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Received 21 November 2018; accepted 13 March 2019

https://doi.org/10.1016/j.abd.2019.03.007

A case of linear atrophoderma of Moulin☆,☆☆

Dear Editor,

A 15-year-old Chinese girl presented with a 10-year history of asymptomatic, unilateral light brown patches affecting the right arm and right side of the trunk. The lesions were asymptomatic. There were no prior skin lesions or inflammation. There was no significant medical or family history. Physical examination found linear hyperpigmented atrophic patches on the right arm and right trunk following Blaschko's lines, involving both the anterior and posterior aspects. The skin was slightly atrophic on palpation. No signs of induration or inflammation were noted (Fig. 1A and B). Laboratory investigations – including full blood count, erythrocyte sedimentation rate, liver function test, renal profile, and antinuclear antibodies – were all negative or within the normal range. Biopsy of a lesion showed a normal epidermis with increased pigmentation of the basal layer, with more compact dermal collagen and mild upper dermal perivascular lymphocytic infiltration (Fig. 2). Dermoscopy found multiple light brown networks with unclear margins. The patient was diagnosed with linear atrophoderma of Moulin (LAM) and started treatment with topical halometasone 0.5% cream and hydroquinone 2% cream for two months, with no improvement.

LAM is a rare and distinct clinical entity characterized by acquired unilateral, hyperpigmented, and atrophic bandlike skin lesions following the lines of Blaschko, without prior inflammation or sclerotic appearance. It is named after Moulin, who, in 1992, reported on five patients with pigmented and more-or-less atrophic bands along Blaschko's lines.[1] LAM usually progresses as a linear atrophic lesion in the first few months; then the lesion ceases to progress and persists. The etiology of LAM remains unclear. All reported cases were so far sporadic. It may be connected with gene mosaicism or autoimmunity. A study of the atrophic component of LAM by ultrasonography revealed that subcutaneous volume reduction was the cause of the atrophic appearance, not dermal atrophy.[2] Even though the clinical manifestation of LAM is rather unique, the histopathology of LAM is quite inconspicuous. Hematoxylin and eosin staining usually shows hyperpigmentation only in basal epidermal layers, without abnormal collagen or elastic fibers in the dermis or any obvious inflammation.[1] There may be some perivascular lymphocytic infiltration, acanthosis, epidermal atrophy, altered collagen in the dermis, and decreased or fragmented elastic tissue.[2]

Lopez et al.[3] proposed the following diagnostic criteria for -LAM, including: (1) Onset during childhood or adolescence; (2) Development of hyperpigmented, slightly atrophic, unilateral lesions following Blaschko lines on the trunk or limbs; (3) Absence of prior inflammation or subsequent scleroderma; (4) A stable, non-progressive clinical course without a pattern of remission; (5) Histologic findings showing hyperpigmentation of the basal epidermis and a normal dermis with unaltered connective tissue and elastic fibers. Up to now, more than 30 cases of LAM have been reported in the literature. However, the condition may be overestimated. If the diagnostic criteria are strictly adhered to, the diagnosis of LAM cannot be reached in some cases, as these authors reported histologic findings that are compatible with other clinical entities.[3]

LAM must be differentiated from atrophoderma of Pasini and Pierini (APP), which also presents with similar configuration, atrophy, and hyperpigmentation, but does not follow Blaschko's lines. In addition, LAM is different from linear morphea, which usually presents preceding inflammation, induration, or scleroderma.

Histopathologically, morphea shows collagen bundles that are closely packed and oriented horizontally, and dermal appendages and subcutaneous fat are progressively lost.

☆ How to cite this article: Zhang L-W, Ma M-S, Chen T, Fu L-X. A case of linear atrophoderma of Moulin. An Bras Dermatol. 2020;95:119–21.

☆☆ Study conducted at the Chengdu Second People's Hospital, Sichuan, China.

