Extragenital lichen sclerosus et atrophicus along the lines of Blaschko

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ABSTRACT

Lichen sclerosus et atrophicus (LSA) is a chronic inflammatory dermatosis of unknown etiology. Extragenital involvement is uncommon and commonly affects the neck, shoulders, and upper portion of the trunk. It is predominant in women with a male-to-female ratio of 1:6 and occurs at any age. Linear pattern along the lines of Blaschko are seen. There is no cure for LSA. Topical corticosteroids and calcineurin inhibitors, such as tacrolimus, pimecrolimus, PUVA antimalarial agents, and topical retinoids have been tried with varying results. A case of a 33-year-old man with LSA over right lower limb along the lines of Blasckho is reported here.

Key words: Extragenital sites, lichen sclerosus et atrophicus, lines of Blaschko

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INTRODUCTION

Lichen sclerosus et atrophicus (LSA) is a rare chronic inflammatory dermatosis with anogenital and extragenital involvement. Extragenital lichen sclerosus is most common on the neck, shoulders, and upper portion of the trunk. It is generally asymptomatic or is occasionally pruritic and presents as flat, white, polygonal papules and slight atrophic white plaques.[1] Linear lesions are uncommon in LSA and very few such cases are reported.[2] There is no known cure for LSA. Standard treatment includes topical corticosteroid and calcineurin inhibitors, such as tacrolimus. We report a case of 33-year-old man with LSA over right lower limb along the lines of Blasckho.

CASE REPORT

A 33-year-old man an industry worker presented with complaints of painful and itchy skin lesions over his right foot, leg, and thigh since six months. Lesions initially appeared over the right foot and gradually progressed upwards to involve the leg and thigh. There was no significant past or family history. On examination, a linear, hypopigmented, atrophic plaque with follicular plugging was noted over the right foot and leg, extending on to the anterolateral aspect of right thigh [Figure 1a-c]. Hemogram, liver, renal, and thyroid function tests were normal. Skin biopsy was drawn from the thigh keeping lichen striatus, localized morphea, and LSA as differentials. Histopathology showed thinning of the epidermis with hyperkeratosis and follicular plugging, with the basal layer showing hydropic degeneration. The upper dermis showed homogenization of the collagen and edema with mild inflammatory infiltrate [Figure 2] consistent with a diagnosis of LSA. The patient was advised topical clobetasol propionate (0.05%) and tacrolimus (0.03%) cream along with a moisturizer, and narrow band UVB phototherapy twice a week. After 4 weeks of phototherapy, lesions regressed dramatically in the form of symptoms and morphology.

DISCUSSION

Lichen sclerosus (LS) originally described by Hallopeau in 1887, is a relatively rare chronic inflammatory dermatosis of unknown etiology. The association of specific HLA types and other autoimmune diseases suggests that LS is an autoimmune process.[3] Recently, immunoreactivity to extracellular matrix protein 1 has been demonstrated in up to 74% of cases.[4]

LS both genital and extragenital has no known racial predilection. A genetic predisposition, based on familial clustering was observed.[5] The exact prevalence is not known. It predominates in women with male-to-female ratio of 1:6 and

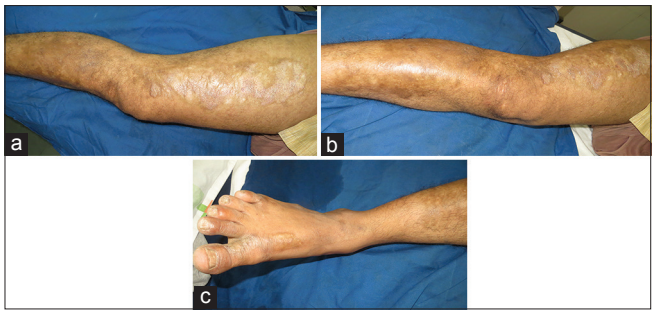

Figure 1: Hypopigmented atrophic plaque with follicular plugging in linear distribution over right side of (a) thigh, (b) leg, (c) foot

occurs at any age. However, the maximum incidence occurs between the 5th and 6th decade of life and there is a second peak in girls between the age of 8 and 13 years.[6] The prevalence of extragenital LSA may be underestimated because it is often asymptomatic.

