Blashckolinear atrophoderma of pasini and pierini with shortenening of foot-a rare entity

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The Hyperpigmented, depressed band-like skin lesions following Blaschko Lines (BL) were described by Moulin, et al and are known as Atrophoderma of Moulin. It is redefined as Blashckolinear Atrophoderma of Pasini and Pierini. Atrophoderma of Pasini and Pierini is a rare type of dermal atrophy described by asymptomatic slightly miserable parts with a distinct 'cliff-drop' edge and is usually situated on the backs. Atrophoderma of Pasini and Pierini generally first appears in puberty or early adulthood. The affected skin seems thinned and discolored, but the consistency and feel of the affected skin remains normal.

Key Words: *Atrophoderma; Blaschko's lines; Linear scleroderma; Lichen striatus; Focal dermal hypoplasia*

INTRODUCTION

Dermatoses with linear distribution include Linear Atrophboderma of Moulin (LAM), Atrophoderma of Pasini and Pierini (APP) and linear morphea [1-2]. Linear Atrophoderma of Moulin (LAM) is a rare linear dermatosis. It is sometimes redefined as Blaschkolinear Atrophoderma of Pasini and Pierini. It usually presents in childhood or early adolescence as hyperpigmented atrophic linear bands along Blaschko's lines without prior inflammation or sclerotic appearance [3].

CASE REPORT

A 14 year old female presented with gradually progressive hyperpigmented, depressed lesions along Blaschko's lines (Figure 1A and 1B) over right upper limb and ipsilateral trunk for last 7 years. There was also history of shortening of right foot (Figure 2) since 4 years.

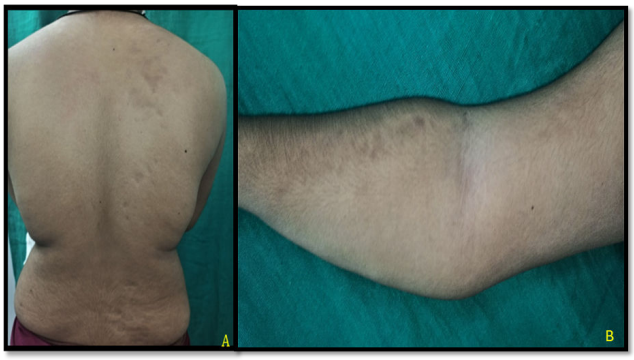

Figure 1) *(A,B) Lesions along Blaschko's lines*

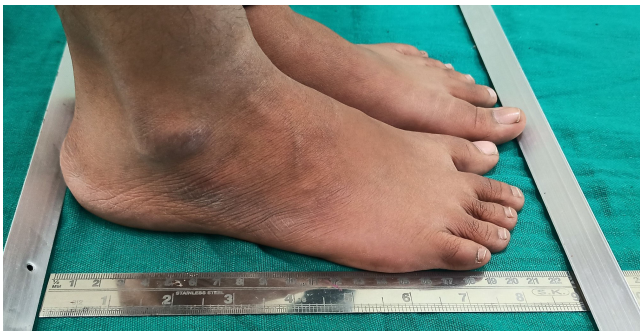

Figure 2) *Shortening of right foot*

On examination, there were multiple depressed, hyperpigmented, anhidrotic lesions over right forearm and ipsilateral trunk. There was muscle wasting of right leg (Figure 3A-3D) and shortening of right foot (approximately 1 cm shorter than left foot) with clinical evidence of anhidrosis around ankle joint. Skin of right leg was fixed to underlying bone. There was no other significant history.

