

## Lichen sclerosus et atrophicus – report of two cases with atypical presentations\*

### *Líquen escleroso e atrófico – relato de dois casos de apresentação atípica\**

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**Abstract:** Lichen sclerosus et atrophicus is a chronic skin disease with multiple clinical presentations. It affects women, and less frequently men and children. The disease is usually located in the anal and genital regions. There is a strong association with autoimmune diseases, and immunogenetic studies demonstrated an association with HLA DQ7. The disease rarely affects extra-genital regions. The authors report two cases of lichen sclerosus et atrophicus in atypical locations (extra-genital), in which different management was used. High potency topical corticosteroid therapy is reported, with risks, however. Topical immunomodulating agents are described, obtaining favorable clinical responses and decreased risk of atrophy.

**Keywords:** Atrophy; Lichen sclerosus et atrophicus; Scleroderma, localized

**Resumo:** O líquen escleroso e atrófico é doença crônica da pele que se apresenta clinicamente de forma variada. Pode acometer mulheres e, menos frequentemente, homens e crianças, sendo a área mais afetada a região anogenital. Há forte associação com doenças auto-imunes, e estudos imunogenéticos têm revelado associação com o HLA DQ7. Raramente acomete áreas extragenitais; entretanto, os autores relatam dois casos da doença, com localizações atípicas (extragenitais) e fazem uma abordagem terapêutica distinta. O uso tópico de corticoterapia de alta potência é relatado, porém, com riscos; enquanto agentes imunomoduladores tópicos são descritos com boa resposta clínica e com menor risco de atrofia.

**Palavras-chave:** Atrofia; Esclerodermia localizada; Líquen escleroso e atrófico

## INTRODUCTION

Lichen sclerosus et atrophicus, described originally by Hallopeau, in 1887,<sup>1,6</sup> is an infrequent benign chronic inflammatory dermatosis affecting both the epidermis and the dermis.<sup>1</sup> Typical findings are white-opalescent papules that may cluster and progressively result in parchment-like skin.<sup>1,3</sup>

Lichen sclerosus et atrophicus is relatively uncommon in adult women, rare in men and girls

and extremely rare in boys.<sup>2</sup> Women are more commonly affected than men in a 6 to 10 ratio.<sup>3,7</sup> The disease is more frequent in Caucasians.<sup>3</sup>

This disease may occur at any age, but there is a peak in prepuberal children, women after menopause and men with an average age of 43 years.<sup>2</sup>

The etiology of lichen sclerosus et atrophicus is still unknown, although evidence suggests a multifac-

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torial disease.<sup>2</sup> Data suggest hormonal<sup>2</sup> and immunological<sup>3,4</sup> alterations related to disease development, such as: disease preference for females, post-menopausal incidence, regression in some cases during puberty, preferential location in the genitals, good response to topical testosterone<sup>2</sup> and reduction of dihydroepiandrosterone, free testosterone and androstenedione levels.<sup>5,7,8</sup> Familial occurrence suggests that genetic factors may also be involved.<sup>2,5,8</sup> The most common associated diseases are alopecia areata and vitiligo,<sup>4,7</sup> however thyroid dysfunction, pernicious anemia, scarring pemphigoid and *diabetes mellitus* were also reported.<sup>2,5,7,8</sup>

Its pathogenesis is not well known, and human papillomavirus has been implied. Aberrant expression of the P53 gene was demonstrated in immunocytochemical tests of genital lesions.<sup>3,7</sup>

Lichen sclerosus et atrophicus occurs mostly in the anogenital area (83% to 98%),<sup>1,3</sup> with extra-genital lesions in 15% to 20% of patients.<sup>7</sup>

The most frequent symptoms in women are pruritus, local irritation, dysuria, dyspareunia, pain on defecation and fissures.<sup>2,4,7,8</sup> Anatomical structures most frequently involved, in order of frequency, are the clitoris, labia minora, vaginal introitus and labia majora.<sup>2,7</sup> Lesions are white, erythematous, opaque, opalescent, located on the inner aspect of the labia majora and vulvar vestibule, with frequent extension to the perineum and the perianal region, in a typical shape of an "eight",<sup>2</sup> in this case the clitoris and the labia majora and minora disappear completely and there is stenosis of the vaginal introitus, a condition named vulvar craurosis.<sup>2,7</sup>

Lichen sclerosus et atrophicus is less frequent in men and is characterized by isolated atrophic papules on the glans and prepuce, with local whitish lesions,<sup>2,6</sup> vesicles and hemorrhagic bullae. The external urethral meatus may undergo gradual narrowing,<sup>2</sup> with local pain and obstruction of the urinary meatus, dysethesia, impaired penis sensitivity and phimosis.<sup>1,3,4</sup> The diseases usually affects the glans and the prepuce, rarely the body of the penis and never the perianal region. The most frequent symptom is difficulty in retracting the prepuce.<sup>6</sup>

