Extra-genital lichen sclero-atrophic

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Abstract

Sclero-Atrophic Lichen (SAL) is a chronic inflammatory dermatosis of unknown etiology that presents with severe pruritus, epidermal atrophy and dermal sclerosis affecting predominantly the anogenital area of post-menopausal females. Extranagenital involvement is uncommon and commonly affects the neck, shoulders and upper portion of the trunk, and is generally asymptomatic. There is no cure for LSA. Topical corticosteroids and calcineurin inhibitors, such as tacrolimus, pimecrolimus, topical retinoids, antimalarial agents, Narrowband UVB and PUVA have been tried with varying results. We report a new observation of sclero-atrophic lichen purely extra genital.

Keywords: Sclero-atrophic lichen; Extra genital; Topical corticosteroids; Narrowband UVB

Introduction

Sclero-Atrophic Lichen (LSA) is a rare, chronic, inflammatory disease of unknown etiology. It mainly affects post-menopausal women in the genital area. In 15-20% of cases extragenital involvement is associated with classic genital LSA and only in 2.5% cases it is found exclusively at an extragenital site [1]. Skin localization is rarely reported in the literature. We report a new observation of sclero-atrophic lichen purely extra genital.

Observation

This is a patient aged 18 years, with no pathological history, who had for 6 years, pigmented lesions slightly pruriginous, gradually increasing in size. Clinical examination showed well-defined pigmented macules, hypo-pigmented and sclerotic in the center, with a highly pigmented border, sitting on the trunk, lower back, upper and lower limbs (Figure 1), skin surface=12%. It had no mucosal involvement. The rest of the clinical examination was normal. This clinical aspect allowed us to evoke the morphea in plaque, the LSA and the sarcoidosis in plaque. However, histology showed an epidermal atrophy associated to basal vacuolization and moderate dermal fibrosis with lymphocytic infiltrate orienting to the diagnostic of LSA cutane. The patient was treated with very strong class of local corticoids for inflammatory and progressive lesions, and topical tacrolimus for atrophic lesions, associated to Narrowband UVB (NBUVB) and we had noted partial improvement of lesions and disappearance of pruritus.

Discussion

First described clinically by Hallopeau in 1887 and histopathologically by Darier in 1892 [2], Lichen Sclerosus (LS) or Sclero-Atrophic (LSA) is a fibrosing inflammatory dermatosis of chronic evolution, affecting mainly the anogenital region (80%). Purely extra-genital localization is only seen in 2.5% of cases [3]. Both sexes are affected with a predilection to females and may occur at any age. However, the maximum incidence occurs between the 5th and 6th decade of life [1,4]. Unlike genital LSA, extra-genital LSA is rarely complicated by malignant transformation. This could be explained by the decrease in the expression of the marker Ki 67 and p53 during the LSA.

The etiology is unknown. Although, there is an association with autoimmune diseases, including thyroid disorders, vitiligo, alopecia areata, and type I diabetes suggests that is an autoimmune process. Several other factors including genetic susceptibility, low levels of androgens, chronic infections, and trauma have been implicated as pathogenic factors [4,5].

Clinically, the lesions are ivory-white or pearly white, “porcelain”, atrophic, plaques, particularly in the trunk, the root of the limbs and the folds. Pruritus is inconstant. Blaschkoinectic and bullous clinical forms have been described. Lichenification may occur secondarily as a result of rubbing. The diagnosis is based on the cutaneous histology, which reveals atrophy of the squamous epithelium with basal horizontalization, follicular hyperkeratosis, and especially the presence of an epithelial band made of fibrous or oedematous collagen in the superficial dermis, devoid of elastic fibers with orceine staining. [1,4,6] Genital SAL leads to progressive atrophy and destructive scarring resulting in dryness, severe pruritus, pain, and often functional impairment. Exogenous LSA is generally asymptomatic [4].

The clinical distinction between LS and morphea in plaque is very difficult, only the presence of typical genital lesions can confirm the diagnosis of lichen sclerosis whose association has a morphea is likely. Histologically, in morphea, there is diffuse sclerosis of the dermis with horizontalization and thickening of the collagen bundles, atrophy of the appendages and disappearance of fatty lobules normally located around the sweat glands [7].

The rate of spontaneous resolution may be lower than 25%. Although there is no definitive or satisfactory treatment for LSA, ultra-potent topical corticosteroids remains the treatment of choice. Therapy for extragenital LSA is warranted only for extensive lesions, bullous and haemorrhagic lesions, symptomatic and cosmetically disfiguring lesions [1,4,5]. Numerous therapies have been used including topical and systemic corticosteroids, topical estrogen and testosterone containing ointments, retinoids, tacrolimus and phototherapy [1]. Several studies have demonstrated that both UVA1 and NB UVB 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, NB UVB is also effective in LSA [5].

In our case, the patient had multiple lesions, hence the combination of local corticosteroids and NB UVB for better synergy. There was a partial improvement with disappearance of pruritus with a follow-up of 3 months.

Conclusion

LSA is a chronic inflammatory dermatosis that usually affects the genital mucosa. Isolated skin involvement is rare and its treatment is not codified. We think that the treatment must be individualized according to the atrophic or sclerotic aspect of the lesions, the extent and the aesthetic gene. Patients should be monitored for the onset of genital involvement that should be treated early in the pruritus stage to prevent sequellar and degenerative complications.

References