# Linear Atrophoderma of Moulin with Facial Involvement: A Case Report and Review of the Literature

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**ABSTRACT:**

**WEERASUBPONG P, SILPA-ARCHA N. LINEAR ATROPHODERMA OF MOULIN WITH FACIAL INVOLVEMENT: A CASE REPORT AND REVIEW OF THE LITERATURE. THAI J DERMATOL 2021;37:69-75.**

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Linear atrophoderma of Moulin (LAM) is a rare, self-limited, linear dermatosis characterized by acquired, unilateral asymptomatic, atrophic, hyperpigmented lesion along the Blaschko's lines. The prognosis is good, since there is no evidence of any systemic involvement or long-term progression. Lesions usually appear on the trunk and extremities, with few incidents on the facial area. The main purpose of treatment is aesthetic. However, unresolved questions still remain regarding the existence of a preceding inflammatory phase, the pathogenesis of the disease, and the most effective therapy. In this study, we report a new case of LAM with facial involvement, along with review the relevant literatures.

**Key words:** Atrophoderma, blaschko's lines, face, linear, moulin

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Received: 30 November 2020 Revised: 1 March 2021 Accepted: 24 March 2021

## Introduction

Linear atrophoderma of Moulin (LAM) is a rare, self-limited entity of linear dermatosis that was first described and reported by Moulin et al. in 1992[1]. The classic presentation is characterized by acquired, unilateral asymptomatic, atrophic, hyperpigmented patches, bands or plaque along the Blaschko's lines with or without preceding inflammation on the trunk and/or limbs. The disease is thought to be due to genetic mosaicism, and it typically affects children and adolescents with no familial or medical history. Since there is no evidence of any systemic involvement and long-term progression, the prognosis is typically excellent. Treatment is sought for aesthetic reasons only. However, some questions remain to be answered regarding the existence of a preceding inflammatory phase, the pathogenesis of the disease, and the most effective treatment modalities. The purpose of this article is to report a new case of LAM with an uncommon presentation of facial involvement and to review the literature after more than 50 such cases were documented.

## Case presentation

A 50-year-old woman from Bangkok presented with shiny, skin-colored plaque on her right forehead, chin, arm, and upper back which she claimed to have since birth. The rashes gradually darkened from childhood on. Subsequently, the lesions on her face became depressed over the past two years. No preceding skin inflammation occurred. She denied of having any other systemic symptoms. No dysphagia, dyspnea, or Raynaud phenomenon was detected. No family history of similar skin disease was noted. She had not received any prior medication for this condition. She has had diabetes mellitus for eight years.


**Figure 1A, 1B:** Right facial hemiatrophy and linear, atrophic, brownish plaque on the right forehead, upper vermillion border, and chin

Skin examination showed right-facial atrophy. There was linear, atrophic, hyperpigmented plaque on her right forehead, upper vermillion border, and chin. Ill-defined hyperpigmented atrophic plaque on her right shoulder and right arm following the Blaschko's lines, no sclerodermoid skin changes nor Raynaud phenomenon were observed. Others were unremarkable. (Figure 1)

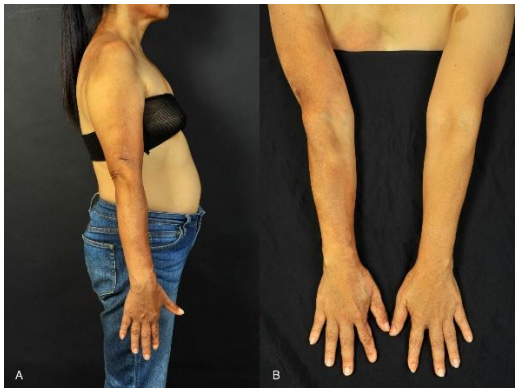

**Figure 2A, 2B** Ill-defined, hyperpigmented, atrophic plaque on the right shoulder and right arm

Histopathology from the patient's right arm showed hyperpigmentation of the basal epidermis with mild, superficial, perivascular lymphocytic infiltrate. Dermal sclerosis was not observed. (Figure 3) We performed elastic staining with Verhoeff-van Gieson stain and found no decrease amount of elastic fiber as compare to periphery of the lesion. (Figure 4)