The incidence of extragenital disease in women varies between 8% and 20%. Lesions are maculas or atrophic papules with a "cigarette paper"<sup>3,7</sup> aspect, located on the upper anterior torso, neck, forearms, dorsal region, ears, mouth and nose.<sup>2,4,7,8</sup> Atypical location would be the palmar and plantar regions, nipples, scalp, vaccination sites and face, when the differential diagnosis should be made with discoid lupus and scleroderma circumscripta.<sup>1,2,9</sup> Disseminated forms of the disease are extremely rare.<sup>4</sup>

The treatment of lichen sclerosus et atrophicus includes symptom control,<sup>7</sup> prevention and treatment of complications, and early diagnosis of malignant lesions.

Topical corticosteroids<sup>2</sup> are used on extragenital lesions. Recent studies have described moderate responses with PUVAtherapy. Topical use of tacrolimus is now being proposed, with the promise of being a highly effective drug with few side effects.<sup>2,10</sup>

## CASE REPORTS

### Case 1

A 55-year-old white, married woman presented with grouped, asymptomatic, atrophic and hypochromic skin lesions measuring two to three millimeters in diameter, with a parchment-like aspect, located on the neck, dorsum, abdomen and thighs, starting two years ago. Some of the lesions had superficial corneum spicules (Figures 1, 2 and 3). The presumptive diagnosis was disseminated lichen sclerosus et atrophicus, which was biopsied and sent to the pathologist. Histopathology revealed hyperkeratosis with follicular plugs, spinous layer atrophy, hydropic degeneration of the basal layer and a mononuclear infiltrate in the middle dermis (Figure 4), confirming the clinical diagnosis.

The work-up – complete blood count, glucose, liver function tests, lipid profile, free T4 and TSH – was within normal limits.

Initial treatment was topical corticotherapy (betamethasone dipropionate at night during 30 days). There was partial improvement and the treatment was continued.



FIGURE 1: Patient (case 1) presenting scleroatrophic, opalescent lesions disseminated in the dorsum





FIGURE 2: Detail of lesions of patient (case 1). Observe the atrophic aspect of lesions and central corneum spicules

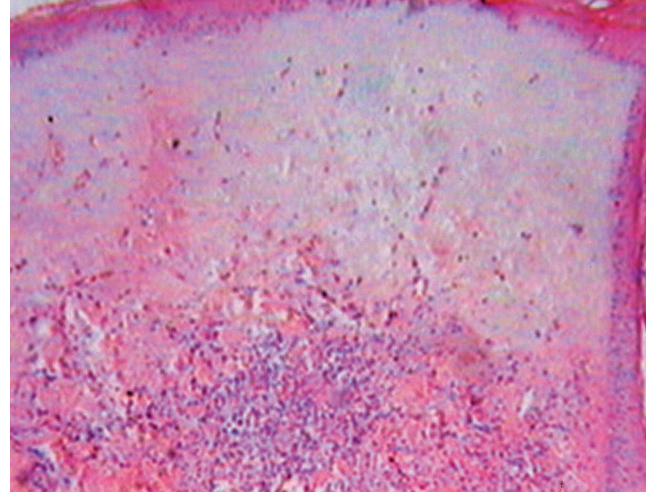


FIGURE 4: Histopathological examination of patient (case 1) showing epidermal atrophy with superficial dermis hyalinization, compatible with lichen sclerosus et atrophicus (HE; 40x)

**Case 2**

A 25-year-old mulatto, married man working as a car body worker, born in and residing in Cariacica (ES) presented with a hypochromic opalescent lesion with slight central atrophy and mild pruritus, distributed linearly and perpendicular to the medial frontal region, measuring approximately five centimeters in length (Figure 5). The differential diagnoses were linear scleroderma (en coup de sabre) and lichen sclerosus et atrophicus. The work-up included a complete blood count, which revealed eosinophilia (16%), the latex test, ANF, VDRL, C-reactive protein, LE cells, free T4 and TSH, which were within normal limits. Histopathology of a biopsy specimen suggested lichen sclerosus et atrophicus.

Initial treatment was topical clobetasol propionate 0.05% for 30 days, but as the condition did not regress, pimecrolimus ointment twice a day was introduced. There was a significant regression of the local atrophy and beginning of repigmentation after 120 days of treatment (Figure 6).

**DISCUSSION**

Lichen sclerosus et atrophicus is an infrequent benign chronic inflammatory dermatosis. It involves mostly white women and men with an average age of 43 years.<sup>2,7</sup> Case 1 corroborated literature data on gender, race and age, but case 2 was a black male patient aged 25 years, an uncommon incidence as compared to literature data.

