Linear Atrophoderma of Moulin: A Case Report

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ABSTRACT:
Linear Atrophoderma of Moulin (LAM) is an uncommon acquired, hyperpigmented, and atrophic resembling band lesions along the Blaschko's lines in the absence of antecedent inflammation or sclerotic appearance. The disease is benign, with no related complications, despite the lack of a successful treatment. The precise etiology and optimal course of treatment are still up for debate. In the present study, a 19-year-old female patient was diagnosed with LAM. Her condition, which highlighted the typical clinical and histological characteristics, improved with topical calcipotriol treatment.
Key words: Linear Atrophoderma of Moulin, Blaschko's lines

Introduction

Linear Atrophoderma of Moulin (LAM), an unusual and distinctive disease, was initially described in 1992[1]. Acquired, unilateral, atrophic resembling band of hyperpigmented lesions following Blaschko's lines that do not appear sclerotic or have any prior inflammation are the hallmarks of the disease[2, 3]. It typically appears in childhood and adolescence, though it can also show up at a later age[4]. Although the exact etiology of the condition is unknown, mosaicism from a postzygotic mutation in an early developmental stage is thought to be the cause of disease[5]. The prognosis is benign, self-limited, and no related consequences have been reported, despite the lack of a successful treatment[2,3,5-7]. Currently, the actual pathogenesis of the illness, its nature, and the best course of therapy are still up for debate.

Case report

A 19-year-old female from Bangkok developed asymptomatic, slow-progressive, unilateral atrophic patches from the left upper arm to the left wrist for 2 years. She denied a previous history of inflammation or chemical use before the development of these skin lesions. No systemic symptoms, including dyspnea, dysphagia, and joint stiffness, were observed. She was previously prescribed unknown topical and oral medications without clinical improvement. She was healthy with an unremarkable family history.

Physical examination revealed multiple discrete unilateral linear brownish atrophic patches with an irregular border extending from the left upper arm to the wrist, following Blaschko's line (Figure 1). There were no sclerodactyly, sclerodermoid skin alterations, Raynaud's syndrome, or aberrant nailfold capillaries. Others were unremarkable.

A differential diagnosis included LAM, atrophoderma of Pasini and Pierini, and linear morphea. Basic investigations, including blood cell count, liver and renal function tests, antinuclear antibody (ANA), and urinalysis, were normal.

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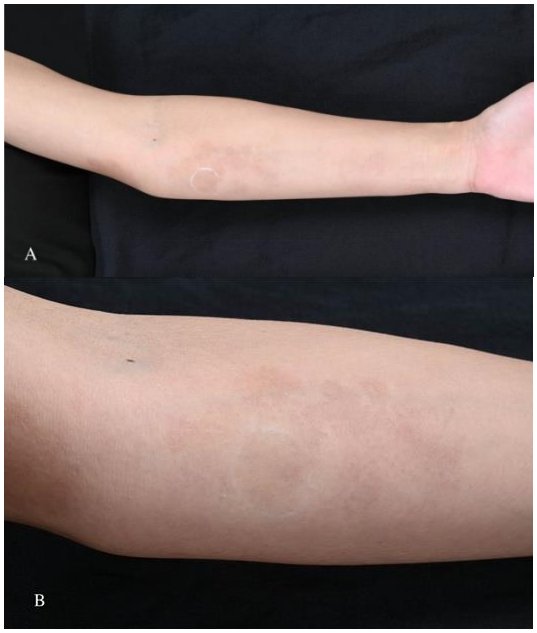

Figure 1 (A and B) Multiple discrete unilateral linear brownish atrophic patches with an irregular border extending from the left upper arm to the wrist, following Blaschko's line

An incisional skin biopsy from the lesion on the left forearm was performed. Histopathology revealed basal hyperpigmentation, along with sparse superficial perivascular lymphocytic infiltration and the presence of focal thickened hyalinized collagen in the dermis (Figure 2).

According to the patient's clinical presentation and histopathology, the final diagnosis was LAM. Treatment with topical 0.05% clobetasol propionate cream was prescribed for one month without clinical improvement; thus, the medication was changed to topical calcipotriol ointment twice daily. After a few months of follow-up, the lesions were partially improved and did not progress.

