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Extragenital Lichen Sclerosus: A Rare Case with Clinical, Dermoscopic, and Histopathological Findings

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Abstract **Case Report**

We report the case of a 65-year-old woman presenting with exclusively extragenital lichen sclerosus, a rare clinical form of the disease. The patient exhibited atrophic whitish plaques and macules on the trunk and limbs, with associated hyperpigmentation. Dermoscopic evaluation revealed an erythematous background, shiny white streaks, dotted vessels, and a pigmented network. Histopathological analysis confirmed the diagnosis, showing an atrophic epidermis, orthokeratotic hyperkeratosis, and a fibrotic dermis with a lymphohistiocytic infiltrate. This case highlights the importance of recognizing extragenital presentations of lichen sclerosus, which may mimic other dermatoses and primarily pose cosmetic concerns, with a low risk of malignant transformation.

Keywords: Extragenital lichen sclerosus, Lichen scleroatrophicus (LSA/LS), Atrophic plaques, Hyperpigmentation, Dermoscopy.

INTRODUCTION

Lichen scleroatrophicus (LSA), also known as lichen sclerosus (LS), is a rare and chronic inflammatory disorder of unknown etiology. Approximately 85% of LS cases involve the genital area, while the extragenital form is observed in only 15–20% of cases. Extragenital lichen scleroatrophicus (ELSA) may occur simultaneously with the genital form; however, in 6% of cases, exclusively extragenital involvement has been reported. We report a case of exclusively extragenital lichen scleroatrophicus.

OBSERVATIONS

Mrs. A.N., a 65-year-old married woman and mother of three, with no significant medical history, presented with diffuse whitish lesions that had been moderately pruritic and evolving over the past three months. The lesions initially appeared on the trunk before progressively extending to the upper and lower limbs, accompanied by the development of cutaneous sclerosis.

Clinical examination revealed pearly white, atrophic plaques with a wrinkled appearance, confluent into large patches in some areas, surrounded by hyperpigmentation, involving the trunk, the anterior aspects of the forearms, and the lower limbs (Figures 1-2).

Dermoscopy showed an erythematous background associated with shiny white streaks, dotted vessels, and a pigmented network in some areas (Figure 3).

The remainder of the clinical examination, including mucocutaneous and general examination, was unremarkable.

Histological analysis revealed an atrophic epidermis with orthokeratotic hyperkeratosis, focal ostiofollicular hyperkeratosis, an edematous papillary dermis, and a fibrotic deep dermis containing a lymphohistiocytic inflammatory infiltrate.

The diagnosis of extragenital lichen scleroatrophicus was established. Biological investigations showed no evidence of systemic disorders, particularly autoimmune diseases.

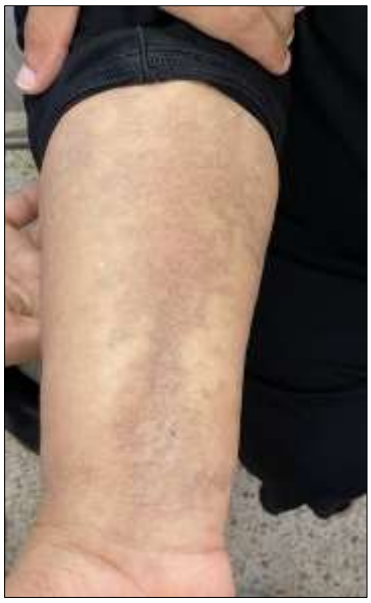

Figure 1: Atrophic whitish plaques surrounded by hyperpigmentation on the anterior aspect of the forearms

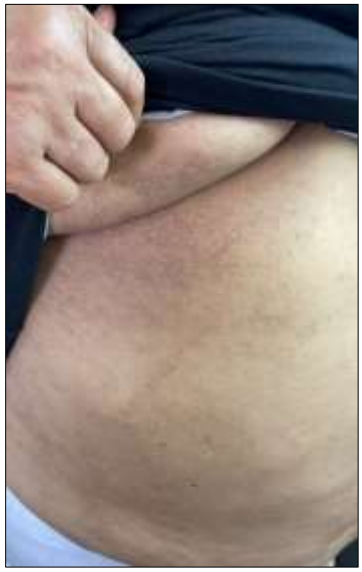

Figure 2: Atrophic whitish macules resembling white spot disease on the trunk

Figure 3: The dermoscopic examination revealed an erythematous background associated with shiny white streaks, dotted vessels, and a pigmented network in some areas

DISCUSSION

First described by Hallopeau in 1887, lichen scleroatrophicus (LSA) is a chronic, fibrosing inflammatory dermatosis with a female predominance, primarily affecting the anogenital region (80%) [1]. Purely extragenital involvement occurs in only 2.5% of cases [2].

Although the exact etiology of the disease remains unknown, its association with autoimmune disorders (such as Hashimoto's thyroiditis, vitiligo, and type 1 diabetes) suggests a possible autoimmune component [3]. However, no such association was observed in our patient. Several other factors, including genetic predisposition, low androgen levels, chronic infections, and trauma (Koebner phenomenon), have also been proposed as pathogenic contributors [4, 5].

Clinically, lesions are mainly located on the trunk, the proximal parts of the limbs, and skin folds [6], which is consistent with the clinical presentation observed in our patient. Unlike genital lichen scleroatrophicus, the extragenital form is generally asymptomatic or mildly symptomatic [7]. The diagnosis is typically based on clinical and dermoscopic examination, and subsequently confirmed by histopathological findings.

CONCLUSION

Lichen sclerosus (LS) is a chronic inflammatory skin disease that typically affects the anogenital region, with isolated cutaneous involvement being less common. Extragenital LS mainly poses a cosmetic concern and rarely leads to malignant transformation.

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