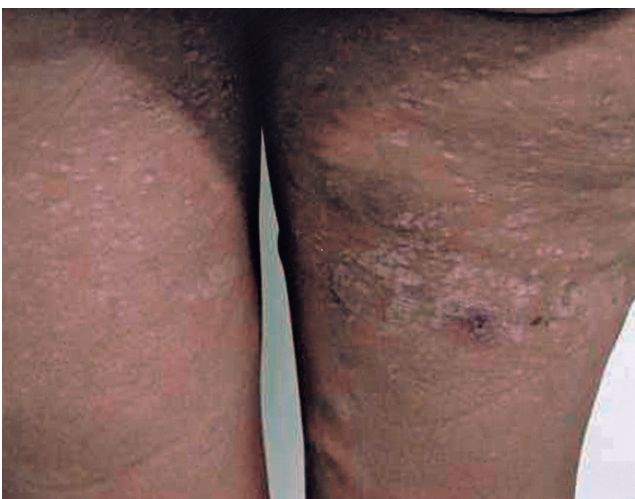


FIGURE 3: Patient (case 1) presenting scleroatrophic lesions disseminated in the proximal thigh region



FIGURE 5: Lichen sclerosus et atrophicus on the face of patient (case 2). Observe facial hypochromic macula, with opalescent, bright aspect, slight central atrophy in the medial frontal region



FIGURE 6: Patient (case 2) with lichen sclerosis et atrophicus on the face after four-month treatment with pimecrolimus cream. Observe macula with matt aspect and discreet central pigmentation islets

The cause of lichen sclerosis et atrophicus remains unknown.<sup>2</sup> According to literature, there is a 21.5% to 34% rate of association between this entity and autoimmune diseases, and 79% of cases had autoantibodies. In 18 familial cases there was an association with HLA B40,8 B44, and AW31.<sup>5,8</sup> In both cases there was no association between lichen sclero-

sus et atrophicus and autoimmune diseases or any family cases.

The disseminated form of lichen sclerosis et atrophicus is poorly described in literature and occurs in 15 to 20% of cases.<sup>1,2,9</sup> The isolated facial lesion was described in few cases and may clinically simulate scleroderma en coup de sabre, requiring careful histopathological study. There is a controversy between clinicians and dermatopathologists on the association between these two diseases and a possible progression of lichen sclerosis et atrophicus to scleroderma. Utitto et al. found 10 patients with lichen sclerosis et atrophicus and a strong connection with morphea, and poor histopathological differential diagnosis between both entities.<sup>1</sup> Thus, case 1 is a rare clinical form of lichen sclerosis et atrophicus and case 2, although initially similar to scleroderma, did not leave room for doubt in the final diagnosis based on the clinical findings and histopathology.

High potency topical corticosteroids are used in the treatment of lichen sclerosis et atrophicus, however prolonged use may worsen local atrophy. In case 1 there was marked improvement with middle potency topical corticosteroids. In case 2 there was significant clinical improvement with partial regression of atrophy and slight repigmentation of the lesions when treated with pimecrolimus. □

## REFERENCES

1. Azevedo LMS, Santos MAL, Pereira Júnior AC. Líquen escleroso e atrófico. *An Bras Dermatol.* 1989;64:179-82.
2. Tasker GL, Wojnarowska F. Lichen Sclerosus. *Clin Exp Dermatol.* 2003;28:128-33.
3. Tremaine R, Adam JE, Orizaga M. Morphea coexisting with lichen sclerosis et atrophicus. *Int J Dermatol.* 1990;29:486-9.
4. García-Bravo B, Sánchez-Pedreño P, Rodriguez-Pichardo A, Camacho F. Lichen sclerosis et atrophicus. A study of 76 cases and their relation to diabetes. *J Am Acad Dermatol.* 1988;19:482-5.
5. Smith SB, Meffert JJ. Lichen Sclerosus: an atypical presentation. *Cutis.* 2002;69:285-6.
6. Tafelkruyer J, Claessens FLE. Lichen Sclerosus et atrophicus and sclerodermia circumscripta. *Proceedings Verhandlungsberichte. Dermatologica.* 1978;156:313-6.
7. Assmann T, Becker-Wegerich P, Grewe M, Megahed M, Ruzicka T. Tacrolimus ointment for the treatment of vulvar Lichen Sclerosus. *J Am Acad Dermatol.* 2003;48:935-7.
8. Shirer JA, Ray MC. Familial occurrence of Lichen sclerosis et atrophicus. *Arch Dermatol.* 1987;123:485-8.
9. Lipscombe TK, Wayte J, Wojnarowska F, Marren P, Luzzi G. A study of clinical and aetiological factors and possible associations of lichen sclerosis in males. *Australas J Dermatol.* 1997;38:132-6.
10. Powell JJ, Wojnarowska F. Lichen sclerosis. *Lancet.* 1999;353:1777-83.

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