

Acral lichen sclerosus et atrophicus - Case report

Líquen escleroso e atrófico acral - Relato de caso

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Abstract: Lichen sclerosus et atrophicus is a chronic disorder of the skin and mucosal surfaces, most commonly affecting the female genitalia. Few cases of lichen sclerosus et atrophicus confined to the feet and/or hands have been reported. We report a case of lichen sclerosus et atrophicus involving first the extremities and then the perigenital area.

Keywords: Sclerosis; Lichen sclerosus et atrophicus; Foot

Resumo: O líquen escleroso e atrófico é uma doença crônica da pele e mucosas, que, usualmente, afeta a área genital de mulheres. Poucos casos de líquen escleroso e atrófico, confinados nos pés e/ou mãos, têm sido relatados. Relatamos um caso de líquen escleroso e atrófico envolvendo as extremidades e, posteriormente, a área perigenital.

Palavras-chave: Esclerose; Líquen escleroso e atrófico; Pé

INTRODUCTION

Lichen sclerosus et atrophicus (LEA) is an uncommon chronic inflammatory disease which particularly affects the genital area of adult women.^{1,2,3} Extragenital forms of the disease are rare, mainly affecting the thoracic region.² We describe a rare case of extragenital LEA which appeared initially in the palmoplantar region.

CASE REPORT

57-year-old female patient, born and resident in Belém, complaining of clear spots on the soles of the feet, accompanied by localized burning, which emerged two years previously. Patient had tried several topical medications such as desonide, calcipotriol, pimecrolimus and clobetasol oral prednisone, without improvement. A dermatological examination

showed the presence of hypochromic papules located on the soles of the feet and also bilaterally in the paranasal sinuses (Figures 1 and 2). Some of the papules were isolated, while others were grouped in plaques, with areas of scaling. Similar lesions could also be seen in smaller numbers on the palms. The disorder was diagnosed as Flat Wart and a biopsy of the lesion was performed. Histopathology revealed epidermal hyperkeratosis, flattened interpapillary crests, scaling of interface areas, a band-like lymphocytic inflammatory infiltrate on the upper dermis and homogenization of the connective tissue with dilated vessels separating the epidermis from the infiltrate (Figure 3). Given that these findings were consistent with LEA, we prescribed tacrolimus 0.1% to be applied in the mornings and clobetasol cream at night. The

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FIGURE 1: Hypochromic papules, some isolated, others in groups of plaques with areas of scaling, located on the soles of the feet and including the bilateral paranasal sinuses

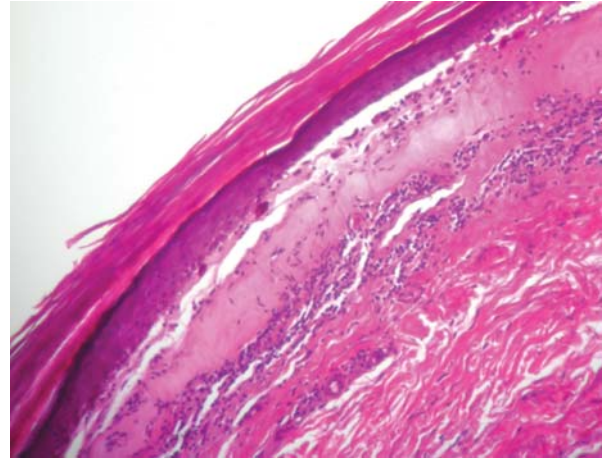


FIGURE 3: Epidermis with hyperkeratosis, flattened interpapillary crests and scaling of interface areas. Band-like lymphocytic inflammatory infiltrate on the upper dermis and homogenization of the connective tissue with dilated vessels separating the epidermis from the infiltrate



FIGURE 2: Hypochromic papules, some isolated, others in groups of plaques with areas of scaling, located on the soles of the feet and including the paranasal sinuses bilaterally



FIGURE 4: Erythematous-hypopigmented lesion with shiny surface, chaffed, well-defined, of varying sizes in groin region E

patient returned three months later showing improved appearance of the lesions on the feet and less burning, but presenting erythematous, hypopigmented, shiny, well-defined, chaffed lesions of varying sizes in the inguinal region (Figure 4). We decided to biopsy the new lesions, the results of which were consistent with LEA.

DISCUSSION

LEA is a chronic, benign and uncommon inflammatory disease. It is more prevalent in females (ratio of 6-10 women to one man).^{1,2} Higher incidence

has been observed in white women aged around 40. The etiology is unknown but it is thought that genetic susceptibility plays a role in the disease. LEA is however closely related to autoimmune diseases and immunogenetic studies have revealed a link with HLA-DQ7.^{4,5,6} Some authors have advanced the theory that the LEA and scleroderma could be facets of the same disease, but the subject remains controversial.⁷ It occurs predominantly in the genital area (in 83-98% of all cases), with extragenital lesions occurring in between 8% to 20% of cases.^{1,2,3,8}

The genital form of the disease in women is

called *craurose vulvar*. This manifests as extremely puriginous lesions forming painful erosions, vulvar atrophy with progressive narrowing of the vaginal opening and dispaurenia.^{1,3} In men, the genital form is known as *balanitis obliterans xerotica*, presenting as painful itching, erosions and fissures.¹ LEA presents clinically as macules, papules and shiny white plaques with follicular corneal plugs. The extragenital forms focus on the upper anterior torso, neck, arms and legs, ear, face, oral mucosa and nose.^{2,9} Other even more unusual locations are the palmoplantar region, nipples and scalp.^{1,10} LEA may be asymptomatic or cause itching and burning at the site.² Diagnosis is clinical and histopathological. Treatment is with high-potency topical steroids, immunomodulators, systemic retinoids, estrogen and progesterone.^{1,3,11} Other options include cryosurgery, vulvectomy, CO2 laser, but these have high recurrence rates.³ Our case of extragenital LEA, involving the palmoplantar region and proceeding to the groin area, is extremely rare. Only one other case has been reported in the literature.¹⁰ □

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