its amount of mycosporine-2-glycine, a natural sunscreen factor among the most efficient in nature, when undergoes to chemical stresses. **Material and Methods:** In two group of volunteers was used a gel made up with 2 g of Aphanotece sacrum powder in 1.5% karaya gum aqueous solution was prepared and gel with 2 g of Aphanotece sacrum powder following the above mentioned method in 1.5% karaya gum aqueous dispersion. Was evaluate the capacity of the cyanobacterium to protect skin from UVB rays under long exposure to artificial sun. **Results:** The cosmetic that reveals a SPF equivalent to 56 is able to minimize at all the chances of burning. Phototype I may be protected exclusively by inorganic powerds (titanium dioxide,barium sulphate,kaolin etc.), and thanks to the application of gel two it is possible to have a minimal tanning. **Conclusion:** We have tried to treat chemically the cyanobacterium (easily available on the market, as in Japan it is considered a common foodstuff) and let it be an exceptional sunscreen, able to protect even phototype I and albinoes.

Key words: Cyanobacterium; Aphanothece sacrum; Karaya gum; Phototype

INTRODUCTION

Since UV-radiation is readily absorbed by some important biomolecules such as DNA, protein and lipids and there has been extensive documentation of adverse effects of UV-B on marine algae, which include increase in mortality, reduction in growth and photosynthetic rates, inhibition of carbon and nitrogen assimilation, destruction of photosynthetic pigments and retardation of reproductive cell motility and so on, it is useful to focus our attention indeed to manifold adaptive ways by which UV-induced damage is mitigated. One of the mechanisms is the presence of UV-absorbing pigments (UVP). Compounds of these types have now been isolated from various marine organisms and identified as a series of mycosporine-like amino acids (MAAs). The MAAs are composed of a cyclohexenone ring attached with an amino acid side group. These compounds absorb in wavelengths ranging from 310 to 365 nm, spanning both UV-B and UV-A (320-400nm) portions of the solar spectrum, but transmit photosynthetically active radiation (PAR; 400-700nm), Seven MAAs have so far been isolated from marine macroalgae and identified as mycosporine-glycine, shinorine, porphyra-334, palythine, asterina-330, palythinol, usujirene and

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palythene. The role of MAAs as UV protectants is inferred from observations that their concentrations are correlated with the environmental UV fluences organisms can experience [1].

For instance a halotolerant cyanobacterium Aphanothece sacrum thrives in extreme salinity with accumulation of a potent osmoprotectant glycine betaine. Recently, this cyanobacterium was shown to accumulate sunscreen molecule mycosporine-2-glycine significantly at high salinity [2].

These A.A. and other researchers [3-5] investigated upon the effects of increase of nitrate and salinity in *Aphanothece sacrum* on the accumulation of glycine betaine and mycosporine-2-glycine. With elevated nitrate concentrations at high salinity, intracellular levels of both metabolites were enhanced. Six-fold high nitrate concentration increased the relative amounts of glycine betaine and mycosporine-2-glycine to be 1.5 and 2.0 folds compared with control condition.

Now, Aphanothece sacrum powder is available on the global market from Japan,it is not expensive since it is commonly employed as foodstuff and is admitted in INCI and is commonly used in Japan in cosmetics as: Absorbent; Emulsion stabilising; Film forming; Viscosity controlling. Albeit some suppliers disclaim its peculiar capabilities like: Moisturizing effect (10-fold higher mosisture retention capacity campared to hyaluronic acid); Function as a barrier to protect skin from external stimulus; Anti-inflammatory effect.

But nobody has hitherto studied the implications of its capacity to absorb the perilous UB rays so that it could be inserted in cosmetic items as well.

Aphanothece sacrum powder is available on the global market from Japan,it is not expensive since it is commonly employed as foodstuff.

Following the suggestions of the above mentioned A.A., Aphanothece sacrum powder was grown photoautotropically (70 μE m^{-2} sec^{-1}) in blue–green liquid (BG,that is a 0.13% bezalkonium chlride aqueous solution) containing 18 mM $NaNO_3$ and Turk Island salt solution, at 30°C for 14 days prior to the stress treatment. For high-nitrate-salt experiment, sodium nitrate concentration was increased from 18 mM (1X) to 54 mM (3X) and 108 mM (6X), respectively, and the concentration of NaCl in growth medium was changed from 0.5 to 2.0 M.

We have not detected the arousal of levels of glycine-betaine and mycosporine-2-glycine, since the paper was exhaustive and A.A. asserted that in this condition, glycine-betaine (GB) content was increased from ~7.5 to 29.5 μmol/g and then, under 6X nitrate condition, GB content was ~1.5 times higher than control condition after 15 days treatment.

The procedure was not complicated and not expensive at all.

MATERIALS AND METHODS

A gel made up with 2 g of Aphanotece sacrum powder (grown photoautotropically in blue–green liquid containing 18 mM $NaNO_3$ and Turk Island saltsolution, at 30°C for 14 days) in 1.5% karaya gum aqueous solution was prepared for the first series of experiments on 6 volunteers, and another gel made up with 2 g of Aphanotece sacrum powder following the above mentioned method in 1.5% karaya gum aqueous dispersion was used for the second series of experiments in order to evaluate the capacity of the cyanobacterium to protect skin from UVB rays under long exposure to artificial sun (owing to a normal sun lamp).

Every series of volunteers guarantees the real scrutinizing of the 6 kinds of phototypes, as in Table 1:

Here follows Table 2 where the Sun Protection factor advisable for each phototype is recorded:

We have selected 12 volunteers (A,B,C,D,E,F,G,H,I,L,M,N)

A and G belonging to phototype I
B and H belonging to phototype II
C and I belonging to photype III
D and L belonging to phototype IV
E and M belonging to phototype V
F and N belonging to phototype VI.
The first series of experiments comprised volunteers A,B,C,D,E,F
The second series of experiments comprised volunteers G,H,I,L,M,N.

The first series underwent to the application of the first gel (idest 2 g of Aphanotece sacrum powder (grown photoautotropically in blue–green liquid containing 18 mM $NaNO_3$ and Turk Island saltsolution, at 30°C for 14 days) in 1.5% karaya gum aqueous solution.

The second series underwent to the application of the second gel (idest 2 g of Aphanotece sacrum powder following the above mentioned method in 1.5% karaya gum aqueous solution).

The calculation of the SPF is obtained resolving the following equation:

$$SPF = \frac{\text{Minimal erythemal dose in sunscreen protected skin (MEDp)}}{\text{Minimal erythemal dose in unprotected skin (MEDu)}}$$

After two single experiments on C and I (corresponding to phototype III: Burns moderately: tans gradually) we have determined the SPF for gel number One, that is 18.

After two single experiments on B and H (corresponding to phototype II. Always burns easily: tans minimally) we have stated that the SPF of gel number Two is 56.

The experiments were carried for seven days (5,10,15,20,25,30 and 35 min of exposure every day) once a day onto the six volunteers of the first series and onto the six volunteers of the second series, by the aids of a normal sun lamp, after application of the gels (number one for the first series and number two for the second series).

RESULTS

In Table 3 it is possible to behold the results obtained after seven days of applications of the two gels and exposure to gradual exposure to the sun lamp.

Table III: Observations after one week of experimentations onto the 12 volunteers.

DISCUSSIONS

It is easy to comprehend that a cosmetic that reveals a SPF equivalent to 56 is able to minimize at all the chances of burning.

It is welknown, indeed, that phototype I may be protected exclusively by inorganic powerds (titanium dioxide,barium sulphate,kaolin etc.), even though thanks to the application of gel two it is possible to have a minimal tanning.

Table 1: The series of phototypes in Man

Phototype I	Always burn easily: never tans
Phototype II	Always burns easily: tans minimally
Phototype III	Burns moderately: tans gradually
Phototype IV	Burns minimally: always tans well
Phototype V	Rarely burns: tans profusely
Phototype VI	Never burns, deeply pigmented

Table 2: Fitzpatrick's Classification of Skin Phototypes

Phototype	Recommended SPF
I	>40
II	20-40
III	7-20
IV	6-15
V	5-10
VI	4

Table 3: Observations after one week of experimentations onto the 12 volunteers

Volunteer (gel n.One)	Observations after one week	Volunteer (gel n.Two)	Observations after one week
A	Burns hard: no tans	G	Burns minimally, tans moderately
B	Burns hard: tans minimally	H	No burns, tans moderately
C	Burns moderately: tans minimally	I	No burns, tans profusely
D	Burns minimally: tans well	L	No burns, tans well
E	No burns: tans well	M	No burns, tans well
F	No burns, already pigmented	N	No burns

This is the most suggestive impression we can argue, after these trials:

There is no way to protect phototype I, using natural or chemical sunscreens.

CONCLUSIONS

Pre-treated Aphanotece sacrum (2%) is sufficient to avoid burns in phototype I and guarantees always a long lasting tanning for almost all the phototypes.

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.

REFERENCES

1. Park JH, Han T. Overview of UV-absorbing Pigments in Marine Algae. Algae. 1999:14:201-12
2. Waditee-Sirisattha R, Kageyama H, Fukaya M, Rai V, Takabe T. Nitrate and amino acid availability affects glycine betaine and mycosporine-2-glycine in response to changes of salinity in a halotolerant cyanobacterium Aphanothece halophytica. FEMS Microbiology Letters. 2015;362:1-3.
3. Balskus EP, Walsh CT. The genetic and molecular basis for sunscreen biosynthesis in cyanobacteria. Science. 2010;329:6.
4. Flores E, Frias JE,Rubio LM: Photosynthetic nitrate assimilation in cyanobacteria.2005; Photosynth Res.83(117)33.
5. Xue L, Zhang Y, Zhang T, An L, Wang X. Effects of enhanced ultraviolet-B radiation on algae and cyanobacteria. Crit Rev Microbiol. 2005;31:79-89.

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Our Dermatology Online

Brief Report

Assessment of knowledge, attitudes and practices about sun exposure and sunscreen usage in outpatients attending a Dermatology Clinic in North India

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ABSTRACT

Background: There has been a significant increase in the cases of skin cancer throughout the world in the last few decades. Sun exposure and photoprotection-related behavior and knowledge are important aspects in the prevention of skin cancer. Despite various recommendations advising individuals to reduce their sunexposure, many people still do not utilize these sunprotection strategies. **Aims and Objectives:** To assess the knowledge, attitude and practices about sun exposure and sunscreen usage in outpatients attending a dermatology clinic **Materials and Methods:** Three hundred consecutive patients attending a dermatology clinic were enrolled for this questionnaire-based cross-sectional, descriptive study. **Results:** A total of 300 patients, comprising of 112 (37.33%) men and 188 (62.67%) women, aged between 14 and 78 (mean 47.13) years, were included in the study. Majority of the patients, 216 (72%) were in the 20-60 year age group and 226 (75.33%) belonged to an urban background. Assessment of the sunexposure revealed that 224 (74.67) respondents reported daily sunexposure whereas 76 (25.33%) had occasional sunexposure. Assessment of the knowledge revealed that 241 (80.33%) patients were aware of the adverse effects of excessive sunexposure but only 139 (46.33%) were aware of the carcinogenic effect of sunlight. A total of 212 (70.67%) respondents were aware of the benefits of sunscreens but only 156 (73.58%) were using the sunscreens. Sixty one respondents (39.1%) reported that they used sunscreens on a daily basis while 95 (60.89%) used it occasionally. Lack of awareness was the most common reason (44.67%). Newspapers (47%) and television (39%) were the most common source of information in our respondents. **Conclusions:** Information, education, and communication activities are imperative to educate people regarding the risks of excessive sunexposure and significance of preventive measures like sunscreens to bridge the gaps in their knowledge. While improvement in individual economic status and education remains highly desirable, mass media can play a pivotal role in creating awareness among masses.

Key words : Skin cancer, UV radiation, Sunscreeen, Photoprotection

INTRODUCTION

The sun is the principal source of environmental ultraviolet radiation (UVR). Excessive UVR exposure to skin leads to widespread epidermal and dermal cellular damage. DNA is probably the primary molecular target of injury, as a result of both direct UVB absorption and also secondary UVA-induced photosensitization reactions. The acute harmful effects of ultraviolet rays on the skin include damage to DNA, apoptosis, erythema, immunosuppression and an increase in pigmentation due to stimulation of melanogenesis, while the long-term effects include photoaging and photocarcinogenesis. Epidemiological studies have reported an increasing prevalence of cutaneous malignancies, which has been attributed to factors like large quantities of UV radiation entering the atmosphere due to the thinning of the ozone layer,

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living and travelling in sunny climates, excessive sunbathing and sun bed use, outdoor sports, and the usage of appliances and devices that emit UV radiation in domestic and industrial settings [1].

Sun exposure and photoprotection-related behavior and knowledge are important aspects in the prevention of skin cancer and photodermatoses. Primary skin cancer prevention strategies include increasing knowledge and awareness in individuals, changing sun protection behavior and implementing environmental policies and interventions. It is estimated that the regular use of photo-protectors during childhood may reduce the incidence of skin cancer by almost 80% [2]. The various protective strategies advocated include appropriate use of sunscreens, avoidance of UV exposure by seeking shade, staying indoors during the hours of peak UV radiation, and wearing protective clothing. Unfortunately, patient adherence to these recommendations has been disappointingly low and various barriers have been identified which include lack of knowledge, misconceptions regarding skin cancer risks, difficulty in initiating behavioral changes, and socioeconomic factors such as time and costs involved, etc [3].

This study was carried out to assess the knowledge, attitude and practices about sunexposure and sunscreen usage in outpatients attending a dermatology clinic.

MATERIALS AND METHODS

Consecutive patients attending outpatient dermatology clinic during July 2015 to June 2016 were enrolled for this questionnaire-based, cross-sectional, descriptive study. Children aged <14 years and severely ill patients were excluded from the study owing to their inability to comprehend or respond to the questionnaire. After informed written consent and assuring confidentiality, they were asked to answer a predesigned, structured questionnaire in their native language. The questionnaire had two parts with the first section for their sociodemographic details and the second section comprised questions aimed at assessing their knowledge, attitude, and perception for sunexposure and photoprotection.

RESULTS

A total of 300 patients, comprising of 112 (37.33%) men and 188 (62.67%) women, aged between 14 and 78 (mean 47.13) years, were included in the study. Their baseline demographic features are shown in Table 1. Majority of the patients, 216 (72%) were in the 20-60 year age group and 226 (75.33%) belonged to an urban background. Among the males, self employed and office workers were the most common occupational group (10.67% and 11.33% respectively), while among the females, homemakers were the most common group (48%). Fitzpatrick skin type III (27.33%) and IV (47.33%) were the most common skin types in our study. Assessment of the sunexposure

Table 1: Baseline characteristics of patients studied

Baseline characteristics	Number of patients (%) *n*=300
Gender	112 (37.33)
Males	188 (62.67)
Females	1 : 1.69
Male: female	28 (9.33)
Age (years)	
<20	28 (9.33)
20-40	102 (34)
41-60	114 (38)
>60	56 (18.67)
Social Background	
Rural	74 (24.67)
Urban	226 (75.33)
Occupation	
Men	
Office Workers	32 (10.67)
Self-Employed	34 (11.33)
Students	26 (8.67)
Farmers and Laborers	16 (5.33)
Defense Person	4 (1.33)
Women	
Homemakers	144 (48)
Students	30 (10)
Office Workers	14 (4.67)
Education Status	
Illiterate/School dropout/<12th standard	56 (18.67)
Graduate	156 (52)
Postgraduate	78 (26)
Professional	10 (3.33)
Fitzpatrick Skin Type	
Type II	12 (4)
Type III	82 (27.33)
Type IV	142 (47.33)
Type V	60 (20)
Type VI	4 (1.33)
Source of information	
Television	117 (39)
Radio	36 (12)
Newspaper	141 (47)
Books	81 (27)
Internet	59 (19.67)
Health personnel	31 (10.33)
Family and friends	42 (14)
No information	38 (12.67)

revealed that 224 (74.67) respondents reported daily sunexposure whereas 76 (25.33%) had occasional sunexposure. Among the respondents who had daily sunexposure, 102 (45.53%) had less than one hour of sunexposure daily whereas 96 (42.86%) had an exposure of 1-3 hours per day and only 26 (11.6%) had sunexposure more than three hours daily.

Assessment of the knowledge revealed that 241 (80.33%) patients were aware of the adverse effects of excessive sunexposure but only 139 (46.33%) were aware of the carcinogenic effect of sunlight. The most frequently identified adverse effects were sunburn (60.33%), blemishes (49%), freckles (42.3%), aging (31.33%) and wrinkles (29.66%), skin cancer (46.33%) and aggravation of acne (19.33%). When questioned regarding the awareness about sunscreens, 212 (70.67%) respondents were aware of the benefits of sunscreens and females (n=140) were more aware than the males (n=72), but only 156 (52%) were using the sunscreens. Of the 156 users of sunscreens, females (n=102) outnumbered the males (n=54). Sixty one respondents (39.1%) reported that they used sunscreens on a daily basis while 95 (60.89%) used it occasionally. When asked about the sunscreens, 74 (47.43%) users were aware of the Sun Protection Factor (SPF) of the product they were using and 64 (41.02%) respondents were using a sunscreen with SPF ≥30. Among the users of sunscreens, only 32 (20.51%) were using them twice a day and only 39 (25%) were using it over all the exposed sites including face, neck, arms and hands while the rest reported applying it over the face and neck only. The amount of sunscreen used per application was significantly less and 76.28% of the users (n=119) were applying less than 5ml sunscreen over the face and neck per usage. When enquired about the reasons for not using sunscreens, lack of awareness was the most common reason (44.67%), followed by whitish discoloration and excessive oiliness of the face post-usage (21.33%), lack of time for application (10.66%) and cost of sunscreens (5.33%). Newspapers (47%) and television (39%) were the most common source of information in our respondents, followed by books and internet but 38 (12.67%) respondents had no idea about the sunscreens.

DISCUSSION

Ultraviolet (UV) radiation has been classified by the International Agency for Research on Cancer (IARC) as a Group 1 carcinogen to humans [4]. Sun protection is therefore an important public health message for skin cancer prevention. Experts advocate the use of sunscreen, as well as other sunprotective measures like wearing protective clothing and sunglasses, wearing widebrimmed hats, and sun-avoidance to protect from sun exposure. Sunscreens have been proven to have protective effects against photoaging and reduce the incidence of skin cancers [5].

This population-based survey documents that the knowledge about risks of solar radiation in general population is suboptimal and even in respondents with adequate knowledge, the sunscreen usage is inadequate. In the present sample, 80.33% patients (n=241) were aware of the adverse effects of excessive sunexposure but only 46.33% (n=139) were aware of the relationship between sunexposure and skin cancers, which is much lower than those of the western populations. In a Brazilian study, 94.3% of the respondents were aware of the risks of sun exposure and 80.8% knew that the sunlight increases the risk of skin cancer [6]. In a study performed at the National Institute of Cancer on the USA, it was found that 77% of the participants knew that the sun increases the risks of skin cancer [7]. This low level of awareness in our study population could be attributed to minimal public awareness campaigns in our setup.

The knowledge regarding the sunscreens and their usage was higher among the females in our study, which was similar to the results of other studies. Devos et al. showed that knowledge regarding sunscreens and their regular use was considerably higher in the female participant group than in the male group [8]. Yurtseven et al also showed that while 90.6% of women used sunscreen, only 57.1% of men were using sunscreens [9]. In our study also, of the 156 users of sunscreens, females (n=102) outnumbered the males (n=54). This can be related to the fact that women are more concerned about cosmetics and skin care. In our study 52% respondents were using sunscreens, out of which only 20.33% (n=61) were using sunscreens on a regular basis. In a study by Fabris et al [6], 74.1% respondents were using sunscreens on a regular basis, whereas in a study by Al-Mutairi *et al*. [10], 80% of the respondents had been using sunscreens regularly and 27% were using repeated applications of sunscreen. In our study, 47.43% (n=74) users were aware of the SPF of the product they were using, while 96% of sunscreen users were aware of the product's SPF in the study by Al-Mutairi [10]. The amount of sunscreen is an important factor in determining the sunprotection

offered by the product. A European study showed that individuals used one-fifth (0.3-0.5 mg/cm2) of the quantity recommended by the manufacturer on the product packaging (2 mg/cm^2) [11]. In our study too, only 23.72% (n=37) users were using an adequate amount of sunscreen. Lack of awareness was the most common reason cited by the respondents for not using sunscreens in our study, while in a Brazilian study, the lack of patience to apply (34.2%), followed by messing up the tan (31.6%) were the most common reasons [6]. Television and the printed media were the most common sources of information in our study, more so than health professionals.

Experience from various countries demonstrates that it is possible to improve the sun protection behaviors and attitudes of a population with public health campaigns. Use of television and the printed media, particularly newspapers, are the key in targeting a large population but campaigns should also incorporate alternative approaches such as healthcare professionals and the internet to make these campaigns more effective.

CONCLUSIONS

As the incidence of skin cancers is on the rise, proper education of the masses regarding the adverse effects of sunexposure and the use of various photoprotective measures including sunscreens is imperative. The respondents evaluated in our study had an acceptable understanding of the risks of sun exposure; however, large majority were unaware of benefits of sunscreens or were not using sunscreens in a proper manner to be of any benefit.

LIMITATIONS

There were several potential limitations in our study. Firstly, the sample size was small and as convenience sample of respondents from only one centre was surveyed; thus, caution must be exercised in extending our findings to the whole population, especially in other geographical regions. Moreover, results of this study rely on self-reported data, which could introduce recall and social desirability biases.

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.

REFERENCES

1. Diepgne TL, Mahler V. The epidemiology of skin cancer. Br J Dermatol 2002;146(Suppl 61):1-6.
2. Balato N, Gaudiello F, Balato A, Monfrecola G. Sun habits in the children of southern Italy. J Am Acad Dermatol. 2007;57:883-7.
3. Melia J, Pendry L, Eiser JR. Harland C, Moss S. Evaluation of primary prevention initiatives for skin cancer: a review from a U.K. perspective. Br J Dermatol. 2000;143:701-8.
4. The International Agency for Research on Cancer Working Group on artificial ultraviolet (UV) light and skin cancer. Int J Cancer. 2006; 120:1116–22.
5. Geller A, Cantor M, Miller D, Kenausis K, Rosseel D, Rutsch L, et al. The Environmental Protection Agency National Sun Wise School Program: Sun protection education in US schools (1999-2000). J Am Acad Dermatol. 2002;46:683-9.
6. Fabris MR, Martignago BC, Fabris TR, Duraes ES, Blanco LF. Assessment of knowledge of skin cancer prevention and its relation with sun exposure and photo protection amongst gym academy members on the south of Santa Catarina, Brazil. An Bras Dermatol. 2012;87:36-43.
7. Gebert B, Johnston K, Bleecker T, McPhee S. Attitudes about skin cancer prevention: A qualitative study. J Cancer Educ. 1996;11:96-101.
8. Devos SA, Baeyens K, Van Hecke L. Sunscreen use and skin protection behavior on the Belgian beach. Int J Dermatol. 2003;42:3526.
9. Yurtseven E, Ulus T, Vehid S, Koksal S, Bosat M, Akkoyun K. Assessment of Knowledge, Behaviour and Sun Protection Practices among Health Services Vocational School Students. Int J Environ Res Public Health. 2012;9:2378-85.
10. Al-Mutairi N, Issa BI, Nair V. Photoprotection and vitamin D status: A study on awareness, knowledge and attitude towards sun protection in general population from Kuwait, and its relation with vitamin D levels. Indian J Dermatol Venereol Leprol. 2012;78:342-9.
11. Autier P, Boniol M, Severi G, Dore JF. Quantity of sunscreen used by European students. Br J Dermatol. 2001;144:288-91.

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Our Dermatology Online

Case Report

A giant groin lipoma mimicking an inguinal hernia: A case report

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ABSTRACT

An inguinal hernia repair operation is a common procedure in general surgery. However, several other conditions such as inguinal lymphadenopathy, groin lipoma, spermatic-cord lipoma and sarcoma may have similar clinical findings with inguinal hernia. Preoperative diagnosis is crucial to make the appropriate treatment plan. An accurate preoperative diagnosis prevents both unnecessary operations and surgical complications. Radiological imaging techniques such as ultrasonography, computed tomography and magnetic resonance imaging should be considered in the differential diagnosis of inguinal masses. Hereby, we present a 41-year-old Caucasian male patient with a giant lipoma in the right groin which mimicked an inguinal hernia. Preoperative ultrasound imaging helped us to make an initial diagnosis of lipoma which was subsequently confirmed by histopathological examination.

Key words: Hernia; Inguinal; Lipoma

INTRODUCTION

Lipomas are benign adipose tumors which usually present in the subcutaneous tissues. Lipomas are characterized as slow-growing, soft, mobile, round and asymptomatic masses on the head, neck, shoulders, and the back. They usually develop between the ages of 40-60. Solitary lipomas are more common in women, whereas multiple lesions usually appear in men. Histopathologically, lipomas consist of mature adipocytes with a fibrous capsule. The differential diagnosis includes epidermoid cyst, subcutaneous tumors, nodular fasciitis, liposarcoma, metastatic disease, erythema nodosum, nodular subcutaneous fat necrosis, Weber-Christian panniculitis, vasculitic nodules, rheumatic nodules, sarcoidosis and hematoma. Since lipomas are generally not painful, they usually do not require any further treatment. Even it is rare, malignancy should be kept in mind especially in fast-growing tumors. Magnetic resonance imaging (MRI) may be useful to differentiate lipomas from liposarcomas. Steroid injections, liposuction, and surgical excision are the possible treatment options *[1]*.

CASE REPORT

A 41-year-old Caucasian male patient with an asymptomatic inguinal swelling presented for further clinical evaluation. The patient stated that the lesion gradually enlarged within the past 5 years. The patient confirmed that he did not receive any prior treatment. Past medical history and family history were both unremarkable. The physical examination revealed a skin-colored, soft, well-defined, subcutaneous mass in the right inguinal region (Figs. 1 and 2). Ultrasonography showed an isoechoic, subcutaneous nodule with regular borders measuring 9x6x4 cm in size. Laboratory tests including complete blood count, C-reactive protein, INR and urinalysis were all in normal limits. Serum levels of hepatitis B surface antigen, antibodies against hepatitis C virus and anti-human immunodeficiency virus antibody were all negative. The biochemistry

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panel was normal except the slightly increased serum glucose; 115 mg/dL (normal range: 70-110 mg/dL) and alanine aminotransferase; 43 u/L (normal range: 0-40 u/L). The lesion in the right inguinal region of the patient was completely removed under local anesthesia with 2% prilocaine (Citanest®) (Figs. 3 and 4). The histopathological examination of the lesion revealed a giant lipoma with mature adipocytes surrounded by a fibrous capsule (Figs. 5 and 6).

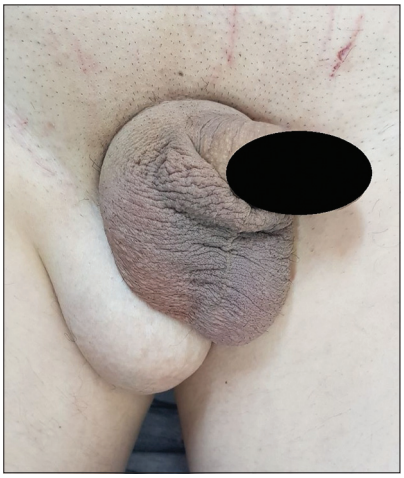

Figure 1: A skin-colored, well-defined, subcutaneous mass in the right groin measuring 9x6x4 cm in size

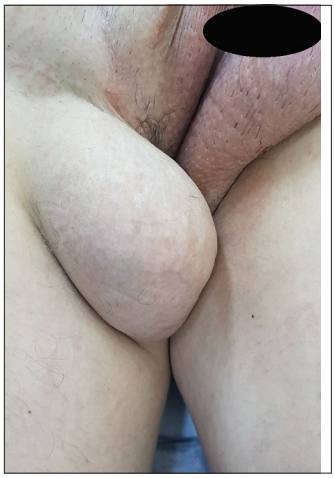

Figure 2: Perioperative view of the lesion

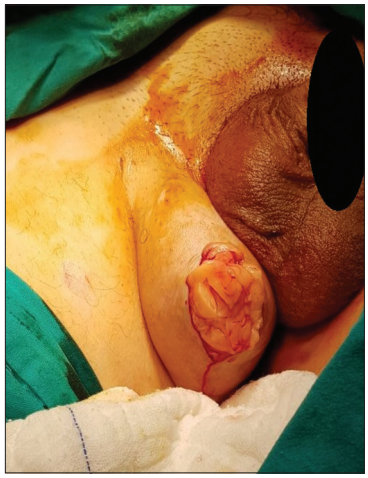

Figure 3: Intraoperative view of the lesion

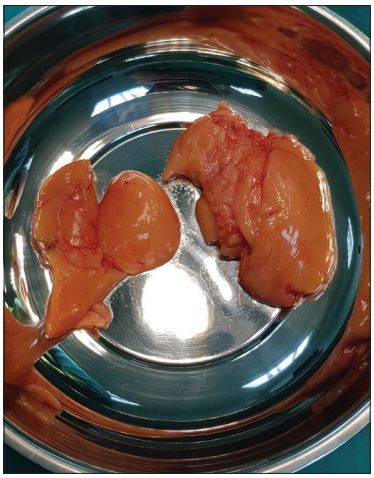

Figure 4: Surgically excised specimen

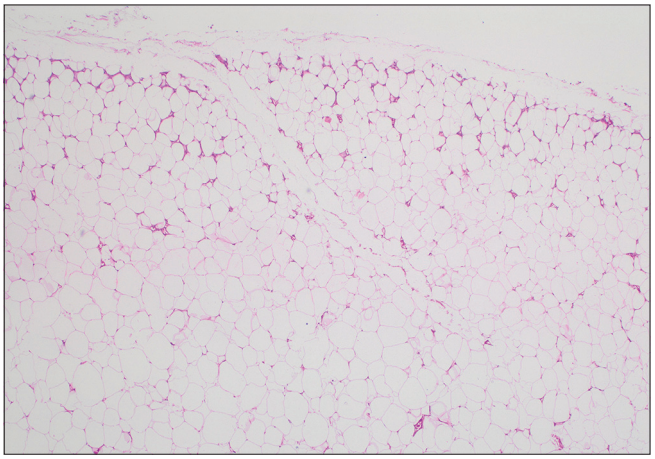

Figure 5: Mature adipocytes separated by a fibrous septa (H&Ex40)

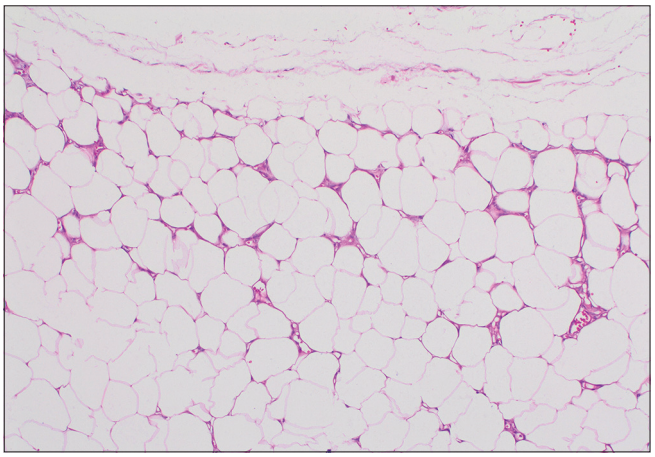

Figure 6: Closer appearance of the adipocytes (H&Ex100)

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

DISCUSSION

A lipoma in the inguinal region can easily be misdiagnosed as an inguinal hernia, since both a lipoma and an inguinal hernia may lead to similar symptoms and physical examination findings. Gerych et al. reported a 70-year-old female patient with a 20x12 cm sized lipoma in the right groin mimicking an inguinal hernia. Gerych et al. stated that they performed preoperative ultrasonography and computed tomography (CT) in order to differentiate a lipoma from an inguinal hernia, aneurysm of great saphenous vein, inguinal lymphadenopathy and a cold abscess *[2]*.

Another clinical entity which presents with inguinal hernia signs and symptoms is a pure spermatic-cord lipoma. Jo et al. reported a 33-year-old male patient with a spermatic-cord lipoma which was initially misdiagnosed as an inguinal hernia. Spermatic-cord lipomas are usually detected incidentally during the hernia repair surgery. Therefore, Jo et al. advised to perform an abdominopelvic CT and MRI in patients with a slowly growing, non-reducible, fixed, relatively hard and non-tender mass *[3]*. Moreover, an encysted spermatic cord hydrocele was described in a male patient as a painful, well circumscribed, 4 cm sized swelling in the left inguinal region which clinically mimicked an incarcerated inguinal hernia *[4]*. In addition, a sarcoma is a rare, malignant, soft tissue tumor which usually occurs in the thigh and retroperitoneum. A sarcoma should also be kept in mind in the differential diagnosis of an inguinal hernia. Therefore, preliminary diagnosis of a lump in the inguinal region is crucial both to offer the best available treatment option and to perform the most appropriate surgery *[5]*.

