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Authors

Ahearn, Ian M
Gittler, Julia
Shvartsbeyn, Marianna
et al.

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Case presentation

Linear atrophoderma of Moulin

Ian M Ahearn MD PhD, Julia Gittler MD, Mariana Shvartsbeyn MD, Shane A Meehan MD, and Miriam Keltz Pomeranz MD

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Abstract

We present a 40-year-old woman with asymptomatic, linear, hyperpigmented atrophic plaques in a Blaschkoid distribution on the right back and right upper extremity that is consistent with a diagnosis of linear atrophoderma of Moulin. Clinical lesions developed with a biphasic pattern in late adolescence and in adulthood. The pathogenesis of this acquired, progressive Blaschkolinear dermatosis may hold insight into the pathogenesis of this rare dermatologic condition, as well as other dermatoses, which include those resulting from post-zygotic genetic mosaicism.

Case synopsis

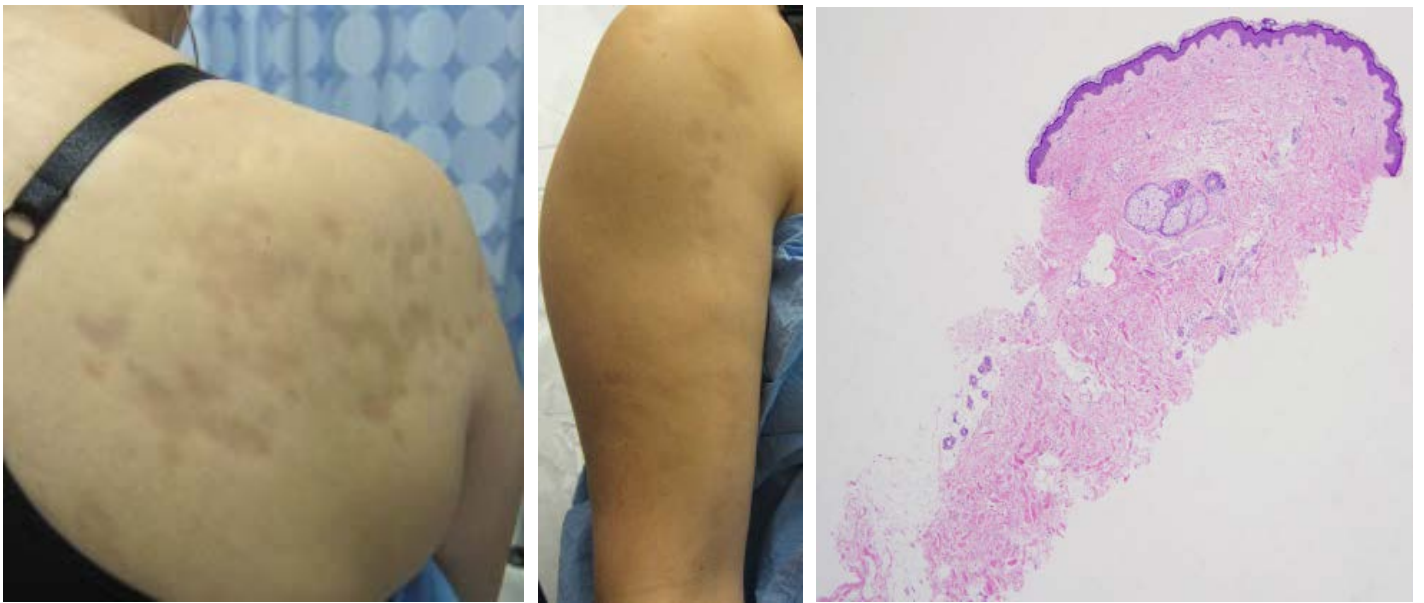
HISTORY: The patient is a 66-year-old woman who presented to the Dermatology Clinic at Bellevue Hospital Center with progressive, dark skin lesions over the right arm and trunk. The patient reported that the lesions first developed over 20-years ago, at the age of 19 over right forearm. Over the next five years, the lesions slowly spread proximally over the arm where they remained stable for over ten years. In the last five years, the lesions again have spread proximally over the right shoulder and back but have remained otherwise asymptomatic.

The patient did not recollect any cutaneous or systemic symptoms at the time when the lesions developed or subsequently. Past medical history was negative, and she took no medications. Review of symptoms was negative for neurologic, musculoskeletal, cardiopulmonary, gastrointestinal, or genitourinary symptoms.

PHYSICAL EXAMINATION: Over the right mid-back and extending from the midline, there were numerous, gray-brown, hyperpigmented, atrophic, round, linear plaques with a subtle recumbent-S arrangement that was oriented toward the right shoulder and distally over the anterior aspect of the right upper arm and dorsal aspect of the forearm. The lesion did not cross the posterior midline and was notable for the absence of any erythema or induration. The right and left upper limbs were of equal size and muscular tone.

LABORATORY: None

HISTOPATHOLOGY: There is a sparse, superficial, perivascular lymphocytic infiltrate. Within the dermis, there are only a few slightly thick collagen bundles and occasional elastic fibers that appear elongated with an Verhoeff-van Gieson stain. There is no loss of CD-34 expression.