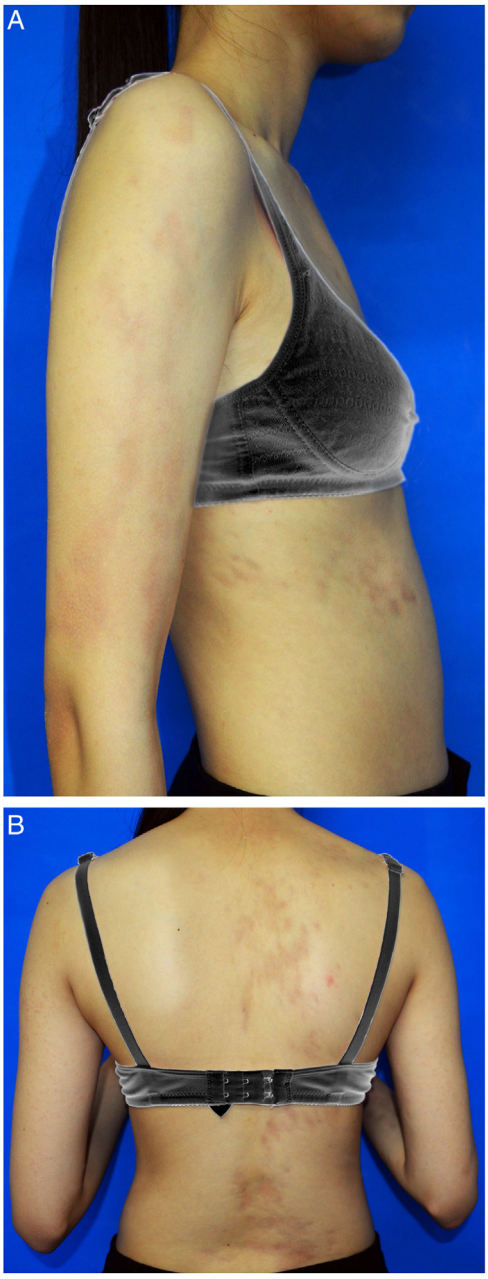

Figure 1 (A) Linear hyperpigmented patches on the right arm and right trunk following Blaschko's lines. (B) Linear hyperpigmented patches on the right arm and right trunk following Blaschko's lines.

However, it is still debated whether LAM is a distinct entity. There are many clinical and histologic similarities between LAM, APP, and morphea, thus some of the literature suggests that these diseases represent part of a disease spectrum, and that LAM may not be a distinct entity.[4] LAM may be a Blaschko-linear variant of APP, and APP may be considered to be an abortive form of morphea.[4]

There is no effective treatment for LAM. Topical corticosteroids and heparin were not helpful. Some trial treatments showed partial response to LAM, including the following: topical calcipotriol, systemic methotrexate or aminobenzoate, and intralesional platelet-rich plasma therapy.[5] The current report presented a case of LAM with classic clinical and histopathological features.

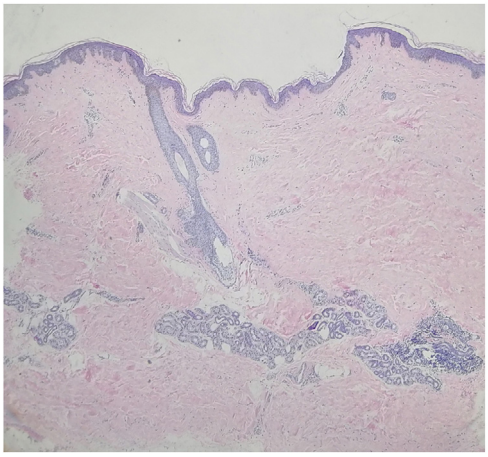

Figure 2 A normal epidermis with increased pigmentation of the basal layer, with more compact dermal collagen and mild upper dermal perivascular lymphocytic infiltration (Hematoxylin & eosin, ×40).

Financial support

None declared.

Authors' contribution

Li-Wen Zhang and Meng-Sha Ma contributed equally to this work. Li-Wen Zhang: Approval of the final version of the manuscript; conception and planning of the study; composition of the manuscript; critical review of the literature; critical review of the manuscript.

Meng-Sha Ma: Collection, analysis, and interpretation of data; critical review of the manuscript.

Tao Chen: Approval of the final version of the manuscript; conception and planning of the study.

Li-Xin Fu: Critical review of the literature.

Conflicts of interest

None declared.

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Received 18 January 2019; accepted 18 March 2019

https://doi.org/10.1016/j.abd.2019.03.005

Spiky follicular mycosis fungoides and hidradenitis suppurativa-like lesions in a patient – complete remission with interferon alpha☆,☆☆

Dear Editor,

Folliculotropic mycosis fungoides (FMF) is a variant of mycosis fungoides (MF) with distinctive clinicopathologic features, in which the neoplastic T lymphocytes display tropism for the follicular epithelium.[1] The spectrum of the clinical manifestations is heterogeneous.[1,2] Among them, acneiform lesions are a common presentation, but hidradenitis suppurativa-like lesions (HSLL) are scarcely described in literature. Moreover, spiky follicular mycosis fungoides is an uncommon clinicopathologic presentation, characterized by multiple hyperkeratotic follicular papules.

