VOLUME - 12, ISSUE - 10, OCTOBER - 2023 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Dermatology

LINEAR ATROPHODERMA OF MOULIN- A RARE CASE REPORT

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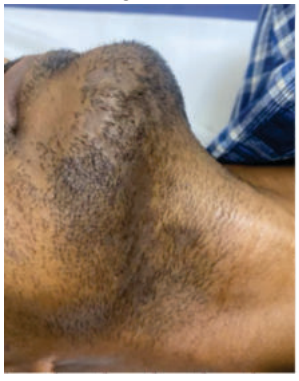
KEYWORDS :

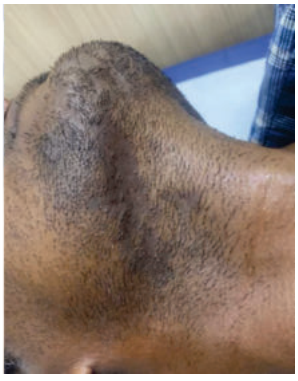
INTRODUCTION :

Linear atrophoderma of Moulin (LAM) is a rare dermatologic disorder characterized by hyperpigmented atrophic plaques following the Blaschko lines (BL).The trunk and limbs are the usual sites affected. Isolated facial involvement is an exceedingly rare entity. The etiology and pathogenesis of LAM are still unclear. All patients that have been reported were sporadic, and it has been postulated that LAM may be mosaic from a postzygotic autosomal mutation in genes due to the linear distribution following Blaschko's lines. Despite a comprehensive medical literature search, this case is presented due to its rarity and to have a clear knowledge about clinical features, diagnosis and treatment of the disease.

Case Report:

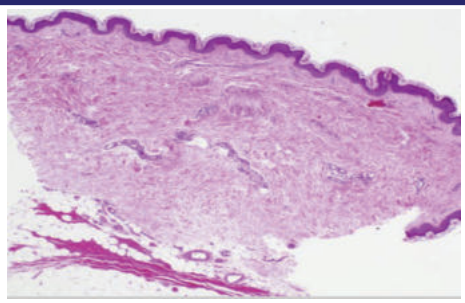
a 22 year old male presented to the out patient department with complaints of dark thick hard linear depressed skin lesion since adolescence. On examination, a purplish brown hyperpigmented linear atrophic band of plaque was seen over the left sub mental and sub mandibulae region of the face. Dermoscopy showed multiple light brown networks with unclear margin.





Laboratory investigations including complete blood count, erythrocyte sedimentation rate, renal profile, liver function test, antinuclear antibodies, double-stranded antinuclear DNA antibodies, anti–SCL-70 antibody, anti-SSA(Ro), anti-SSB(La), anti–Jo-1, and anti-RNP antibodies were all negative or within the normal range. A 3mm punch biopsy was made and sent for histopathilogical examination.

Histopathologic examination of the affected skin found a mild upper dermal perivascular lymphocytic infiltration. When compared with the adjacent normal skin, the dermal thickness was reduced and the dermal collagen appeared more compact. The sweat glands, pilosebaceous units, and appendages were not affected. Histological confirmation was done and the patient was advised various treatment modalities and expected outcome of results. Its clinical appearance varies and may closely resemble that of atrophoderma of Pasini and Pierini (APP) and linear scleroderma.



CONCLUSION:

So far no effective treatment options have been discovered for LAM. High-dose penicillin, topical steroids, heparin and oral potassium benzoate have all been tried without success. Recently, methotrexate was reported to partially improve the clinical appearance of widespread LAM in a 20 year old female. However, its true effectiveness still remains to be ascertained. In contrast, the lesions of linear scleroderma are routinely treated with systemic steroids and methotrexate with measurable clinical improvement. This therapeutic distinction is important as LAM is a benign self-limited disorder, confined to the skin, with mainly cosmetic concern whereas linear scleroderma may extend beyond the skin and affect the underlying muscle and bone. Consequently, unlike linear scleroderma, LAM lesions that overly joints are not worrisome as they do not lead to joint contractures due to the lack of sclerosis and subsequent tightening of overlying tissues.

DISCUSSION:

Although different from the original report of Moulin et al, the most common histologic finding is a perivascular lymphocytic inflammatory infiltrate in the superficial dermis combined with abnormal collagen fibers.

The etiology of LAM remains unknown. All reported cases were so far sporadic. Dermatoses that follow Blaschko's lines are thought to be caused by a somatic mutation that takes place early in embryogenesis, resulting in a genotypic and phenotypic mosaicism. Danarti et al postulated that LAM may reflect the action of an autosomal lethal gene surviving by mosaicism. A postzygotic mutational event may lead to loss of the corresponding wild-type allele at the atrophoderma locus, and this gives rise to a homozygous cell clone which becomes

manifested along the lines of Blaschko later in life.

The differential diagnosis of LAM includes congenital dermopathy following Blaschko lines such as linear and whorled nevoid hypermelanosis, incontinentia pigmenti, lichen striatus, and epidermal nevi. LAM should also be differentiated from atrophoderma of Pasini and Pierini, which also presents with similar configuration, atrophy, and hyperpigmentation but does not follow Blaschko lines. Atrophoderma of Pasini and Pierini is considered an abortive variant of morphea. In our patient, the initial clinical diagnosis was linear morphea. However, the clinical presentation, absence of preceding inflammation, induration, or scleroderma and absence of dermal sclerosis on histopathologic examination, led to a diagnosis of LAM. It is important to differentiate LAM from linear morphea, as the prognosis is different. The prognosis of LAM is favorable, as there are no associated complications.

There is no effective treatment for LAM. Topical corticosteroids and heparin were not helpful. High dose penicillin and potassium aminobenzoate were noted to be ineffective as well. A partial response to topical calcipotriol was reported. A case of LAM successfully treated with methotrexate 20 mg/wk for 6 months with an improvement of pigmentation and atrophy was reported. The authors suggested that LAM, atrophoderma of Pasini and Pierini, and linear scleroderma may be a spectrum of a common disease entity in view of the response to methotrexate, which is an effective therapeutic option for morphea. However, some cases of LAM do not respond to potent topical steroids and topical calcipotriol, which are the first-line treatment for morphea.

Since the original description, much debate has occurred regarding whether atrophoderma of Pasini and Pierini is a distinct entity or an abortive, nonindurated variant of morphea. Subsequent reports of the co-occurrence of morphea and occasionally lichen sclerosus with atrophoderma of Pasini and Pierini suggest a close relationship between atrophoderma of Pasini and Pierini and morphea. However, Yokoyama et al reported that skin glycosaminoglycans extracted from atrophoderma of Pasini and Pierini lesions are different from those in typical morphea lesions.

It remains controversial whether LAM is a distinct entity or on a spectrum of a common disease entity with atrophoderma of Pasini and Pierini and linear scleroderma. Although some similarities exist between these 3 conditions, there are also differences in the age of onset, distribution, histology, origin, development, and prognosis of the lesions.

This case highlights the importance of recognizing the clinical presentation of this disease with atrophic hyperpigmented patches along Blaschko lines. When atrophoderma is suspected, skin biopsies should be taken from both normal skin and lesional skin, as atrophoderma often shows very subtle features.

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