Case for diagnosis

Caso para diagnóstico

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CASE REPORT

A 67-year-old woman, without remarkable medical or family background, presented with a 6-month history of a pruritic sclerotic lesion on her abdomen. The sclerotic lesion had been accompanied by a blister on the back of the right foot for 4 months and there had been no previous trauma. Physical examination revealed several 2-4 cm, ivory colored, shiny, sclerotic lesion on the abdomen (Figure 1) and soft plaque on the back of the right foot (Figure 2). There were no other lesions on her body. An incisional biopsy specimen revealed hyperkeratosis, atrophy of the epidermis, marked edema in the upper dermis resulting in a subepidermal blister and homogenization of collagen in the papillary dermis (Figure 3).

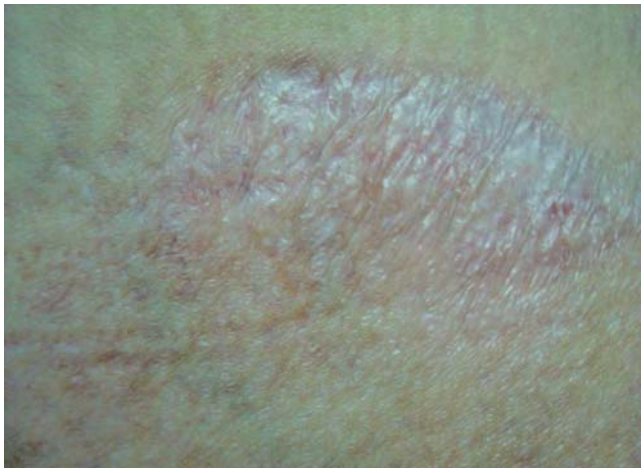

FIGURE 1: Ivory colored, shiny, sclerotic lesion on the abdomen

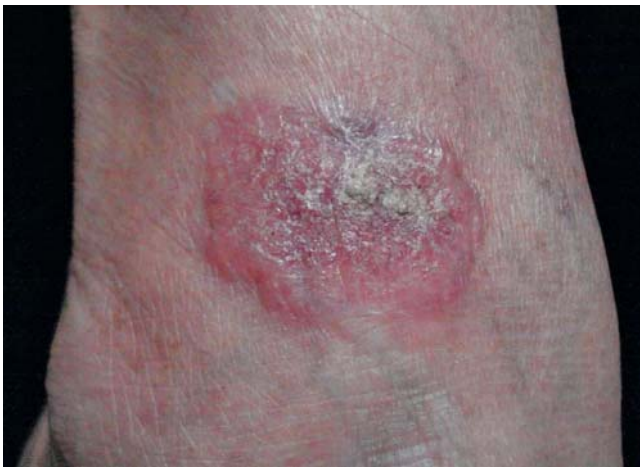

FIGURE 2: Blister on the back of the right foot

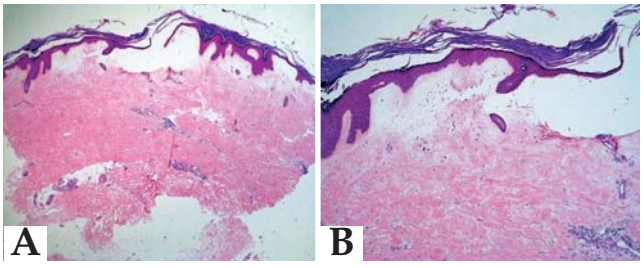

FIGURE 3: (A) Skin biopsy showing hyperkeratosis, atrophy of the epidermis, marked edema in the upper dermis resulting in a subepidermal blister and homogenization of collagen in the papillary dermis (HE 40x). (B) On detail, the marked edema in the upper dermis resulting in a subepidermal blister

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After the diagnosis of Bullous lichen sclerosus et atrophicus was made, we started treatment with topical corticosteroid and achieved stabilization and slight improvement.

DISCUSSION

The first clinical description of lichen sclerosus (LSA) as an inflammatory dermatosis commonly affecting the female genitalia was given by Hallopeau. [1] Darier gave the first account of its histological features in 1892. Lichen sclerosus may affect all areas of the body, but it is most frequently found in the genital area of post-menopausal women. The etiology is unknown but it is believed that genetic susceptibility plays a role in the disease.[2]

Lesions are white porcelain-like sclerotic commonly involving the female anogenital region. Extragenital lesions with similar morphology may be occasionally present in the absence of genital involvement. Soreness and pruritus are the most usual symptoms found.

Bullous lichen sclerosus et atrophicus (BLSA) is an uncommon form of the disease and the exact etiology and prevalence is uncertain. Bullous formation is observed more frequently in the extragenital area than in the genital one.

Several reports about the mechanisms of blister formation suggest that BLSA results from extensive vacuolar degeneration of the epidermal basal layer and edema in the upper dermis. It is often accompanied by disruption and loss of collagen support of the dermal capillaries, specially type VII collagen, that may result in haemorrhage within the bullae, but the pathogenesis is still controversial.[1] Some studies also have suggested that the findings of follicular keratosis and blister formation are very important in the diagnosis of extragenital LSA.

The differential diagnosis includes bullous pemphigoid, epidermolysis bullosa acquisita and traumatic blisters.

Numerous therapeutic modalities have been used in LSA and BLSA, including topical and systemic corticosteroid, testosterone and other hormonal treatments, topical calcineurin inhibitors, topical and systemic retinoids, ciclosporin, methotrexate and other immunosuppressive agents.[3] The bullous lesions are usually transient and heal before onset of typical plaques of the disease, which are more resistant to treatment.[4] ❑

Abstract: Lichen sclerosus is a chronic inflammatory mucocutaneous disorder of unknown etiology that most commonly affects the female genitalia. Cutaneous involvement with nonhaemorrhagic bullous is very unusual. We describe a case of bullous lichen sclerosus.
Keywords: Blister; Chronic disease; Lichen sclerosus et atrophicus

Resumo: Líquen escleroso é uma doença mucocutânea inflamatória, crônica, de etiologia desconhecida, que afeta mais comumente a genitália feminina. O envolvimento cutâneo com bolhas não hemorrágicas é muito raro. Descrevemos um caso de líquen escleroso bolhoso.
Palavras-chave: Doença crônica; Líquen escleroso e atrófico; Vesícula

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