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PHOTOLETTER TO THE EDITOR

Linear atrophoderma of Moulin progressing slowly over 46 years

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Abstract

Linear atrophoderma of Moulin is a rare acquired disorder arising most commonly during childhood or adolescence, occurring equally in both sexes and characterized by hyperpigmented atrophoderma in a unilateral bandlike distribution along the lines of Blaschko. Since Moulin *et al* described the condition in 1992, only a few dozen cases have been reported. It has been postulated that linear atrophoderma of Moulin may be due to mosaicism.

A 66-year-old man presented with a 46-year history of evolving tan soft atrophic confluent plaques in a strikingly Blaschkoid distribution, involving the left upper back, shoulder, upper arm, chest and flank. Initial onset, at age 20, consisted of a single mildly pruritic pink patch on the left back that was unresponsive to topical antifungals. Each new lesion arose similarly as a pink pruritic patch, subsequently becoming depressed, hyperpigmented, and asymptomatic over several years. Lesions were never scaly, firm, or indurated. Punch biopsy specimens were obtained. The clinical and histopathological features confirmed the diagnosis of linear atrophoderma of moulin.

Our present case has the characteristic clinical and histopathological features of linear atrophoderma of Moulin, but is the first reported case with mild pruritus at the onset of each new lesion and progressing slowly over 46 years. The lack of any systemic symptoms or other complications in our patient reaffirms the benign nature of this skin disease. (*J Dermatol Case Rep*. 2012; 6(4): 125-126)

Key words:

atrophy, plaque, pruritus

A 66-year-old man presented with a 46-year history of evolving tan soft atrophic confluent plaques in a strikingly Blaschkoid distribution, involving the left upper back, shoulder, upper arm, chest and flank (Fig. 1). Initial onset, at age 20, consisted of a single mildly pruritic pink patch on the left back that was unresponsive to topical antifungals. Each new lesion arose similarly as a pink pruritic patch, subsequently becoming depressed, hyperpigmented, and asymptomatic over several years. Lesions were never scaly, firm, or indurated. Punch biopsy specimens were

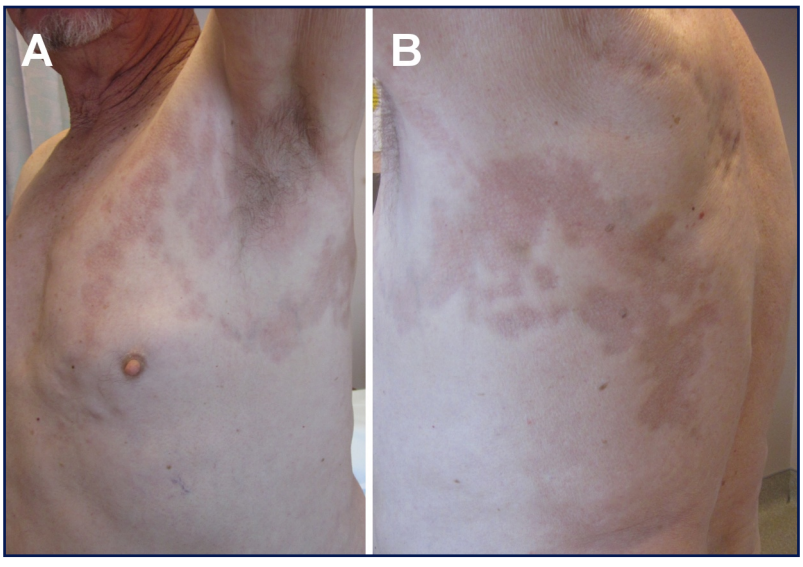

Figure 1

Hyperpigmented atrophic confluent plaques in a Blaschkoid distribution involving the left (A) chest, flank, (B) shoulder, and upper back.

obtained both from the most recent lesion, located on the left medial back (Fig. 2A), as well as from normal adjacent skin (Fig. 2B).

Histological comparison of the biopsy specimens showed the lesional skin with a mildly atrophic epidermis overlying notable dermal atrophy with thinned collagen bundles (Fig. 2, original magnification x 2). Fragmented elastic fibers were seen in the superficial reticular dermis as highlighted by Verhoeff-Van Gieson staining. The results were consistent with atrophoderma. The patient was reassured of the benignity of his skin condition, and no treatment was desired.

Linear atrophoderma of Moulin (LAM) is a rare acquired disorder arising most commonly during childhood or adolescence, occurring equally in both sexes and characterized by hyperpigmented atrophoderma in a unilateral bandlike distribution along the lines of Blaschko.[1,2] Since Moulin *et al* described the condition in 1992, only a few dozen cases have been reported.[3] The trunk and limbs are primarily affected, with no history of preceding induration, inflammation, or scleroderma.[2] Lesions typically develop over a limited time period and then remain present.[2] New lesions may arise. The prognosis is favorable, with no systemic manifestations or damage to underlying structures reported. The pathogenesis of LAM is unknown. All cases in the literature have been sporadic, and it has been postulated that LAM may be due to mosaicism from a postzygotic autosomal mutation in genes such as lamin A or collagen III (COL3A1).[2,4] The differential diagnosis for LAM primarily includes idiopathic atrophoderma of Pasini and Pierini (APP), plaque type morphea, and linear scleroderma. APP can be differentiated from LAM by its bilateral, symmetric pattern not following Blaschko's lines, and older lesions having central induration.[2] Plaque type morphea, in contrast to LAM, occurs predominantly in females, and lesions are ivory or white with a lilac margin, and densely sclerotic.[2] Linear scleroderma is distinguished from LAM by the presence of dermal sclerosis.[1]

In conclusion, we report the first case of LAM associated with mild pruritus at the onset of each new lesion and progressing slowly over 46 years. The lack of any systemic symptoms or other complications in our patient reaffirms the benign nature of this skin disease. Treatments used to date — UV-light, high-dose penicillin, topical steroids and heparin — for LAM have been ineffective.[1,3-5]

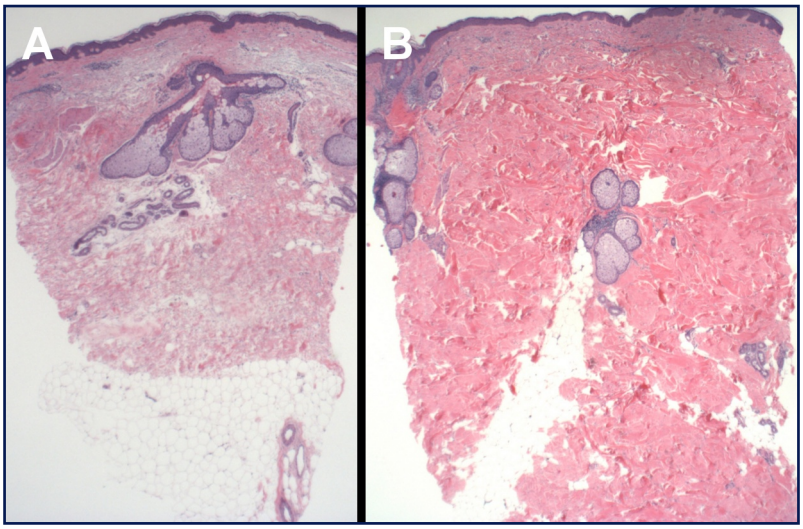

Figure 2

(A) Punch biopsy of lesional skin of left medial back showing a mildly atrophic epidermis overlying dermal atrophy, with thinned collagen bundles, compared to biopsy of (B) normal adjacent skin (original magnification x 2).

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