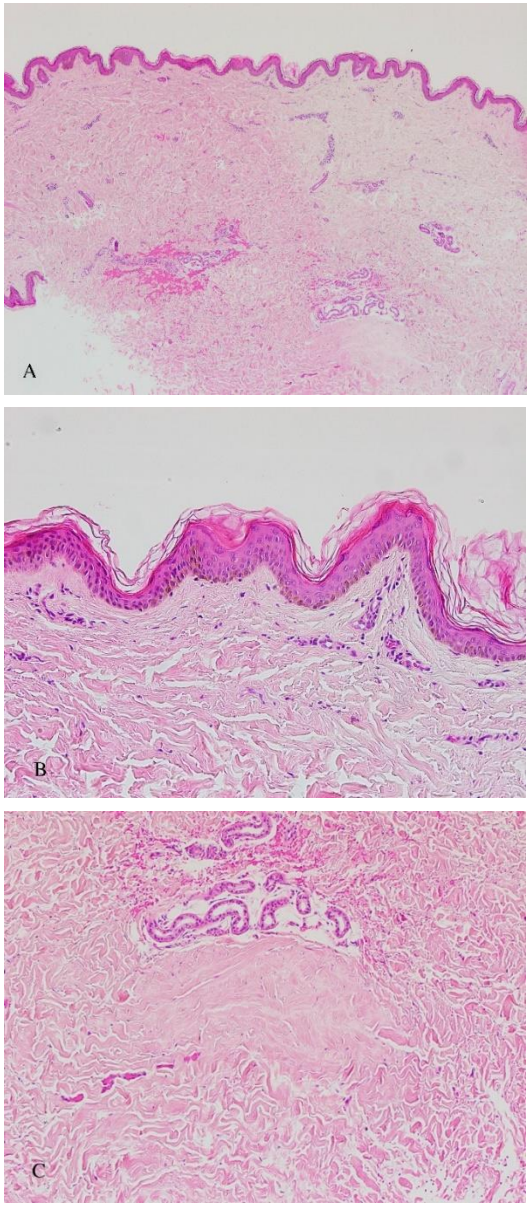

Figure 2 The hematoxylin and eosin staining of the lesion from the left forearm. (A-B) Basal hyperpigmentation, along with sparse superficial perivascular lymphocytic infiltration. (C) The presence of focal thickened hyalinized collagen in the dermis. (A: 4X; B: 20X; C: 10X)

Discussion

LAM is a rare disease characterized by unilateral, atrophic, and hyperpigmented band-like cutaneous lesions along the Blaschko's lines without preceding inflammation or subsequent sclerotic appearance[1]. Clinically, LAM presents very distinctive features that are unique to identified and differentiated from other conditions. Our patient's clinical presentation closely resembles those previously described[1].

LAM may be a late-onset skin development instance that manifests between the ages of 2 and 37[4]. The condition is self-limiting and can remain stable for an entire 30-year observation period[8]. The exact cause of LAM is still unclear, and specific genetic associations are currently unknown[5]. Since LAM, atrophoderma of pasini and pierini, and morphea share a lot of clinical and histological characteristics, some study indicates that these conditions are part of a larger spectrum of disorders and that LAM itself may not be a separate entity[9]. Another theory stated that LAM was an autoimmune disease, because ANA was positive in several cases of LAM[9, 10]. To date, all reported cases have been unpredictable. The Blaschko lineage is thought to be the source of the mosaicism resulting from a postzygotic mutation during an early developmental stage[5].

The differential diagnosis includes pigmentary dermatoses along Blaschko's line, such as lichen striatus, linear and whorled nevoid hypermelanosis, epidermal nevi, and incontinentia pigmenti, in addition to atrophoderma of Pasini and Pierini and linear morphea. The lesions of atrophoderma of Pasini and Pierini are characterized by bilateral, symmetric, gray-brown, depressed patches with a cliff-drop border. These lesions do not follow Blaschko's line, which differentiate them from LAM. A lack of cutaneous inflammation in the initial stages, which ultimately results in skin atrophy and induration, helps distinguish LAM from linear morphea. The distinction between LAM and linear morphea is crucial since the former is a benign condition, while the latter may harm the underlying muscle and bone[11].

As previously mentioned[1,3,4], the histopathological characteristics of LAM are diverse and nonspecific. The most frequent histologic finding is perivascular lymphocytic infiltration in the superficial dermis associated with aberrant collagen fibers[6]. According to recent ultrasound imaging investigations, the atrophic feature in the majority of LAM patients is caused by a decrease in dermal thickness, although subcutaneous tissue may also be involved in some cases[10,12,13].

Currently, the proposed criteria for the diagnosis of LAM include[6]

- Onset during childhood or adolescence
- Formation of unilateral, atrophic, hyperpigmented lesions on the trunk or extremities that resemble Blaschko's lines
- Lack of prior inflammation and subsequent induration or scleroderma
- Constant course without a remission pattern
- Histopathologic results show the hyperpigmented basal layer of the epidermis and intact connective tissue and elastic fibers of the dermis

Thus far, LAM has no established course of treatment. Topical corticosteroids, heparin, high-dosage penicillin, and potassium aminobenzoate have all previously proved to be unsuccessful options of treatment[2, 14-16]. On the other hand, topical calcipotriol, topical tacrolimus, oral methotrexate, and intralesional platelet-rich plasma therapy have been shown to provide some degree of improvement[7,10,17,18]. In contrast, treating linear morphea with methotrexate and corticosteroids frequently yields excellent result. The primary goal of treatment is often to address cosmetic concerns, with alternatives including self-tanning lotion and filler augmentation[19].