A healthy 65-year-old man presented a six-month history of generalized cutaneous lesions, alopecia, and severe itching. Dermatological examination revealed numerous spiky follicular papules and alopecia of the scalp (Fig. 1), with follicular keratotic lesions in trichoscopy. Patchy alopecia of the body hair and multiple millimetric hyperkeratotic spicules the on trunk and limbs were present, giving the sensation of rough skin at palpation. Moreover, HSLL were observed in the axillary area. Palpable lymphadenopathy and visceromegalies were not present. The biopsy of scalp showed an infiltrate of atypical lymphocytes in the follicular epithelium, with epidermotropism (Fig. 2). Immunohistochemically, follicular lymphocytes showed positivity for CD3 and CD4, with partial loss of CD7; CD30 was negative. Molecular analysis of TCR revealed a monoclonal population of lymphocytes. Laboratory tests were within normal limits (blood cell count, Sézary cells, biochemistry, electrophoresis, immunoglobulins, β-2 microglobulin) and no systemic involvement was detected in the body scan. A diagnosis of FMF was made. The patient received interferon alpha (IFN-α, 3,000,000 units three times weekly) and topical clobetasol, achieving complete remission one year later without recurrences after three years of follow-up (Fig. 3).

FMF represents less than 10% of patients with MF. This variant is more common in men, with an age of presentation similar to classic forms (around 55–60 years). Typically, it presents as hairless indurated plaques and tumors mainly on the head and neck, with severe pruritus. However, FMF is characterized by a broad clinical spectrum that comprises a variable combination of follicular lesions that may coexist.[1,2] Among them, spiky FMF has recently been well-described in a series of eight cases.[3] This peculiar clinical

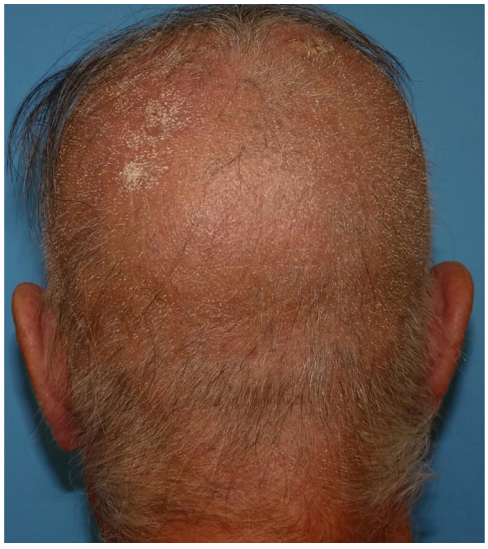

Figure 1 Extensive scalp involvement, with numerous whitish and spiky hyperkeratotic follicular papules, and alopecia.

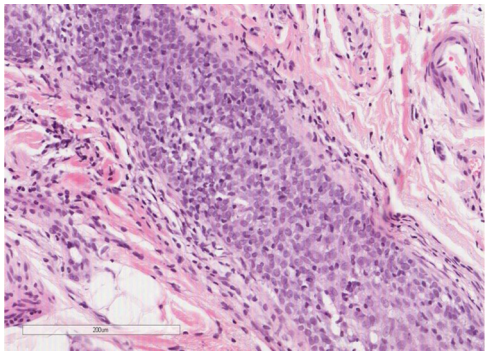

Figure 2 Biopsy of scalp: infiltrate of small-to-medium-sized lymphocytes with mild atypia, around and within follicular epithelium. No follicular mucinosis was present (Hematoxilin & eosin, ×20).

☆ How to cite this article: Garcia-Arpa M, Flores-Terry MA, Franco-Muñoz M, Lara-Simón IM. Spiky follicular mycosis fungoides and hidradenits suppurativa-like lesions in a patient – complete remission with interferon alpha. An Bras Dermatol. 2020;95:121–3.

☆☆ Study conducted at the Department of Dermatology. Hospital General Universitario de Ciudad Real, Ciudad Real, Spain.