In conclusion, inguinal hernia repair is a common surgical procedure in general surgery. However, several other conditions such as a giant lipoma can clinically mimick an inguinal hernia. In such cases, radiological examinations such as ultrasonography, CT and MRI may be helpful to reach a preliminary diagnosis prior to the surgical intervention.

CONSENT

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

REFERENCES

1. Salam GA. Lipoma excision. Am Fam Physician. 2002;65:901-4.
2. Gerych I, Ivankiv T, Ogurtsov O, Kalynovych N. Giant right groin lipoma mimicking inguinal hernia. Int J Surg Case Rep. 2015;12:106- 7.
3. Jo DI, Choi SK, Kim SH, Kim CK, Chung H, Kim HS. The case of huge pure lipoma of the spermatic cord misdiagnosed as inguinal hernia. Urol Case Rep. 2017;13:10-2.
4. Manimaran D, Karthikeyan TM, Khan DM. Encysted spermatic cord hydrocele in a 60-year-old, mimicking incarcerated inguinal hernia: A case report. J Clin Diagn Res. 2014;8:153-4.
5. Valeshabad AK, Walsh A, Lloyd GL. An important mimic of inguinal hernia. Urology. 2016;97:e11.

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Our Dermatology Online

Case Report

Urticaria multiforme: A commonly misdiagnosed entity

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ABSTRACT

Cutaneous hypersensitivity reactions in paediatric age group have varied presentations. Urticaria multiforme is a characteristic morphologic type of self resoluting hypersensitivity reaction secondary to drugs or infections, characterised by acute onset annular, polycyclic wheals with ecchymotic centres. We present an eighteen month female and a 5 years male child who presented to us with itchy erythematous blanching annular wheals with dusky centre and were diagnosed as urticaria multiforme. It resolves completely with antihistamines and therefore need to be differentiated from severe reaction patterns.

Key words: Urticaria multiforme; Hypersensitivity; Dusky centre

INTRODUCTION

Urticaria multiforme (UM) is also known as acute annular urticaria. It is characterized by annular, arciform, polycyclic erythematous blanchable wheals with violaceus dusky centre [1]. These lesions are sudden in onset and subside in an hour or two, otherwise resembling erythema multiforme. There is associated acral edema [1]. We report two cases of urticaria multiforme to emphasize that UM is a distinct variant of urticaria and should be differentiated from other disorders with similar morphology.

CASE REPORT

Case 1

An eighteen months female child presented with history of fever and cough for 5 days and itchy erythematous lesions all over the body for past 2 days. The lesions were persisting for 24 hours with onset of new lesions at other locations. The child was otherwise active. On examination child was febrile with a temperature of 100.4^0 F. There was no lymphadenopathy or joint swelling. Cutaneous examination showed multiple wheals which were erythematous annular and polycyclic, present all over the body with dusky purpuric centre (Fig. 1a) and associated edema of hands and feet (Fig. 1b). Dermographism was positive. Haematological examination, urine microscopy, erythrocyte sedimentation rate were within normal limits. Antstreptolysin O, blood and urine culture were non-contributory. C-reactive protein was slightly raised. As the clinical presentation of patient was quite characteristic she was diagnosed to be having urticaria multiforme and prescribed antihistamines; the wheals disappeared within few hours but few new lesions appeared at different location. Antihistamines were continued and the child was completely asymptomatic after 2 weeks follow up, with no pigmentation or scarring.

Case 2

A 5 years male child with history of discharge from left ear for 4 days, for which he was taking amoxicillin presented with severely itchy erythematous raised plaques all over the body for 2 days. On examination child was afebrile, but irritable due to pruritus. Cutaneous examination showed multiple variable sized erythematous annular wheals present all over

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the body (Figs. 2a and 2b) and positive (Fig. 2c). All routine investigations were normal. Child was given antihistamines; lesions subsided within 24 hours of medication but reappeared next day. The dose of antihistamine was doubled and child was completely asymptomatic after 2 weeks (Fig. 2d).

DISCUSSION

In 1997 Tamayo-Sánchez et al observed acute onset of annular urticarial and named the entity as acute annular urticarial(AAU) [2]. Ten years later Shah et al described acute urticarial lesions with dusky centre and due to its similarity with erythema multiforme, named it as urticaria multiforme(UM) [3]. Both these studies had similar diagnostic criteria except for few differences. The latter studies considered UM to be a variant of acute urticaria with dermographism [4]. Both the terms AAU and UM are now used synonymously. Terminology UM is debatable because of the absence of variable forms and the nomenclature of erythema multiforme like urticaria was proposed [1].

Urticaria multiforme (UM) is a morphological variant of acute urticaria. It is seen most commonly in children older than 4 months but less than 4 years of age [5]. Occasionally UM in neonates and adolescents have also been reported [1]. UM is a hypersensitivity reaction usually secondary to viral infections or certain drugs. Patient usually has a history of fever, sore throat, otitis media, upper respiratory, gastrointestinal infection preceding the episode. There may be history of onset of lesions after intake of drugs like amoxicillin, cephalosporins or macrolides, furazolidones, aspirin or after immunization [6]. Like any urticaria, the lesions start as erythematous macule, papule which progress to itchy, blanchable wheals which are transient (<24 hours), healing without pigmentation or scarring. Morphology of the lesion in erythema multiforme (EM) and UM is similar with annular, polycyclic lesions and dusky centre. In contrast to target lesions in EM lesions, they are targetoid in UM. The lesions in EM persist for few days while they are transient in UM. Also UM is associated with dermographism, pruritus and acral edema which are absent in EM. Other cutaneous hypersensitivity responses with similar presentation are urticarial vasculitis and serum sickness like illness [7]. Urticarial vasculitis(UV) lesion is annular but not targetoid, persisting for more than 24 hours, burning is the common complaint rather than pruritus. UV is uncommon in paediatric age group less than 5 years. UV unlike UM heals with hyperpigmentation. In

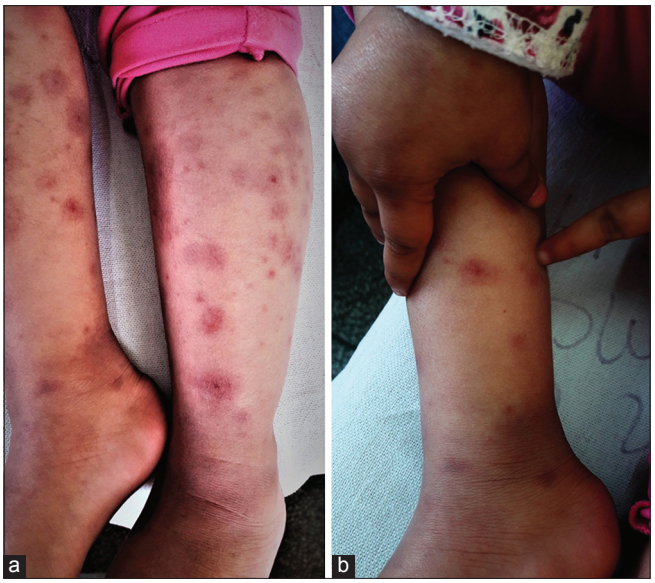

Figure 1: (a) Erythematous, targetoid wheals on both lower limbs (b) Acral edema with targetoid lesions.

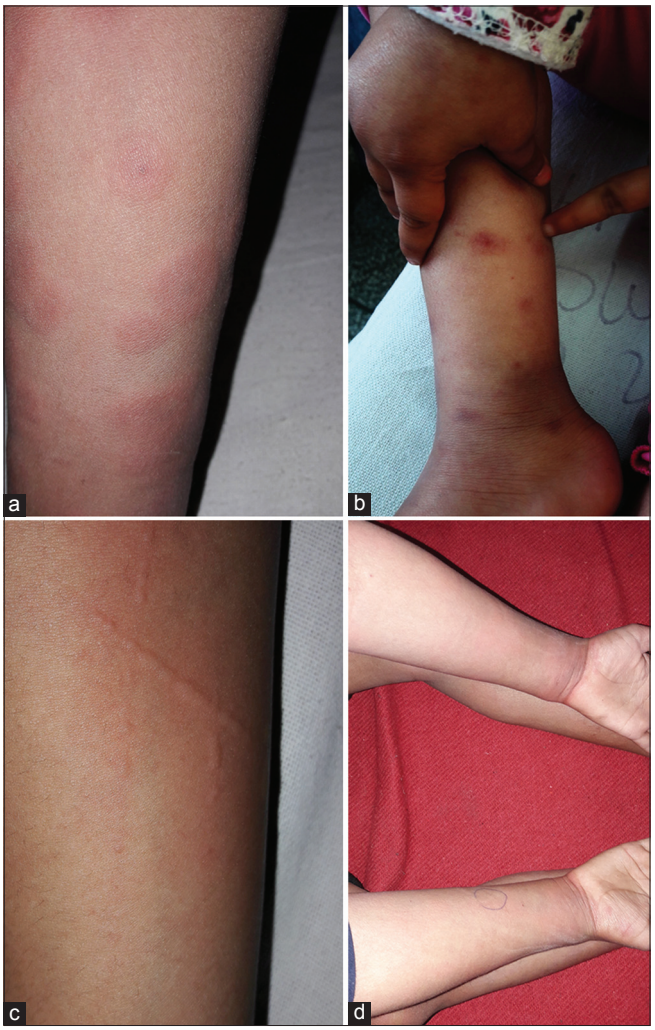

Figure 2: (a) Erythematous wheals with dusky purpuric centre over forearm, (b) Lesions showing targetoid morphology over bilateral upper and lower limbs (c) Remission of lesion with medication within 24 hours (d) Dermographism after stroking forearm of child.

serum sickness like reaction child is sick with fever, lymphadenopathy, arthralgia and acral oedema. Acute haemorrhagic edema of infancy may also present as a targetoid lesion but is purpuric and leaves hyperpigmentation [8].

Clinical history and examination are sufficient to diagnose UM. Histopathology of the lesions show dermal edema and perivascular lymphocytic with eosinophilic infiltrate, with a normal epidermis in contrast to basal layer necrosis and interface dermatitis seen in EM and no inflammatory infiltrate in urticaria. Routine haematological tests, C-reactive proteins, IgE levels are normal.

UM being a hypersensitivity response is not an uncommon disease in children and is self limiting in 10-14 days [9]. It needs to be clinically differentiated from other similar but severe reaction pattern, so that the load of investigations can be minimised. Antihistamines are the only treatment required.

CONSENT

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

REFERENCES

1. Tarnes L, Patel T, Skinner RB. Urticaria multiforme - a case report. Pediatr Dermatol. 2011;28:436–8.
2. Tamayo-Sanchez L, Ruiz-Maldonado R, Laterza A. Acute annular urticaria in infants and children. Pediatr Dermatol. 1997;14:231-4.
3. Shah KN, Honig PJ, Yan AC. "Urticaria multiforme": a case series and review of acute annular urticarial hypersensitivity syndromes in children. Pediatrics. 2007;119:e1177–e83.
4. Sempau L, Martín-Sáez E, Gutiérrez-Rodríguez C, Gutiérrez-Ortega MC. Urticaria Multiforme: A Report of 5 Cases and a Review of the Literature. Actas Dermosifiliogr. 2016;107:e 1-5.
5. Guerrier G, Daronat J-M, Deltour R. Unusual Presentation of Acute Annular Urticaria: A Case Report. Case Rep Dermatol Med. 2011;2011:604390.
6. Fung IN, Berger EM, Castelo-Soccio L, Brown Whitehorn TF. Urticaria multiforme in an 18-year-old girl. Allergy Clin Immunol Pract. 2013:1;520-1.
7. Mathur AN, Mathes EF. Urticaria mimickers in children. Dermatol Ther. 2013:26; 467-75.
8. Emer JJ, Bernardo SG, Kovalerchik O, Moneeb Ahmad BS. Clin Aesthet Dermatol. 2013:6;34–9.
9. Myers SR, Lavelle J. Picture of the month—quiz case. Pneumonia with associated urticaria multiforme rash. Arch Pediatr Adolesc Med. 2009:163;1157.

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Our Dermatology Online

Case Report

Fixed cutaneous sporotrichosis

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ABSTRACT

The sporotrichosis is the most common sub cutaneous mycosis in our country and the lymphocutaneous type is the most frequent clinical presentation. Case report: a case of cutaneous fixed sporotrichosis has been reported in a woman of 46 years old with good response to itraconazole. Conclusion: the fixed cutaneous type is less well recognized. In this type, the lesions are restricted to the site of inoculation without any lymphatic spread. Important clues from the clinical history, such as travel and occupation, can help to raise the suspicion of this infection in the differential diagnosis. tissue culture are necessary to confirm the mycological diagnosis.

Key words: Cutaneous fixed sporotrichosis, *Sporothrix schenckii*, *itraconazole*

INTRODUCTION

Sporotrichosis is a subcutaneous fungal infection first described by Benjamin Schenks in 1898. It is caused by the dimorphic fungi complex Sporotrhix [1,2] as the result of traumatic inoculations with contaminated material, usually spindles and thorns. Thus, this disease is popularly known as "rose gardener´s disease", though it may also be transmitted by cats [1,2]. It is considered as a mycosis of global distribution, but its observed with more frequency in tropical countries, having higher incidence in Latin America [1,3]. The clinical types of sporotrichosis are lymphocutaneous sporotrichosis, fixed cutaneous and extracutaneous. Moreover, the heterogeneous morphology of lesions often makes the clinical diagnosis difficult, thereby leading to delayed treatment [1-3].

CASE REPORT

Female patient, 46 years, who consulted a private doctor due to her dermatological condition in her face region was sent for diagnosis. Upon examination, a dermatosis was found on the left cheek, a plaque with ulcerations on an erythematous base (Fig. 1). The rest of the physical exam was within normal limits.

Familiar and personal history was found negative.

The patient indicated that the disease began 3 months ago with a small asymptomatic "pimple" on the left cheek, discharging pus, and afterwards, slowly increased and ulcerated. She received different antibiotics and non-specific creams with no effective results Three clinical diagnosis were made based on the clinical data: inflammatory, neoplasic or infectious disease. A biopsy was performed, and the histologic sections of skin stained with HE, showing epidermis with hyperkeratosis and irregular acanthosis. From the papillary dermis to the deep reticular dermis, a diffuse granulomatosis infiltrate present at cutting surface was observed (Fig. 2).

At a higher magnification, the infiltrate was formed by granulomas with suppurative center, lymphocytes, histiocytes, plasma cells and abundant giant cells of Langhans type. Moreover, large number of yeast was observed presenting an elongated and oval shape, which was evident on PAS staining (Fig. 3). Afterwards the clinical diagnoses of fixed cutaneous sporotrichosis was performed. Upon re-interrogation, the patient revealed receiving a blow on her left cheek with a

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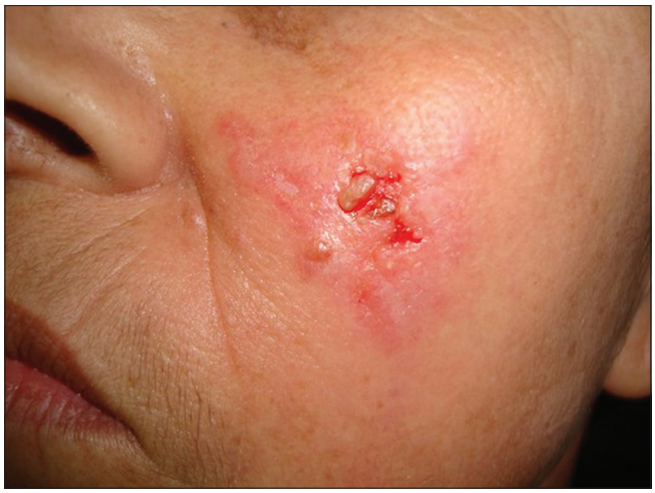

Figure 1: Ulcerations on a well-defined erythematous plaque of 2 x 2 cm.

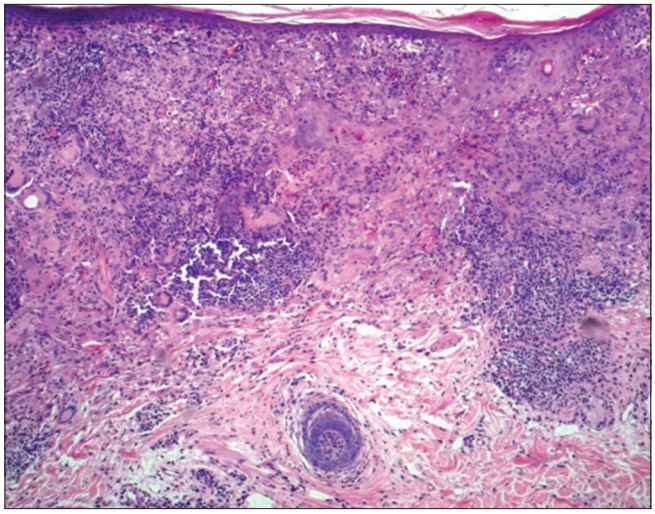

Figure 2: Epidermis with hyperkeratosis, irregular acanthosis. From the papillary dermis to the deep reticular dermis, a diffuse granulomatosis infiltrate.

can of gaseous water about 9 months ago. A fungus cultivation was carried in Sabouraud´s glucose agar, surface growth was brown, glabrous and yeast like, with reverse also brown (Fig. 4). The microscopy confirmed *Sporothrix schenkii* (Fig. 5), the microscopic morphology of the colony was septate, branching hyphae and cluster spores. Moreover, conidia are in a flowerette arrangement at the single conidiophores (Fig. 6). The final diagnosis of the patient was fixed cutaneous sporotrichosis and itraconazole 200 mg daily was prescribed for 6 weeks, resulting in clinical cured (Fig. 7).

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

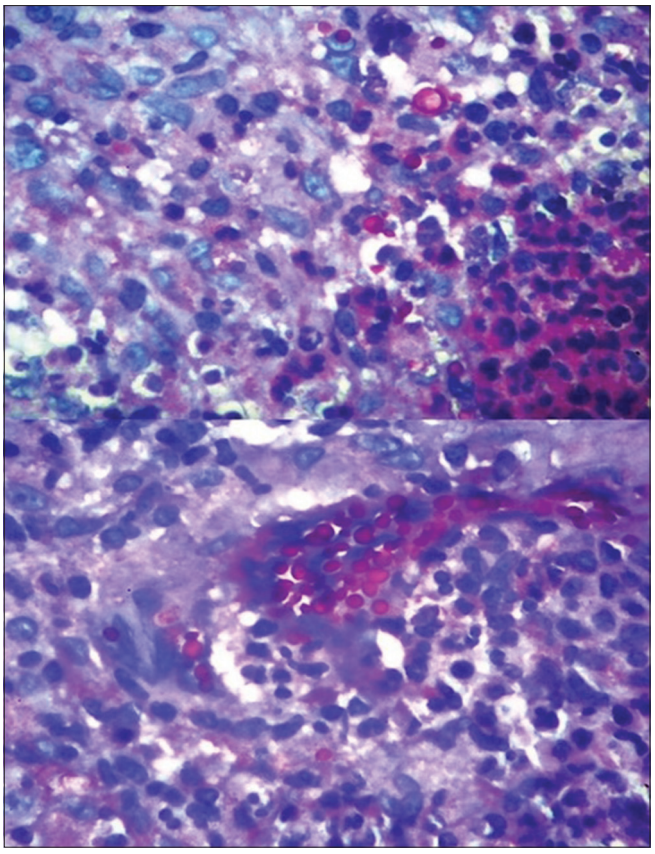

Figure 3: Close up the infiltrate were formed by granulomas with suppurative center, lymphocytes, histiocytes, plasma cells and abundant Langhans type giant cells, a large number of yeast are observed in elongated and oval form, which were evident on PAS staining.

DISCUSSION

Sporotrichosis, or rose gardener´s disease [1], was first described in 1898 by Benjamin Schenk at Johns Hopkins Hospital and was named by Hektoen and Perkins in 1900 [1,2]. The disease is a polymorphic subacute or chronic subcutaneous mycosis. Sporotrichosis is dispersed worldwide and more prevalent in tropical and subtropical areas, with the highest prevalence in Central and South America, Africa and Japan [1,3,4]. Sporotrichosis is caused by a complex group of dimorphic fungus species called *Sporothrix* [1,5-7], the major species includes *Sporothrix brasiliensis*, *S. schenckii*, *S. globosa* and *S. albicans* and *S. mexicana* [8-11]. *Sporothrix* is present on the natural environment in soil, dead wood (Splinter and thorns favor the growth), mosses and cornstalks [12]. But has never been observed as plant pathogen, probably due to the antifungal activity of plants [10].

It grows in the environment in a mold-like form at temperatures between 25-30°C. Moreover, it develops

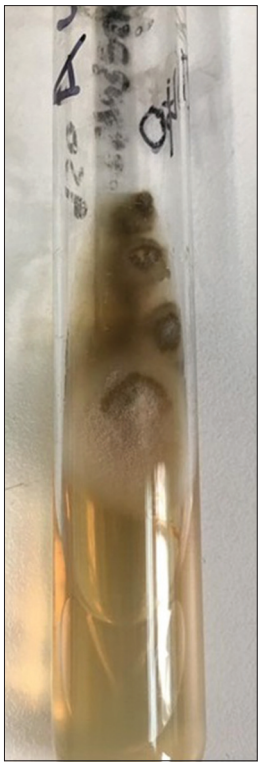

Figure 4: Sabouraud´s glucose agar, surface growth is brown, glabrous and yeast like, and reverse is brown.

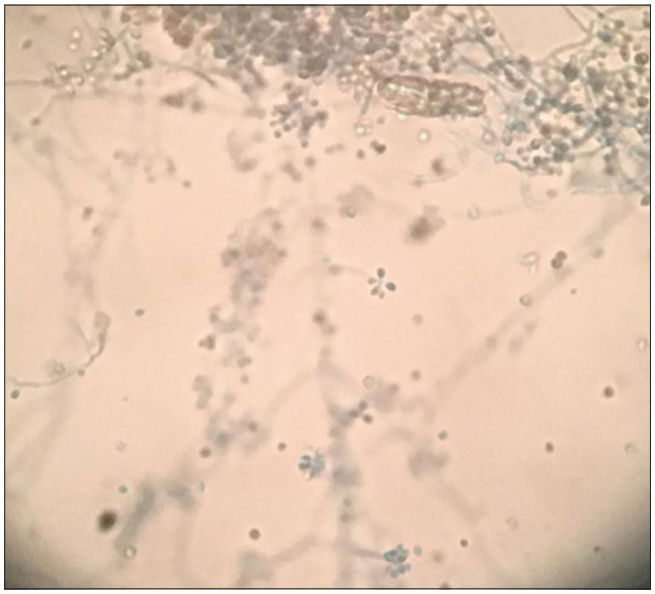

Figure 5: Microscopic morphology of the colony septate and branching hyphae, cluster spores.

into yeast-like form in host tissues at 35-37°C [5,13]. It is considered an occupational disease of particular risk for agriculturists, gardeners, florists and foresters [2,5]. Furthermore, alcoholism and diabetes have also been correlated as risk factors associated to it [2].

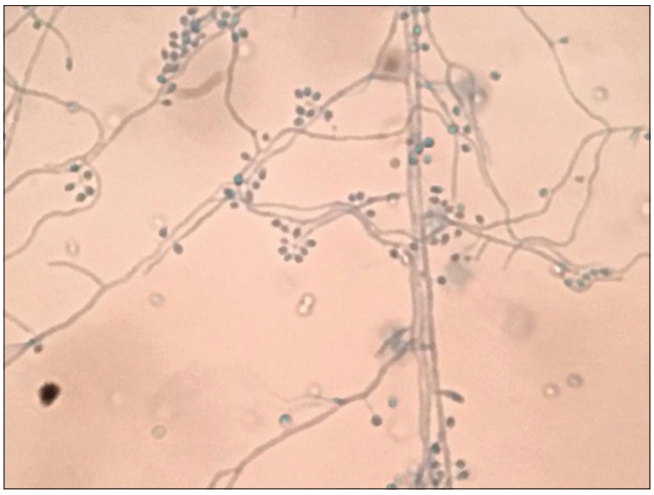

Figure 6: Conidia are in a flowerette arrangement at the single conidiophores.

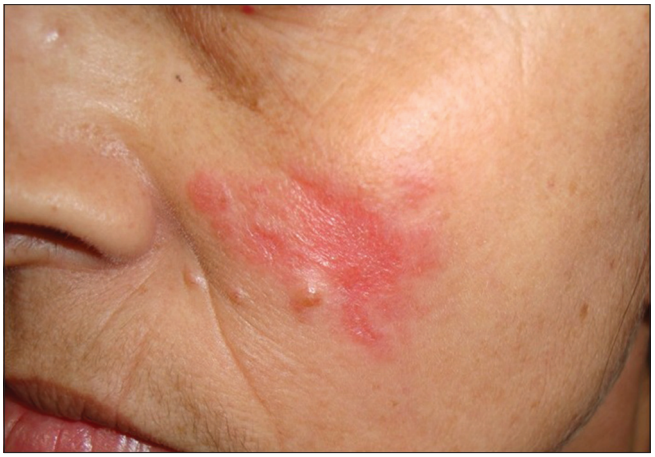

Figure 7: Control post six weeks of treatment with atrophic scar.

This mycosis is more commonly seen in tropical and subtropical countries, principally in Mexico, Central America and Peru. However, the region of Abancay in Perú is considered hyperendemic with an incidence of 48 to 60 cases per 100 000 population [11].

This subcutaneous mycosis strongly depends on exposure [12]. It affects individuals regardless of age and gender and it is easily acquired by in children and young adults. Traumatic inoculation is the typical mode for acquisition of cutaneous infection in immunocompetent hosts [5,7], and typically affects the face region and extremities. More recently, it has also been found to be transmitted via domestic cats and rodents in Brazil and Japan [1,8,14].

Several factors such as host immunity, virulence of the inoculated strain and the depth of traumatic inoculation may determinate the different types of presentation [3].

Conventionally, three clinical forms are described: 1) Fixed Cutaneous (nodulopapular, ulcerative, verrucouse and furunculoide) [10,11], 2) Lympho-Cutaneous (most commonly) and 3) Disseminated [1,5].

Fixed cutaneous variety of sporotrichosis remains localized and is less common than the lymphocutaneous type [5,6]. Clinically, initial lesion develops at the site of skin inoculation [10], commonly on the face region in children and on the arms in adults [14]. This form usually presents a single and painless infiltrated, erythematous or violaceous plaque that may become ulcerated or verrucous without lymphatic involvement [2,6]. Also it may start with an acneiform or nodular form of variable duration [5,14].

The lymphocutaneous form is the classical presentation of sporotrichosis and the systemic disease is the product of the conidia inhalation or hematogenous dissemination from primary sites. Pulmonary sporotrichosis displays a form radiographically indistinguishable from tuberculosis and histoplasmosis in patients with sever underlying chronic obstructive pulmonary disease. Osteoarticular sporotrichosis result from the direct inoculation or hematogenous dissemination, with involvement of multiple visceral organs, this occurs almost exclusively in persons with AIDS [10].

Differential diagnoses include cutaneous chronic infections such as cutaneous leshmaniasis in its localized form, but the typical ulcer is painless with erythematous base and well defined high edges [6]. It must be differentiated from tuberculosis, sarcoidosis, paracoccidioidomycosis, chromoblastomycosis, leprosy and other mycobacterial diseases [2,9]. Clinical suspicion is essential for diagnosis. Moreover, direct examination of exudate is not suitable due the structures not being observed [2]. The histopathological examination of tissue stained with conventional HE (hematoxylin & eosin) lacks sensitivity [5]. The gold standard for diagnosis is the isolation of the fungus in Sabouraud dextrose agar with chloramphenicol [2,11].

Prognosis is mainly excellent when proper pharmacological treatment is instituted. Currently, oral itraconazole has become the drug of choice in a recommended dosage of 100-200mg daily, to be administered for 3-6 months [5]. However, the choice of therapy depends upon the location and form of the disease. Potassium iodide concentrated solution in dosage of 3-6 grams daily for 3-4 months, has been described more affordable and equally effective option in cutaneous forms, especially in endemic areas of developing countries [2,3]. Another therapeutic option is terbinafine, which shows similar results to those of itraconazole [4]. Heat application to lesions may help since low temperatures are preferred by fungus [10].

CONSENT

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

REFERENCES

1. Larson K, Pandey S, Hoover W, Sun N. Sporotrichosis in the nail – An unusual location and presentation. JAAD Case Rep. 2018;4:47-9.
2. Vásquez-del-Mercado E, Arenas R, Padilla-Desgarenes C. Sporotrichosis. Clin Dermatol. 2012;30:437-43.
3. Franco-Marques G, Prazeres-Sousa J, Wachholz P. Characterization of sporotrichosis cases treated in a dermatologic teaching unit in the State of Sao Paulo – Brazil, 2003-2013. An Bras Dermatol. 2015;90:273–5.
4. Macedo P, Lopes-Bezerra L, Bernardes-Engemann A, Orofino-Costa R. New posology of potassium iodide for the treatment of cutaneous sporotrichosis: study of efficacy and safety in 102 patients. J Europ Acad Dermatol Venereol. 2015;29:719-24.
5. Sudheer N, Venkatakrishna A, Nagasetha M, Monmahan G. Sporotrichosis – A Case Report. J Evol Med Dental Scien. 2015;4:16124-5.
6. Antonio L, Fernandes-Pimentel M, Rosandiski-Lyra M, Madeira M, Campos-Miranda L, Almeida-Paes R, et al. *Sporothrix schenckii* Sensu Lato identification in fragments of skin lesion cultured in NNN medium for differential diagnosis of cutaneous leishmaniasis. Diag Microbiol Infect Dis. 2017;87:118-20.
7. Lee H, Young-Kim D, Hoon-Lee K, Soo-Choi J, Kyu-Suh M. Deformity of the earlobe caused by fixed cutaneous sporotrichosis in a pediatric patient. Int J Dermatol. 2015;54:187-9.
8. De Araujo M, Rodrigues A, Ferdandes G, De Camargo Z, De Hoog Sybren. Human sporotrichosis beyond the epidemic front reveals classical transmission types in Espírito Santo, Brazil. Mycoses. 2015;58:485-90.
9. Fischman-Gompertz O, Rodrigues A, Fernandes G, Bentubo H, Pires-De-Camargo Z, Petri V. Atypical Clinical Presentation of Sporotrichosis Caused by *Sporothrix globosa* Resistent to Itraconazole. Am J Trop Med Hyg. 2018;94:1-6.
10. Bimbi C, Brzezinski P. Cutaneous sporotrichosis as an occupational disease: Case report. Our Dermatol Online. 2017;8:37-9.
11. Ramírez Soto MC. Facial Sporotrichosis in children from endemic area in Peru. Our Dermatol Online. 2013;4:237-40.
12. Hakrabarti A, Bonifaz A, Gutierre-Galhardo M, Mochizuki T, Li S. Global epidemiology of sporotrichosis. Med Mycol. 2015;53:3-14.
13. Messias-Rodrigues A, De Hoog S, Zhang Y, Pires-Camargo Z. Emerging sporotrichosis is driven by clonal and recombinant *Sporothrix* species. Emerg Microbes Infect. 2014;3:1-3.
14. Gyu-Song J, Bum-Song Y, Youl-Yun S, Kyu-Suh M, Yim-Ha G, Ran-Kim J, et al. Cutaneous Sporotrichosis Presenting as Clinical Feature of Facial Cellulitis in an Adult. Ann Dermatol. 2016;28:507-8.

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Our Dermatology Online

Case Report

Systemic drug related intertriginous and flexural exanthema (SDRIFE) or intertriginous drug eruption: A matter of semantics

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ABSTRACT

Systemic drug-related intertriginous and flexural exanthema (SDRIFE) is a recently coined term to describe an uncommon adverse cutaneous drug reaction described previously as intertriginous drug eruption or flexural drug exanthema. Characteristic flexural erythema notably of the axillae and groins following systemic administration of offending drug may eventuate to TEN-like eruption with relative absence of systemic involvement. Beta-lactam antibiotics like amoxicillin remain the most common offending drug. A1-year-old boy developed SDRIEF overnight after oral amoxicillin(125mg) given for upper respiratory infection.The eruptions deteriorated evolving to TEN-like skin tenderness and exfoliation despite withdrawal of offending drug and treatment with oral cetirizine and prednisolone. It subsided within 1-2days after intravenous immunoglobulin treatment. The putative drugs and various pathogenetic mechanisms proposed for this very unusual adverse cutaneous drug reaction reflects that the nomenclature SDRIFE, intertriginous drug eruption or flexural drug exanthema is just semantics.

