Linear Atrophoderma of Moulin over Face: An Exceedingly Rare Entity

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Sir,

A 16-year-old girl presented to us with asymptomatic hyperpigmented lesions over the left side of her chin since the past 6 months. According to the patient, it started as a small black discoloration of the skin which gradually increased in size. There was no history of preceding trauma, redness or tightness of the skin, associated systemic complaints, or family history of similar illness. Cutaneous examination revealed three broad unilateral linear hyperpigmented atrophic lesions, with depressed margins along Blaschko’s lines without any sign of inflammation or induration [Figure 1a and b]. The surface of the atrophic lesions was wrinkled. A 4 mm punch biopsy was taken from the margin of the lesion. Histopathologic examination (HPE) with hematoxylin and eosin staining showed epidermal atrophy along with dense melanin deposition along the basal layer with apparently normal subcutaneous tissue [Figure 2]. Sparse perivascular and periappendageal lymphocytic infiltrate with slight thickening of collagen bundles was present in the dermis. There was no evidence of sclerosis or atrophy of the appendages [Figure 3]. The difference with the normal epidermis could be seen in the HPE [Figure 4]. Verhoeff–van Gieson stain showed normal elastic tissue [Figure 5]. On the basis of the clinical and histopathologic findings, we diagnosed the case as linear atrophoderma of moulin (LAM).

LAM is a rare dermatosis characterized by a hyperpigmented atrophoderma that follows Blaschko’s lines with onset usually during childhood and adolescence.\(^1\)
Postzygotic mutation in lamin A gene has been postulated as a possibility for developing this disorder.\(^2\) The trunk and limbs are usually involved without any preceding inflammation or subsequent induration and sclerosis.\(^3\) Presentation over the face, as was seen in our case, is rarely reported in literature. The clinical atrophy is reported to be induced by a reduction of subcutaneous tissue but not dermal tissue as observed by ultrasound imaging.\(^4\) However, subcutaneous tissue is not routinely mentioned to be reduced according to other reports and text.\(^1,2\) LAM is a self-limited disease. Progression of the lesions usually stops within a few months without any pattern of remission.\(^1\) Hematoxylin and eosin staining usually show normal or atrophic epidermis, along with hyperpigmentation of the basal layer. The collagen bundles in the mid and deep dermis may be edematous or slightly homogenized in appearance. Although elastic fibers are usually normal, occasionally some clumping and loss of fibers may be seen in the deep dermis. Adnexal structures are usually preserved. Mild perivascular infiltrate is seen in the upper dermis and somewhat heavier in the deep dermis consisting of lymphocytes and a few macrophages and rarely, plasma cells. Some superficial vessels may be mildly dilated. There may be a few melanophages in the superficial dermis.\(^2\)

The differential diagnosis includes atrophoderma of Pasini and Pierini, linear scleroderma, and porokeratosis. Clinically, the preceding inflammation, sclerosis, and induration that accompany linear scleroderma are usually absent in LAM. Histopathology of the former reveals thickened and closely packed collagen bundles with atrophic eccrine glands, hair follicles, and periappendageal fat. Atrophoderma of Pasini and Pierini clinically and histologically resembles LAM except that it does not follow Blaschko’s lines.\(^5\) Porokeratosis was excluded clinically and histopathologically by the absence of keratotic ridge with central groove and cornoid lamella, respectively.\(^2\)

Effective treatment with methotrexate 20 mg/week for a duration of 6 months has been reported.\(^6\) Due to esthetic nature of the disorder, use of self-tanning cream has been advised.\(^4\) The partial improvement was reported with topical calcipotriol, intravenous penicillin together with topical PUVA, oral Potaba (potassium para-aminobenzoate), high-dose Vitamin E (400 IU/day), and topical clobetasol propionate.\(^1\) Since none of these medications are uniformly effective for this rare dermatosis, especially over face, we started topical tretinoin (0.05%) cream empirically once at night. If we do not see any appreciable improvement after 3 months, we would consider starting methotrexate. We report this case of LAM because of paucity of its clinical and histopathological description in the literature, especially from India.

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**Conflicts of interest**
There are no conflicts of interest.

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Hemifacial Microsomia and Accessory Auricles in an Adolescent Boy

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Sir,

Goldenhar syndrome is a rare disorder with developmental defects involving the first and second branchial arches being named in the Online Mendelian Inheritance of Man (OMIM) as "hemifacial microsomia" also known as "oculoauriculovertebral syndrome or spectrum."

A 17-year-old boy presented with multiple bilateral preauricular nodules since birth and swelling in the left eye since childhood which was progressively increasing in size, obscuring the vision. His hearing was normal. There was no maternal illness during pregnancy. On examination, he had epibulbar dermoid in the left eye [Figure 1a] and showed high-arched palate [Figure 1b] and multiple accessory auricles [Figure 1c and d]. Linear and rectangular atrophic macules were seen over cheeks [Figure 1c and d]. The atrophic macules appeared spontaneously without any history of inflammation or trauma. Flattening of the face was noted on the left side with micrognathia and macrostomia [Figure 2]. Audiogram was normal.

X-ray and computed tomography scan of the cervical spine showed block vertebra between C5, C6 level [Figure 3a].

X-ray of the thoracolumbar spine showed hemivertebra at L5 level with left-sided lumbar scoliosis [Figure 3b].

There were no cardiac anomalies, and ultrasonogram of the abdomen was normal.

"Oculoauriculovertebral spectrum" was described by Goldenhar in 1952 with the triad of ear defects, epibulbar dermoids, and vertebral anomalies. [1]

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