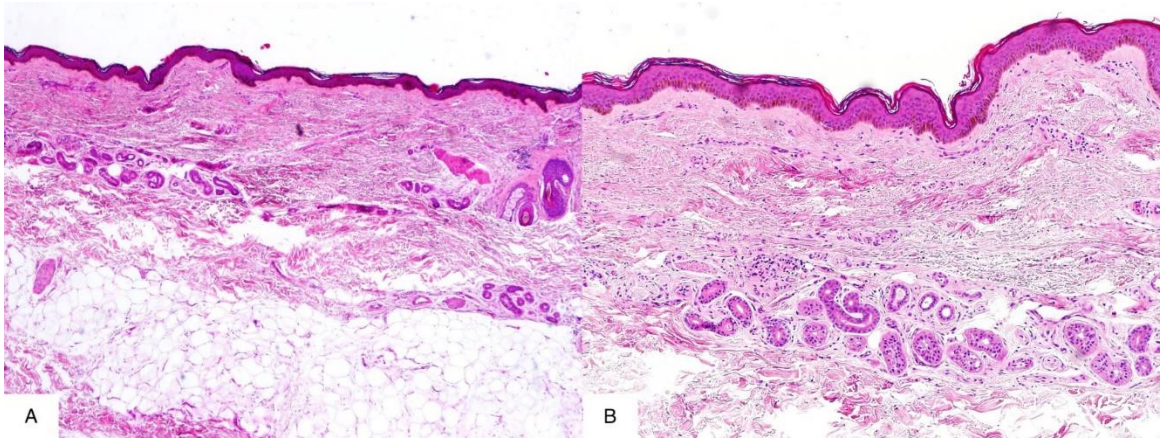

**Figure 3 A, B** Hyperpigmentation of the basal epidermis with mild, superficial, perivascular lymphocytic infiltrate. No dermal sclerosis was observed (Fig 3A: H&E X4, Fig 3B: H&E X10)

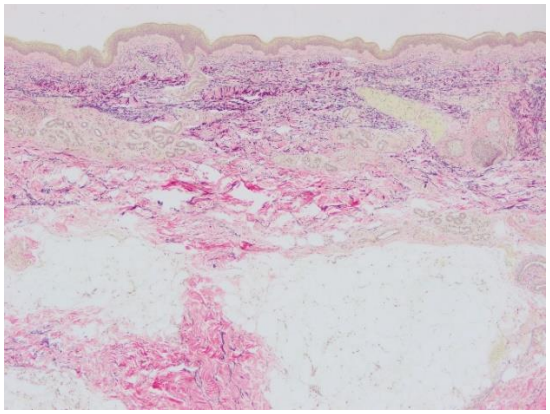

**Figure 4** The elastic stain does not show decrease amount of elastic fiber as compare to periphery of the lesion (H&E X4)

## Discussion

LAM is a rare, self-limited entity of dermatosis characterized by acquired, unilateral, asymptomatic atrophic patches, bands, or plaque along the Blaschko's lines, with or without preceding inflammation on the trunk and/or limbs.[2]

The initial five cases described by Moulin et al[1] were characterized by unilateral, hyperpigmented, atrophic plaques following the Blaschko's lines. The histopathology typically describes a moderate basal hyperpigmentation with normal collagen and elastic fibers without inflammation. Clinical variants of the disease have included bilateral[3,4], telagiectatic[3], and lentiginous lesions[4].

Thirty years after the first description of this entity, with more than 50 reported cases of linear atrophoderma of Moulin, some issues remain unresolved. The exact pathogenesis of LAM is still unknown. However, inferred from its distribution, it is presumed to reflect genetic mosaicism[5]. Table 1 summarizes the reported cases of linear atrophoderma of Moulin with facial involvement.

The diagnostic for LAM[5] can be made clinically and has been proposed as follows: (1) hyperpigmented, slightly atrophic, unilateral lesions following the Blaschko's lines on the trunk or limbs; (2) an absence of preceding inflammation and subsequent induration or scleroderma; (3) the onset is usually during childhood or adolescence; (4) the disease's course is stable, non-progressive clinically, and without a pattern of remission; (5) histologic findings show hyperpigmentation of the basal epidermis and a normal dermis with unaltered connective tissue and elastic fibers, slight thickening of collagen fibers, and sparse perivascular lymphocytic infiltration in the dermis. Some acanthosis, epidermal atrophy, and decrease or fragmented elastic tissue may be found with no clear signs of dermal atrophy[6]. Dermoscopy findings of multiple light-brown networks with unclear margins may suggest LAM[7]. Our case matches the characteristic clinical and histopathological features of LAM without any systemic symptoms. The distribution on the chin is similar to what Darung et al[8]. reported in 2017, which is a rarity.