Key words: Amoxicillin; Baboon syndrome; Flexural drug eruption; Intertriginous drug eruption; SDRIFE; Systemic contact dermatitis

INTRODUCTION

The term 'baboon syndrome' was first described in 1984 by Andersen to describe a mild systemic cutaneous reaction characterized by diffuse bright red erythema resembling red bottoms of baboons after oral exposure to allergens such as nickel, mercury or drugs [1]. The "symmetrical drug related intertriginous and flexural exanthema" (acronym; SDRIFE) is a recent term coined by Häusermann et al [2] in 2004 for a similar entity induced by systemic drugs that was invariably described as 'systemically induced allergic contact dermatitis', 'systemic contact dermatitis', 'drug-induced intertrigo', 'eczema rubrum', 'flexural drug eruption', 'intertriginous drug eruption' or 'paraptic eczema'. It is an unusual yet characteristic drug-related eruption that typically involves the flexural folds and gluteal areas without any predilection for age or gender. Clinically, well delineated erythema of the perigenital and perianal area and maculopapular eruptions involving the large folds such as the neck-fold, axillae, cubital fossae and groins appears within hours to 2 days of drug exposure. Palms, soles, face and mucosae are usually spared. Rarely the lesions may consist of pustules, vesicles or bullae. Systemic features like fever, lymphadenopathy and internal organ involvement usually do not occur [2-4]. The most common offending drugs are antibiotics, especially beta-lactams (benzylpenicillin, phenoxymethylpenicillin, ceftriaxone, cefuroxime, and ceftazidine) with amoxicillin topping the list. The other implicated drugs include pseudoephedrine, roxithromycin, allopurinol, barium sulfate-containing contrast media, omeprazole, hydroxyzine, cimetidine, deflazacort, hydroxyurea, heparin, immunoglobulin, mitomycin C, naproxen, oxycodone, terbinafine, cetuximab, valacyclovir, risperidone, and very rarely clindamycin [2,5-8].

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CASE REPORT

A1-year-old boy developed a symmetric, confluent, erythematous, mildly pruritic, and macular eruptions affecting both axillae and inguinal folds overnight after oral amoxicillin (125mg) given for upper respiratory infection. Similar lesions over the cubital fossa, buttocks and the lower abdomen appeared during next 1-2 days. Parents denied using any topical treatment earlier. His family and past medical history was unremarkable, he was immunized for his age and weighed 10kg. He was uncomfortable, irritable, and has no fever, pallor, lymphadenopathy or other systemic disease. A review of his laboratory parameters for complete blood counts, hepato-renal function tests, urinalysis, chest x-ray and electrocardiogram revealed no abnormality. No fungal hyphae were visualized in KOH mounts from skin lesions. Parents did not consent for skin biopsy. A diagnosis of SDRIFE was made based on clinical criteria (Table1) proposed by Hausermann et al. [2]. Amoxicillin was stopped and he was prescribed oral cetirizine 5mg/d. Oral prednisolone 10mg/d was added when his irritability, skin tenderness and eruptions deteriorated to TEN-like exfoliation particularly over flexurals but without benefit (Fig. 1). At this point treatment with intravenous immunoglobulin (IVIG, 2.5gm/day for 4 days) initiated. His eruptions resolved over next 1- 2 days without any residual pigmentation or scarring. Parents did not consent for oral re-challenge and were advised to avoid amoxicillin in future.

Table 1: Clinical diagnostic criteria for systemic drug-related intertriginous and flexural exanthema (SDRIFE)

S. No	Diagnostic criteria
1.	Exposure to a systemically administered drug either following the first or subsequent dose (excluding contact allergens)
2.	Sharply demarcated erythema of the gluteal/perianal area and/or V-shaped erythema of the inguinal/perigenital area
3.	Involvement of at least one other intertriginous/flexural localization
4.	Symmetry of affected areas
5.	Absence of systemic symptoms and signs

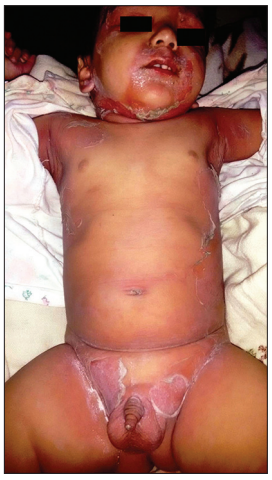

Figure 1: Erythematous and TEN-like exfoliating skin rash predominantly involving periocular and perioral skin, and neck, axillary and inguinal folds.

DISCUSSION

As in our patient the diagnosis of SDRIFE remains clinical in most cases not requiring extensive investigative work up. On the other hand, histologically it is characterized by superficial perivascular infiltrate primarily of mononuclear cells and in some cases neutrophils and eosinophils. Rarely, there may be EM- or TEN-like histologic features of necrotic keratinocytes with vacuolar and hydropic degeneration of the basal cell layer [2]. Drug re-challenge is diagnostic but has potential to trigger severe reactions. Patch and prick tests have low sensitivity and diagnostic value. The utility of in-vitro lymphocyte transformation test that may detect the T-cell mediated delayed hypersensitivity response, macrophage migration or indirect rat mast cell degranulation tests remains limited as a research tool [6]. SDRIFE carries a good prognosis with complete resolution following withdrawal of offending drug. Topical corticosteroids and systemic antihistamines may provide symptomatic relief. Although severe form has been treated with systemic corticosteroids, our patients recovered only after IVIG administration that apparently is more effective than corticosteroids [9].

Several pathomechanisms for this baffling entity have been proposed in last few years. It was initially speculated to be a type IV delayed hypersensitivity to the offending drug involving macrophages and Th1 response (type IVa reaction) and cytotoxic CD8 T cells (type IVc reaction) [2,4]. Daito et al [7] and Wolf et al [10] considered it a type of recall phenomenon means a recall of previous dermatitis such as diaper dermatitis in childhood. However, the concept as proposed by Mahajan et al [9] appears more plausible. It suggests that the offending drug or its metabolites first get concentrated in the apo-eccrine glands preferentially localized to flexural skin and produce a direct toxicity to them. The concentrated drug/metabolites then gets excreted on the skin surface in a gradient fashion, highest concentration in the upper epidermal layers than in the deeper layers, and inflicts a non-immunologic direct toxicity and necrosis of keratinocytes in outer layers while occlusion, sweating

and friction aggravate it further. Their hypothesis has also been ratified in a recently proposed p-i (pharmacologic interaction with immunoreceptors) concept by Miyahara et al. [5] According to them, certain drugs are able to bind directly and non-covalently to T-cell receptors without first being presented by major histocompatibility complex (MHC) molecules and without prior metabolism (direct recognition).However, the putative drugs and various pathogenetic mechanisms proposed for this very unusual adverse cutaneous drug reaction reflects that the nomenclature SDRIFE, intertriginous drug eruption or flexural drug exanthema is just semantics.

CONSENT

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

REFERENCES

1. Andersen KE, Hjorth N, Menne T. The baboon syndrome: systemically-induced allergic contact dermatitis. Contact Dermatitis 1984;10:97-100.
2. Häusermann P, Harr T, Bircher AJ. Baboon syndrome resulting from systemic drugs: is there strife between SDRIFE and allergic contact dermatitis syndrome? Contact Dermatitis. 2004;51:297-310.
3. Häusermann P, Bircher AJ. SDRIFE - another acronym for a distinct cutaneous drug exanthema: do we really need it? Dermatology. 2007;214:1-2.
4. ÖzkayaE, Babuna G. A challenging case: symmetrical drug related intertriginous and flexural exanthem, fixed drug eruption, or Both? Pediatr Dermatol. 2011;28:711-4.
5. Miyahara A, Kawashima H, Okubo Y, Hoshika A. A new proposal for a clinical-oriented sub classification of baboon syndrome and a review of baboon syndrome. Asian Pac J Allergy Immunol. 2011;29:150-60.
6. Kardaun SH, Tupker RA. Symmetric drug-related intertriginous and flexural exanthema (Baboon syndrome) induced by omeprazole. Int J Dermatol. 2012;51:1134-7.
7. Daito J, Hanada K, Katoh N, Katoh S, Sakamoto K, Asai J, et al. Symmetrical drug-related intertriginous and flexural exanthema caused by valacyclovir. Dermatology. 2009;218:60-2.
8. Mahajan VK, Sharma RC. Intertriginous drug eruptions. Indian J Dermatol. 2005;50:146-9.
9. Mahajan VK, Sharma NL, Jindal R. Intertriginous drug eruption: report of a case and proposed pathogenetic mechanism. Int J Dermatol. 2008;47:1310-1.
10. Wolf R, Orion E, Matz H. The baboon syndrome or intertriginous drug eruption: a report of eleven cases and a second look at its pathomechanism. Dermatol Online J. 2003;9:2.

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Our Dermatology Online

Case Report

Giant congenital melanocytic nevus with neurocutaneous melanosis

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ABSTRACT

Giant congenital melanocytic nevi (CNM), present at birth and greater than 40 cm in size, exhibit an increased risk of malignant transformation and may be associated with neurocutaneous melanosis. We present the case of a twelve month old female who presented with a giant congenital melanocytic nevus involving the upper, middle, and a portion of the lower back with multiple satellite lesions on the face, trunk, and extremities. An MRI of the brain displayed areas of hyperintensity in the cerebral hemispheres, right thalamus, and right cerebellum, consistent with neurocutaneous melanosis, however the patient was asymptomatic. Awareness of the clinical features of giant CMN is important for early detection of neurocutaneous melanosis and appropriate treatment.

Key words: Congenital; Melanocytic; Neurocutaneous

INTRODUCTION

Congenital melanocytic nevi (CMN) are typically present at birth, harbor mutations in NRAS, and can vary greatly in size. Small CMN are less than 1.5 cm, medium-sized CMN are 1.5 to 20 cm, large CMN are greater than 20 cm, and giant CMN are greater than 40 cm [1]. Large CNM, which have a greater association with NRAS, have a 2-7% increased risk of malignant transformation, often occurring before the patient reaches puberty [1]. Neurocutaneous melanosis (NCM) is a rare neuroectodermal syndrome, defined as a single giant congenital nevus or multiple congenital nevi, accompanied by meningeal melanosis or CNS melanoma with the absence of cutaneous melanoma or meningeal melanoma [2].

CASE REPORT

A 12 month old Vietnamese female with no significant past medical history presented for evaluation of a congenital melanocytic nevus that had been present since birth. Physical exam showed a darkly pigmented, hypertrichotic plaque involving the upper, middle, and a portion of the lower back, with a rim of erythema along the inferior border of the plaque and several large satellite lesions near the inferior aspect of the plaque (Fig. 1). A large, compressible pedunculated nodular growth was noted on the posterior neck within the plaque as well as numerous pigmented satellite lesions of varying size scattered on the trunk, upper and lower extremities, and face (Figs. 1 and 2). All laboratory investigations were within normal limits. An MRI of the brain showed multifocal, patchy areas of cortical hyperintensity of the cerebral hemispheres, right thalamus, and right cerebellum, consistent with NCM. The patient's family denied that the patient ever had seizures or other neurologic manifestations. There is no family history of a similar condition. Spinal MRI revealed a central enhancing nodule within the large nodule on the posterior neck, and biopsy was considered. After review by radiology at Texas Children's Hospital, the decision was made to defer biopsy and follow with MRI. The patient was then seen by pediatric hematology-oncology, who also recommended repeat MRI along with regular neurology and dermatology follow up.

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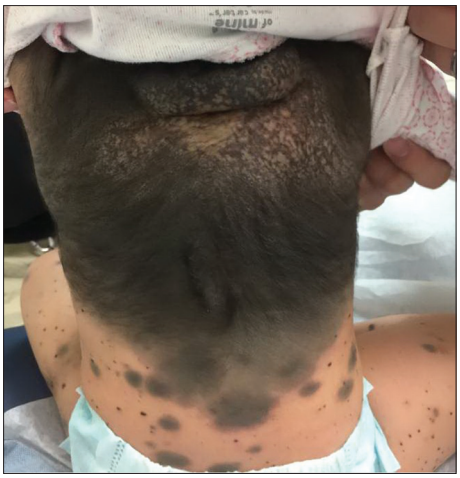

Figure 1: Giant congenital melanocytic nevus involving the upper, middle, and a portion of the lower back with multiple satellite lesions.

Figure 2: Multiple satellite lesions on the face, trunk, and extremities.

DISCUSSION

Large/giant CMN are rare, with an estimated incidence of 0.005%. Symptomatic NCM may affect 6% to 11% of patients with large CMN [3]. NCM may be symptomatic or asymptomatic. While seizures are the most common initial manifestation of NCM, other possibilities include hydrocephalus, developmental delays, psychiatric disorders, cranial nerve palsies, and intracranial hemorrhage [2,3]. The prognosis is poor, with treatment being palliative, including ventriculoperitoneal shunts and anticonvulsants. Some reports have shown death within 2 to 3 years of symptom onset [3]. In addition to the size of CMN, anatomic location and presence of satellite nevi may be associated with a higher risk of development of NCM. In a series of 33 patients with NCM, all had a posterior axial location, and 31 had satellite nevi [4]. Therefore, MRI of the CNS should be considered in all patients with large/giant CMN with multiple satellite nevi, in addition to frequent monitoring for signs of CNS involvement [5].

The risk of melanoma in patients with CMN is likely related to the size, with larger nevi having a higher risk of melanoma. Large CMN have a 2.5% risk of development of melanoma, and giant CMN have a risk of 3.1%. When melanoma does occur in smaller CMN, it typically begins in the epidermis, whereas melanoma arising within larger nevi may involve the dermis or subcutaneous tissue and may be more difficult to detect early [5].

Lifelong examination every 6-12 months, including palpation, of giant CMN is required if not completely excised, along with baseline photography. Biopsy is indicated for enlarging firm papulonodules and new areas of induration or ulceration. Surgical options include dermabrasion, curettage, Lasers (Q-switched ruby, Q-switched alexandrite), and staged excision. If surgical excision is performed, it is recommend to wait until at least six months of age to decrease the risk of adverse effects from general anesthesia [5].

CONSENT

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

REFERENCES

1. Barnhill RL, Cerroni L, Cook M. State of the Art, Nomenclature, and Points of Consensus and Controversy Concerning Benign Melanocytic Lesions: Outcome of an International Workshop. Adv Anat Pathol. 2010;17:73-90.
2. Kadonaga JN, Frieden IJ. Neurocutaneous melanosis: Definition and review of the literature. J Am Acad Dermatol. 1991;24:747-55.
3. Agero ALC, Benvenuto-Andrade C, Dusza SW. Asymptomatic neurocutaneous melanocytosis in patients with large congenital melanocytic nevi: A study of cases from an Internet-based registry. J Am Acad Dermatol. 2005;53:959-65.
4. Lovett A, Maari C, Decarie J-C. Large congenital melanocytic nevi and neurocutaneous melanocytosis: One pediatric center's experience. J Am Acad Dermatol. 2008;61:766-74.
5. Balin S, Barnhill R. Benign Melanocytic Neoplasms In: Bolognia JL, Schaffer JV, Cerroni L. Dermatology. 4rd ed. Philadelphia, PA: Elsevier Saunders; 2018:chap 112:1976.

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

Our Dermatology Online

Case Report

Erythroderma due to iatrogenic immunosuppression: A case of Norwegian scabies

Mohammad Adil[1], Syed Suhail Amin[1], Mohd Mohtashim[1], Sabha Mushtaq[1], Shagufta Qadri[2], Iti Varshney[1]

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ABSTRACT

Norwegian scabies is a rare type of scabies characterised by hyperkeratotic and crusted plaques and is usually seen in immunologically and neurologically impaired patients. An elderly female presented to us with erythema and scaling over the body along with thick plaques covered with yellow crusts over the flexor aspects of the wrists and forearm. There was family history suggestive of scabies in other family members. The patient had received multiple injections of triamcinolone and was applying topical ointment containing steroids for tinea corporis. The HIV ELISA was non reactive. A potassium hydroxide mount showed multiple mites, eggs and faecal pellets. A diagnosis of Norwegian scabies was made and patient treated with oral ivermectin and topical permethrin. We present this case to highlight the possibility of this rare cause of erythroderma in a patient who has been receiving corticosteroids.

Key words: Scabies, Erythroderma, Parasitic disease

INTRODUCTION

Erythroderma, defined as erythema and scaling over more than 90% body surface area is usually the result of eczematous group of diseases (40%), psoriasis (15%), drugs (10%) and malignancies (15%) in elderly patients. Rarely, it may result from other conditions such as pemphigus foliaceous (0.5%) dermatomyositis, scabies and lichen planus (0.5%) [1]. Scabies, an ectoparasitic infestation, is a common disease and can present in various forms. A very rare but highly contagious form is the Norwegian scabies, where the host immune response in modified. The unhindered multiplication of the mite on the skin occurs as a result of absence of pruritus [2]. Norwegian scabies presents as thick crusted and hyperkeratotic plaques and rarely may lead to generalised involvement leading to erythroderma [3]. Herein, we describe a patient of Norwegian scabies occurring after long duration systemic and topical steroid therapy and presenting as erythroderma.

CASE REPORT

A 60 year old female presented to us with complaints of red raised itchy lesions over body since 6 months and generalised redness, scaling and crusting over body since 20 days. She gave history of receiving multiple injections of Triamcinolone in the last five years as well as application of an ointment containing clobetasol, gentamicin and miconazole for itchy annular skin lesions in axillae and groins (possibly Tinea cruris and corporis), prescribed to her by a quack. All members of her family had itchy lesions, predominantly present over the webs of fingers, genitalia, axillae and abdomen suggestive of scabies. On examination, she was found to have generalised erythema and scaling over most of her body (Fig. 1) including the face (Fig. 2) but sparing the palms and soles. There were ill defined plaques with thick, yellow, hyperkeratotic crusts with deep fissures present over the flexor aspect of wrists, forearms, cubital fossa (Fig. 3) and upper medial aspect of

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thighs. Excoriated papules surmounted by crusts were also seen over the trunk and extremities. Examination of nails, hair, mucosa showed no abnormality. The systemic examination was normal with no evidence of any neurological deficit.

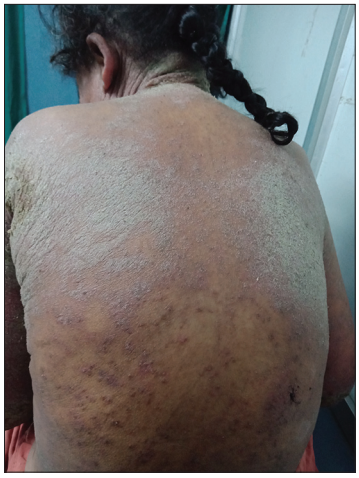

Figure 1: Erythema and scaling and few excoriated papules over the back.

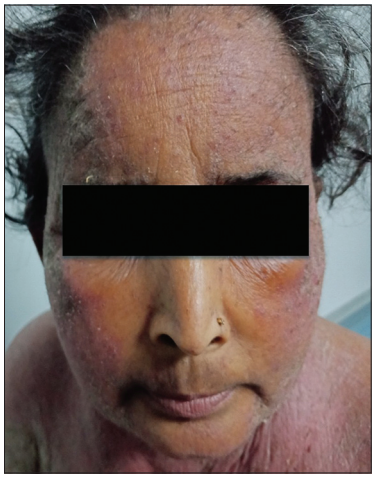

Figure 2: Erythema and scaling over the face

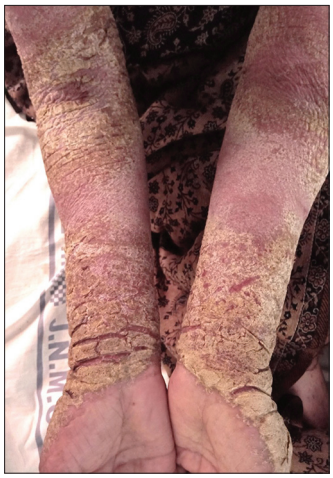

Figure 3: Crusted plaques over the flexor aspect of wist, forearm and cubital fossae

A potassium hydroxide mount revealed the presence of numerous scabies mite, eggs and scybala (Fig. 4). Routine investigations including blood counts and serum biochemistry was within normal limits, except for anaemia (Hb-8.8mg%). HIV ELISA was non-reactive. The chest radiograph, ultrasonography of abdomen and pelvis was normal. Skin biopsy from a papule over the back showed an intra-corneal vesicle with scabies mite and its faecal matter. There was polymorphonuclear infiltrate in the dermis (Fig. 5). Based on the clinical picture, KOH mount and histopathology, a diagnosis of erythroderma due to Norwegian scabies resulting from immunosuppression of systemic steroid abuse was made. The patient was admitted in isolation ward and treated with weekly Ivermectin 18mg and topical permethrin 5% cream with all her family members. She

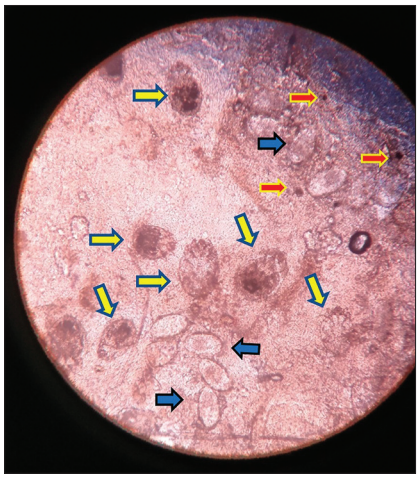

Figure 4: Potassium hydroxide mount showing multiple scabies mites (yellow arrows), eggs (blue arrows) and scybala (red arrows)

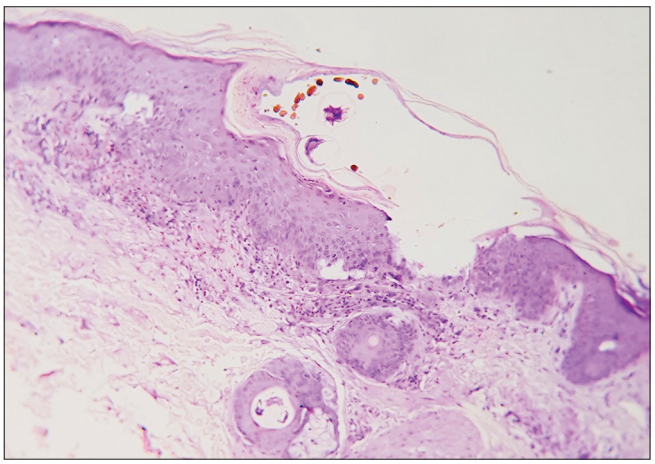

Figure 5: Histopathology of the papule shows intra-corneal vesicle containing scabies mite, faecal pellets and infiltrate in the dermis.

responded very well to treatment and KOH mount was negative for mites after 15 days.

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

DISCUSSION

Crusted scabies is a rare manifestation of infestation by *Sarcoptes scabei* mite which was first described in leprosy patients in Norway, hence the name Norwegian scabies. Crusted scabies is seen in immunosuppressed and mentally debilitated or neurologically impaired patients. While the number of mites in a patient of classical scabies is usually in the range of 10-20, millions of mites are present in a patient of Norwegian scabies, which is attributed to the lack of hypersensitivity and no itching seen in such patients. An altered immunity is responsible for the excessive multiplication of mites in immunosuppressed patients. Itching is a protective response that destroys the burrows and keeps the population of mites in check. Itching is a result of hypersensitivity and is thus less prominent in patients with immune deficiency. Also neurologically impaired patients do not perceive itch and are predisposed to itching [4].There have been very few reports of crusted scabies in patients receiving immunosuppressive drugs including systemic and topical steroids [5-7]. The disease may present as crusted plaques, psoriasiform lesions or as erythroderma [4,8].The diagnosis requires a high index of suspicion. A simple potassium hydroxide mount of the skin scrapping reveals numerous mites in various stages of development, eggs and scabala, the faecal pellets of the mite [9]. Treatment is challenging due to the heavy load of mites on the body as well as the presence of thick plaques. Treatment consists of the application of keratolytic agents to diminish the hyperkeratotic plaques and requires repeated oral ivermectin administration with multiple applications of topical scabicidal agents such as permethrin 5% [10].

We present a rare case of erythroderma due to Norwegian scabies resulting from immunosuppression due to systemic and topical steroids. This case is being presented to highlight the need to consider the possibility of this rare disease entity, particularly in patients who have received long term steroids.

CONSENT

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

REFERENCES

1. Das A, Bar C, Patra A. Norwegian scabies Rare cause of erythroderma. Indian Dermatol Online J. 2015;6:52–4.
2. Ebrahim KC, Alves JB, Tomé LA, Moraes CF, Gaspar AD, Franck KF, et al. Norwegian scabies - rare case of atypical manifestation An Bras Dermatol. 2016;91:826-8.
3. Burns DA. Diseases caused by arthropods and other noxious animals. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. Rook's Textbook of Dermatology. 8th ed. Oxford: Wiley-Blackwell; 2010. pp. 38.40–l4.
4. Chang P, Quijada Ucelo ZM. [Norwegian scabies in an immunocompromised patient]. Our Dermatol Online. 2017;8:484-6.
5. Binic I, Jancovic A, Jovanovic D, Ljubenovic M. Crusted scabies following systemic and topical corticosteroid therapy. J Korean Med Sci. 2010;25:188-91.
6. Lima FCR, Cerqueira AMM, Guimarães MBS, Padilha CBS, Craide FH, Bombardelli M. Crusted scabies due to indiscriminate use of glucocorticoid therapy in infant. An Bras Dermatol. 2017;92:383-5.
7. Sivasubramanian G, Siddiqui MF, Tangella KR. Scabies crusts following corticosteroid therapy in an elderly patient. Am J Med Sci 2012;343:248-9.
8. Anbar TS, Raouf HA, Shalaby S, Abdel-Rahman AT, Ahmed SS. Erythroderma, Scaly Scalp and Nail Dystrophy: A Misleading Association. J Clin Exp Dermatol Res. 2014;5:219.
9. Kamath MV, Gupta RA, Nadkarni N, Sonavane S. Scrape or Perish: The importance of skin scraping in erythroderma. Indian Dermatol Online J. 2011;2:107-8.
10. Costa JB, Rocha de Sousa VL, da Trindade Neto PB, Paulo Filho Tde A, Cabral VC, Pinheiro PM. Norwegian scabies mimicking rupioid psoriasis. An Bras Dermatol. 2012;87:910-3.

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Our Dermatology Online

Case Report

Multiple brown nodules in an infant

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ABSTRACT

Mastocytosis is a clonal disorder of mast cells. Systemic symptoms are typically a result of mast-cell mediator release. Urticaria Pigmentosa is the most common type of children mastocytosis. We report a case of a 9-month-old boy who presented for a widespread eruption since the age of 2 months. Physical examination revealed numerous red to brown slightely hyperpigmented papules and plaques on the trunk, the neck, the proximal upper and lower extremities and the genitalia. Complete blood count, hepatic and renal functions, abdominal ultrasonography were normal. A skin biopsy was performed and the histological exam confirmed the diagnosis of urticaria pigmentosa. The avoidance of chemical and physical stimuli was provided. He had only residual pigmentation at the lesional sites with no new lesion formation. The prognosis of childhood UP is good.

Key words: Mastocytosis; Urticaria Pigmentosa; Infant

INTRODUCTION

Mastocytosis is defined as a heterogeneous group of disorders characterized by an accumulation of mast cells in one or more organs, particularly in the skin, bone marrow, liver, spleen and lymph nodes [1]. Urticaria Pigmentosa (UP) is the most common presentation of cutaneous mastocytosis in children and represents 70-90% of the cases [2]. We report a new case of UP in a 9-month-old boy.

CASE REPORT

A healthy 9-month-old boy born at full term after an uncomplicated pregnancy, presented for a widespread eruption since the age of 2 months. He was otherwise, afebrile with a normal development. He was asymptomatic although sometimes some of his lesions became red and pruritic. There were no systemic symptoms such as flushing, vomiting, diarrhea, respiratory distress or irritability. Physical examination revealed a well-appearing infant. He had numerous red to brown slightely hyperpigmented papules and plaques on the trunk, the neck, the proximal upper and lower extremities and the genitalia (Fig. 1). There were no other skin lesions. Complete blood count, hepatic and renal functions, abdominal ultrasonography were all normal. A skin biopsy showed hyperpigmentation of the basal layer with dermal grouping of lymphocytes and mast cells (Fig. 2). The mast cells were identified with Giemsa staining and CD117 antibody (Figs. 3 and 4). The patient was regularly followed up in our department. General recommendations such as avoidance of chemical and physical stimuli were provided. He had only residual pigmentation at the lesional sites with no new lesion formation.

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

DISCUSSION

Urticaria pigmentosa (UP), also known as maculopapular mastocytosis, is the most common presentation of cutaneous mastocytosis in children

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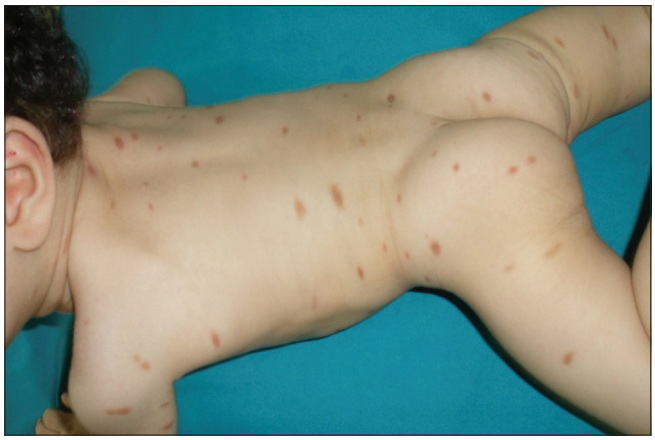

Figure 1: Multiple brownish nodules and plaques on the trunk and limbs

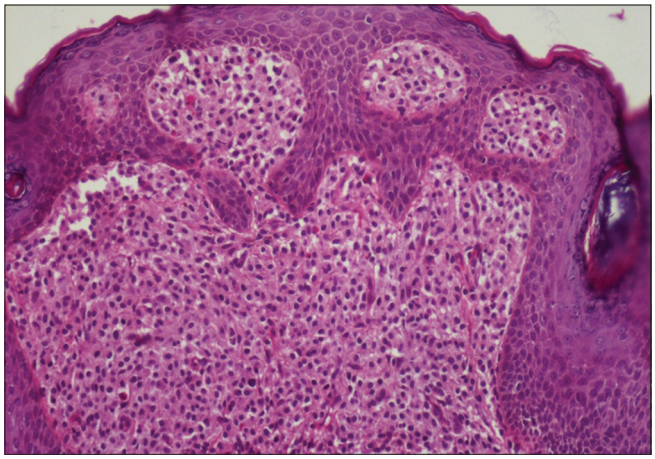

Figure 2: Dense dermal infiltrate constituted of fusiform cells with rounded nuclei and eosinophilic cytoplasm

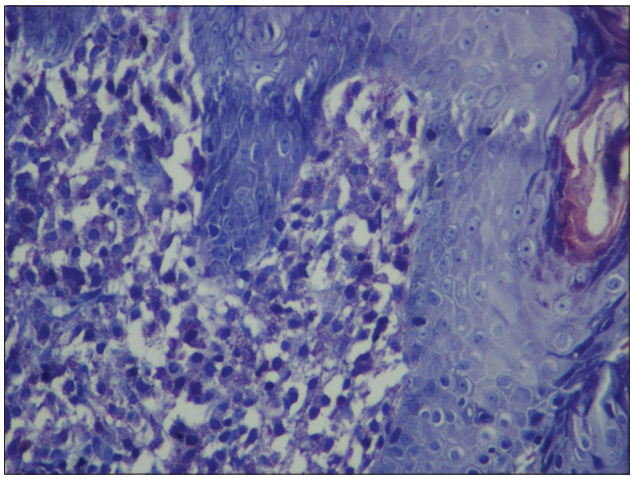

Figure 3: Giemsa staining showed abundant cytoplasmic metachromatic granules

and represents 70-90% of the cases [2]. Urticaria pigmentosa, is the most common form of mastocytosis disorder characterized by mast cell proliferation and accumulation within various organs, most commonly

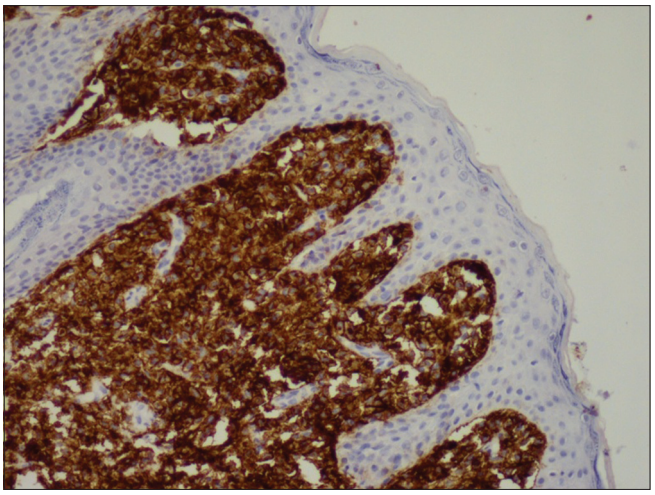

Figure 4: Positive staining with CD117 antibody

the skin. It is defined by multiple ovale or round red-brown hyperpigmented macules, papules, or nodules measuring a few mm to 1-2 cm in diameter. In 55% of patients, lesions appear before the age of 2 years, and in 10% of patients, it occurs between ages 2 and 15 years [3]. The affected areas include the trunk and extremities. The palms, soles, scalp and face are usually free of lesions [2]. The Darier's sign is typically present. The pathogenesis of UP is unknown. Increased mast cell growth factors in skin lesions of UP are thought to stimulate mast cell proliferation, melanocyte proliferation and the production of melanin pigment. In addition, molecular studies demonstrated that UP is caused by several mutations in the KIT gene [4]. Histologically, the number of mast cells in the papillary dermis is increased. They are round or spindle shaped with abundant eosinophilic cytoplasm, distinct cytoplasmic boundaries, and large pale nuclei. They aggregate around blood vessels, and are sometimes associated with eosinophils. The basal layer is hyperpigmented. Nodular infiltrates extending to the subcutis may be seen, especially with special stains like toluidine blue and chloracetate esterase [5]. The treatment of UP is mainly symptomatic. It relies mainly on topical corticosteroids, oral antihistamines and oral cromolyn sodium therapy for gastrointestinal tract symptoms [2,3]. Education of parents to avoid precipitating factors is essential.