Extragenital lesions occur in 15%-20% of patients.[7] In the first stage, it presents as interfollicular, pearly, polygonal papules, which merge to form atrophic, sclerotic plaques. In more advanced stages, follicular hyperkeratosis and telangiectasias are seen.[8] It occurs on the palms of the hand, soles of the feet, face, scalp, and mouth sometimes distributed following lines of Blaschko.[9]

In disorders that affect skin areas corresponding to lines of Blaschko, it is believed that two distinct cell clones arise early in embryogenesis, often produced by genetic mosaicism. Lichen striatus, linear psoriasis, linear lichen planus, linear scleroderma, and linear atrophoderma are dermatoses seen along the lines of Blaschko.[10] Linear extragenital LS represents an exceptionally rare form of LS.

In 1995, Izumi *et al*. were the first to describe a linear form of LS extending from the left upper back and along the left arm, probably following the lines of Blaschko. Okamoto *et al*. described another case of linear LS in a 23-year-old woman who developed initial lesions at the age of 18 years.[9,11]

The Koebner phenomenon occurs in LS and scarring or trauma may induce typical skin lesions of the disorder, and extragenital lesions commonly occur in pre-existing scars and damaged areas.[7] Lesions preferentially occurring on left side of body in most of the reported cases has been attributed to stronger cell-mediated immune hypersensitivity in the left side of the body than the right in healthy young subjects and it is speculated that the cellular immune responsiveness might influence the confinement of the Blaschko-linear LS to the left side of the body.[1,12] This is in contrast to our case where lesions were on right side of the body.

Histologically, LSA has a characteristic pattern. Hyperkeratosis, follicular occlusion, thinning of the epidermis, and vascular alterations in the basal layer are seen in the epidermis. There

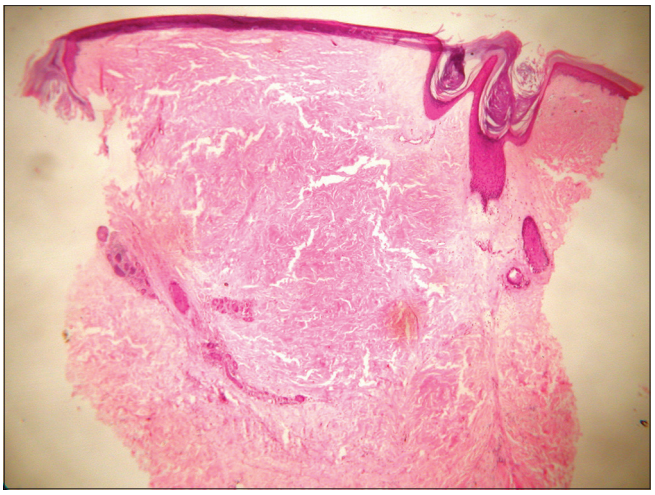

Figure 2: Hyperkeratosis and follicular plugging with basal layer showing hydropic degeneration with homogenization of the collagen and edema with mild inflammatory infiltrate in dermis [H and E x10]

is a large area of subepidermal edema with homogenization of collagen, sclerosis, and dilation of the small vessels with hemorrhage. A diffuse perivascular infiltrate of lymphocytes appears under the edema in the middle third of the dermis.[13] Reticular dermal changes of fibrosis and inflammation in morphea is in contrast with edema and loss of elastic tissue in LS, which helps to differentiate both conditions.

The rate of spontaneous resolution may be lower than 25%.[14] Currently, there is no effective treatment available for LS. Most patients are initially treated with potent topical corticosteroids. Various treatments including PUVA,[15] topical testosterone and estrogen, topical tacrolimus[16] or pimecrolimus, antimalarial agents, penicillin, topical retinoids, and vitamins, have been tried. NBUVB has both anti-inflammatory and immunosuppressive effects. Several studies have demonstrated that both UVA1 and NBUVB increase matrix-metalloproteinase levels in human skin and cultured dermal fibroblasts, which explains the effectiveness of UVA in sclerosing skin diseases, but LSA affects only the epidermis and superficial dermis, so along with UVA, NBUVB is also effective in LSA.[17]

Our case was unique with unilateral involvement over right side of lower limb along the lines of Blaschko in a male who responded well to NBUVB therapy.

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