The growing literature has revealed that the onset is not limited to childhood or adolescence,

but the condition may also be congenital[9] or present later in life. Lesions may be bilateral, but in a linear distribution along the Blaschko's lines. There have also been unusual cases of LAM with telangiectatic[3,10], lentiginosis[4], hypopigmented,[9] or initial lesions with papules[11].

Thus, if the diagnostic criteria or clinical characteristics are strictly adhered to, a diagnosis of LAM cannot be made in some cases. The histologic findings may be beneficial in making a diagnosis of LAM.

The differential diagnoses of LAM have included linear atrophoderma of Pasini and Pierini (APP) in which lesions are often bilateral, symmetric, and do not follow a blaschkoid pattern. Pain, pruritus, or even paresthesia may occur. In Morphea, the lesions are ivory, sclerotic with a lilac margin lesion, and with a presence of sclerosis. Linear scleroderma also presents in sclerosis.

Histopathologically, APP resembles LAM in terms of the hyperpigmentation of the basal cells, altered collagen fibers, and a sparse perivascular lymphocytic infiltrate; however, APP may present with dermal thinning, as well. The histologic features of linear morphea include homogenization of the papillary dermis, thickened collagen bundles, an absence of appendageal structures, and a presence of inflammatory cells.

Thus far, there are no standard regimen or proven effective treatment options for LAM. Many treatment modalities had been tried on LAM lesions, including high-dose penicillin, topical corticosteroids, topical calcineurin inhibitors, heparin and oral potassium benzoate, oral methotrexate, PUVA therapy, and platelet-rich plasma, but there has been little success. As LAM is a benign, self-limited disorder, with a stable clinical course, and is confined to the skin, the treatment has mainly been to improve how one looks. A report of En coup de sabre, a variant of linear morphea, involving the use of a dermal hyaluronic injections showed satisfying outcome, with no complications[12]. There was also a reported case of Parry–Romberg syndrome, a variant of localized scleroderma which is characterized by facial hemiatrophy and treated with a calcium hydroxylapatite filler[13]. Filler and a lipofilling injection were given in the chin in our patient for cosmetic correction. Unfortunately, she was not available for follow-up.

## Conclusions

The diagnosis of LAM is clinical and includes the existence of acquired, hyperpigmented, linear atrophoderma which follows the Blaschko's lines, along with an absence of sclerosis, and an onset during childhood or adolescence, with a stable disease course. Facial involvement has rarely been reported, thus should be. No certain

effective therapy is currently available. The treatment for LAM is mainly for aesthetic reasons, and long-term follow-up should help guide us in understanding the evolution of the disease and the best treatment options.

Hyaluronic acid is the most prominent glycosaminoglycan in the skin. When injected into the skin, it volumizes, softens, and hydrates the skin by potently binding to water. Apart from being widely used to enhance aging skin, it may also be beneficial off-label for patients who experience atrophic changes due to a disease.

**Acknowledgement**

The authors would like to thank Associate Professor Penvadee Pattanaprichakul for her kind assistance in histopathology.

**Table 1** List of reported cases of Linear atrophoderma of Moulin with facial involvement

| Author, year | Sex | Age of onset (year) | Duration | Location | Skin biopsy/Histology | Treatment |
|---|---|---|---|---|---|---|
| Miteva[10], 2002 | F | 16 | 4 years | Face, leg | Psoriasiform epidermal hyperplasia with hyaline eosinophilic bodies; perivascular lymphocytes in the dermis; increased collagen in the dermis | N/A |
| Cecchi[14], 2008 | M | 8 | 1 year | Face, neck | Hyperpigmentation of the lower epidermis; perivascular lymphocytes in the dermis | N/A |
| Tukenmez Demirci[15], 2011 | F | 17 | 22 years | Face, neck | Hyperpigmentation of the lower epidermis; perivascular lymphocytes in the dermis | N/A |
| Darung[8], 2017 | F | 16 | 6 months | Face | Epidermal atrophy; dense melanin deposition along the basal layer with sparse perivascular and peri appendageal lymphocytic infiltrate with slight thickening of collagen bundles in the dermis | Topical tretinoin (0.05%) cream once at night |
| Our case, 2020 | F | At birth | 50 years | Face | Hyperpigmentation of the basal epidermis with a mild superficial, perivascular lymphocytic infiltrate | Filler and lipofilling injection |

Abbreviations: F: female, M: male, N/A: not applicable

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