CONCLUSION

The diagnosis of UP can be made clinically but a definitive diagnosis requires a skin biopsy. In UP, systemic involvement is rare as in our patient and

most childhood cases will experience a spontaneous resolution by adolescence.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles. Written informed consent was obtained from the child's parents for publication of this article.

REFERENCES

1. Dhar S, Maji B, Roy S, Dhar S. Diffuse Cutaneous Mastocytosis with Bullous Lesions and Pulmonary Involvement: A Rare Case. Indian J Dermatol. 2015;60:179-81.
2. Castells M, Metcalfe DD, Escribano L. Guidelines for the diagnosis and treatment of cutaneous mastocytosis in children. Am J Clin Dermatol. 2011;12:259-70.
3. Bajoghli AA, Blankenship CM. Picture of the month. Urticaria pigmentosa. Arch Pediatr Adolesc Med. 2008;162:383-4.
4. Macri A, Cook C. Urticaria Pigmentosa (Cutaneous Mastocytosis). Treasure Island (FL): Stat Pearls Publishing; 2018.
5. Vasani RJ, Medhekar SV. Urticaria pigmentosa. Indian Dermatol Online J. 2015;6:464-5.

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Our Dermatology Online

Case Report

Acquired partial lipodystrophy: Barraquer –Simons syndrome: A rare case report

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ABSTRACT

Acquired partial lipodystrophy (APL) or Barraquer-Simons syndrome is a rare form of progressive lipodystrophy of unknown etiology. Metabolic complications are less common than with other lipodystrophies. Patients usually have renal involvement in the form of membrenoproliferative glomerulonephritis which can lead to end stage renal disease. Here we report a case illustrating the importance of recognizing the clinical features of lipodystrophic syndrome, which may present with potentially severe consequences and psychological distress. A brief overview is made, addressing the disease, course, and its consequences.

Key words: Lipodystrophy, Barraquer simons syndrome, Membranoproliferative Glomerulonephritis

INTRODUCTION

Lipodystrophies are heterogenous group of disorders with selective loss of adipose tissue which can either be partial or complete. Barraquer Simons Syndrome is the first acquired partial lipodystrophic (APL) disorder described by Mitchell (1885), Barraquer (1907) and Simons (1911). It is also known as *lipodystrophia progressive* or *Cephalothoracic lipodystrophy* [1]. Gellis, Senior and Colleagues have described association of Acquired partial lipodystrophy (APL) with Membranoproliferative Gomerulonephritis (MPGN).

CASE REPORT

A 35 year old female presented to our skin OPD with the complaints of loss of fat over bilateral cheeks since 4 years. It was Insidious in onset which gradually progressed to involve bilateral arms, upper back & both the hips in a span of about 1 year. She also complained of increased loss of hair from the scalp and right eyebrow since 3 months. She was the second child of non-consanguineous, healthy parents. She had regular menstrual cycles. Her past medical history was insignificant and no intake of any drug. There was no history suggestive of recent viral infection, insulin resistance or any significant weight loss. There was no family history of the same condition.

On examination there was bilateral loss of buccal fat pads (Fig. 1) and prominent zygomatic arches. There was loss of subcutaneous fat over bilateral arms, (Fig. 2) upper back and bilateral waist. Furthermore, there were multiple nodular swellings, each measuring about 1x2 cm palpable in the submandibular region and solitary firm mass with ill-defined margins palpable in the lower inner quadrant of right breast. Scalp showed ill-defined areas of hair loss with thickened skin and loss of hair from the lateral aspect of right eye brow with thick underlying skin. The subcutaneous fat was preserved in other anatomic regions. Thyroid was normal on palpation. Hepatosplenomegaly, umbilical hernia, acanthosis nigricans, clitorimegaly, hirsutism or acromegalic features were absent. Systemic

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examinations were unremarkable, including her Ophthalmic and neurological status.

Her complete blood count, biochemical parameters (with renal and liver function tests), urine analysis with urinary albumin excretion, thyroid function tests revealed no abnormalities. Her fasting glucose and HbA1c presented normal values. There was reduced HDL-cholesterol with normal LDL cholesterol and triglycerides levels. The C3 levels were normal but towards lower level (94 mg/dL). ANA was negative with serum markers being nonreactive. Histopathology was suggestive of lipoatrophy (Fig. 3). Ultrasound of neck and breast suggested multiple lipomas and breast mass suggestive of BIRADS grade IV with moderate suspicion of malignancy respectively. Furthermore, Excision biopsy of breast mass reported chronic mastitis. The patient had neither renal involvement, nor any severe metabolic disorders until our last observation. She was counseled about the disease and was started on Tab Rosiglitazone 4mg OD and called for follow up.

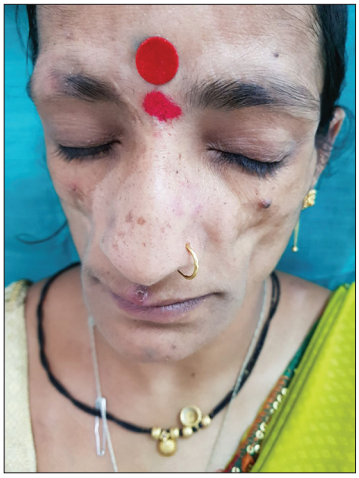

Figure 1: Loss of Buccal pad of fat.

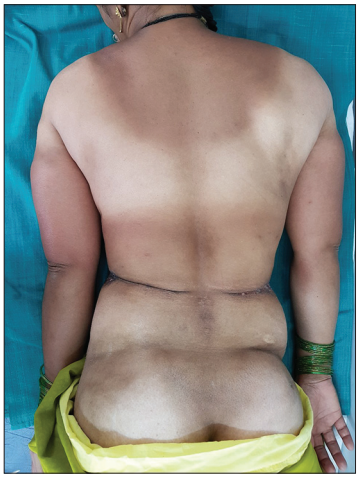

Figure 2: Loss of fat over upper third of arm and bilateral hip.

DISCUSSION

There are four major categories of Lipodystrophies: Congenital generalized lipodystrophy (Berardinelli Seip syndrome), Familial partial Lipodystrophy, Acquired Generalised Lipodystrophy (Lawrence syndrome) and Acquired partial Lipodystrophy (APLD, Barraquer – Simmons syndrome. The new entity is HAART induced lipodystrophy in HIV patients [2].

Our patient had clinical features suggestive of APL fitting into essential diagnostic criteria proposed by Misra et al. (Table 1) and one of supportive criteria that is absence of family history. The etiology of APL is unclear. Most cases are sporadic with Female to Male ratio 4:1. Mutations in LMNB2 gene has been identified in few patients [3].

The onset of APL is in childhood or adolescence age group. Clinically it starts with fat loss in a cephelo-caudal distribution starting from face, neck, upper extremities and trunk sparing lower abdomen, thighs and legs [4]. The fat loss is because of Complement

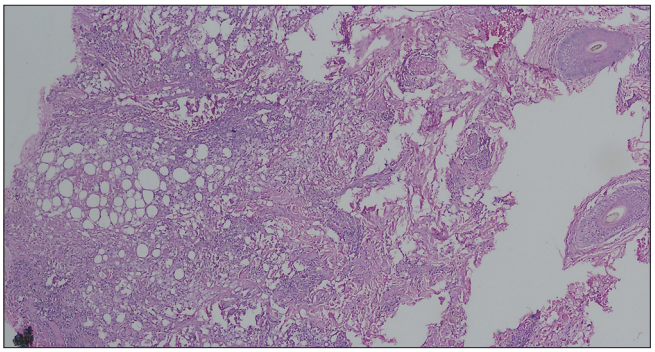

Figure 3: Histopathology showing decrease in fat with atrophy.

Table 1: Diagnostic critertia for acuired partial lipodystrophy (proposed by misra et al[1])

1. Essential criteria: Gradual onset of bilaterally symmetrical subcutaneous fat loss from face, neck, upper extremities, thorax and abdomen but sparing lower extremities.
2. Supportive criteria:
A: Clinical
1. Onset of subcutaneous fat loss during childhood and adolescence
2. Absence of family history of lipodystrophy
3. Presence of autoimmune diseases.
B: Laboratory
1. Low serum levels of complement 3
2. Presence of serum C3NeF
3. Proteinuria
4. MPGN on renal biopsy
5. Characteristic body fat distribution as documented by skinfold thickness measurements, computerized axial tomography or MRI (confirmatory).

3 Nephritic factor that blocks degradation of C3 Convertase enzyme and lysis of adipocytes that expresses factor D [5]. Approximately 20% of patients develop MPGN and some of these can progress to end stage renal disease requiring renal transplantation [6]. Metabolic complication is not seen in APL. Infections and autoimmune diseases have been linked with APL.

Our patient had essential criteria of fat loss in cephelocaudal distribution and supportive criteria absence of family history. Her metabolic profile was within normal limits. She had atypical presentation of loss of fat from waist; such presentation is unusual in APL.

Mangement of APL is diet and exercise. Metreleptin therapy is not useful in APL as serum leptin levels are not deranged. Thiazolidinediones has been tried in APL which improves insulin resistance and hyperglycemia and stimulates the growth of adipocytes [7,8]. Cosmetic procedures like autologous fat transfer, dermal fillers for fat loss has been tried.

CONCLUSION

APL is a very rare form of lipodystrophy. We report this case because of its atypical presentation and its importance of regular follow up and management to prevent metabolic and renal complications.

CONSENT

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

REFERENCES

1. Misra A, Peethambaram A, Garg A: Clinical features and Metabolic and Autoimmune derangements in Acquired Partial lipodustrophy. Report of 35 cases and Review of literature: Medicine. 2004;83:18-34.
2. Fiorenza CG, Chou SH, Mantzoros CS. Lipodystrophy: Pathophysiology and Advances in Treatment. Nat Rev Endocrinol. 2011;7:137-50.
3. Hegele RA, Cao H, Liv DM, Costain GA, Charlton-Menys V, Rodger NW, et al. Sequencing of the Reannotated LMNB2 Gene Reveals Novel Mutations in Patients with Acquired PaRTIAL Lipodystrophy. Am J Hum Genet. 2006;79:383-9.
4. Brown RJ, Chair C, Vilar DA, Chaung PT, Dunger D, Garg A, et al. The diagnosis and management of lipodystrophy syndrome: A Multi-Society Practice Guidelines. J Clin Endocrinol Metab. 2016;101:4500-11.
5. Garg A. Lipodystrophy: Genetic and Acquired Body Fat Disorders. J Clin Endocrinol Metab. 2011;96:3313-25.
6. Hussain I, Garg A. Lipodystrophy Syndromes. Dermatol Clin. 2008;26:569-81.
7. Oliveria J,FreitasP,Law E, Carvalho D. Barraquer Simons Syndrome: a rare form of acquired lipodystrophy. BMC Res Notes. 2016;9:175-9.
8. Parker VE,Semple RK. Genetic forms of Severe insulin resistance: what endocrinologists Should Know. Eur J Endocrinol. 2013;169:R71-R80.

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

Our Dermatology Online

Case Report

Localized annular lichen planus on foot mimicking tinea pedis

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ABSTRACT

Lichen Planus (LP) is an inflammatory mucocutaneous disease characterized by typically itchy, pink-purple, polygonal, flat, papules and plaques. The genital region, the oral mucosa, and the flexor surfaces of the extremities are the most common locations of lichen planus. LP usually appears as diffuse lesions. In the literature, single liken planus lesion cases were re ported on the lower lip, esophagus, and breast. Localized foot annular lichen planus is a very rare and extraordinary manifestation of LP. Herein, we present two cases of a single plaque LP on the foot that had been diagnosed as tinea pedis and nummular dermatitis in external family medicine clinics. We want to underline the significance of this uncommon entity in the differential diagnosis of plantar dermatoses in primary care.

Key words: Lichen planus; Tinea pedis; Histopathology; Family physician; Primary care

INTRODUCTION

Lichen Planus (LP) is an inflammatory mucocutaneous disease characterized by typically itchy, pink-purple, polygonal, flat, papules and plaques [1]. The genital region, the oral mucosa, and the flexor surfaces of the extremities are the most common locations of lichen planus [2]. LP usually appears as diffuse lesions [3]. In the literature, single liken planus lesion cases were reported on the lower lip, esophagus, and breast [4-6]. Localized foot annular lichen planus is a very rare and extraordinary manifestation of LP.

Herein, we present two cases of a single plaque LP on the foot that had been diagnosed as tinea pedis and nummular dermatitis in external family medicine clinics. We want to underline the significance of this uncommon entity in the differential diagnosis of plantar dermatoses in primary care.

CASE REPORT

Case Report One

A 50-year-old woman presented with erythematous violaceous plaque over the medial side of her left foot (Fig. 1). This lesion had first appeared one year ago. She was diagnosed with tinea pedis and nummular dermatitis in external clinics and was treated with topical antifungal and corticosteroid therapy. The cutaneous examination revealed erythematous violaceous plaques over the medial side of her left foot. There were no additional features in her general physical examination.

In terms of the patient's medical history, she had had type 2 diabetes mellitus for seventeen years and essential hypertension for two years. Laboratory studies were unremarkable.

The histopathological examination revealed focal parakeratosis, an increase in the granular cell layer, and

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a band-like lymphocytic infiltrate in the dermis (Fig. 2). A periodic acid-Schiff [PAS] stain was negative. The final diagnosis was lichen planus.

0.05% clobetasol 17-propionate was given twice daily for ten days, and the patient was advised to take it in the form of occlusion in the evening. After 10 days, tacrolin pomade was recommended to be applied twice a day.

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

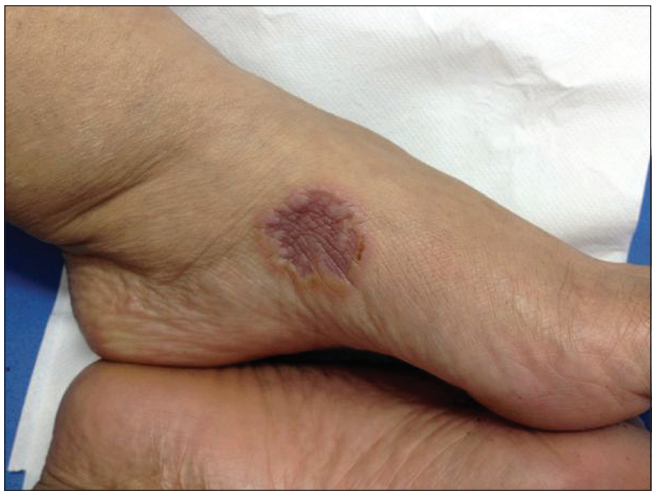

Figure 1: Erythematous violaceous plaque over the medial side of her left foot

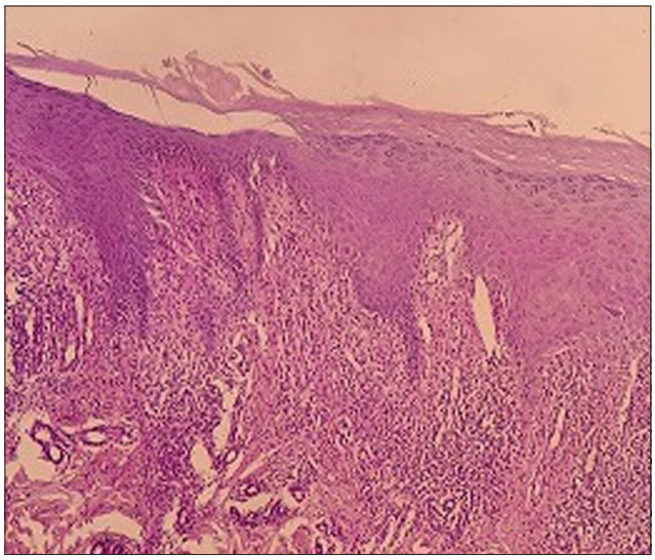

Figure 2: The histopathological examination revealed focal parakeratosis, an increase in the granular cell layer, and a band-like lymphocytic infiltrate in the dermis.

Case Report Two

A 43-year-old man presented with erythematous violaceous plaque over the medial side of his right foot for six months. He was diagnosed as having tinea pedis in external clinics. The cutaneous examination revealed 3x4 cm erythematous violaceous plaques over the medial side of his right foot (Fig. 3). His other body areas and mucous membranes were examined and found to be normal. There were no characteristics in the patient's medical history. A native (%10 KOH) study was negative for fungal disease.

The histopathological examination revealed focal parakeratosis, an increase in the granular cell layer, and

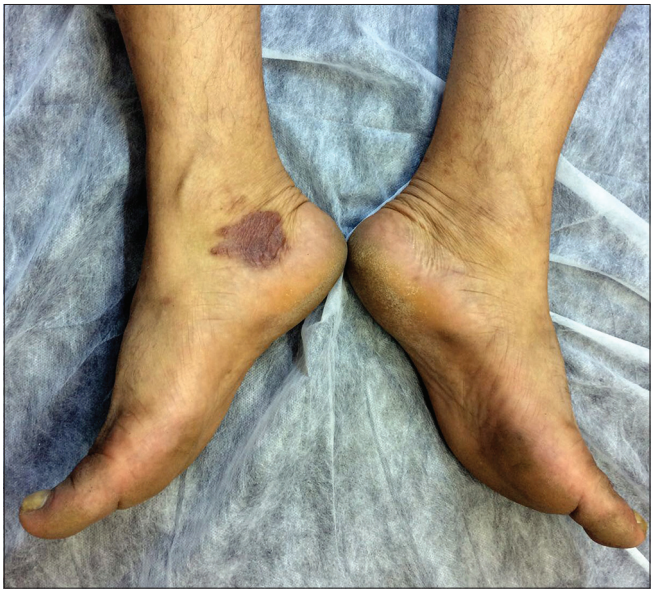

Figure 3: 3x4 cm erythematous violaceous plaques over the medial side of his right foot

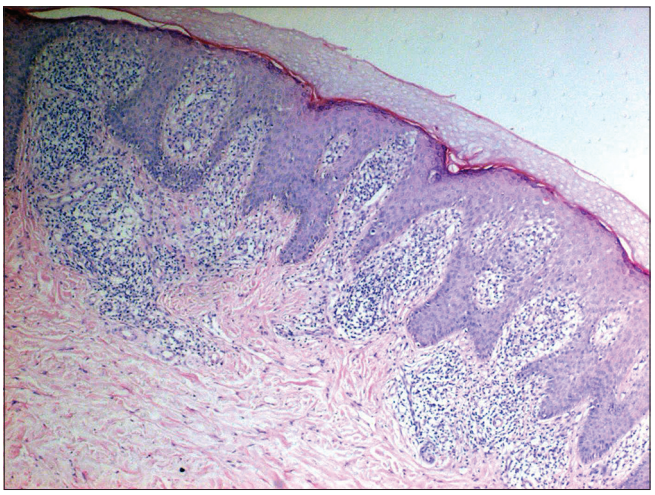

Figure 4: The histopathological examination revealed focal parakeratosis, an increase in the granular cell layer, and a band-like lymphocytic infiltrate in the dermis

a band-like lymphocytic infiltrate in the dermis (Fig. 4). A periodic acid-Schiff [PAS] stain was negative. The final diagnosis was lichen planus.

0.05% clobetasol 17-propionate was given to the patient twice daily for ten days, and he was advised to take it in the form of occlusion in the evening. After 10 days, tacrolin pomade was recommended to be applied twice a day.

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

DISCUSSION

Lichen planus lesions can emerge in many different forms: annular, linear, hypertrophic, atrophic, bullous, ulcerative, and pigmented [3]. The lesions in our cases were compatible with annular lichen planus. LP is generally localized to flexures, extremities, genitalia, and oral mucosa [2]. Palmoplantar lichen planus is an uncommon limited variant of lichen planus. In our cases, there was only a single plaque lichen planus on the foot, a rather rare variant of lichen planus [7]. Although the lesion was only on the foot and not on the hands, our cases was thought to be a single plaque lesion as a rare variant of palmoplantar lichen planus.

Lichen planus usually occurs with diffuse lesions anywhere in the body [5]. Lichen planus is mostly acute at its onset [8]. It is quite rare for lichen planus to appear as a single plaque LP. In the literature, single liken planus lesion cases were reported on the lower lip, esophagus, breast, and eyelids [4-6,9]. We did not find a case of a single plaque lichen planus on the foot previously in the literature; thus, our cases were the first reported cases. It was remarkable that the patients were followed for such a long time in primary care with the incorrect diagnosis of tinea pedis. These types of cases can be easily misdiagnosed when they are first referred to the family physician [10].

The differential diagnosis of palmoplantar LP includes psoriasis, callus, tinea, hyperkeratotic eczema, secondary syphilis, verruca, nummular dermatitis, mycosis fungoides, and granuloma annulare [11]. The histopathological examination plays a key role in the differential diagnosis [12]. In our cases, the native examination was negative, and the histopathological examination was compatible with lichen planus.

Acitretin, enoxaparin, cyclosporine, topical cyclosporine, systemic corticosteroid, topical corticosteroid, topical tacrolimus, surgery, UVA1, db-UVB, metronidazole, retinoic acid, sulfasalazine, hydroxychloroquine, mycophenolate mofetil, and thalidomide are the treatment modalities for palmoplantar LP In the literature [13,14]. There was also a case of a positive response with topical calcineurin inhibitor therapy reported in the literature [15,16]. In our cases, we added tacrolimus cream to a topical steroid therapy, and clinical follow up of our cases continue.

CONCLUSION

We reported here two cases of a single plaque lichen planus on the foot that had been diagnosed as tinea pedis and nummular dermatitis in external clinics. We present our cases to emphasize the importance of the native examination and histopathology in a differential diagnosis of palmoplantar dermatoses and the rare occurrence of single plaque lichen planus on the foot. Since clinical properties may not be suggestive of LP, a skin biopsy is important for the diagnosis.

CONSENT

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

REFERENCES

1. Sharma A, Białynicki-Birula R, Schwartz RA, Janniger CK. Lichen planus: an update and review. Cutis. 2012;90:17-23.
2. Schilling L, Vogt T. Lichen ruber planus. Der Hautarzt. 2018;1-7.
3. Gorouhi F, Firooz A, Khatami A, Ladoyanni E, Bouzari N, Kamangar F, et al. Interventions for cutaneous lichen planus (Protocol). Cochrane Database of Systematic Reviews 2009,Issue 4.Art.No: CD008038.
4. Petruzzi M, De Benedittis M, Pastore L, Pannone G, Grassi FR, Serpico R. Isolated lichen planus of the lip. Int J Immunopathol Pharmacol. 2007;20:631-5.
5. Sica M, Zulli C, Manta R, Villanacci V, Conigliaro R, Bassotti G. One-shot balloon dilation of esophageal stricture due to unusual lichen planus localization. J Gastrointestin Liver Dis. 2016;25:427.
6. Palleschi G, Bruscino N, Corradini D, Bassi D, Vega P, Pimpinelli N. Annular lichen planus on the mammary areola: an unusual localization. G Ital Dermatol Venereol. 2016;151:114-5..
7. Ucmak D, Azizoglu R, Harman M. Palmoplantar lichen planus- a report of four cases. J Clin Exp Invest. 2011;2:80–4.
8. Boyd AS, Neldner KH. Lichen planus. J Am Acad Dermatol. 1991;25:593-619.
9. Huang YYM, Wang CM, Potenziani S, Hsu S. Lichen planus of the eyelids: a case report and review of the literature. Dermatol Online J. 2017;23.pii: 13030/qt1c04h08s.
10. Özyurt K, Sucaklı MH, Çölgeçen E, Çelik M. Knowledge of family physicians on common dermatological diseases and their diagnosis

and management trends. Turkderm Arch Turkish Dermatol Venerol. 2014;48:254-62.
11. Kim MJ, Choi M, Na SY, Lee JH, Cho S. Two cases of palmoplantar lichen planus with various clinical features. J Dermatol. 2010;37:985-9.
12. Landis M, Bohyer C, Bahrami S, Brogan B. Palmoplantar lichen planus: A rare presentation of a common disease. J Dermatol Case Rep. 2008;2:8.
13. Feily A, Yaghoobi R, Nilforoushzadeh MA. Treatment modalities of palmoplantar lichen planus: a brief review. Adv Dermatol Allergol. 2016;33:411.
14. Karakatsanis G, Patsatsi A, Kastoridou C, Sotiriadis D. Palmoplantar lichen planus with umbilicated papules: an atypical case with rapid therapeutic response to cyclosporin. J Europ Acad Dermatol Venereol. 2007;21:1006-7.
15. Ojeda T, Rodríguez-Rey E, Camacho FM. Ulcerative lichen planus of the sole treated with tacrolimus, 0.1% Actas Dermosifiliogr. 2011;102:383–4.
16. Al-Khenaizan S, Al Mubarak L. Ulcerative lichen planus of the sole: excellent response to topical tacrolimus. Int J Dermatol. 2008;47:626–8.

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Our Dermatology Online

Case Report

Garlic: From treatment to disease

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ABSTRACT

Garlic belongs to the family Alliaceae. As a condiment, garlic is added to food to improve its flavor. Therapeutic uses have also been reported for some of garlic components that are allyls with disulfide and thiol groups. Case reports have highlighted the possibility that garlic use may cause allergic reactions. We report a new case of irritative dermatitis due to the application of garlic.

Key words: Garlic; Allergic, Reaction

INTRODUCTION

Garlic is one of the best-researched/best-selling herbal remedies and is also commonly used as a food and a spice. Pharmacological actions of garlic include antibacterial, antiviral, antifungal, antihypertensive, blood glucose lowering, antithrombotic, antimutagenic and antiplatelet actions. Traditionally, garlic has been used both orally and topically most consistently perhaps to prevent and treat infections and as a way of maintaining general health [1]. Case reports have highlighted the possibility that garlic use may cause allergic reactions. We report a new case of irritative dermatitis due to the application of garlic.

CASE REPORT

A 22 years old patient was presenting a history of personal atopy (allergic rhinitis and conjunctivitis). After having a "prescription" from a radio show, he applied a Garlic juice on the face without solar exposure. One day later, she developed a burning sensation, pain, itch and erythema, he subsequently developed multiple bullae (Fig. 1). A diagnosis of dermatitis irritative was made, with a complete resolution of her symptoms after symptomatic treatment (Fig. 2).

DISCUSSION

Garlic belongs to the family Alliaceae. The allergenic potential of garlic is well recognized, and allergens have been identified as diallyl disulfide (which is considered to be the primary allergen), allylpropyl sulfide and allicin (the latter may be an irritant) [1]. Garlic also contains a not-yet identified high molecular-weight protein that presumably leads to systemic allergic [2].

Case reports of allergic reactions associated to garlic use include allergic contact dermatitis, generalized urticaria, angiedema pemphigus and anaphylaxis [1]. Allergic contact dermatitis is a delayed type IV allergic reaction of the skin with varying degrees of erythema, edema, and vesiculation resulting from cutaneous contact with a specific allergen [2].

The possibility that garlic may cause irritant contact dermatitis (known as "garlic burns") has been highlighted by a number of case reports/case series, both in infants and in adults [1]. Compared to infants, a longer exposure time seems to be generally needed for causing burns in adults. The cases involved the feet, wrists, hand, trunk, breast, and forehead. It is responsible for a particular dermatitis: pulpitis of the first 3 fingers. The localization at the level of the face is rare. Atopic terrain is frequently found. The garlic was applied to treat asthma, skin lesions, pain, and fever, and in some instance. Garlic

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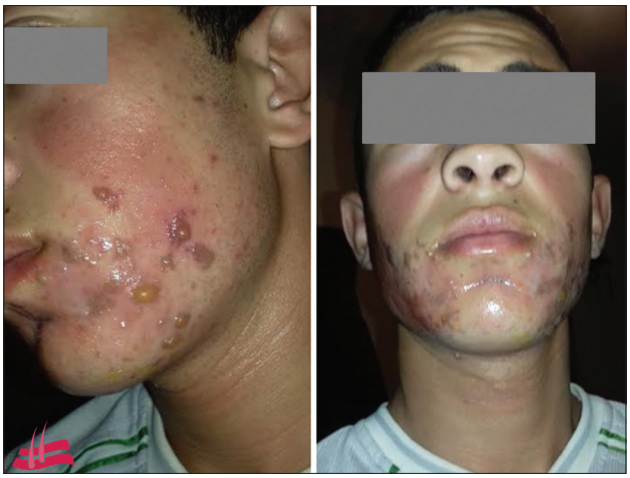

Figure 1: Erythematous and bullous lesions after application of garlic.

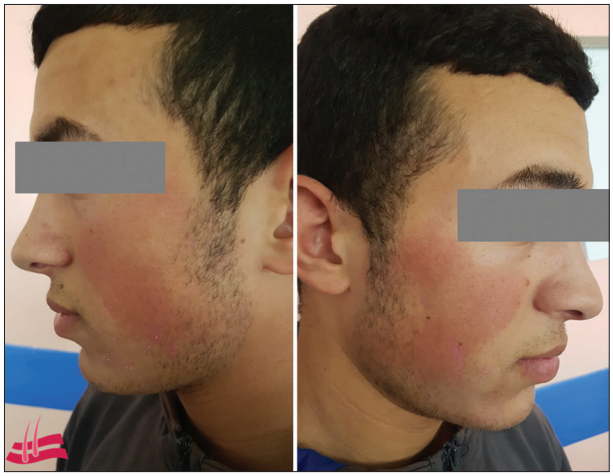

Figure 2: Clinical aspect after treatment.

burns have been also induced for self-mutilation in order to avoid military duty or without a precise rational motive [3,4]. In all cases, the burns were successfully managed with conservative treatment alone [4].

CONCLUSION

Despite a very high consumption and use of garlic in the world, published cases of irritative dermatitis are rare.

Consent

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

REFERENCES

1. Borrelli F. Garlic (*Allium sativum* L.): Adverse effects and drug interactions in humans. Mol Nutr Food Res. 2007;51:1386–97.
2. Beaumont P, Moneret-Vautrin DA, Dzviga C, Grand J-L. Severe anaphylaxis to onion and garlic: Lack of efficiency of prick tests and specific IgEs to the allergenic source. A series of five cases. Rev Fran d'Allergol. 2013;53:446-9.
3. Lachter J, Babich JP, Brookman JC, Factor AY. Garlic: a way out of work. Mil Med. 2003;168:499–500.
4. Friedman T, Shalom A, Westreich M. Self-inflicted garlic burns: our experience and literature review. Int J Dermatol. 2006;45:1161–3.

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

Our Dermatology Online

Case Report

Multiforme erythema complicating an orf

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ABSTRACT

Orf, also known as ecthyma contagiosum, is a spontaneously zoonotic epitheliotropic skin infection caused by a parapox virus. The disease develops in sheep and goats. The virus can also infect humans who come into contact with active lesions, or with their products. Erythema multiforme is considered to be a rare complication of Orf's disease. We report the case of a young Moroccan woman, who presented an Orf complicated of multiforme erythema, appeared 8 days after the Feast of Sacrifice, having well heal under symptomatic treatment. Orf is capable of causing epidemics after the Sacrifice Feast in Muslim countries, therefore information on the disease as well as the precautions to be taken, should be provided to groups at risk.

Key words: Orf, Multiforme erythema, Virus sacrifice, Feast animals

INTRODUCTION

Orf, also known as contagiosum ecthyma, is a spontaneously zoonotic epitheliotropic skin infection caused by a parapox virus of the group of DNA viruses. The name of the disease is an Irish word for sheep disease. It develops in sheep and goats generally in spring and summer [1]. Orf virus is usually transmitted through the breaks and abrasions on the skin. Farmers, butchers, sheep shearers and veterinarians are prone to diseas [2]. Multiforme Erythema is often associated with Herpes simplex virus infection or Mycoplasma pneumoniae [3]. It rarely complicates Orf's disease [4]. We report the case of a young Moroccan woman with multiforme erythema generated by an Orf, appeared few days after the Sacrifice Feast which has well heal under symptomatic treatment.

CASE REPORT

She was a 31-year-old woman, of Moroccan origin, without significant pathological history, who was presented to the emergency department, for acral cutaneous lesions, appeared 8 days after a skin break with a knife used during the Feast of Sacrifice. The patient did not report any recent herpes infection or recent pulmonary symptomatology.

She was stable, apyretic (37.2°). Clinical check has found multiple pseudocockades and some cockades lesions, with 3 concentric areas: an peripheral erythematous disc, a paly middle disc and a central zone sometimes vesicular and sometimes crustal, affecting both hands and feet (Fig. 1). We had also noted the presence of a rounded nodule, purplish- red, well-defined, 1.5 cm long axis,with hard consistency, painful on palpation, on the 2nd phalanx of the 3rd finger of the left hand (Fig. 2).