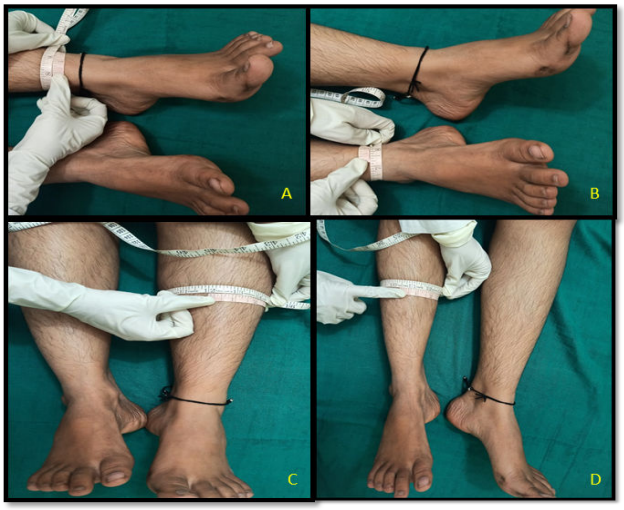

Figure 3) *(A-D) Muscle wasting of right leg*

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We considered linear scleroderma, Atrophoderma of Moulin, Atrophoderma of Pasini and Pierini as differential diagnoses. Histopathological examination revealed findings consistent with scleroderma. Other lab investigations including anti-nuclear antibody and anti-ds DNA antibody came out to be negative. Correlating the clinical picture with histological examination, a probable diagnosis of linear atrophoderma of Moulin was made. Patient was treated with oral mini pulse therapy of Tab. Prednisolone 40 mg OD twice per week. There was significant clinical improvement in the lesions of right upper limb within first 1 month of therapy and then patient lost to follow up.

DISCUSSION

Dermatoses with linear distribution include Linear Atrophboderma of Moulin (LAM), Atrophoderma of Pasini and Pierini (APP) and linear morphea [1]. Moulin first described linear atrophoderma of Moulin in 1992, as an acquired unilateral hyperpigmented atrophic band along Blaschko's lines [4]. Linear atrophoderma of Moulin is a rare skin condition characterized by unilateral, hyperpigmented and atrophic bandlike skin lesions following the lines of Blaschko. Usually, the disease begins in childhood or adolescence on the trunk or limbs, with no preceding inflammation or subsequent sclerodermatous changes. It then progresses for a few months in a blaschkoid pattern and then persists as an atrophic plaque. There are two variants of the same disease: an inflammatory and a non-inflammatory linear Atrophoderma of Moulin [5]. The etiology of LAM remains unclear [3]. 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. Hyperpigmented atrophoderma that follows Blaschko's lines, with onset usually occurring during childhood and adolescence. Etiologically it is unknown form of dermal atrophy. It usually presents as oval or round atrophic, nonsclerotic, hyperpigmented patches following Blaschko's lines. These patches are usually located on the trunk and the upper and lower extremities. This case study discusses a patient that had the classical form of LAM with the initial lesions presenting along Blashcko's lines [6].

The differential diagnosis of LAM includes APP, linear scleroderma, lichen striatus, and focal dermal hypoplasia. Linear scleroderma and APP may show a segmental atrophic plaque similar to LAM [7]. Histologically, linear atrophoderma of Moulin is characterized by hyperpigmentation of the basal cells along with slight thickening of the collagen fibers in the dermis No clear signs of dermal atrophy are seen. Rather, atrophic appearance on clinical examination is secondary to reduction in subcutaneous tissue is present [8]. In this case report no signs of sclerosis noted and there was apparent shortening of right lower limb along with restriction of movements at the involved site due to atrophic changes.

There is no effective treatment for linear atrophoderma of Moulin. Topical steroids and high-dose penicillin are reported to give good results. Oral potassium aminobenzoate 12 g/day, platelet rich plasma therapy have been used successfully [9]. Wongkietkachorn, et al.[3] used topical calcipotriol. Zaouak, et al. [7] reported a case successfully treated with methotrexate [10].

CONCLUSION

Linear Atrophoderma of Moulin is considered to be a variant of morphea. Linear trophoderma of Moulin presents along Blaschko's lines. It is very rare variant of morphea. It arises without preceding irritation, subsequent induration or scleroderma. Linear Atrophoderma of Moulin (LAM) usually grows as a linear atrophic lesion in the first few months; then the injury ceases to progress and persists. Several treatments were done but none was consistently fruitful. Linear Atrophoderma of Moulin takes a chronic course. The disease results in important cosmetic impairment and this might cause emotional stress. Till now there is no effective and lasting treatment available for treatment of atrophoderma of Moulin.

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