

CASE REPORT

EXTRAGENITAL LICHEN SCLEROSUS ET ATROPHICUS MASQUERADING AS DISCOID LUPUS ERYTHEMATOSUS

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ABSTRACT: Lichen sclerosis et atrophicus is rare chronic inflammatory dermatoses of unknown yet multifactorial etiology. The disease tends to affect the anogenital region either alone or in combination with other areas. Extragenital areas are affected in the form of depressed and atrophic, whitish macule or plaques. The lesions tend to affect the upper half of the trunk, neck and upper limbs. We are reporting a case of 45 years old female patient who presented with multiple hypopigmented to depigmented plaques with hyperpigmented border over the anterior, posterior and lateral aspect of both legs since five years associated with moderate grade itching. Dermoscopic examination shows homogenous white areas with mixed blood vessels. Histopathology examination showed features suggestive of lichen sclerosis et atrophicus. This case has been reported for its rarity and atypical presentation.

KEYWORDS: Lichen sclerosis et atrophicus (LSA).

INTRODUCTION: Lichen sclerosus et atrophicus (LSA) also known as Csillag's disease¹ is an uncommon disease of unknown etiology although hereditary, endocrine, and autoimmune factors are known to be involved.^{2,3} It is characterized by small, porcelain white, sclerotic areas occur at any site on the skin including mucosa.⁴ Although the anal and genital regions are predominantly affected, 2.5% of patients only presents with extragenital lesions particularly over the trunk, neck, and upper limbs. The wrists, palmoplantar region, nipple, and face are less commonly involved.^{2,3} It mainly affects post-menopausal women but can occur in young women. F: M varies from 10:1 to 6:1.⁵

Extra-genital LS needs to be distinguished from discoid lupus erythematosus, morphea and atrophic lichen planus.⁶

We are reporting a case of 45 year old female having lichen sclerosus et atrophicus masquerading as discoid lupus erythematosus.

CASE REPORT: A 45 year old female presented with multiple symmetrical lesions over lower extremities since five years, these lesions were progressive in nature associated with moderate grade of itching. There was no history of lesions elsewhere, photosensitivity, muscle weakness or weight loss.

General and systemic examination did not reveal any abnormality.

Dermatological examination showed multiple, discrete, bilateral, hypopigmented to depigmented plaques with hyperpigmented borders over the anterior, posterior and lateral aspect of both legs. [Figure 1, 2 & 3]. There were no genital or oral lesions.

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Investigations showed normal hematocrit, blood sugar, thyroid profile, liver function tests and renal function tests. Peripheral smear showed no sezary cells. ANA, VDRL and antibodies against HIV were negative.

Skin biopsy revealed thinned out epidermis, hyperkeratosis, mild spongiosis and focal parakeratosis. Focal exocytosis and basal vacuolar degeneration seen. Superficial dermis shows prominent dilated blood vessels with homogenization of superficial collagen bundles. A sparse perivascular lymphocytic infiltrate is noted. Thus a diagnosis of extra-genital lichen sclerosis was made. [Figure 4 & 5]

Dermoscopic examination revealed homogenous white areas with mixed blood vessels suggestive of atrophy. [Figure 6]

DISCUSSION: Hallopeau⁷ first describe lichen sclerosus in 1887, and then Darier⁸ reported the histological changes in 1892. They consider the disorder to be a type of lichen planus; other thought that the condition was related to localized scleroderma. But now it is regarded separate entity because of its distinct clinical signs and pathological changes.⁴ Lichen sclerosus is an inflammatory disease with a worldwide prevalence of 0.03%. The underlying cause is unknown with genetic predisposition, trauma, infections, vaccinations, autoimmune, and hormonal factors being described as the most likely causes. Immunogenetic studies have demonstrated links with HLA A29, B44, DQ-7, 8, and 9.⁹ Autoantibodies to glycoprotein extracellular matrix protein 1 have been demonstrated in few cases.¹⁰ Csillag's disease affects anogenital area in 85–98% cases, with additional extragenital lesions being reported in 15–20% of patients.¹¹ Extragenital lesions alone occur in 2.5% and are found usually on the back and shoulders.¹² They are generally asymptomatic and less common presentations include involvement of the palms, soles, infraorbital and scrotal regions, appearance in acrochordons, and scarring alopecia.¹³

Our patient, in addition had hypopigmented atrophic plaques on both legs. These lesions have rarely been described earlier in literature.

Investigations include skin biopsy and autoimmune workup. Histopathology of lichen sclerosus et atrophicus is specific with thin epidermis, basal cell degeneration, upper dermal homogenisation of collagen and mid dermal chronic inflammatory infiltrate, all of which were present in our case.¹⁴

Dermoscopy reveal whitish homogeneous areas associated with presence of yellow circles (comedo-like appearance).¹⁵

First line treatment of lichen sclerosus et atrophicus includes super potent topical corticosteroids which has a high response rate. Second line therapies include topical calcineurin inhibitors. Systemic agents like oral steroids, etretinate and cyclosporine may be of some benefit.¹⁶

NB-UVB can be used in patient having extragenital of lichen sclerosus et atrophicus.¹⁷

Our patient responded well to potent topical corticosteroids and topical emollients.

The diagnosis of Lichen sclerosus et atrophicus was made with the aid of histopathology. This case has been reported for its rarity and atypical presentation.

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Figure 1: Multiple, discrete, bilateral hypopigmented to depigmented plaques with hyperpigmented borders over the anterior aspect of both legs.



Figure 1

Figure 2: Multiple, discrete, hypopigmented to depigmented plaques with hyperpigmented borders over the anterior aspect of right leg.



Figure 2

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Figure 3: Multiple, discrete, hypopigmented to depigmented plaques with hyperpigmented borders over the lateral aspect of left leg.



Figure 3

Figure 4: Microscopy, 10x shows thinned out epidermis and homogenization of collagen in upper dermis.

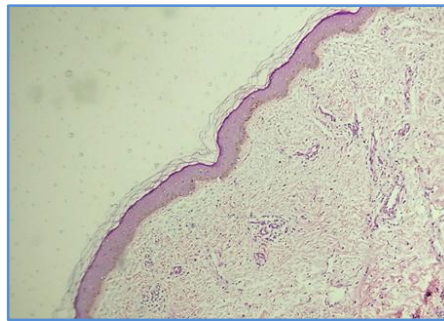


Figure 4

Figure 5: Microscopy, 40x shows basal vacuolar degeneration with homogenization of collagen in upper dermis.

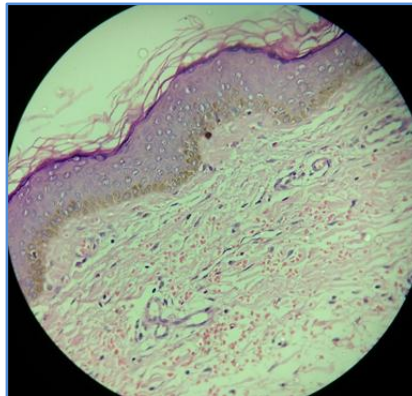


Figure 5

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Figure 6: Dermoscopy- homogenous white areas with mixed blood vessels suggestive of atrophy.



Figure 6

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