The biological assessment revealed an inflammatory syndrome with a slight hyperleucocytosis dominant by PNN, and a CRP at 32g/l. Local care and symptomatic analgesic treatment were prescribed for the patient. The evolution of the cas has been marked by the patient recovering with a total disappearance of the lesions after about twenty days.

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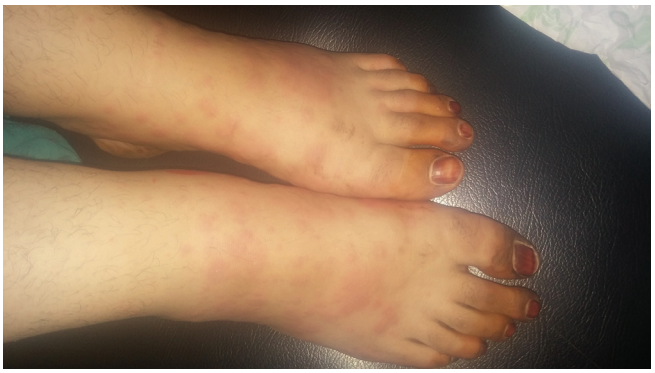

Figure 1: Clinical image showing feet'spseudocockades.

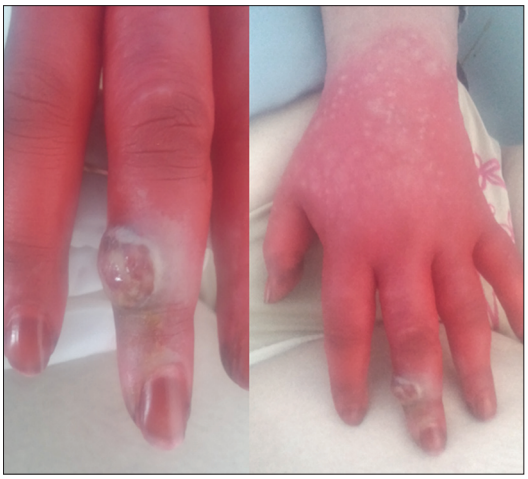

Figure 2: Clinical image of Orf nodule in the left hand.

DISCUSSION

Orf is not an endemic disease, but it may cause epidemics after the Sacrifice Feast in Muslim countries [1]. It is more common among butchers (24.4%) before the Sacrifice Feast, and among those who slaughter animals (40.5%) after. With a male predominance (30.4%) before the Feast, and femininepredominance (32.7%) after that. The results were consistent in several countries, notably Turkey and Tunisia [5].

The incubation period goes from 3 to 10 days, with single or multiple lesions affecting mainly hands and the face [6]. For humans, the Orf goes through six clinical stages, each lasting nearly a week. The disease begins with the appearance of an erythematous papule that turn to be a nodule " of target " type, red in the center and white in the borders, surrounded by a red ring, then the lesion regresses leaving place for a thick crust that develops on the lesion before it disappears without a trace [1]. The diagnosis is clinical, but in some cases the virus isolation, tissue culture or polymerase chain reaction (PCR) may be useful, however they still be expensive and not common [6]. In our patient, the appearance of lesions during the Feast of Sacrifice, after contact with a sheep, as well as the typical clinical and evolutionary aspect of the lesion, was sufficient to retain the diagnosis.

Lesions might be complicated by lymphangitis or secondary bacterial infection, but systemic complications such as multiforme erythema and generalized lymphadenopathy are rare [6]. Multiforme Erythema is an acute eruptive syndrome characterized by the acral lesion in the roundel. It is most often associated with Herpes simplex virus infection or Mycoplasma pneumoniae infection [3]. The association of an Orf and an multiforme erythema has rarely been described in the literature. This complication is related to an immune response caused by contagiosum ecthyma [4]. In our case it was a minor multiforme erythema complicating an Orf, having evolved well under symptomatic treatment.

In general, conservative therapy and local antiseptic are recommended to prevent bacterial infection, but cryotherapy or topical cidofovir cream can be used for large lesions. Low-dose of systemic steroids and antihistamines are useful in the treatment of multiforme erythema [6].

CONCLUSION

Orf is a zoonotic viral infection, able of causing epidemics after the Sacrifice Feast in Muslim countries [1].

Therefore, care should be taken when coming into contact with sheep or goats with ecthyma, through the use of gloves and the disinfection of contaminated areas. We believe that more information about the disease, including vaccination and treatment of affected animals should be provided to groups at-risk, such as breeders, as well as to the general population, especially during Sacrifice days [1].

CONSENT

The examination of the patient wasconductedaccording to the Declaration of Helsinki principles.

REFERENCES

1. Saçar H, Uyar B. Investigation of the complications and incidences of orf disease during and after the Feast of the Sacrifice period. Dermatol Sinica. 2015;33:191-5.

2. Tamer F, Yuksel ME. The spectacular presentation of orf disease. Our Dermatol Online. 2018;9:152-3.
3. Saada D, Velasco S, Vabres P, Guillet G. Érythème polymorphe majeur et infection à *Chlamydia pneumoniaE*. Ann Dermatol Vénéréol. 2006;133:1001-4.
4. Larquey M, Mahé E. Echtyma contagiosum compliquè d'érythéme polymorphe: à propos d'un case. Arch Pédiatr. 2016;23:1184-90.
5. Khaled A, Robbana F, Hammami H, Kharfi M, El Fekih N, Fazaa B, et al. Orf of the han. Tunis Med. 2009;87:352-3.
6. Biazar T, Shokri M. Erythema Multiforme as a Result of Orf Disease; a Case Report. Emerg (Tehran). 2016;3:163–5.

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Our Dermatology Online

Case Report

Extra genital HPV-6

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ABSTRACT

The buschke lowenstein tumor is a rare sexually transmitted disease. We report a case of a buschke lowenstein disease sitting onthe neck, the back in addition to genital area. A 58 years old patient, without any significant pathological antecedents, presented for more than 3 years genital tumors gradually getting bigger. Dermatological examination found several ulcerated, exophytic, cauliflower tumors taking all the urogenital area, two tumors setting in the neck and one another on the back. Cytological sampling was performed, and the search for HPV returned positive. Other serologies have returned negative. The patient was subsequently treated by the urologist and plastician team. Complete exeresis was performed. An anatomopathological study was done which confirmed the diagnosis. HPV typing was done and confirmed HPV 6. The patient has evolved well and presented no complication. Buschke-lowenstein disease is a curable sexually transmitted urogenital disease. We report for the an unusual three location of this disease, which can shed light on the mode of transmission.

Key words: Buschke lowenstein, Condyloma acuminate, Extra genital, HPV 6

INTRODUCTION

The buschke lowenstein tumor, also called the giant condyloma, is a rare sexually transmitted disease [1]. Men are affected, but case reports of women and children have been reported [2].

Extra genital cases are very rare but not impossible. We report a case of a buschke lowenstein disease sitting onthe neck, the back in addition to genital area.

CASE REPORT

A 58 years old patient, married, without any significant pathological antecedents, presented for more than 3 years genital tumors gradually getting bigger. And soon, tumors also appeared in the neck and back. On examination the patient was conscious, hemodynamically stable. Dermatological examination found several ulcerated, exophytic, cauliflower tumors taking all the urogenital area, two tumors setting in the neck and one another on the back (Fig. 1).

Cytological sampling was performed, and the search for HPV returned positive.

Other serologies have returned negative.

The patient was subsequently treated by the urologist and plastician team. Complete exeresis was performed. An anatomopathological study was done which confirmed the diagnosis. HPV typing was done and confirmed HPV 6 (Fig. 2).

The follow-up didn't reveal any complications (Fig. 3).

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

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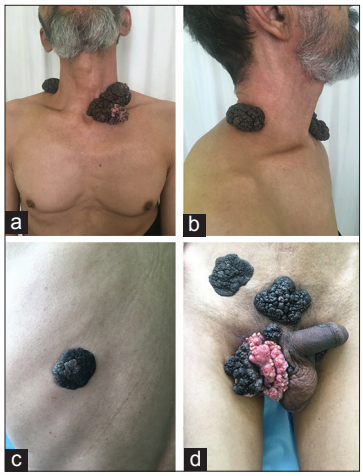

Figure 1a-d: Cawliflower tumors.

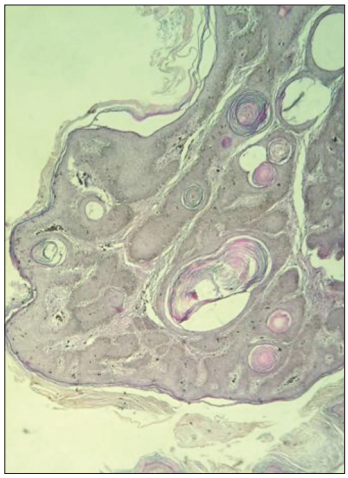

Figure 2: Histopathology: Geant condyloma acuminata.

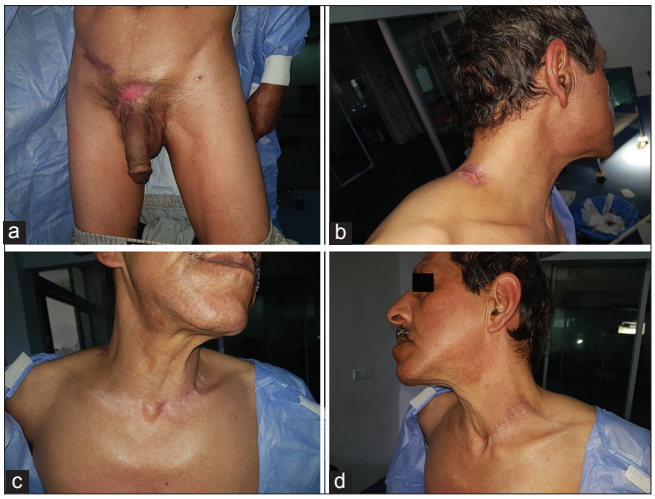

Figure 3a-d: Three months after surgery.

DISCUSSION

Buschke lowenstein disease, described for the first time by ludwig loweinstein and abraham buschke as a malignant acuminous condyloma [3]. Subsequently, it was classified as a verrucous carcinoma [4]. It is a rare enough sexually transmitted disease affecting mainly men between the age of 40 and 60 years as in our patient [5,6].

This tumor usually affects the ano-genital area, the inner sides of the thighs.

Anogenital giant condyloma usually appears in young patients as a slow-growing mass, present for months or years before the patient seeks medical attention. Its development and growth rate have been associated with impaired immunity, particularly that caused by human immunodeficiency virus infection, as well as with pregnancy and multiple comorbidities

Viral cytopathic changes and HPV types 6 and 11 are consistently identified in cases of giant condyloma, whereas this association is less consistent in verrucous carcinoma [7]. In addition, giant condyloma lacks the broadbased appearance and altered keratinocyte differentiation of verrucous carcinomas [8,9]. Nonetheless, others believe that both lesions represent the same entity, given that giant condyloma can sometimes be locally destructive [10].

The particularity of our observation lies in the extra-genital location, and more particularly in the neck and back. Our patients share clinical and pathologic features typically seen in anogenital giant condyloma.

The clincal presnetation is very classic, but when found on extra genital area, some questions must be asked [7]. For many reason we sought to make HPV typing trying to confirm the nature of the extra genital tumors, and concluded that the same HPV was responsible for the tumors on both locations. A previously reported case of cervical giant condyloma also occurred in a young woman during pregnancy [10].

This could be explained by the manuportage, and the transmission of HPV by scratching.

Several therapeutic eventualities exist, the gold standard is surgery.

Other treatments such as Photodynamic therapy, CO2 laser, podophyllin, methotrexate and cryotherapy are reserved for small tumors and reccurent lesions.

CONCLUSION

Buschke-lowenstein disease is a curable sexually transmitted urogenital disease. We report for the an unusual three location of this disease, which can shed light on the mode of transmission.

CONSENT

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

REFERENCES

1. Spînu D, Rãdulescu A, Bratu O, Checheriâã IA, Ranetti AE, Mischianu D. Giant Condyloma Acuminatum – Buschke-Löwenstein Disease – a Literature Review. Chirurgia. 2014;109:445-50,
2. Gole GN, Shekhar TY, Gole SG, Prabhala S. Successful treatment of Buschke-Löwenstein tumor by surgical excision alone. J Cutan Aesthet Surg. 2010;3:174-6.
3. Steffen C. The men behind the eponym – Abraham Buschke and Ludwig Lowenstein: giant condyloma (Buschke-Loewenstein). Am J Dermatopathol. 2006;28:526-36.
4. Singh P, Nathani D, Ranjan S, Issar R. A Giant Cutaneous Horn Projecting from Verrucous Carcinoma of Buccal Mucosa: A Rare Case Report. J Clin Diagn Res. 2017;11:ZD04-5.
5. Qarro A, Ait Ali A, Choho A, Alkandry S, Borki K. Ann Chirur. 2005;130: 96–100.
6. Ottavioli A, Campana F, Catherine JH, Massereau E, Del Grande J, Ordioni U. [Proliferative verrucous leukoplakia: Three cases and literature review]. Ann Dermatol Venereol. 2016;143:187-96.
7. Parra-Herran C, Herfs M, Doria M, Crum CP, Nucci MR. Giant Condyloma of the Cervix An Uncommon Entity Associated With Low-risk Human Papilloma Virus Infection. Am J Surg Pathol. 2013;37:33-4.
8. Crum Christopher P, Nucci Marisa R, Lee Kenneth R. Diagnostic Gynecologic and Obstetric Pathology. 2nd ed. Philadelphia, PA: Elsevier; 2011:201–203. 9. Trombetta.
9. Shenoy S. Perianal giant condyloma acuminatum (Buschke-Löwenstein tumor). Skinmed. 2014;12:114-5.
10. Longacre TA, Kong CS, Welton ML. Diagnostic problems in anal pathology. Adv Anat Pathol. 2008;15:263–78.

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Our Dermatology Online

Case Report

Multiple skin metastases in a patient with acute myelomonocytic leukemia

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ABSTRACT

Metastatic skin tumors may occur before or at any time before the diagnosis of malignancy. Skin metastases are thought to be indicative of advanced stage malignancy or non-treatment response. Cutaneous metastases are a worse prognostic mark, especially in patients with cancer of the lung, ovary, upper respiratory tract, or upper digestive tract. Acute myelomonocytic leukemia is a malignant-hematopoietic clonal sickness of bone marrow and disrupts production of normal blood cells. Although there are many and various dermatological findings in acute leukemia, they are mostly due to cytopenia and haemostasis disorders and skin metastasis is not frequent. Here, we present a case of skin involvement with a diagnosis of acute myelomonocytic leukemia, which is uncommon in the literature and nodules on the whole body.

Key words: Acute leukemia; Acute myelomonocytic leukemia; Skin metastas

INTRODUCTION

Metastatic skin tumors may occur before or at any time before the diagnosis of malignancy [1]. Skin metastases are thought to be indicative of advanced stage malignancy or non-treatment response [2]. Cutaneous metastases are a worse prognostic mark, especially in patients with cancer of the lung, ovary, upper respiratory tract, or upper digestive tract [3].

Acute myelomonocytic leukemia (AML M4) is a malignant-hematopoietic clonal sickness of bone marrow and disrupts production of normal blood cells [4]. Although there are many and various dermatological findings in AML, they are mostly due to cytopenia and haemostasis disorders and skin metastasis is not frequent [5].

Here, we present a case of skin involvement with a diagnosis of AML M4, which is uncommon in the literature and nodules on the whole body.

CASE REPORT

A 19-year-old woman was referred from the hematology polyclinic to the dermatology polyclinic because of the multiple purple nodular lesions on the skin. Dermatologic examination revealed purple colored infiltrative nodular lesions in the trunk and extremities (Fig. 1). It was identified that the lesions of the case developed within one month and covered the whole body. It was learned that the case was followed up with AML M4 diagnosis. In the laboratory review; WBC: 30,9/ul Neu: 1,48/ul Monocyt: 23,8/ul Hgb: 10,5 g/dL Plt: 35,4/uL LDH: 1333 U/L CRP: 201,7 mg/L.

In terms of differential diagnosis, skin biopsies were taken with preliminary diagnosis (AML skin metastasis, lichenoid drug reaction, papular mucinosis, B cell lymphoma). Histopathologic examination, diffuse tumor infiltration in the dermis, strong CD33 (+) in the tumor cells due to weak CD117 and CD4 (+); myeloid originated (especially M4) leukemia infiltration (Figs. 2A – 2E).

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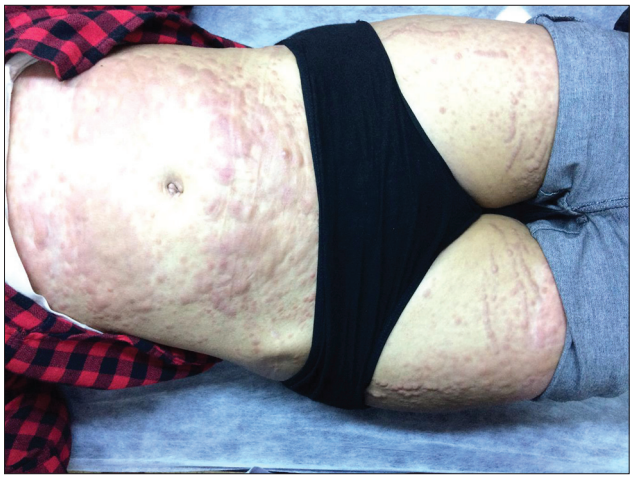

Figure 1: Purple colored infiltrative nodular lesions on the whole body.

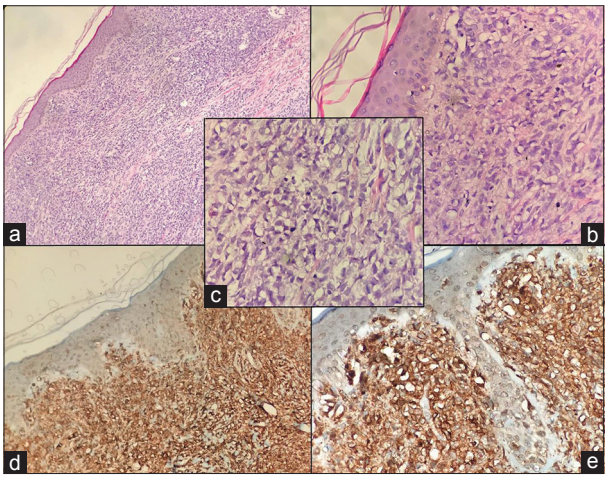

Figure 2: (a)X10, H&E. (b)X20, H&E. (c)X40, H&E; papillary and reticular dermis are seen as infiltrating with atypical blastic cells. (d)X20, LCA (leukocyte common antigen). (e)X40, CD33; immunocytochemically, blastic cells were positive with LCA and CD33.

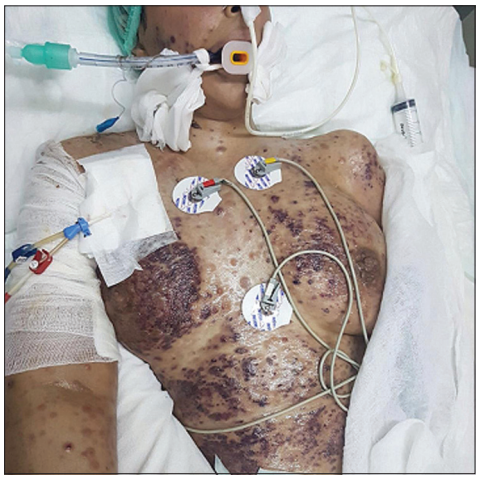

Figure 3: Hemorrhagic infiltrative nodular lesions on the whole body.

In the present case, sepsis was considered by hematology, the patient was hospitalized. The patient was admitted to intensive care unit and the multiple skin metastases were found to be hemorrhagic (Fig. 3). The patient died within one week due to sepsis in the intensive care unit. Informed consent was obtained from the case for the publication of this case report and images.

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

DISCUSSION

Cancers of the lung, kidney and gastrointestinal tract in males, breast cancer in females are the most common skin metastases [6]. Clinically, skin metastases are usually seen as nonspecific nodule, zosteriform pattern or carcinoma erysipeloides. More rarely, skin metastases may be seen in neoplastic alopecia, annular erythema and ulcer [7]. Lookingbill et al. have demonstrated that the most common lesion in cutaneous metastasis is the nodule [3]. In our case, multiple nodular lesions were observed throughout the whole body.

Histopathological examination is the most important property in the diagnosis of cutaneous metastases, as it is similar to that of the primary tumor [8]. In our study, histology of the sections showed acute myelomonocytic leukemia.

Most cases of cutaneous metastases are indicative of extensive spreading of the disease and death with worse prognosis [9]. The factor that influences the survival is the time elapsed between diagnosis and the occurrence of the cutaneous metastases. The treatment for most patients is palliative, and although chemotherapy and radiotherapy are frequently used in these patients, they are inconclusive in many cases [10].

Skin metastases can lead to a diagnosis of primary unknown cancer. However, skin metastases usually occur during the course of an existing cancer [11]. The presence of skin metastases is important in terms of showing the failure of treatment with cancer. When skin metastases are the first distant metastases, recognition of it is of great importance in terms of treatment change [12]. In our case, skin metastasis was the first metastasis detected.

Leukemia cutis (LC) is a condition presenting various cutaneous lesions such as papules, macules, plaques, nodules, ecchymoses, palpable purpura and ulcerative lesions, which are formed by skin invasion of neoplastic

cells [13]. Survive from diagnosis of leukaemia cutis to death ranged from 3 to 24 months. In 13% of LC cases, the underlying disease is AML, most commonly in the monocytic subtype of AML [14]. So in our case, Acute myelomonocytic leukemia was present.

Leukemia cutis lesions are generally localized, and can influence any part of the body. Along with being variable in the studies, it has been reported that the most common affected region is the trunk [15]. In our case it was interesting to find that AML M4 skin involvement is widespread. In the literature, there were no cases as wide as skin involvement of our case.

CONCLUSION

Leukemia cutis is an uncommon condition, its occurrence entails worse prognosis, as it is corporated with disease progression and survival reduce in these patients. We wanted to note that leukemia cutis should not be forgotten in the differential diagnosis of nodular skin lesions.

Consent

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

REFERENCES

1. Chopra R, Chhabra S, Samra SG, Thami GP, Punia RP, Mohan H. Cutaneous metastases of internal malignancies: a clinicopathologic study. Indian J Dermatol Venereol Leprol. 2010;76:125-31.
2. Song Z, Lin B, Shao L, Zhang Y. Cutaneous metastasis as a initial presentation in advanced non-small cell lung cancer and its poor survival prognosis. J Cancer Res Clin Oncol. 2012;138:1613-7.
3. Lookingbill DP, Spangler N, Helm KF. Cutaneous metastases in patients with metastatic carcinoma: a retrospective study of 4020 patients. J Am Acad Dermatol. 1993;29(2):228-36.
4. Douet-Guilbert N, Chauveau A, Gueganic N, Guillerm G, Tous C, Le Bris MJ, et al. Acute myeloid leukaemia (FAB AML-M4Eo) with cryptic insertion of cbfb resulting in cbfb-Myh11 fusion. Hematol Oncol. 2017;35:385-9.
5. Martínez-Leboráns L, Victoria-Martínez AM, Torregrosa-Calatayud JL, de Miquel VA. Leucemia cutis. Serie de 17 casos y revisión de la literatura. Actas Dermo-Sifiliograf. 2016;107:e65-9.
6. Alcaraz I, Cerroni L, Ruetten A, Kutzner H, Requena L. Cutaneous metastases from internal malignancies: a clinicopathologic and immunohistochemical review. The Am J Dermatopathol. 2012;34:347-93.
7. Wong CYB, Helm MA, Kalb RE, Helm TN, Zeitouni NC. The presentation, pathology, and current management strategies of cutaneous metastasis. North Am J Med Scien. 2013;5:499.
8. Kanyilmaz G, Aktan M, Koc M, Findik S. Cutaneous metastases of the synchronous primary endometrial and bilateral ovarian cancer: an infrequent presentation and literature review. Case Rep Oncol Med. 2016;2016:4568653.
9. Wick MR. Primary Lesions That May Imitate metastatic tumors histologically: A selective review. Semin Diagn Pathol. 2018;35:123-42.
10. Leonard R, Hardy J, Van Tienhoven G, Houston S, Simmonds P, David M, et al. Randomized, double-blind, placebo-controlled, multicenter trial of 6% miltefosine solution, a topical chemotherapy in cutaneous metastases from breast cancer. J Clin Oncol. 2001;19:4150-9.
11. Kara A, Belli AA, Alatas ET, Tanriverdi O, Dere Y. Widespread cutaneous metastasis from ovarian serous adenocarcinoma. Dermatol Online J. 2016;22:pii:13030/qt9mx8r32z.
12. Atış G, Tükenmez Demirci G, Kıvanç Atunay İ, Sakız D. The clinical characteristics and the frequency of metastatic cutaneous tumors among primary skin tumors. Turkderm. 2013;47:166-9.
13. Cronin DM, George TI, Reichard KK, Sundram UN. Immunophenotypic analysis of myeloperoxidase-negative leukemia cutis and blastic plasmacytoid dendritic cell neoplasm. Am J Clin Pathol. 2012;137:367-6.
14. Kang YS, Kim HS, Park HJ, Lee JY, Kim HO, Cho BK, et al. Clinical characteristics of 75 patients with leukemia cutis. J Korean Med Scien. 2013;28:614-9.
15. Peña-Romero AG, Domínguez-Cherit J, Méndez-Flores S. Leukemia cutis: clinical features of 27 Mexican patients and a review of the literature. Gac Med Mex. 2016;152:629-35.

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Our Dermatology Online

Case Report

Chemotherapy induced Beau's and Mee's line simultaneously: A case report and review of literature

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ABSTRACT

Chemotherapy affect skin and its appendages. Nail changes due to chemotherapeutic agents are common asymptomatic and usually resolve after discontinuation of therapy. There are various chemotherapeutic agents implicated for the changes like vincristine, hydroxyurea, etoposide, daunorubicin, bleomycin, cyclophosphamide etc. We report a case undergoing chemotherapy with etoposide and cyclophosphamide who had beau's and Mee's line simultaneously. Clinical acumen including history taking, knowledge about chemotherapy induced nail changes, counselling of the patient are prerequisite to diagnose such cases deferring unnecessary diagnostic workup and treatment.

Key words: Beu's line; Etoposide; Mee's line

INTRODUCTION

Chemotherapeutics agents can have not only systemic but as well as mucocutaneous side effects. Nail changes are common like nail dystrophies, chromonychia, leukonychia (including Mee's and Muehrcke's lines), Beau's lines, paronychia and onycholysis following anticancer drugs [1,2]. Few case reports has been mentioned in the literature of simultaneous appearance of the Mee's lines and Beu's lines in same patient.

CASE REPORT

A 20 year girl, diagnosed as Ewing sarcoma receiving chemotherapy including cyclophosphamide and Etoposide presented with multiple white continuous transverse bands with regular intervals covering whole width of nail plates with transverse depressions of fingernails and toenails as shown in Fig 1. During chemotherapy cycle white transverse lines moves distally as the nail grew, and new stripes developed after each cycle. She noticed this nail changes after starting of chemotherapy and completed 4 cycle till now. There were no history of any arsenic exposure or any medical illness in our patient. This white lines are called Mee's line and transverse depression are called Beu's lines. Patient was counseled about the nail changes due to chemotherapy.

DISCUSSION

Nail abnormalities due to result from drug toxicity leads to the matrix, nail bed, periungual tissues or digital blood vessels involvement. Chemotherapy agents implicated are – vincristine, hydroxyurea, etoposide, daunorubicin, bleomycin, cyclophosphamide, dacarbazine, 5-fluorouracil and methotrexate [2-4].

Mee's lines and Beu's lines occur due to temporary arrest of proliferative function of the nail matrix with cytotoxic chemotherapeutic agents. Mees' lines are signs of toxicity to the distal nail matrix, disorganized keratinization of the nail matrix leads to parakeratosis of the nail plate, which becomes white and opaque. True leukonychia (Mees' lines) due to drug toxicity shows as one or several parallel transverse white bands affecting all nails at the same level and which moves distally with nail growth. Leukonychia is seen in arsenic and thallium intoxication and also had been reported in various medical diseases such as myocardial infarction,

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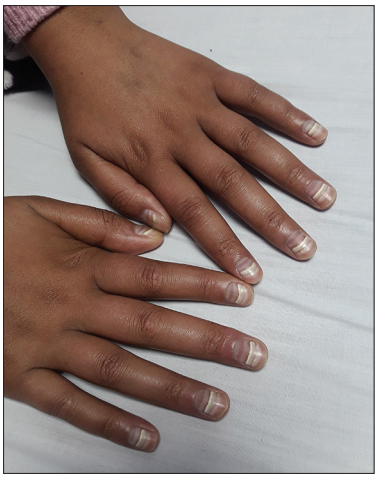

Figure 1: Bilateral fingernails of hands shows Mee's lines and Beu's lines.

acute and chronic renal failure, kidney allograft rejection, systemic lupus erythematosus, immune hemolytic anemia and Hodgkin's disease [5]. Beu's lines are signs of acute toxicity to the nail matrix with transient arrest in nail plate production due to decrease in matrix cell proliferation which are associated with temporary cessation of nail growth according to cycles of chemotherapy shows a transverse depression that migrates distally as the nail grows.

There is difference of proliferative potential between fingernails and toenails which leads to different clinical manifestations as per Kim et al. Rapid mitotic activity having organs are likely to damaged more severely from chemotherapy in much the same way fingernails are more vulnerable to anticancer drugs than toenails but in his case report rapidly proliferating fingernails were less affected by chemotherapeutic nail matrix damage; consequently, less severe depression and less opaque Mees' lines manifested. The depth of the depression indicates the degree of the damage, and the width indicates the duration of the insult [6]. We are reporting nail changes like Beau's line or Mees' line occurring in a same patient after chemotherapy for the first time from our part of the world. There are only two case reports, having both Mees lines and Beus lines had been previously reported in the literature till date [6,7]. The most common nail finding observed was melanonychia which was seen in 26 (78.7%) patients, followed by Muehrcke's lines, Mee's lines, and Beau's lines [8].Nail changes can be the only symptoms after chemotherapy so a good history, knowledge about nail changes following chemotherapy, counselling of the patient and good clinical acumen is necessary to diagnose such cases alleviating unnecessary diagnostic workup and treatment.

Consent

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

REFERENCES

1. Gupta A, Parakh A, Dubey AP. Chemotherapy induced nail changes. Indian J Dermatol. 2008;53:204-5.
2. Hinds G, Thomas VD. Malignancy and cancer treatment- related hair and nail changes. Dermatol Clin. 2008;26:59–68.
3. Dasanu CA, Vaillant JG, Alexandrescu DT. Distinct patterns of chromonychia, Beau's lines and melanoderma seen with vincristine, adriamycin, dexamethasone for multiple myeloma. Dermatol Online J. 2006;12:10.
4. Chang P, Vásquez Acajabón MV. Beau´s lines due to cytostatic drugs in a patient with breast cancer. Our Dermatol Online. 2014;5:198-200.
5. Piraccini BM, Iorizzo M, Tosti A. Drug-induced nail abnormalities. Am J Clin Dermatol. 2003;4:31–7.
6. Kim IS, Lee JW, Park KY, Li K, Seo SJ, Hong CK. Nail change after chemotherapy: simultaneous development of Beau's lines and Mees' lines. Ann Dermatol. 2012;24:238-9.
7. Kinjo T, Shibahara D, Higa F, Fujita J. Beau's Lines and Mees' Lines Formations after chemotherapy. Intern Med. 2015;54:2281.
8. Pavey RA, Kambil SM, Bhat RM. Dermatological adverse reactions to cancer chemotherapy. Indian J Dermatol Venereol Leprol. 2015;81:434.

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Our Dermatology Online

Case Report

Diffuse melanosis cutis secondary to metastatic malignant melanoma: Case report

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ABSTRACT

Diffuse melanosis cutis (DMC) is an exceptional presentation of metastatic melanoma characterized by progressive blue-gray discoloration of the skin and mucous membranes. It attests a generally advanced metastatic tumor extension. We report an additional case of diffuse melanosis associated with widespread cutaneous and visceral metastases from malignant melanoma. The prognosis are poor in patients with diffuse cutaneous melanosis associated with malign melanoma.

Key words: Diffuse melanosis cutis – melanoma - metastatic

INTRODUCTION

Diffuse melanosis cutis (DMC) is an exceptional presentation of metastatic melanoma characterized by progressive blue-gray discoloration of the skin and mucous membranes [1]. It attests a generally advanced metastatic tumor extension. The discoloration represents the abnormal presence of pigment within the dermis causing scattering of light [1], and is usually more intensive on sun-exposed areas, including head and upper torso [2]. Usually, dark urine associated with melanüri, darkening of the serum and peritoneal fluid can be also seen [3].

Histological findings are relatively uniform in DMC with the most consistent feature reported being the presence of dermal pigment [1].