Our patient was treated with topical calcipotriol ointment twice daily. The synthetic

vitamin D3 analogue calcipotriol binds to the vitamin D receptor with a high affinity[20]. Therefore, it is possible to hypothesize that calcipotriol might improve disorders through anti-inflammatory and anti-proliferative effects as well as enhanced terminal differentiation.

In conclusion, we reported herein that a teenage girl presented with classic LAM and showed partial response to topical calcipotriol therapy.

References

1. Moulin G, Hill MP, Guillaud V, Barrut D, Chevallier J, Thomas L. Bandes pigmentées atrophiques acquises suivant les lignes de Blaschko [Acquired atrophic pigmented band-like lesions following Blaschko's lines]. Ann Dermatol Venereol 1992;119:729-36.
2. Tan SK, Tay YK. Linear atrophoderma of Moulin. JAAD Case Rep 2016;2:10-2.
3. Zhang LW, Ma MS, Chen T, Fu LX. A case of linear atrophoderma of Moulin. An Bras Dermatol 2020;95:119-21.
4. Villani AP, Amini-Adlé M, Wagschal D, Balme B, Thomas L. Linear atrophoderma of moulin: report of 4 cases and 20th anniversary case review. Dermatology 2013;227:5-9.
5. Danarti R, Bittar M, Happle R, König A. Linear atrophoderma of Moulin: postulation of mosaicism for a predisposing gene. J Am Acad Dermatol 2003;49:492-8.
6. López N, Gallardo MA, Mendiola M, Bosch R, Herrera E. Atrofodermia lineal de Moulin: presentación de un caso [A case of linear atrophoderma of Moulin]. Actas Dermosifiliogr 2008;99:165-7.
7. Kharkar VD, Abak BA, Mahajan SA. Linear atrophoderma of Moulin: A rare entity. Indian J Dermatol Venereol Leprol 2018;84:591-4.
8. Patsatsi A, Kyriakou A, Chaidemenos G, Sotiriadis D. Linear atrophoderma of Moulin: a case report and review of the literature. Dermatol Pract Concept 2013;3:7-11.
9. de Golian E, Echols K, Pearl H, Davis L. Linear atrophoderma of Moulin: a distinct entity? Pediatr Dermatol 2014;31:373-7.
10. Ertop Doğan P, Topuz M, Hazinedar E, Doğan Gün B, Türk Y, Koca R. Clinical and dermoscopic findings of linear atrophoderma of Moulin: a case with an autoimmune and inflammatory background. Int J Dermatol 2024;63:1102-4.
11. Zahedi Niaki O, Sissons W, Nguyen VH, Zargham R, Jafarian F. Linear atrophoderma of Moulin: an underrecognized entity. Pediatr Rheumatol Online J. 2015;13:39.
12. Zhang LW, Li CH, Fu LX, et al. Linear atrophoderma of Moulin is due to the decreased dermal thickness. Skin Res Technol 2022;28:646-8.
13. Li C, Zhang L, Zheng L, Lu Y, Liu Y. Linear atrophoderma of Moulin: dermal or subcutaneous atrophy? Australas J Dermatol 2020;61:e365-e6.
14. Baumann L, Happle R, Plewig G, Schirren CG. Atrophodermia linearis Moulin. Ein neues Krankheitsbild, das den Blaschko-Linien folgt [Atrophodermia linearis Moulin. A new disease picture, following the Blaschko lines]. Hautarzt 1994;45:231-6.
15. Wollenberg A, Baumann L, Plewig G. Linear atrophoderma of Moulin: a disease which follows Blaschko's lines. Br J Dermatol 1996;135:277-9.
16. Artola Igarza JL, Sánchez Conejo-Mir J, Corbí Llopis MR, Linares Barrios M, Casals Andreu M, Navarrete Ortega M. Linear atrophoderma of Moulin: treatment with Potaba. Dermatology 1996;193:345-7.
17. Wongkietkachorn K, Intarasupht J, Srisuttiyakorn C, Aunhachoke K, Nakakes A, Niumpradit N. Linear atrophoderma of moulin: a case report and review of the literature. Case Rep Dermatol 2013;5:11-4.
18. Zaouak A, Ghorbel HH, Benmously-Mlika R, et al. A case of linear atrophoderma of Moulin successfully treated with methotrexate. Dermatol Ther 2014;27:153-5.
19. Utikal J, Keil D, Klemke CD, Bayerl C, Goerdt S. Predominant telangiectatic erythema in linear atrophoderma of Moulin: novel variant or separate entity? Dermatology 2003;207:310-5.
20. Holm EA, Jemec GB. The therapeutic potential of calcipotriol in diseases other than psoriasis. Int J Dermatol 2002;41:38-43.