Figures 1, 2. Hyperpigmented atrophic plaques Figure 3. Thickened collagen and occasional elongated elastic fibers

Discussion

Diagnosis: Linear atrophoderma of Moulin

Comment: Linear atrophoderma of Moulin (LAM) is a rare, acquired atrophoderma, which is characterized by linear, hyperpigmented, atrophic, unilateral plaques that are oriented along the lines of Blaschko. Typically, the lesions develop in childhood or adolescence and remain stable and asymptomatic thereafter. The differential diagnosis of LAM includes atrophoderma of Pasini and Pierini, linear morphea, epidermal nevus, inflammatory linear verrucous epidermal nevus, lichen striatus, linear psoriasis, and linear lupus erythematosus. Clinically, LAM may be distinguished from these entities by its Blaschkoid distribution, atrophic character, and the absence of inflammatory changes or induration.

LAM was first reported in 1992 in a series of five individuals.[1] Subsequently, over 30 cases have been reported.[2-4] Most reports have consistently described characteristic, acquired, linear, hyperpigmented, and atrophic lesions that were distributed over Blaschko's lines, but several clinical variations also have been described. These variants include lesions with prominent telangiectases [5], a congenital lesion [6], a lesion associated with anti-nuclear antibodies, [7] anatomic distribution over the neck, [8,9] lesions with preceding inflammation,[10] and lesions associated with lentiginosis [11-13]. Whereas the initial description of this dermatosis by Moulin identified only basal layer hyperpigmentation as a consistent histopathologic feature, subsequent reports have identified perivascular, lymphocytic infiltrates, thick or increased dermal collagen, and dermal atrophy.[7,14] Consequently, debate as to whether these reports are fully consistent with LAM has ensued. [5,15-17]

Ultrasonographic investigation of lesional skin in one patient with LAM demonstrated that the atrophy is secondary to the loss of subcutaneous adipose tissue [18] although other reports have described dermal atrophy.[7,14] Decreased dermal dermatan sulfate content has been purported to underlie atrophy in idiopathic atrophoderma of Pasini and Pierini,[19,20] but this finding has not been investigated in LAM.

Some clinicians consider LAM to represent one pole on one spectrum of putatively related conditions that include both idiopathic atrophoderma of Pasini and Pierini (IAPP) and linear morphea.[15] Regardless, LAM is not associated with lesional or systemic symptoms and portends a favorable prognosis. Treatment is aimed at cosmesis. Multiple, partially effective treatment modalities have been reported, which include the use of penicillin [15,21], psoralen plus ultra violet A (PUVA) photochemotherapy, [15] potassium aminobenzoate, [22] glucocorticoids,[23] heparin, [24] calcipotriene,[4] and methotrexate.[25]

The linear distribution of LAM over the lines of Blaschko highlights the fact that this dermatosis reflects cutaneous evidence of genetic mosaicism. Although candidate genes have been postulated,[14,17] no candidate has been confirmed. The evolution of lesions in childhood and adolescence also raises the possibility that an endogenous or exogenous trigger, perhaps hormonal, infectious, or environmental, may initiate lesion initiation and progression.[15]

Our patient first noted lesions over the right forearm with spread proximally over the arm in late adolescence, which was followed by ten years of quiescence during which the lesions remained stable. A prolonged progression of a LAM lesions has been reported previously, [14] but in our case a reported second-phase of lesion progression years later occurred after increased actinic exposure. The curious, biphasic evolution of LAM as an adolescent and as an adult in our patient raises the consideration that solar radiation was a putative environmental trigger.

In the context of unresolved genetic and pathophysiologic processes that underlie LAM, this case report may highlight several, albeit highly hypothetical, models for disease pathogenesis. As previously suggested, when a mutation is inherited in mosaic pattern as an acquired, somatic, post-zygotic event, perhaps damaging radiation may generate secondary mutations that resulted in either loss-of-heterozygosity or synthetic lethality.[14] In LAM, hypothetically, the consequences of such a genetic feature may lead to abnormal cutaneous tissue regeneration and the evolution of clinically apparent lesions. In this scenario, the putative complementary genetic insult could occur in a mitotically active, potentially multipotent cell type that is fundamental to tissue regeneration, which may occur preferentially along embryonic planes that are marked by Blaschko's lines. Alternatively, cutaneous tissues that inherit mosaic mutations in LAM may instead possess a fundamentally restricted regenerative potential, either inherently or after a subsequent triggering event, such as increased cell turnover. This possibility is independent of a role for pluripotent progenitors or even second-hit loss-of-heterozygosity or synthetically lethal events.

Beyond tissue regeneration, other distinct hypotheses for the pathogenesis of LAM exist. One example is the possibility that tissues that inherit a mosaic mutation either inherently, following an environmental trigger, or via additional synthetically lethal or loss-of-heterozygosity mutations, present an altered self-antigen in a Blaschkolinear distribution, which may subsequently illicit a skin-damaging autoimmune response.

Regardless of the merit of these speculations or whether LAM exists on a clinical spectrum with IAPP and linear morphea, the identification of a genetic causes or phenotypic triggers may hold insights for other challenging dermatologic conditions.

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