Due to the rarity of the observed phenomenon the pathogenesis of DMC is yet to be fully understood.

In the literature, approximately 72 cases of DMC have been described until now, most of them being associated with multiple organ metastases [2].

Patients with melanosis cutis have a bleak prognosis with survival times of only weeks or months after the onset of melanosis [4].

We report an additional case of diffuse melanosis associated with widespread cutaneous and visceral metastases from malignant melanoma

CASE REPORT

A 32-year-old woman, phototype IV (Fig. 1), with history of familial melanomas, had been operated for non-metastatic melanoma of the left thigh. Five years later, the patient presented a local recurrence of her melanoma with homolateral ganglionic involvement. The results of the extension showed cutaneous, subcutaneous, ganglionic and pulmonary metastases. The patient had palliative surgery associated with chemotherapy with dacarbazine followed by paclitaxel. The evolution was marked by a change of phototype with diffuse pigmentation. The clinical examination found a patient with phototype V (Figs. 1a and 1b), a diffuse and homogeneous melanoderma which interest the skin, the nails and the mucous membranes, more pronounced in photo exposed zones (Figs. 2a, 2b and 2d). Dermoscopy showed an homogeneous pigmented pattern (Fig. 2c), with diffuse regular melanonychic bands on all the nails (Fig. 2e). A skin biopsy, taken from his face area revealed melanin deposition in the dermis. Urine analysis showed a brown-amber color

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(Fig. 3). The metabolic and endocrine blood tests were negative. The patient was dead 15 months later.

DISCUSSION

Diffuse cutaneous melanosis (DCM) is rarely encountered condition in malign melanoma. It is often associated with liver, lung and lymph nodes metastases. DCM also can be seen in Addison disease, porphyria cutanea tarda and hemochromatosis [3].

Until 2017, more than 70 cases of DMC were reported, however, mechanisms of the condition have not been elucidated [2]. So far, Different pathogenic mechanisms leading to diffuse melanosis have been discussed.

Bohm et al suggested that cutaneous diffuse melanosis is linked to an excessive production of melanocytic growth factors (MSH (melanocyte stimulating hormone), Hepatocyte growth factor (HGF), Endothelin-1 (ET-1)). Excessive production of MSH (from tumor), HGF and ET-1 (from distinct site of metastasis) induce normal and malignant melanocytes resulting in enhanced proliferation, melanogenesis and melanin releasing [3].

Other several lines of evidence suggest that the direct cause of the skin darkening is melanin derived from the melanoma micrometastases in the dermis [2].

On the other hand, other reports indicate that it is plausible that cytolytic melanoma deposits liberate vast quantities of melanin precursors, as well as free melanin and melanosomes into the circulation. This may occur as a result of rapid turnover of neoplastic deposits, ischemia, immunological responses, or oncologic therapies [1]. Circulating precursors would have the capacity to traverse the glomerulus and be excreted in the urine, causing melanuria [1-6].

Subsequently, an other hypothesis is the peripheral oxidation of melanin precursors. These precursors generated in the tumor pass through the capillary membrane into the dermis and are converted into melanin by oxidizing systems 4]. This theory has been supported by the documented observations of melanin precursors, melanin, melanosomes, and melanophages in patients' serum [2].

In our patient as well, diffuse melanosis appeared after beginning chemotherapy.

The findings of histomorphological examinations are described differently [4]. The most often described

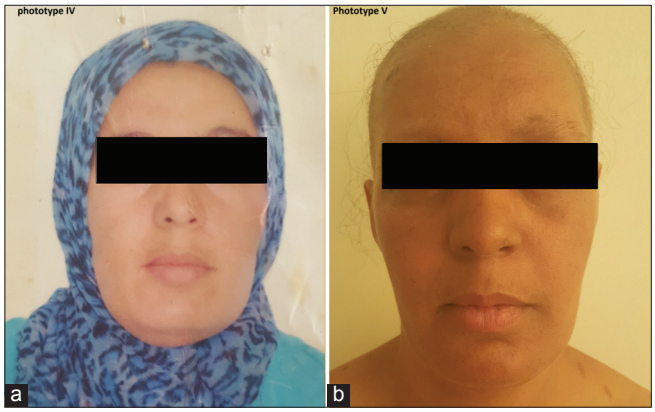

Fig. 1: Phototype of the patient before and after disease.

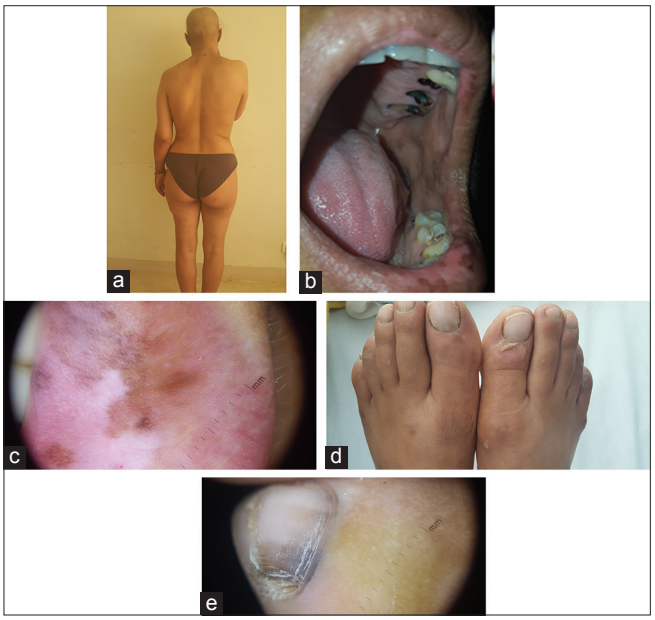

Fig 2 : a: Diffuse and homogeneous melanoderma which interest the skin, more pronounced in photoexposed zones b: Diffuse and homogeneous melanoderma which interest the mucous membranes c: Dermoscopy showed an homogeneous pigmented pattern d: Diffuse and homogeneous melanoderma which interest the nails e: Dermoscopy showed an homogeneous pigmented pattern, with diffuse regular melanonychic bands on all the nails.

histological finding, was the presence of pigmented granules but not of malignant cells [5]. In most of the cases, biopsy specimens revealed that the hyperpigmentation resulted from a marked deposition of pigment in the dermis,Another finding was giant melanosomes in melanocytes, keratinocytes and melanophages [3,4]. Micrometastases or single metastatic melanoma cells, as rarely have been described.

Treatment of diffuse melanosis cutis involves treating the underlying melanoma. There have been no reports of diffuse melanosis cutis reversing [7].

Fig 3: Urine analysis showed a brown-amber color

The prognosis are poor in patients with diffuse cutaneous melanosis [3,4]. The mean duration between diagnosis of melanoma and the onset of melanosis is less than a year. And the survival time from the onset of DMC is approximately 4 months [2].

DCM should be considered in the differential diagnosis list of skin discoloration after excluding other common differential diagnoses.

CONCLUSION

Melanosis cutis is a rare event in advanced metastatic melanoma whose exact mechanisms of development are still unclear. However, the role of micrometastases, as well as melanin precursors, released during lysis of MM metastases, and growth factors play a role. It is associated with a grim and poor prognosis of preterminally affected patients with a mean survival of approximately 4 months

CONSENT

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

REFERENCES:

1. Sebaratnam DF, Venugopal SS, Frew JW, McMillan JR, Finkelstein ER, Martin LK, et al. Diffuse melanosis cutis: A systematic review of the literature. J Am Acad Dermatol. 2013;68:482-8.
2. Maj J, Jankowska-Konsur A, Gruber J, Woźniak Z, Nockowski P, Hryncewicz-Gwóźdź A. Diffuse melanosis cutis related to dermal micrometastases as the first clinical symptom of distant metastatic malignant melanoma. Medicine (Baltimore). 2017;96:e6470.
3. Oruç, Kaplan MA, Yerlikaya H, Urakçı Z, Küçüköner M. A metastatic malign melanoma case with diffuse cutaneous melanosis. J Oncol Scien. 2016;2:71-2.
4. Hofmann M, Kiecker F, Audring H, Grefer K, Sterry W, Trefzer U. Diffuse Melanosis cutis in Disseminated Malignant Melanoma. Dermatology. 2004;209:350-2.
5. El Hadj OE, Bouhajja L, Goucha A, Rekik W, El May A, Gamoudi A. Dubreuilh's melanosis or malignant lentigo. Our Dermatol Online. 2017;8:231-2.
6. Guérin M, Chappard D, Giard C, Dompmartin-Blanchière A, Martin L. Mélanose et mélanurie au cours d'un mélanome multimétastatique. Imag Dermatol. 2012;5:1.
7. Minocha R, Kefford R, Uribe P, Sebaratnam DF, Fernandez-Penas P. Diffuse melanosis cutis in the setting of BRAFV600E mutant melanoma and treatment with targeted therapies. Australas J Dermatol. 2015;56:128-30.

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

Our Dermatology Online

Case Report

A case report of familial combined hypercholesterolemia

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ABSTRACT

Xanthomas are the characteristic cutaneous presentation in hyperlipoproteinemias. Familial combined hyperlipoproteinemia/type II b presents with high plasma cholesterol, high plasma low density lipoprotein (LDL) cholesterol, moderately high plasma triglycerides. Cutaneous findings include tendinous, tuberous and intertriginous xanthomas and xanthelasma palpebrarum. A 5 year old boy presented with multiple yellowish lesions over the joints, gluteal cleft, popliteal fossa. Family history of second degree consanguinity was present with hyperlipidaemia in father, mother and brother. Early diagnosis and treatment mitigate the excess risk of premature atherosclerotic cardiovascular disease that occurs with familial hypercholesterolemia.

Key words: Familial combined hypercholesterolemia; Hyperlipidaemias; Xanthoma

INTRODUCTION

Familial combined hypercholesterolemia is an autosomal dominant disorder characterized by increase in LDL, VLDL, TG levels. The primary defect is a reduction in LDL receptors and increased apoB lipoprotein. Xanthomas are not frequently seen, but can present with tendon, tuberous and plane xanthomas.

CASE REPORT

A 5 year old male child was brought by his mother with complaints of multiple yellowish to skin coloured lesions present over the joints since 2 years, which progressed to involve other sites. There was no history of pain over the lesions, trauma, weight gain, hoarseness of voice, intolerance to cold, lethargy, chest pain, easy fatigability, breathlessness.

Family history of second degree consanguinity was present with hyperlipidaemia in father, mother and brother. Mother additionally had xanthelasma.

On Examination

Cutaneous - Multiple yellowish to skin coloured, firm, mobile, non-tender, lobulated plaques were present over both knee and ankle joints.(figure-1). Single skin coloured nodule was present on each elbow joint, base of right great toe (Fig. 1). Skin coloured to yellowish plaque was present over the gluteal cleft, popliteal fossa (Fig. 2). Arcus juvenalis was present in the eyes (Fig. 3). Oral cavity, hair and mucosa were normal.

Investigations

Routine investigations including blood counts, blood sugar, chest X-ray, thyroid profile, liver and renal function tests were within normal limits [Table 1]. ECG and ECHO was normal. Ultrasound of the abdomen was normal. Histopathology of the section studied from skin showed epidermis with marked thinning (Fig. 4a). The dermis predominantly had foamy histiocytes in aggregates and sheets. Touton type of giant cells interspersed with fibroblasts giving a storiform arrangement was observed at places (Fig. 4b).

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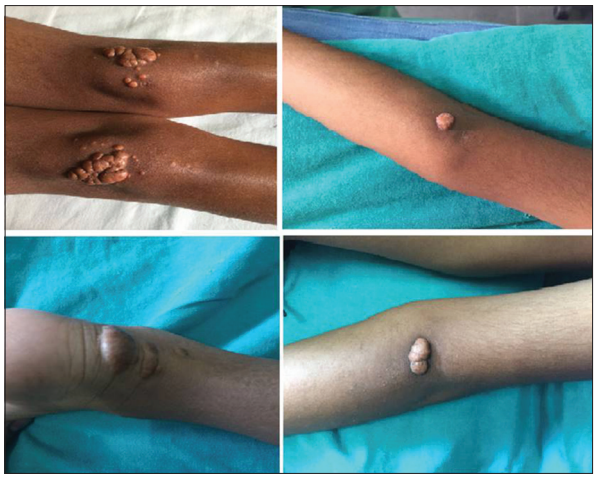

Figure 1: Multiple tuberous xanthomas over knee, elbow and ankle joints.

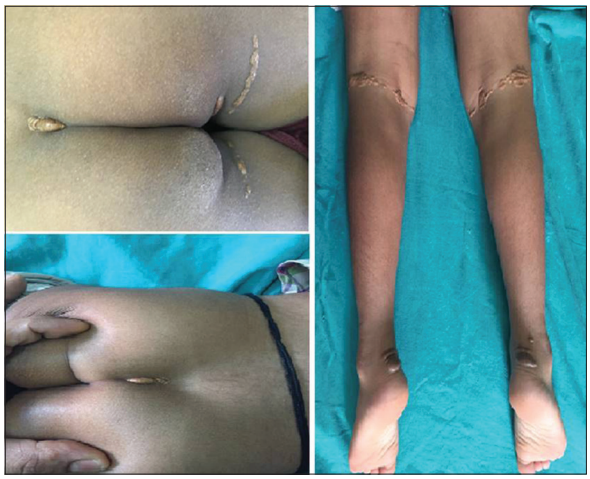

Figure 2: Plane xanthoma over popliteal fossa, gluteal cleft.

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

DISCUSSION

Disorders of lipid metabolism are heterogeneous. They range from monogenic diseases with high penetrance through polygenic disorders.

Circulating lipids are cholesterol, cholesterol esters, triglycerides and phospholipids. They are insoluble and therefore have to be solubilized by combination with proteins like lipoproteins and the proteins they contain are generally called apolipoproteins. The hyperlipoproteinemias are disturbances of lipid transport of cholesterol and triglycerides through plasma [1].

Figure 3: Arcus juvenalis.

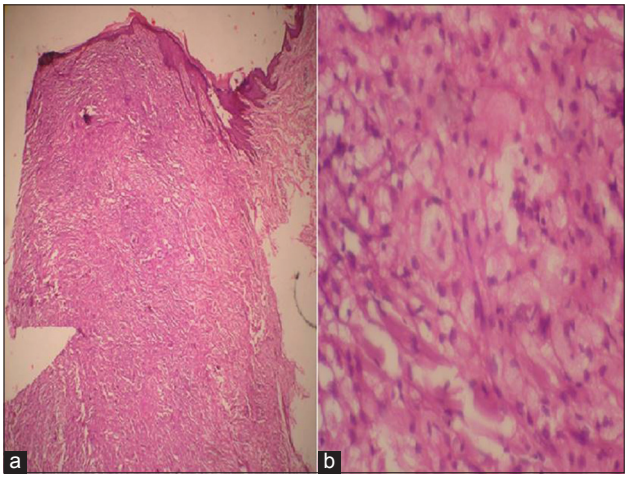

Figure 4: (a) Epidermis and Dermis and (b) showing foamy histiocytes in aggregates and sheets.Touton type of giant cells.

They are classified as WHO/Fredrickson's classification of hyperlipoproteinemia/hyperlipidemia [2]:

Type Lipoprotein abnormality
- I Hyperchylomicronemia
- IIa Elevated LDL (familial hypercholesterolemia)
- IIb Elevated LDL and VLDL (familial combined hypercholesterolemia)
- III Broad β-VLDL (familial dysbetalipoproteinemia)
- IV Elevated VLDL (familial hypertriglyceridemia)
- V Elevated chylomicrons and VLDL (mixed hyperlipidemia)

Xanthomatas are commonly caused by a disturbance of lipoprotein metabolism [3]. They can be tuberous, tendinous, eruptive, plane (palmar, intertriginous, diffuse, xanthelasma), others (corneal arcus, tonsillar) [4,5].

Familial combined hypercholesterolemia is characterized by the finding of hypercholesterolemia and hypertriglyceridemia within the same kindred

Table 1: Fasting lipid profile of family members

Test	Patient mg/dl	Father mg/dl	Mother mg/dl	Brother mg/dl	Normal range
Triglycerides	196	663	222	179	Up to 150 mg/dl
Total cholesterol	1045	358	284	316	Up to 200 mg/dl
HDL cholesterol	25	52	40	31	30-60 mg/dl
LDL cholesterol	969	215	174	249	Up to 100 mg/dl
VLDL cholesterol	39	38	44	36	Up to 20 mg/dl

and with-kindred members having either one of these abnormalities or both. Patients of familial combined hyperlipoproteinemia type II b present with high plasma cholesterol, high plasma low density lipoprotein (LDL), moderately high plasma triglycerides, tendinous, tuberous and intertriginous xanthomas and xanthelasma palpebrarum [3].

A diet low in cholesterol and saturated fats and high in polyunsaturated fats is recommended. Drugs like gemfibrozil, clofibrate are the first choice. Statins may also be used [4,6].

CONCLUSION

This case is being presented to highlight the rarity of tuberous, tendon, arcus juvenalis, plane xanthoma with familial combined hypercholesterolemia.

Consent

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

REFERENCES

1. Flynn PD. Xanthomas and abnormalities of lipid metabolism and storage. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. Rook's Textbook of Dermatology. 8th ed. Oxford: Blackwell Science; 2010. p. 59.81-59.103.
2. Pai VV, Shukla P, Bhobe M. Combined planar and eruptive xanthoma in a patient with type lla hyperlipoproteinemia. Indian J Dermatol Venereol Leprol. 2014;80:467-70.
3. Bhagwat PV, Tophakhane RS, Kudligi C, Noronha TM, Thirunavukkarasu A. Familial combined hypercholesterolemia type II b presenting with tuberous xanthoma, tendinous xanthoma and pityriasis rubra pilaris-like lesions. Indian J Dermatol Venereol Leprol. 2010;76:293-6.
4. Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Hamlin, Leffell DJ, Wolff K. Xanthomatoses and Lipoprotein disorders. Fitzpatrick's Dermatology in General Medicine. 8th edition. Mc Graw Hill companies: 2012;1600-12.
5. Hilal Ayvaz H, Çelik G, Gönül M, Kılıç A, Özcan N, Çolak A. A case of adult onset dısseminated juvenile xanthogranuloma. Our Dermatol Online. 2016;7:66-8.
6. Elsy B, Khan AA, Maheshwari V. Effect of vitamin E isoforms on the primary intention skin wound healing of diabetic rats. Our Dermatol Online. 2017;8:369-75.

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Our Dermatology Online

Case Report

Rare case of Behcet's disease in an African patient: A case report and review of the literature

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ABSTRACT

Behcet's disease was first described by Hulusi Behcet, is a rare immune-mediated small-vessel vasculitis with multisystemic manifestations. Patients with this disease usually present with recurrent oral aphthous ulcers, genital ulcers and eye lesions. A 61 year old African man presented to the dermatology clinic with recurrent oral and penile ulcers of 26years duration, associated with grittiness of the eye and joint pains. Pathergy test was positive and ESR was elevated. Diagnosis of Behcet's disease was made on clinical grounds. Behcet's disease though rare could occur in patient with Fitzpatrick type VI skin and a high index of suspicion coupled with innovative approach is required to optimize patient outcome in a poor resource limited African setting.

Key words: Behcet's disease; Fitzpatrick type-6 skin; Resource-Limited setting; Skin of color

INTRODUCTION

Behcet's disease, first described by Hulusi Behcet, is a rare immune-mediated small-vessel vasculitis. It often presents with a triple-symptom complex of recurrent oral aphthous ulcers, genital ulcers and eye lesions [1]. This condition has seldom been reported in Africans [2]. This article reports a case of Behcet's disease occurring in an African, and showcases the challenges of complex medical dermatology practice in resource-limited settings. It also presents a concise review of the literature on Behcet's disease.

CASE REPORT

A 61year old African man presented to the dermatology clinic with recurrent oral and penile ulcers, of 26years duration. There was associated grittiness, redness of the eye and eye pain, as well as joint pain. There was no history of hair loss or nail abnormality, photosensitivity, proximal muscle weakness or muscle pain.

On examination, there were tender, punched-out ulcers on the tongue and penis, as well as erosions on the palate and buccal mucosa (Figs. 1 and 2). There were several areas of post inflammatory hyperpigmentation in these regions.

Appropriate investigations were requested. ESR was elevated at 62mm/hr. Full blood count was within normal limits. HIV screening, syphilis tests and hepatitis panel were negative. Pathergy test was positive. The patient declined biopsy of ulcers due to financial constraints.

Based on clinical judgment and laboratory findings, a diagnosis of Behcet's disease was made.

Over the course of four months, he has been treated with methylprednisone cream, oral prednisone, clarithromycin, amoxicillin, arthrotec (Diclofenac/misoprostol), azathioprine. He was also referred to the ophthalmologist. Follow up visit is conducted monthly. Despite, inconsistent use of medications due to financial constraints, a progressive healing of ulcers,

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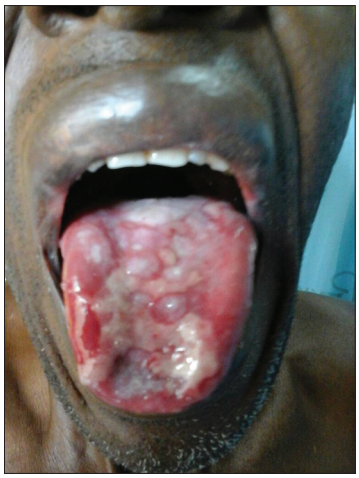

Figure 1: Mouth ulcers.

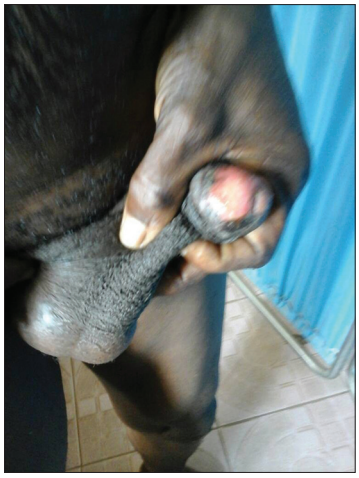

Figure 2: Penile ulcers.

and reduction in ulcer occurrence, reduction in eye and joint symptoms are being observed.

DISCUSSION

Behcet's disease is primarily a clinical diagnosis. It is a multi-system disease that manifests with orogenital ulceration [1]. Other findings may include arthritis, gastrointestinal lesions, neurological involvement, aneurysm and thrombophlebitis, erythema nodosum etc.

Behcet's disease has a higher prevalence in Mediterranean region, Middle East and East Asia [2].

It is rare in Northern Europe, Africa and America. Disease onset is usually in the third and fourth decade of life.

The pathogenesis is hypothesized to be a combination of hereditary and environmental factors [3].

Familial cases demonstrate genetic anticipation. IL-6 and TNF-alpha has been shown to be elevated and IL-10 decreased, compared with controls. There is an association with HLA-B51.

Biopsy of the ulcers reveal neutrophilic, lymphocytic and plasma cell infiltrates. Occasionally necrotising vasculitis is observed [4]. Pathergy test is typically positive, with elevation of inflammatory markers such as ESR and CRP [3,4]. Based on the systems involved, appropriate investigations should be conducted such as endoscopy, angiography etc.

Two sets of criteria are commonly used for diagnosis of Behcet's disease- the International criteria for Behcet's disease, and the O'Duffy criteria [5].

Differential diagnosis includes traumatic ulceration, syphilis, Chancroid, Herpes simplex ulcers, systemic lupus erythematosus, reiter disease.

The choice of treatment depends on organs affected and disease severity [3,6]. Therapy include potent topical and systemic steroids, cytotoxic medications (e.g azathioprine, dapsone, Colchicine methotrexate, cyclophosphamide, thalidomide,), biologics (e.g etanercept, infliximab), oral antibiotics, pain relief agents, antiseptics.

Behcet's disease has an undulating course of exacerbations and remissions, with worse prognosis in males. Complications include blindness from ocular involvement, vascular aneurysm rupture and hemorrhage, progression to dementia from Neuro-Behcet syndrome, and side effects from long-term treatment regimens.

CONCLUSION

This case report demonstrates that, although rare, Behcet's disease could manifest in Fitzpatrick type VI skin. It also highlights the challenges of managing complex medical dermatology cases in resource-limited settings.6 Pertinent issues include unaffordability and unavailability of essential medications and diagnostic tests. An innovative approach, coupled with high clinical acumen, is required to optimize patient outcomes.

Consent

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

REFERENCES

1. Ideguchi H, Suda A, Takeno M, Ueda A, Ohno S, Ishigatsubo Y. Behçet disease: evolution of clinical manifestations. Medicine (Baltimore). 2011;90:125-32.
2. Leonardo NM, McNeil J. Behcet's Disease: Is There Geographical Variation? A review far from the silk road. Int J Rheumatol. 2015;2015:945262.
3. Mazzoccoli G, Matarangolo A, Rubino R, Inglese M, De Cata A. Behçet syndrome: from pathogenesis to novel therapies. Clin Exp Med. 2016;16:1-12.
4. Gündüz O.Histopathological Evaluation of Behçet's disease and identification of new skin lesions. Patholog Res Int. 2012;2012:209316.
5. Davatchi F. Diagnosis/classification criteria for Behcet's disease. Patholog Res Int. 2012;2012:607921.
6. Asemota E, Slaught C, Micheletti R, Kovarik C. Optimizing "best available" medical options when practicing complex medical dermatology in resource-limited settings. J Am Acad Dermatol. 2016;75:e171-2.

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

Our Dermatology Online

Case Report

Atypical presentation of pemphigus vulgaris - A rare case report

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ABSTRACT

Pemphigus vulgaris is an autoimmune blistering disorder affecting the skin and mucous membrane with characteristic intraepithelial blistering. It commonly involves the oral mucosa and skin, Oral mucosal involvement may proceed the skin involvement. We here in report a case, where in the oral and skin lesion occurred concurrently in a 20 year old male with a 1 month history of multiple fluid filled lesions over the body, who is a known case of psychosis and epilepsy treated at many places as case of drug rash finally diagnosed in our hospital as case of pemphigus vulgaris.

Key words: Pemphigus vulgaris; Auto immune; Blisters; Drug rash

INTRODUCTION

Pemphigus derives its name from greek 'pemphix' means blister or bubble. It is an auto immune blistering disorder affecting the skin and mucous membrane of oral cavity, characterised by supra basal split in the epidermis. Nearly all patients have mucosal involvement and pemphigus vulgaris presents as oral lesions in 50 to 70% patients [1-3]. These may precede the cutaneous lesions by months or may be the only feature of the disease. Pemphigus vulgaris is most common form of pemphigus accounting for 70% cases of cases [3]. It is the commonest auto immune blistering disorder in Eastern countries like India, Malaysia, China and Middle east. It is a disease of the middle age, but patients are at a younger age at presentation in India compared to western countries and both sexes are equally affected.

CASE REPORT

A 20 year old male patient since 1 month presented complaints of multiple filled lesions on the body. Patient was dumb and mentally retarded. History was obtained from parents. Initial lesions started in both upper limbs followed by oral cavity and lower limbs for which medication was taken but was not relieved. There is no history of constitutional symptoms. Patient is a known case of psychosis and epilepsy since the age of 7 years. Patient was on anti epileptic drug phenobarbitone 60 mg, has stopped taking medication since 8 months as he developed aggression. Patient was taken to various hopitals where he was treated as a case of drug rash, with intravenous antibiotics, anti histaminics, topical antibacterial cream.

His vital signs were temperature 99° F, pulse 90 beats per min, respiration 16 cycles/min, blood pressure 110/70 mm Hg, height 5'3", weight 50 kg. On examination, patient was ill, moderately built and moderately nourished. Skin is characterised with multiple tense fluid filled lesions over the upper limbs, lower limbs abdomen, chest and back were spared and erosions were present over the oral cavity as shown in Fig. 1 and 2. Conjunctiva were congested. Routine laboratory examination revealed Haemoglobin 11g/dl, ESR 40 mm/hr, PCV 24%, RBS 92.8 mg/dl, Albumin 3.09g/dl, Blood urea 13.1mg/dl, Serum creatinine, Serum bilirubin, Alkaline phosphatase levels were with in normal limit. Tzanck smear was done for the patient

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DOI: 10.7241/ourd.20191.25

which showed few giant cells, Biopsy was done from the lesion and histopathology report revealed the following features: On microscopy (Fig. 3), epidermis shows suprabasal clefting with vesicle formation and tomb stone appearance, Vesicle is filled with neutrophils, eosinophils and acantholytic cells. Dermis shows mild perivascular lymphocytic infiltrate, Features are suggestive of Pemphigus vulgaris.

Patient was given given started on Steroids, Antibiotics, Saline compresses, Topical Antibacterial cream, H2 receptor blockers, Multivitamin capsules, Antihistaminics Tetracycline mouth gargles, Antioxidants, Topical steroid for lips, Oral analgesic gel, Ophthalmic eye drops. On follow up patient was responding well to the treatment.

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

DISCUSSION

Pemphigus vulgaris is a blistering disorder of skin and mucous membrane. Predisposition to pemphigus is linked to genetic factors. The diagnosis of this disease is clinical and confirmed by histopathological examination. Nearly all patients may have mucous membrane involvement and oral cavity is commonly involved, more commonly patients have ill defined, irregularly shaped buccal or palatal erosions which are slow to heal lesions are painful which affects the appetite of the patient. Intact bullae are rare in oral cavity. Other mucous membranes involved are conjunctiva, nasal, pharynx, larynx, oesophagus, urethra, vulva and cervix [4]. Most patients develop cutaneous lesions which may remain localised or more commonly wide spread. The disease has a predilection for scalp, face, axilla, groin and pressure points. Flaccid blisters with clear fluid either arise on a normal or an erythematous base. The blisters are easily broken forming epidermal rings. Histopathological examination reveals eosinophilic spongiosis, suprabasal split with groups of acantholytic cells in the cavity, basal layer keratinocytes shows loss of adhesion with adjacent keratinocytes and characteristic row of tomb stone appearance. The basic therapy options consists of local or systemic corticosteroid therapy. Adjuvant therapies like cyclophosphamide and azathioprine are commonly used, dapsone, cyclosporine, mycophenolate mofetil and methotrexate have also been tried. Other drugs like colchicine, thalidoamide and retinoids have also been used in mild to moderate cases. Recent trials have shown an advantage that with use of low dose methotrexate to be more efficacious and relatively few side effects. Intralesional injections of

Figure 1: Erosions on the lips.

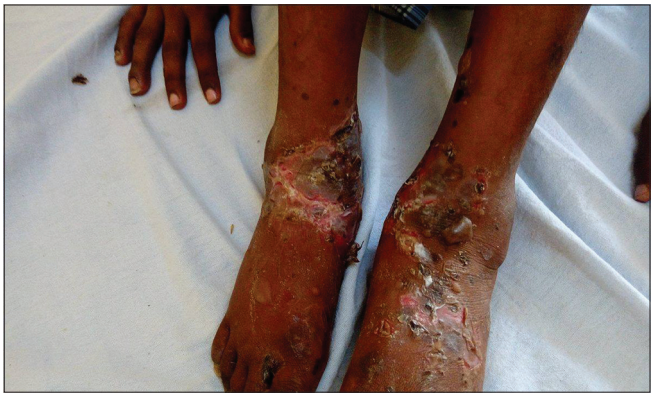

Figure 2: Skin lesions on the left forearm.

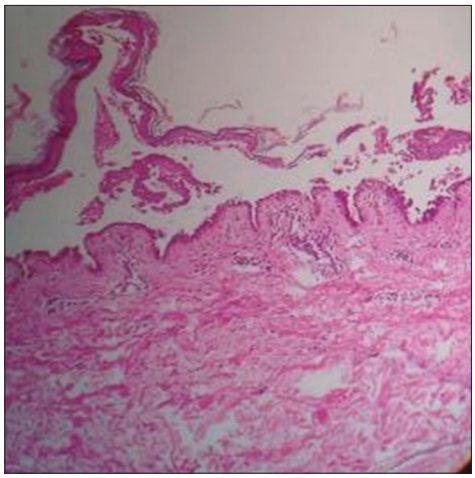

Figure 3: Intraepidemal bulla in the basal layer and dermis showing mononuclear cells.

corticosteroids have also been used in treatment of persistent oral lesions [5]. Traditional drugs combined with antioxidants, calcium and vitamin supplements should be given, antiseptic mouth washes should be given to improve local oral hygiene.

CONSENT

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

CONCLUSION

Pemphigus vulgaris is an autoimmune disorder the mucous membrane and skin, that is often misdiagnosed, there is an increase in morbidity rate. The diagnosis is confirmed by clinical and histopathological examination. Therapeutic and adjuvant treatments help the patients to relieve from symptoms.

REFERENCES

1. Arpita R, Monica A, Venkatesh N, Atul S, Varun M. Oral Pemphigus Vulgaris: Case Report. Ethiop J Health Sci. 2015;25:367-72.
2. Matias AB, Ferreira Roselino AM. Pemphigus: a disease stamped in the skin. Our Dermatol Online. 2013;4(Suppl.3):601-5.
3. Rama Rao GR, Koteswara Rao NR, Sridevi M, Amareswar A, Chowdary AP. Pemphigus vulgaris with squamous cell carcinoma of the tongue: An uncommon association. Our Dermatol Online. 2017:8:286-8.
4. Kabra V, Pai K, Pai S, Shenoi S, Rao R. Pemphigus vulgaris masquerading as subcorneal pustular dermatoses-a case report. Our Dermatol Online. 2014:5:157-9.
5. Benhiba H, Hamada S, Guerouaz N, Saidi A, Senouci K, Hassam B. Pemphigus vulgaris: an unusual clinical presentation. Ann Dermatol Venereol. 2013:140:116-9.

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Our Dermatology Online

Clinical Image

Acne fulminans: A rare form of acne

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An 18 year-old male patient visited our hospital seeking dermatological care. He had history of mild acne vulgaris treated occasionally by topical treatments with little improvement, and had never taken isotretinoins. There was no family history of severe acne or isotretinoin use. He complained of rapid flare up of his acne lesions, within 2 months, with development of severe and extensive lesions, involving his face, back, shoulders and chest. Furthermore, he reported the onset of fever, along with chills and arthralgia of the shoulders which limited his daily activities. Physical examination showed multiple inflamed nodulocystic lesions (Fig. 1), painful on palpation, along with erythematous papules and extensive ulcerations with tick and adherent melliceric and hemorrhagic crusts on his face, back, shoulders and chest (Figs. 2 and 3). Initial testing revealed a white blood count of 17 000 without liver abnormalities. Radiographs didn't show any bone lesions. The diagnosis of acne fulminans was made and the patient was put on prednisone 30 mg daily with antibiotics. Four weeks later, isotretinoin was started at a low dose of 20 mg/day and corticosteroids were gradually tapered. A good clinical response with healing of the fever and arthralgia and progressive amelioration of his nodulocystic lesions was found in our patient.

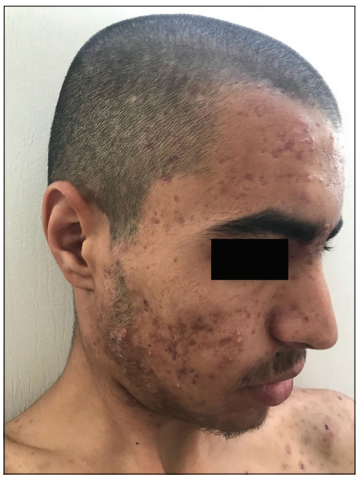

Figure 1: Multiple, scaly and some crusted erythematous papules and pustules, with confluent nodules and hemorrhagic ulcers on the forehead and both cheeks.

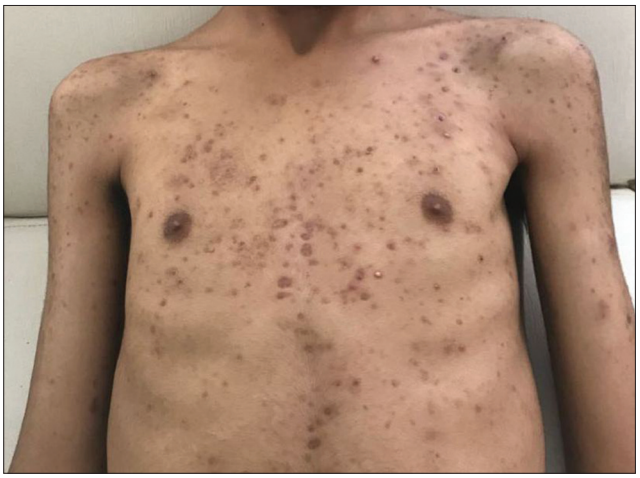

Figure 2: Papules, pustules, nodules and ulcerations on the chest.

Acne fulminans is a rare and severe ulcerative form of acne with an acute onset and systemic symptoms, mainly affecting young male aged 13-22 years, with history of acne vulgaris [1]. It is characterized by rapid onset of painful inflammatory nodules in the habitual areas of acne, which become suppurative, ulcerated and covered by hemorrhagic crusts [2]. The trunk is strongly affected, especially the back, but also the shoulders and face [3]. Systemic symptoms are essential for the diagnosis, most commonly fatigue, malaise, arthralgias, myalgias and fever [1]. Laboratory tests show blood count abnormalities, with leukocytosis and neutrophilia, and elevated erythrocyte sedimentation rate [3]. The treatment has been challenging and must be aggressive. It consists

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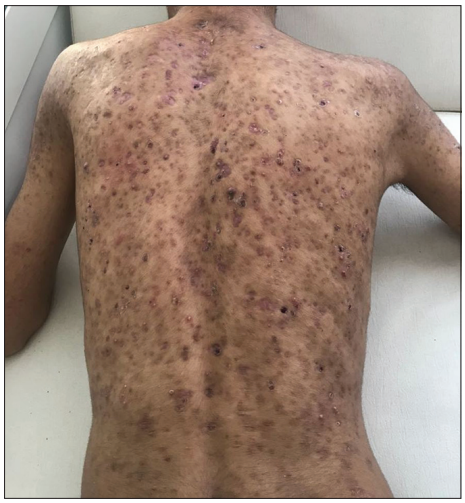

Figure 3: Diffuse ulceronecrotic lesions on the back with crusts and multiples erythematous papules and pustules.

of the use of oral steroids alone then in combination with oral isotretinoin [1].

CONSENT

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

REFERENCES

1. Zaba R, Schwartz RA, Jarmuda S, Czarnecka–Operacz M, Silny W. Acne fulminans: explosive systemic form of acne. J Eur Acad Dermatol Venereol. 2011;25:501-7.
2. Pereira MF, Roncada EM, Oliveira CM, Monteiro R, Abreu MA, Ortigosa LC. Acne fulminans and isotretinoin - Case report. An Bras Dermatol. 2011;86:983-5.
3. Proença NG. Acne fulminans. An Bras Dermatol. 2017;92 (5 Suppl 1):8-10.

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Clinical Image

Acrokeratosis verruciformis of Hopf

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A 30-year-old female presented with multiple nodules, grouped, hyperkeratotic, brownish and malodorous on his feet (Fig. 1), which had been present for more than 3 years. Histopathological examination showed classical feature of "church spires" appearance without dyskeratosis (Fig. 2). Acrokeratosis Verruciformis of Hopf is a rare autosomal dominant genodermatosis. It usually develops during early childhood affecting both sexes equally. Typically, the lesions are warty to convex, brownish to skin-colored papules on the dorsa of the hands and feet, forearms and legs. Histopathologically, the lesion shows considerable hyperkeratosis, acanthosis, and papillomatosis, mimicking a "church spire", and a thickened granular layer. It arises in early life, often at birth or infancy.

The only effective treatment of AKV is superficial ablation.

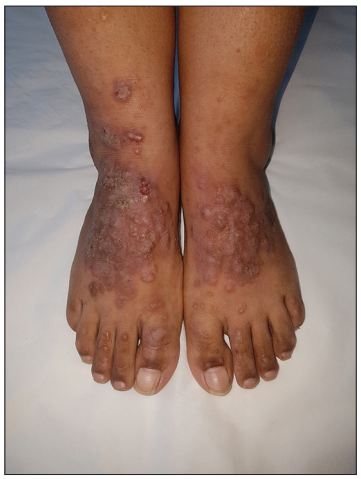

Fig 1: Multiple nodules brownish verrucous on the dorsum of the feet.

CONSENT

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

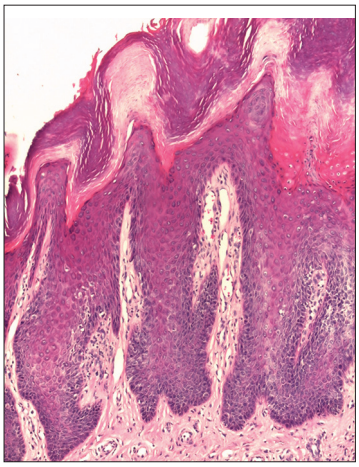

Fig 2: Hyperkeratosis and hypergranulosis with a "church spire" (H&E, ×100).

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

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Letter to the Editor

Raynaud´s phenomenon

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Sir,

Female patient 35 years old in treatment due to systemic sclerosis since1 year ago treated with azathioprine 50 mg/day, felodipine 5 mg/day who came to the emergency room due to her fingertips discoloration and pain. During her examination her fingertips was affected predominantly on her right hand with cyanosis (Figs. 1-3), digital scars on fingertips (Fig. 4).

The diagnosis of secondary Raynaud´s phenomenon associated with her systemic disease. The rest of the examination showed acrosclerosys.

She began 4 days ago with pain and fingertips discoloration associated to the weather cold in the winter, the clinical diagnosis of secondary Raynaud´s phenomenon associated to her systemic disease.

The secondary Raynaud´s phenomenon can be the first clinical manifestation of collagen diseases or present themselves during them. From 2015 to 2017, were studied in the Dermatology and Rheumatology departments a total of 90 patients of these 26 (28.8%) had Raynaud's phenomenon in systemic sclerosis 12 (46.15%), dermatomyositis 6 (23.07%), SLE 5 (19.23%) were studied in the Dermatology and Rheumatology departments.), mixed connective tissue disease2 (7.69%), rheumatoid arthritis 1 (3.84%). In our cases the most frequent association of the secondary Raynaud´s phenomenon was with systemic sclerosis.

On 1862 the French physician Maurice Raynaud described in his thesis an entity characterized by local asphyxia and symmetric gangrene that affected extremities, caused by an overactive neurological reflex [1-3]. Since then, the term Raynaud´s phenomenon (RF) has been used to name these vasospastic episodes, which manifest themselves with cyanosis or pallor in fingers and toes [1,2,4,5].

The Raynaud phenomenon manifests as transient and reversible episodes of peripheral ischemia, which affect fingers and toes in exaggerated vasoconstriction of peripheral arterioles and arteries. Triggers such as cold (the most common), emotional stress and medications, such as beta-blockers, ergot derivates, chemotherapeutic drugs, cyclosporine, bromocriptine, interferon alfa, beta amphetamines, cocaine, nicotine exposure, among other triggers like smoking, vibration or spontaneously [3-7]. It has a general distribution and affects approximately 3 to 5% of the global population with a shift in prevalence toward colder climates [2,3].

It is thought this phenomenon is an abnormal and exaggerated endothelial vascular response to a stimulus (neurogenic, neural, hormonal, vasodilators and vasoconstrictors), generated by an endothelial damage within the microvasculature [4]. The endothelial cells release diverse substances, including endothelin 1 (ET-1), which has a vasoconstrictor effect, regulated by the nitric oxide vasodilator action product of cyclic guanosine monophosphate (cGMP) increments. Within the Raynaud´s phenomenon patient, the cGMP is diminished whenever imbalance between the vasoconstriction and vasodilation exists [4].

When this condition occurs in disease absence, it is known as primary/idiopathic Raynaud´s phenomenon (PRP) or Raynaud disease (80% of cases) [5]. Whereas patients with secondary Raynaud phenomenon (SRP) may occur in association with autoimmune conditions (up to 80 to 95% of the cases) as rheumatoid arthritis (20% of the

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DOI:10.7241/ourd.20191.28

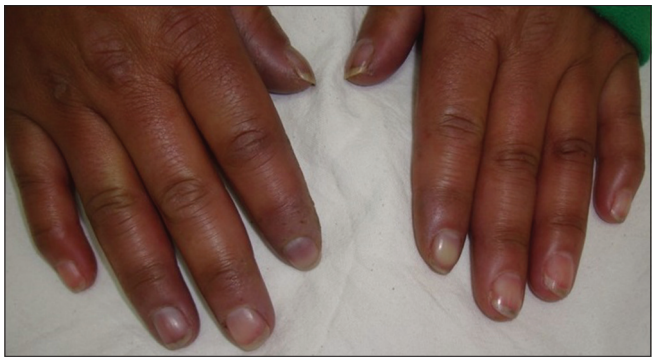

Figure 1: Dorsal face of the hands with cyanosis.

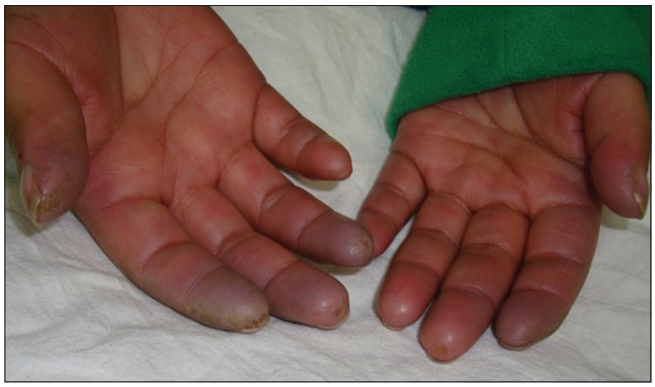

Figure 2: Palmar face of the hands with cyanosis and scars on fingertips.

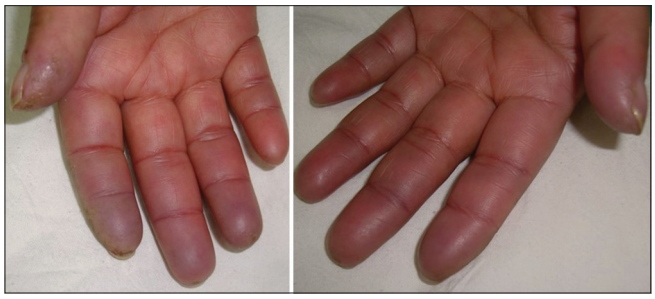

Figure 3: Close up the Raynaud´s phenomenon

cases) scleroderma (90%), systemic erythematous lupus (10-45%), Sjögren syndrome (30%) [2], mixed connective tissue disease, dermatomyositis, arteries diseases [3,4,6,7]. It has been reported in association with neoplasms, including solid tumors and hematologic diseases such as polycythemia vera, essential thrombocytosis leukemias, lymphomas, myeloid metaplasia, Multiple myeloma and cryoglobulinemias, due to the blood viscosity increase [5,7]. Other causes had been described such as endocrinopathies (hypothyroidism, carcinoid syndrome, pheochromocytoma) and arterial disease (Arteriosclerosis, Peripheral embolism, Horton´s vasculitis, thromboangiitis obliterans, Prinz-metal's angina) [7] (Table 1).

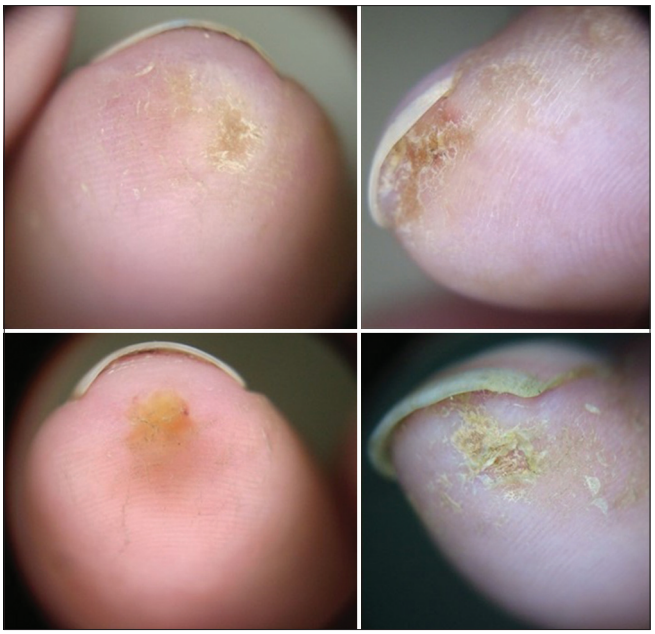

Figure 4: Digital scars on fingertips.

Major risk factors of PRP include female gender, family history of PRP, migraine, smoking and estrogen replacement therapy. The set middle age is 14 years old and only 27% of the cases begins after 40 years old [5]. Clinically, it is characterized by the coloration change of skin on the hands, feet, nose, nipples or ears. It has a "triphasic color pattern": Initially pallor, due the constricted blood-flow and ischemia; Then becoming cyanosis, a sign of tissue hypoxia due the presence of deoxyhemoglobin; and lastly turning red, secondary to the reflex vasodilation (reactive hyperemia) [2,4-6]. This phenomenon most be differentiated from acrocyanosis. This last one characterized by hand and feet continuous cyanosis, exacerbated by low temperatures [5].

The risk of autoimmune disease in patients with Raynaud disease is approximately 6% to 12%. After two years follow-up, if the patient does not present clinical or laboratories signs suggestive of systemic disease the risk dramatically decrease [5]. Complications of primary Raynaud phenomenon are extremely rare. In patients older than 30 years old presenting intensive cyanotic episodes, painful, asymmetric or associated with ischemic cutaneous lesions, in presence with clinical evidence of connective tissue disease, it is suggestive of secondary Raynaud phenomenon [5]. The SRP tends to be asymmetric the attacks are prolonged and Up to 50% of patients with scleroderma may have multiple, bilateral, painful and disabling digital ulcers that heal slowly and tend to relapse [6,7].

Table 1: Raynaud×s disease and secondary phenomenon

	Raynaud disease	Secondary Raynaud×s phenomenon
Prevalence (%)	3.5 – 4	0,4
Women: men	20:1	4:1
Set age (years)	< 15-20	> 25-30
Family history (%)	25	NO
Systemic disease association	NO	Yes
Ulcers/necrosis	Rare	Frequent
Symmetric	Yes	NO
Capillaroscopy	Normal	Capillary dilatations, zones without capillaries, hemorrhages zones
Autoantibodies	Negative or low titles	Frequent
		Causes
		Autoimmune diseases
		Systemic scleroderma, mixed connective tissue disease, systemic erythematous lupus (10-45%), Sjogren syndrome (30%) (8), dermatomyositis, rheumatoid arthritis
		Drugs and toxics
		Ergot derivates, betablockers, chemotherapeutic drugs, cyclosporine, bromocriptine, interferon alfa, beta amphetamines, cocaine, nicotine exposure
		Endocrinopathies
		Hypothyroidism, carcinoid syndrome, Pheochromocytoma
		Arterial Diseases
		Arteriosclerosis, Peripheral embolism, Horton × s vasculitis, thromboangeitis obliterans, Prinz-metal's angina
		Hematologic diseases neoplams
		Polycythemia vera, essential thrombocytosis, leukemias, lymphomas, myeloid metaplasia, multiple myeloma, cryoglobulinemias, solid neoplasms

The diagnosis is made in base different criteria: cold sensibility history, presence of pallor on finger or cyanosis episodes after cold exposition. This may be further corroborated by pictures of hands during the attack. Note that It is not necessary to confirm the diagnosis by provocative tests [2,5].

The evaluation of RF patients must include clinic history, complete physical examination and complementary studies including: hemogram with erythrocyte sedimentation, general biochemistry, thyroid hormones, antinuclear antibodies and specific antibodies, rheumatoid factor and capillaroscopy [2,4,5]. The latter one having a negative predict value of 93% under normal conditions. Additionally, patients with Raynaud disease associated with abnormal capillary patterns increase risk of transition to an autoimmune condition of 47% [4].

The objective of Raynaud phenomenon treatment is reducing vasoconstriction and alleviation of patient symptoms. The treatment is classified in general (no pharmacological) measures and pharmacological measures [4]. General measures such as cold protection, tobacco and vasoconstrictor pharmacy avoidance, and prevention of vibration/stress, are usually enough to control Raynaud disease [4,6]. Only in secondary cases and/or in association with digital ulcers pharmacologic treatment is necessary [4,5-7].

Calcium antagonists (nifedipine, amlodipine, nicardipine) are first-line drugs whenever general measures fail. This treatment presents slight effects on primary Raynaud phenomenon, but moderate benefits regarding secondary forms of scleroderma, decreasing the frequency and intensity of the episodes [2,5,6].

When calcium channel blockers are not effective, therapy with phosphodiesterase 5 inhibitors such as, sildenafil and vardenafils may be substituted; if there is severe vascular involvement (digital ulcers or critical ischemia), infusions of prostaglandin analogs may be used [5,6]. The interdigital and palmar injections of botulinum toxin A improve clinical, especially alleviating pain in patients with Raynaud's phenomenon and digital ulcers [6].

Surgical treatment is reserved for patients with poor response to pharmacological treatment with severe ischemia or active digital ulcers, and are reduced to debridement of necrotic or infected tissue, open thoracoscopic and digital surgical sympathectomy,

electrical stimulation of bone marrow and vascular reconstruction [2].

CONSENT

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

REFERENCES

1. Salazar-López R. Historia del tratamiento de la vasculitis. Rev Colomb Cir Plást Reconstr. 2017;23:75-8.
2. Joven B, Carreira P. Síndrome de Raynaud: etiología y manejo. Reumatol Clin. 2008;4:59-66.
3. Maverakis E, Patel F, Kronenberg D, Chung L, Fiorentino D, Allanore Y, et al. International consensus criteria for the diagnosis of Raynaud´s phenomenon. J Autoinmmun. 2014;48-49:60-5.
4. Guerrero-García A. Fenómeno de Raynaud. Biociencias. 2017;12:93-9.
5. Wigley F. Raynaud´s Phenomenon. N Eng J Med. 2002;347:1001-4.
6. Cervigón-González I, Sánchez-Neila N, Montes-García S, Palomo Arellano A. Algoritmo terapéutico del fenómeno de Raynaud y de las ulceraciones digitales isquémicas de la esclerodermia sistémica. PIEL (Barc). 2016;32:1.
7. Conde-Baena P, Vargas-Hitos J, Sabio J, Navarrete-Navarrete N, Zamora-Pasadas M, Jiménez-Alonzo J. Varón de 62 años con fenómeno de Raynaud. Actual Med. 2015;100:98-100.

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Our Dermatology Online

Letter to the Editor

Onychomadesis due to hand-foot-mouth disease

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Sir,

Case I

Male patient, 2 years of age, who is taken to the clinic for disseminated dermatosis to the peribucal region, lips, back of hands and palms, constituted by vesicles and blood crusts (Figs. 1a-d), and is diagnosed with mouth-hand-feet disease 5 weeks after re-consultation due to onycopathy disseminated to both hands fingernails, and the right foot, manifested by a separation of the nail plate from the proximal fold (Figs. 2a and b). The rest of the physical examination was within normal limits and there is no significant family or personal background.

Case II

Female patient, 7 years old, coming to the clinic because of a four-week-old localized onycopathy in fingernails due to "fungi", presenting alterations in fingernails at index and ring fingers of the right hand and index of the left, manifested by separation of the nail plate from the proximal fold (Figs. 3a – 3e). The rest of the physical examination was within normal limits.

The patient's mentions that prior to this picture the girl presented disease in mouth, hands, and feet soles, with vesicles. There is no significant family or personal background.

Diagnosis is made of onychomadesis secondary to mouth-hand-foot disease.

Onychomadesis is the proximal detachment of the nail plate [1]. It is the result of temporary matrix growth interruption [1-3]. It is associated with autoimmune, systemic diseases; trauma, use of medications, hereditary factors and infections [1-4] (Table 1).

Among the infectious causes, the one that is most frequently associated with onychomadesis is the mouth-hand-foot disease [1,3,5]. It is an infection caused by viruses of the Enterovirus family [5,6]. It usually affects children under 10 years of age, although cases have been reported in adults [5]. In some regions, seasonal outbreaks are reported in spring and autumn [6]. This family of viruses consists of 10 species. The serotypes of human enteroviruses are divided into four species: A-D [7]. Coxsackie virus A16 and Enterovirus 71 are the two serotypes most frequently associated with the disease [1,5,7]. It characterizes by fever, malaise, odynophagia and vesicles that affect palms, plants, buttocks and oral mucosa [7], as well as oral ulceration of the mucous and gingival membranes, the soft palate and tongue [1,5]. Blisters usually appear on the back and side edges of the limbs, on an erythematous basis [1]. The initial picture resolves between 3 and 10 days, with an average of 6 days [1,6]. Subsequent to the initial picture, onychomadesis has been reported.

The first report of onychomadesis secondary to mouth-to-foot disease was in 2000, in five children from Chicago, Illinois. In 2004, four cases were reported in Europe, and since 2008, isolated cases and outbreaks have been reported in different cities of Spain [7]. Cases have also been reported in France, Belgium, Italy, Finland, the United States, Japan and Taiwan, all in children under 7 years of age [1]. Onychomadesis occurs in a range between 4 and 10 weeks after the resolution of the initial symptoms [1,5-8]. And, although it has been proposed that infections by multiple enterovirus serotypes or specific serotypes (A6, A10, B1) are responsible for this manifestation, no study has been

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Table 1: Causes of onychomadesis

Type	Diseases
Autoimmune	Pemphigus vulgaris, alopecia areata, nail psoriasis, pustular psoriasis, pemphigoid gestationis, linear IgA disease, telogen effluvium, bullous pemphigoid, Guillain-Barré syndrome, epidermolysis bullosa acquisita, among others.
Systemic	Stevens-Johnson syndrome, major depressive disorder, immunodeficiency, diabetes mellitus, severe sepsis, myocarditis, pancreatitis, myocardial infarction, erythema nodosum, myelomatosis, Raynaud disease, erythroderma, contact dermatitis, epilepsy, hypopituitarism, rheumatic fever, chronic kidney disease, Kawasaki disease, mycosis fungoides, peritoneal dialysis, zinc deficiency, gastrointestinal bleeding, postcardiac arrest, hyperparathyroidism, reflex sympathetic dystrophy, carpal tunnel syndrome, hemodialysis, severe dysmenorrhea, chronic paronychia, and hypocalcemia.
Trauma	Fractured olecranon or wrist, manicure, high-altitude expeditions, deep saturation dives, and road running.
Medication	Chemotherapeutic agents, antiepileptics, radiation therapy, some antibiotics, retinoids, lithium, octreotide, lead, corticosteroids, azathioprine, itraconazole, and arsenic.
Hereditary	Heimler syndrome, hyper-IgM syndrome, and idiopathic sporadic onychomadesis.
Neonatal	Stresses in utero
Infections	Diphtheria, measles, mumps, varicella virus, syphilis, typhoid fever, scarlet fever, malaria, *Candida spp, Trichophyton tonsurans,* and hand-foot-mouth disease.

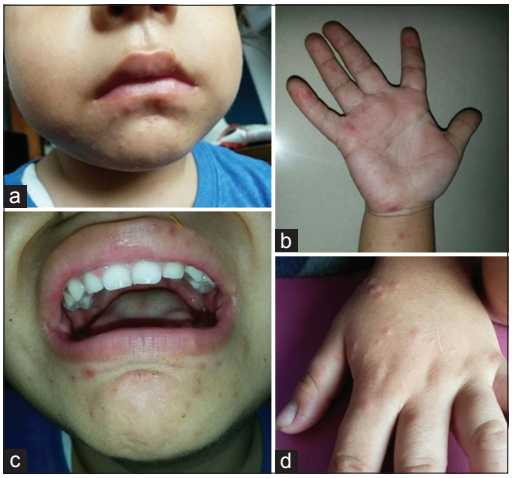

Figure 1: Hand-foot-mouth disease cutaneous lesions

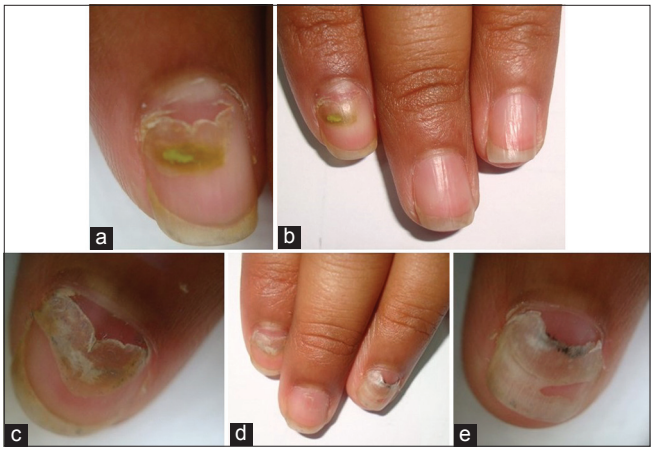

Figure 3a-e: Onychomadesis of fingernails 4 weeks after hand-foot-mouth disease

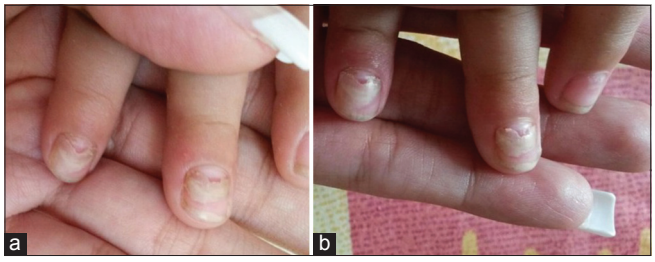

Figure 2: Separation of the nail plate from the proximal nail folds on fingernails 5 weeks after hand-foot-mouth disease

able to confirm this relationship with statistically significant evidence [1,7]. What has been shown is that serotype A6 has been reported more frequently in onychomadesis outbreaks secondary to this disease. The reason, it is believed, is that this serotype causes a more severe and widespread disease [1,6].

The mechanism by which onychomadesis arises as a sequel to mouth-hands-feet disease is also not known with certainty, but several hypotheses have been raised. One of the most accepted theories is that of Bettoli *et al*, which argues that matrix growth interruption is due to viral molecules or immune complexes deposits that cause embolism [9,10]. Cabrerizo and colleagues propose that it is secondary to direct damage by replication of the virus in the matrix [5,10]. This theory is based on two studies that identified the Coxsackievirus A6 in the scales of the lamina detached by PCR with reverse transcription [5,6,10]. It is also proposed that it is due to periungual inflammation or damage secondary to the appearance of vesicles near the nails (6). Other less popularized theories mention that it may be secondary to febrile illness, but this is discredited by some who note that not all patients report fever and that temperature rise is not high or persistent [6].

It being secondary to the use of drugs during the initial symptoms of the disease is not accepted either, has because no relationship has been reported between commonly used drugs, especially analgesics, and onychomadesis [6,10].

CONSENT

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

REFERENCES

1. Hardin J. Haber R. Onychomadesis: literature review. BJD. 2015;172:592-6.
2. Tosti A. Piraccini B. Biology of Nails and Nails Disorders. In: Goldsmith L. Katz S. Gilchrest B. Paller A. Leffell D. Wolff K. Fitzpatrick's Dermatology in General Medicine. 8th edition. United States: Mc Graw Hill; 2008: Volume 1. 1009-1014.
3. Braswell M. Daniel R. Brodell R. Beau lines, onychomadesis, and retronychia: A unifying hypothesis. J Am Acad Dermatol. 2015;73:849-55.
4. Chang P. Escalante K. Onicomadesis. Twelve-case report. Dermatology CMQ. 2013;11:89-93.
5. Mortada I. Mortada R. Al Bazzal M. Onychomadesis in a 9-month-old boy with hand-foot-mouth disease. Int J Emerg Med. 2017;10:1-2.
6. Shin J. Cho B. Park H. A Clinical Study of Nail Changes Occurring Secondary to Hand-Foot-Mouth Disease: Onychomadesis and Beau's Lines. Ann Dermatol. 2013;26:280-3.
7. Bracho M. González-Candelas F. Valero A. Córdoba J. Salazar A. Enterovirus Co-infections and Onychomadesis after Hand, Foot, and Mouth Disease, Spain, 2008. Emerg Infect Dis. 2011;17:2223-31.
8. Gan X. Zhang T. Onychomadesis after hand-foot-and-mouth disease. CMAJ. 2017;189:E279.
9. Bettoli V. Zauli S. Toni G. Virgili A. Onychomadesis following hand, foot, and mouth disease: a case report from Italy and review of the literature. Int J Dermatol. 2013;52:728-30.
10. Jeelani S. Lanker A. Jeelani N. Onychomadesis Following the Outbreak of Hand Foot Mouth Disease in Children: A Study from North India. IJTDH. 2017;27:1-6.

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Letter to the Editor

Köbner phenomenon in systemic lupus erythematosus

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Sir,

The Köbner phenomenon is the newly development of isomorphic lesions in the mechanically stimulated or injured skin [1,2]. This phenomenon can be seen in various disorders, such as psoriasis, lichen planus, and vitiligo; however, cases of systemic lupus erythematosus (SLE) showing Köbner phenomenon have been rarely reported [3,4].

A 40-year-old woman, suffering from SLE and lupus nephritis for over 5 years, was followed at Tsuchiura Kyodo General Hospital. A biopsy specimen taken from the malar rash showed slight liquefaction of the basal layer of the epidermis, and mild mononuclear cell infiltration in the papillary dermis. Examination by direct immunofluorescence showed linear deposition of IgM, IgG and C3 in the epidermal basement membrane. She was treated with oral prednisolone, and leukopenia and serum hypocomplementemia were improved; however, erythematous lesions on the hands were resistant to therapies. Physical examination revealed infiltrative erythemas on the fingers and dorsa of hands, predominantly involving the previously operated sites (Fig. 1). The second finger showed dactylitis (diffuse swelling). Serum antinuclear antibodies (ANA) (1:320, homogenous), anti-double stranded DNA antibodies (25.3 IU/ml; normal <10), and anti-Sm antibodies (33.1 index; normal <6.9) were detected, whereas anti-SS-A, SS-B, Jo-1, and cardiolipin antibodies were within normal ranges. Because she refused a skin biopsy again, we did not carry out a biopsy from the digital erythema.

The pathogenesis of the Köbner phenomenon is still not fully elucidated as yet. Ueki [2] proposed a second-step theory, a first non-specific inflammatory step and a second disease-specific step, in Köbner phenomenon. In the first step, many environmentally induced factors such as cytokines, stress proteins, adhesion molecules, or autoantigens translocated from intracellular areas are involved in the inflammatory phase. Subsequently, in the second step, there may be disease-specific reactions mediated by T-cells, B-cells, autoantibodies and immune complex deposition under the susceptible backgrounds. Recent studies suggest that upon epidermal injury, alarmins are released from keratinocytes, which subsequently induces activation of the innate immune systems leading to activation of acquired immunity *via* toll-like receptors [5].

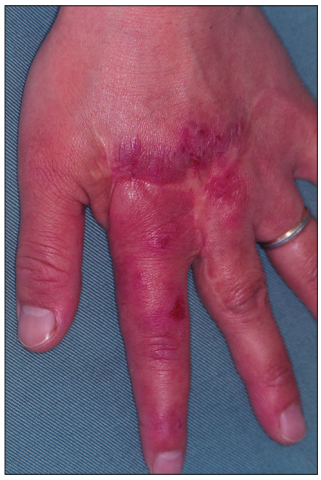

Figure 1: Infiltrative erythema predominantly observed on the operated scar. Dactylitis is also seen in the second finger.

Isomorphic response of Köbner is rarely reported in association with SLE [3,4]. One case is the development of discoid lupus erythematosus (DLE) on the recent herpes zoster scar in a patient with SLE [3], and another case is disseminated linear DLE lesions at the site of trauma [4]. Köbner phenomenon is occasionally observed in DLE [5,6]; however in the present case, cutaneous lupus erythematosus was

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observed involving the previously operated sites in a patient with SLE.

Another interesting feature in the present case is the development of dactylitis. Dactylitis is sometimes seen in association with various diseases such as psoriatic arthritis, tuberculosis, injury, gout, and sarcoidosis. Although the pathomechanism of dactylitis is still unclear, dactylitis is speculated to be caused by minor biomechanics, which may be due to deep Köbner phenomenon. Thus, previous physical stress may have induced both cutaneous lupus lesion and dactylitis in this case.

CONSENT

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

REFERENCES

1. Boyd A, Neldner K. The isomorphic response of Koebner. Int J Dermatol. 1990;29:401-10.
2. Ueki H. Koebner phenomenon in lupus erythematosus with special consideration of clinical findings. Autoimmun Rev. 2005;4:219-23.
3. Longhi BS, Centeville M, Marini R, Appenzeller S. Koebner's phenomenon in systemic lupus erythematosus. Rheumatol Int. 2011;32:1403-5.
4. Yadav S, Kumar R, Sharma A, Saikia UN, Dogra S. Isomorphic phenomenon in discoid lupus erythematosus with review of reported cases. Rheumatol Int. 2013;33:1651-2.
5. Jolly M. Discoid lupus erythematosus after tattoo: Koebner phenomenon. Arthritis Rheum. 2005;53:627.
6. Berger E, Robinson M, Patel R, Franks AG Jr. Koebner phenomenon to heat in cutaneous lupus erythematosus (lupus ab-igne). Dermatol Online J. 2012;18:17.

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Letter to the Editor

'Red starburst' pattern: A new dermoscopic indicator in discoid lupus erythematosus

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Sir,

We would like to draw attention to a peculiar dermoscopic pattern in discoid lupus erythematosus (DLE) that we have noticed- we shall refer to it as the 'red starburst' pattern.

A 32 year old male presented with an asymptomatic red lesion on the right side of his neck, of 6 months duration. He had received no prior treatment for it. Clinical examination revealed an erythematous plaque with a central adherent crust, on the right side of the neck (Fig. 1). The clinical differential diagnoses were seborrheic dermatitis, psoriasis, DLE, and actinic keratosis.

Dermoscopic examination of the lesion showed thick white scales in the centre, with blurred/out of focus radial streaks of red, along the periphery ('red starburst' pattern), with a surrounding whitish halo (Fig. 2). Histopathological examination confirmed the diagnosis of DLE (Fig. 3).

The second patient was a female aged 21 years, who presented with small erythematous scaly plaques on photo-exposed areas, of 5 months duration (Fig. 4). Dermoscopy revealed central scaling, red radial streaks, and a peripheral whitish halo. Biopsy confirmed a diagnosis of DLE.

DLE is the most common subtype of cutaneous lupus erythematosus. It occurs most frequently on the scalp, but can also involve the face, trunk and limbs [1]. The morphology of DLE varies at different stages of disease progression[2].

Characteristic variables observed in dermoscopy of DLE [2] are listed in table 1.

The most common dermoscopic diagnostic criteria of these are perifollicular white halo, follicular keratotic plugs and telangiectasia [2].

The 'starburst' pattern was initially used to describe the dermoscopic hallmark feature of Spitz/Reed nevus. It is characterized by the presence of pigmented sharply focused streaks radially distributed at the periphery of a lesion, which correspond to confluent junctional melanocytic nests and histological radial growth [3].

A 'white starburst' pattern [4,5] has been described in dermoscopy of prurigo nodularis as radially arranged whitish lines on a reddish-brown background. Histopathologically, the radially arranged whitish lines correspond to papillary dermal fibrosis (the thickened collagen fibres in the papillary dermis are arranged perpendicular to the skin surface).

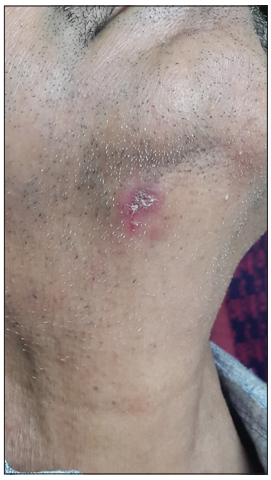

Figure 1: Erythematous plaque with central adherent scales on the neck.

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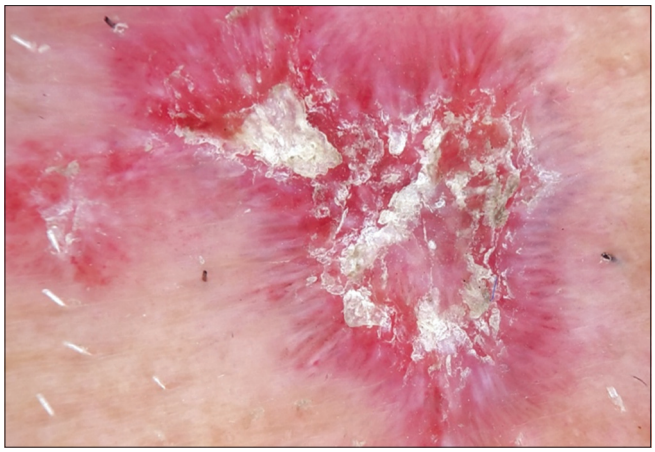

FIgure 2: Dermoscopy reveals a 'red starburst' pattern. The combination of telangiectasia, epidermal hyperplasia and underlying dermal fibrosis may result in this red starburst pattern. Thick white scales and follicular plugging are also seen.

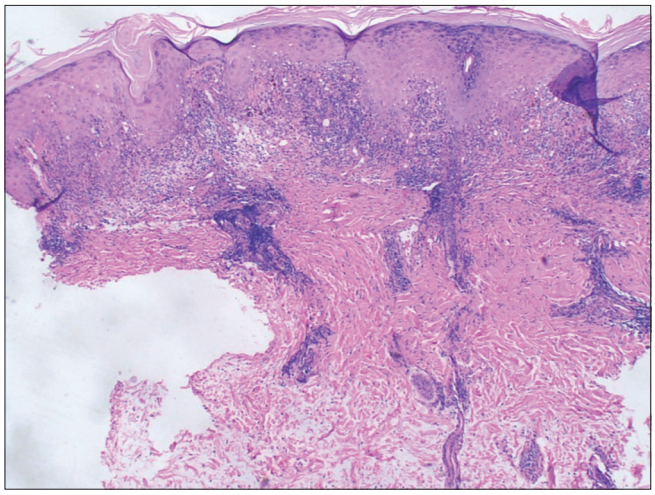

Figure 3: Histopathology- The dermoepidermal junction is smudged and shows occasional necrotic keratinocytes. The papillary dermis is edematous and shows scattered colloid bodies. Moderately dense superficial and mid perivascular infiltrate of lymphocytes, with a lichenoid appearance in the papillary dermis. Thickened collagen bundles are present in the reticular dermis, and scant mucin is seen.

In the cases of DLE described above, dermoscopy revealed radially arranged red lines on an erythematous background that appear as a 'red starburst' pattern. Few pigment dots and globules may also be interwoven with the radial red lines.

Histopathology of long standing cases of DLE shows telangiectasia, pigmentary changes and diffuse dermal fibrosis [6]. The presence of these features are possibly reflected as a red starburst pattern dermoscopically.

Thus, we have observed that the red starburst pattern is found in long standing cases of DLE, and could be one of the specific indicators for late stage of DLE.

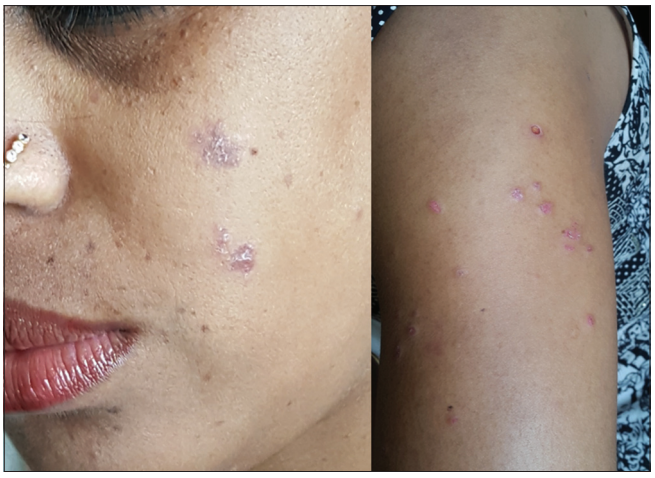

Figure 4: Dermoscopy of one of the lesions on the arm reveals a red starburst pattern, linear and dotted vessels, and white scales.

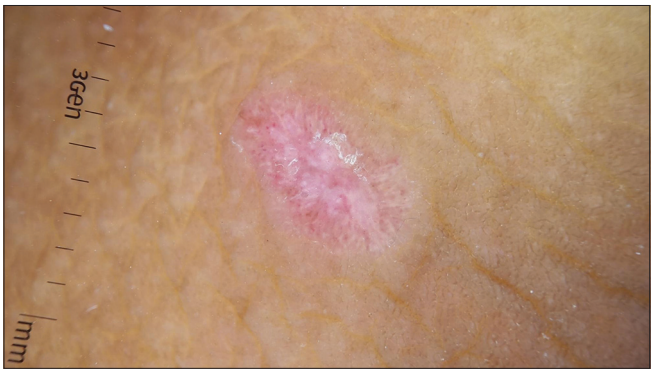

Figure 5: Dermoscopy of one of the lesions on the arm reveals a red starburst pattern, linear and dotted vessels, and white scales.

Table 1: Variables observed in dermoscopy of discoid lupus erythematosus

Early changes	Late changes
Perifollicular white halo	Telangiectasia
Follicular plugs	Structureless white areas
White scales	Honeycomb pigmentation

CONSENT

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

REFERENCES

1. Tsung-Ming T, Kuo-Chia Y. Dermoscopic features of discoid lupus erythematosus. Dermatol Sinica. 2012;30:78-80.
2. Lallas A, Apalla Z, Lefaki I, Sotiriou E, Lazaridou E, Ioannides D, et al. Dermoscopy of discoid lupus erythematosus. Br J Dermatol. 2013;168:284-8.
3. Maione V, Errichetti E, Roussel SL, Lebbé C. Pigmented Bowen's disease presenting with a "starburst" pattern. Dermatol Pract Concept. 2016;6:47-9.
4. Errichetti E, Piccirillo A, Stinco G. Dermoscopy of prurigo nodularis. J Dermatol. 2015;42:632-4.
5. Ankad B S, Beergouder S L. Hypertrophic lichen planus versus

prurigo nodularis: a dermoscopic perspective. Dermatol Pract Concept. 2016;6:9–15.
6. McKee P, Calonje JE, Granter S. Idiopathic connective tissue disorders. Pathology of the Skin with Clinical Correlations (Calonje JE, Brenn T, Lazar A, McKee P), 3rd ed. Amsterdam: Elsevier Mosby, 2005;775–803.

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Letter to the Editor

Palisade pigmentation - describing a new dermoscopic finding

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Sir,

Discoid lupus erythematosus (DLE) represents the most common subtype of cutaneous lupus erythematosus. There is a morphological diversity seen in DLE, at different stages of progression of the disease. Based on these features, dermoscopic criteria have been formed for both early and late stage DLE, and their histopathological correlations studied [1]. We would like to describe a new dermoscopic feature that we have observed in late stage DLE- palisade pigmentation.

Case 1: A 26year old male presented with a lesion over the right pre-auricular area, of nine months duration. Examination revealed an irregularly shaped, depigmented atrophied plaque with a central erythematous area and overlying adherent scales, hyperpigmented border, and absence of hair follicles (Fig. 1). Dermoscopy revealed white shiny structures, telangiectasia, white scales, and palisade pigmentation (Fig. 2).

A biopsy from the lesion showed features confirming the diagnosis of DLE (Fig. 3).

Case 2: A 35year old man noticed multiple lesions on his right cheek, over the past six months. They were asymptomatic. Cutaneous examination revealed multiple irregular violaceous atrophic plaques on the right cheek, with an erythematous border. Dermoscopy revealed white shiny structures, thick white overlying scales, telangiectasia, and palisade pigmentation (Fig. 4). Biopsy confirmed the diagnosis of DLE.

A 'palisade' is a fence of wooden stakes or iron railings fixed in the ground, forming an enclosure or defence. In dermatopathology, the term has been used to describe the typical arrangement of basaloid cells in basal cell carcinoma, and in granuloma annulare, for histiocytes surrounding an area of altered collagen [2].

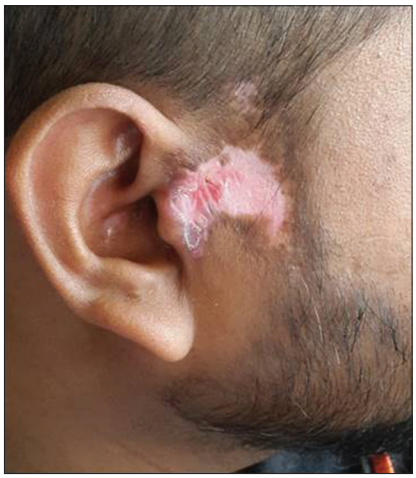

Figure 1: Depigmented atrophied plaque with scaling, over the right pre-auricular area.

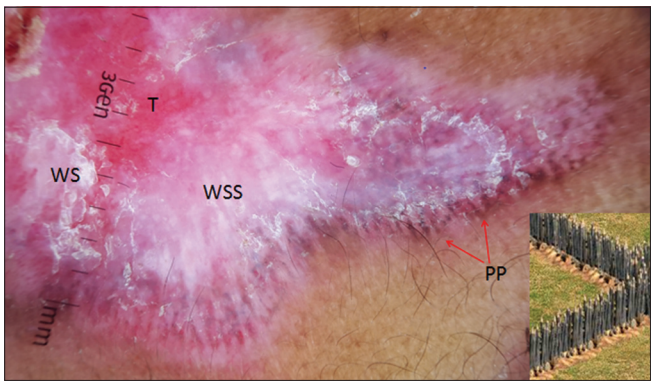

Figure 2: Palisade pigmentation (PP) seen on dermoscopy of the lesion. Note other features of the disease such as white shiny structures (WSS), telangiectasis (T) and white scales (WS), which are findings in late stage DLE. Inset shows a picture of palisade arrangement.

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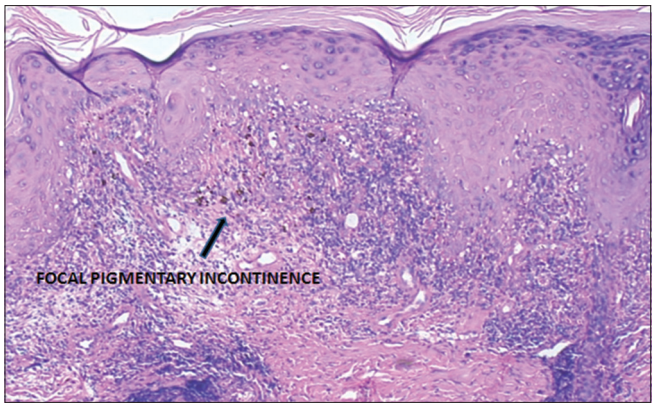

Figure 3: Focal pigmentary incontinence on histopathology corresponds to palisade pigmentation on dermoscopy.

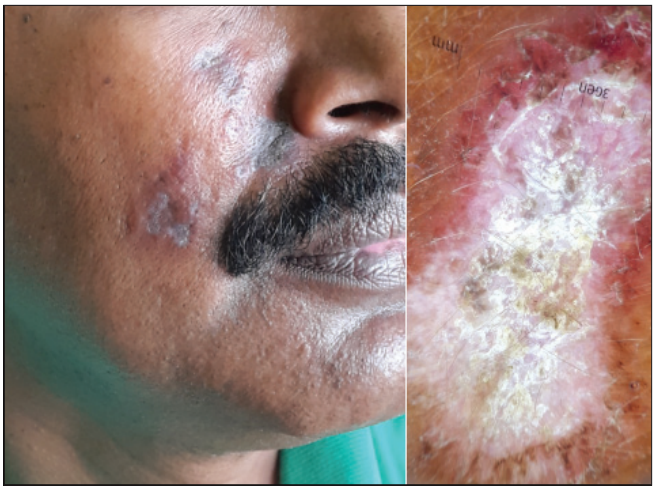

Figure 4: Erythematous plaques with a violaceous tinge, on the right cheek. Dermoscopy showing white shiny structures, thick white scales and palisade pigmentation.

'Palisade pigmentation' observed on dermoscopy here, is formed by the arrangement of pigment structures at the periphery of the lesion in late stage DLE. It appears as linear hyperpigmented streaks. They may be present focally or segmentally, and may extend for a distance, outwards, from the periphery of the lesion.

Histopathologically, it corresponds to pigmentary incontinence, which was confirmed in our case. The incontinence may not involve the entire dermis, but is present focally or segmentally, and is due to focal interface vacuolar degeneration of the basal layer.

Hyperpigmentation has been described in dermoscopy of late stage DLE [1,3] in the form of honeycomb network, perifollicular pigmentation, radial pigment streaks or pigmentation arranged in unspecified patterns. In a study by Lallas et al [1], hyperpigmentation was observed on dermoscopy in 43.6% cases of DLE, and pigmentary incontinence was seen in 49.1% of the corresponding biopsies.

We would like to suggest this characteristic finding of 'palisade pigmentation' as a dermoscopic indicator of late stage DLE. Recognition of this pattern, in conjunction with other already established dermoscopic features, can help clinch the diagnosis.

CONSENT

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

REFERENCES

1. Lallas A, Apalla Z, Lefaki I, Sotiriou E, Lazaridou E, Ioannides D, et al. Dermoscopy of discoid lupus erythematosus. Br J Dermatol. 2013;168:284-8.
2. Madke B, Chougule BD, Kar S, Khopkar U. Appearances in clinical dermatology. Indian J Dermatol Venereol Leprol. 2014;80:432-47.
3. Errichetti E, Stinco G. Dermoscopy in General Dermatology: A Practical Overview. Dermatol Ther. 2016;6:471-507.

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Letter to the Editor

Hailey-hailey disease presenting at an unusual site

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Sir,

A 37-year-old patient presented with painful crusted skin lesions on the scalp for one year. The condition started as small blisters that ruptured spontaneously to form crusted erosions. He had similar lesions in the axillae and groin from two years past, which later developed into warty plaques. There was no family history of the same condition nor was relevant medical history. The patient experienced more exacerbation in the summer months as he worked as a taxi driver and spent most of the day outdoors. Dermatological examination showed four crusted lesions on the scalp, a big one of 4×5 cm near to the vertex, and the remaining three about 1×2 cm each located around it (Fig. 1 a). Moreover, there was a warty plaque measuring 2×3 cm in the right axilla (Fig. 1 b). Oral mucosa and nails revealed no abnormality. Swabs for bacteriological and fungal analysis showed negative results. Blood routine investigations, chemistry tests, and CXR all were normal. Histopathological study revealed intraepidermal cleft and bullous formation containing aggregates of acantholytic cells that arranged in a "dilapidated brick wall appearance" (Fig. 2). Direct immunofluorescence test was negative for immune reactants. A correlation between the clinical picture and the histopathology findings confirmed the diagnosis of Hailey-Hailey disease. The patient was prescribed systemic antibiotics flucloxacillin 500mg 8 hourly, oral Acitretin 25mg/day and topical calcipotriol cream. A good response was observed after four weeks of treatment. (Fig. 3). No signs of recurrence were reported within six months of follow-up.

Hailey-Hailey disease (HHD), also known as benign familial pemphigus, is an autosomal dominant condition in which mutations in the ATP2C1 gene result in abnormal intracellular calcium signaling $Ca2^{+}/Mn2^{+}$ -ATPase isoform 1 (hSPCA1) on the Golgi membrane [1]. Normally this gene plays a part in ensuring that the cells in the outer layer of the skin (the epidermis) stick together. In HHD this adhesion of cells is weakened, resulting in acantholysis in areas of skin prone to friction [2,3]. Thereby, heat, sweating, infection and contact irritants can exacerbate skin lesions, for which reasons the disease has a relapsing and remitting course. The rashes are often itchy but painful when scratched [4].The disease usually manifests in adulthood (the 30s and 40s) and is characterized by vesicular lesions, painful erosions, and scaly erythematous plaques that occur at sites of friction such as the armpits and groins. The current case is being reported due to the rare occurrence of the disease on the scalp. Moreover, an extraordinary presentation has rarely been described in the literature. A case of persistent crusted lesion due to HHD was reported on the face of a 25-year-old woman [5]. One more patient

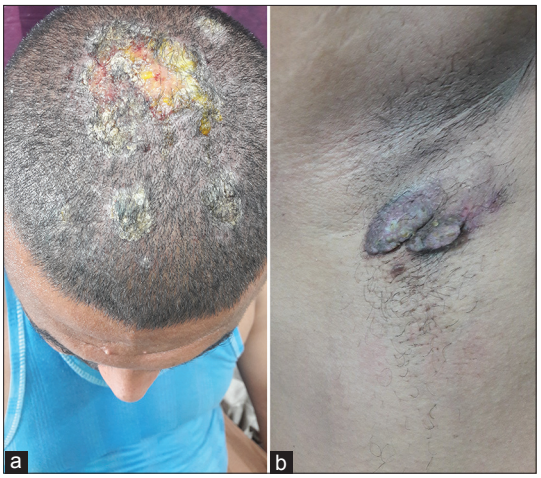

Figure 1: (a and b)A-Four crusted lesions on the scalp, a big one 4×5 cm near to the vertex, and the other three about 1×2 cm surrounding the big one. B- A warty plaque measuring 2×3 cm in the right axilla.

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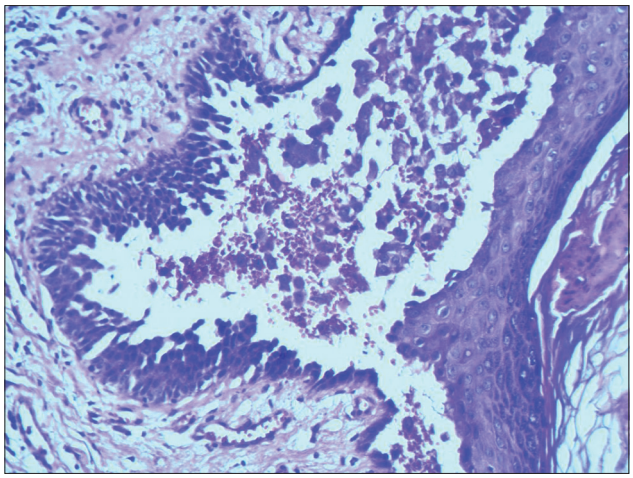

Figure 2: Hematoxylin and eosin (H&E) 40X, stained skin section showed epidermal cleft and bullous formation containing aggregates of acantholytic cells that arranged in dilapidated brick wall appearance, with papillary protrusion of the basal layer.

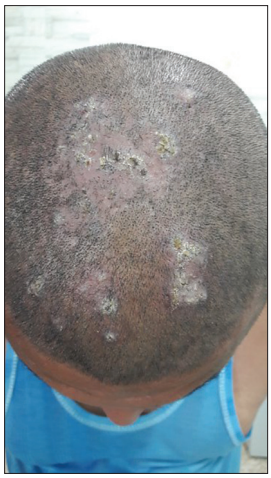

Figure 3: Noticeable response four weeks after treatment

was diagnosed as HHD presented with a solitary bulla on the left forearm that had spontaneously ruptured to form an erosion [6]. The clinical differential diagnosis of Hailey-Hailey disease includes intertrigo, candidiasis, inverse psoriasis, tinea cruris, contact dermatitis, and seborrheic dermatitis. In this case, the painful crusted erosion with the history of exacerbation during summer months, suggests the diagnosis of HHD. Histologic differential diagnosis includes other disorders with intraepidermal acantholysis such as pemphigus vulgaris and Darier's disease. The characteristic dilapidated wall appearance with a papillary protrusion of the basal layer are consistent with HHD (Fig. 2). The absence of dyskeratosis like "corp ronds" and "grains" in the suprabasal layer excluded Darier's disease in this case.

Traditional treatments of HHD include topical and systemic antibiotics, topical and oral corticosteroids, dapsone, retinoid, cyclosporin, and methotrexate for patients with recalcitrant disease. For widespread recalcitrant HHD, laser ablation (with CO2 or Erbium: YAG), radiofrequency, dermabrasion, and botulinum toxin type (BTX -A) have been reported as useful in the literature [7]. We have to consider the possibility of HHD when there are characteristic clinical signs, regardless of the location of the lesion to avoid misdiagnosis. Early diagnosis and treatment can reduce the incidence of complications and improve the patient's quality of life.

REFERENCES

1. Micaroni M, Giacchetti G, Plebani R, Xiao GG, Federici L. ATP2CI gene mutations in Hailey-Hailey disease and possible roles of SPCA1 isoforms in membarane trafficking. Cell Death Dis. 2016;7:e2259.
2. Chourabi M, H' mida-Ben Brahim D, Bonnard C, Aounallah A, Yu Ng A, Tohari S, et al. A novel nonsense ATP2CI mutation causes Hailey-Hailey disease in a Tunisian family. Our Dermatol Online 2018:9:110-3.
3. Hassan I, Keen A. Hailey-Hailey disease: a case report. Our Dermatol Online. 2012;3:116-8.
4. Engin B, Kutlubay Z, Celik U, Serdaroglu S, Tuzun Y. Hailey-Hailey disease: A fold (intertriginous) dermatoses. Clin Dermatol. 2015;33:452-5.
5. Das D, Das A, Gharami R, Bandyopadhyay D. Hailey-Hailey disease: A presentation out of the ordinary. J Turk Acad Dermatol. 2014;8:1484c7.
6. Vasudevan B, Verma R, Badwal S, Neema S, Mitra D, Sethumadhavan T. Hailey -Hailey disease with skin lesions at unusual sites and a good response to acitretin. Indian J Dermatol Venereol Leprol. 2015;81:88-91.
7. Farahnik B, Collin M. Blattner, Micheal B Mortazie, Dirk M. Elston, et al. Interventional Treatment for Hailey –Hailey disease. J Am Acad Dermatol. 2017;76:551-8.

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Our Dermatology Online

Historical Article

Eponyms in dermatology linked to pigmented purpuric dermatoses

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ABSTRACT

Pigmented purpuric dermatoses (PPDs) are chronic benign dermatoses characterized by petechiae, purpura, and increased skin pigmentation. The hallmark of pigmented purpuric dermatoses is their orange-brown, speckled, cayenne pepper-like discoloration. It occurs, most commonly on on the lower extremities and may be asymptomatic or pruritic. The aim in this short communication is to shed some lights on the eponyms in dermatology linked to PPDs.

Key words: Dermatology, Diseases, Eponyms.

Pigmented purpuric dermatoses (PPDs), also known as capillaritis, purpura simplex, and inflammatory purpura without vasculitis, include a spectrum of vascular diseases, usually of unclear etiology [1].

Various conditions have been mentioned under this group. These include:
- Schamberg's disease
- Purpura annularis telangiectodes
- Pigmented purpuric lichenoid dermatitis of Gougerot and Blum
- Lichen aureus
- Eczematid-like purpura of Doucas and Kapetanakis
- Itching purpura
- Unilateral linear capillaritis
- Granulomatous pigmented purpura

PPDs share some common histopathological features such as red blood cells extravasation, hemosiderin deposition (mainly within the dermal macrophages), narrowing of small vessel lumen, endothelial edema and lymphocytic perivascular infiltrate [1].

Stain for hemosiderin (storage iron granules) is very useful to detect iron overload in the tissue.

Perl's iron stain (also known historically as, Perls' Prussian blue), is the classic method for demonstrating iron in tissues. The section is treated with dilute hydrochloric acid to release ferric ions from binding proteins. These ions then react with potassium ferrocyanide to produce an insoluble blue compound (the Prussian blue reaction).

Hemosiderin may be present in areas of old hemorrhage or be deposited in tissues with iron overload. Hemosiderosis refers to the state in which the stored iron does not interfere with organ function.The latter is in comparison to hemochromatosis where iron overload is associated with organ failure.

Perl's iron stain is named after its inventor, German pathologist Max Perls (1843-1881).

PPDs occur predominantly in the lower limbs of adults, but can affect children. Some particular clinical aspects allow the division of PPD into eponymous variants.

In table 1 we listed eponymous conditions in dermatology linked to PPDs.

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Table 1: Eponymous conditions in dermatology linked to pigmented purpuric dermatoses:

Eponymous conditions linked to pigmented purpuric dermatoses	Remarks
Eczematid-like purpura of Doucas and Kapetanakis[2]	It is distinguished from other forms of PPD by the concomitant presence of eczematous features. Some authors have considered eczematid-like purpura of Doucas and Kapetanakis and itching purpura as a single entity. It is named for 2 Greek physicians, Christoforos Doucas (1890-1974) and Ioannis Capetanakis (1913-1987).
Majocchi disease (Purpura annularis telangieciodes) [3]	It presents with nonblanchable, annular, 2 to 20 cm, symmetrical, purpuric, telangiectatic patches. It is named for Domenico Majocchi (1849–1929), [Figure 1], who was an Italian dermatologist. Majocchi, also, characterized Fungal folliculitis (known as Majocchi granuloma).
Pigmented purpuric lichenoid dermatitis of Gougerot-Blum[4]	In this type of PPD, the patient develops polygonal or round lichenoid purpuric papules that coalesce to form red-brown to violaceous plaques. It was characterized in 1925 by 2 French dermatologists; Paul Blum (1878-1933) and Henri Gougerot (1881-1955), [Figure 2].
Schamberg disease [5]	Also known as progressive pigmented purpuric dermatitis, and progressive pigmented purpura .It is characterized by non- blanchable, red-brown purpuric patches. Close inspection of the patches reveals non-palpable pinpoint petechiae. It is named for an American dermatologist, Jay Frank Schamberg (1870-1934), [Figure 3], who first described it in 1901.

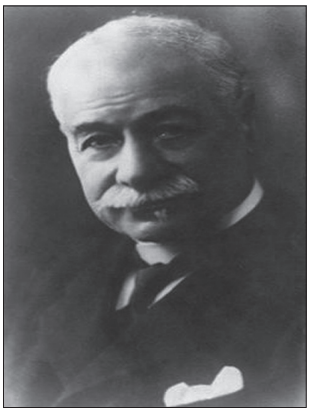

Figure 1: Domenico Majocchi (1849–1929).

Figure 2: Henri Gougerot (1881-1955).

Figure 3: Jay Frank Schamberg (1870-1934).

REFERENCES

1. Cavalcante MLLL, Masuda PY, Brito FF, Pinto ACVD, Itimura G, Nunes AJF. Schamberg's disease: case report with therapeutic success by using colchicine. An Bras Dermatol. 2017;92:246-8.
2. Al Aboud K, Al Aboud A.Eponyms in the dermatology literature linked to Greece. Our Dermatol Online. 2013;4(Suppl. 2):435-6.
3. Brzezinski P, Bourée P, Chiriac A, Bouquot JE, Schepis C, Hofer T, et al. Dermatology Eponyms – Sign – Lexicon – (M). Our Dermatol Online. 2014;5:312-26.
4. Al Aboud A, Al Aboud K. A mini-review on eponyms in the dermatology literature linked to France. Our Dermatol Online. 2013;4(Suppl. 2):440-3.
5. Al Aboud A, Al Aboud K.A mini-review on eponyms in the dermatology literature linked to United States of America (USA). Our Dermatol Online. 2013;4(Suppl. 1):409-13.

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