

Lichen Sclerosus in the Oral Mucosa: a Rare Form of Presentation

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SUMMARY Lichen sclerosus is a chronic inflammatory disease of unknown origin, which affects mostly women in the fifth and sixth decades of life, but can also occur in men and children. The involvement of the oral mucosa alone or together with other forms of presentation is extremely rare, requiring a differential diagnosis with other diseases of the oral cavity, particularly lichen planus. There are less than 30 cases of lichen sclerosus in the oral mucosa described in the literature and there are no reports on malignant transformation so far. We describe a patient with skin, oral and genital lesions of lichen sclerosus.

KEY WORDS: lichen sclerosus, mouth

INTRODUCTION

Lichen sclerosus (LS) is an uncommon inflammatory disease that affects the skin and sometimes mucosal membranes. It seems to have a predilection for involvement of the genital area, especially in women, as well as of the perianal skin (1-4). Extragenital involvement occurs in 15%-20% of patients, with lesions in the oral mucosa being extremely rare (1,4,5). It was first described in 1887 by Hallopeau, who used the name "lichen planus atrophicus". In 1892, Darier published histopathologic findings of a similar lesion that he called "lichen planus sclerosus". Both lesions were considered a variant of lichen planus. Montgomery and Hill have shown that these two lesions belong to the same disease, considering the term "lichen sclerosus et atrophicus" as the best terminology.

Presently, only lichen sclerosus is used because not all LS lesions present atrophy on histopathologic examination (4,6-10).

Its etiology has not yet been fully clarified but a genetic susceptibility and an autoimmune mechanism seem to be involved. There are described associations with disorders of the thyroid, alopecia areata, vitiligo, pernicious anemia and diabetes mellitus (9-11).

Any age group may be affected, particularly women in pre- or postmenopause. There is also a peak of incidence in girls aged 1-13 years. The occurrence in men is usually at age 30-50 and it is not uncommon to find features of lichen sclerosus in boys with phimosis, thus the incidence in men may possibly be underestimated (5).

Skin lesions are asymptomatic and appear as small stains or plaques, pearly white, with follicular keratosis on the surface, telangiectasia and purplish color being commonly found. Atrophy occurs progressively and the surface of the lesion becomes chafed and may even become depressed. Genital lesions appear as atrophic plaques, shining white, and may present erythema and purpuric lesions. Atrophy is an important characteristic that can lead to tissue loss with fusion of the labia minora and difficult exposure of the clitoris. Vulvar itching and dyspareunia are common complaints. In men there is frequent phimosis of the prepuce, with limited exposure of the glans, ulceration and telangiectasia. Erection can be painful. Sometimes lesions in the oral cavity can be found, which consist mostly of white plaques located on the lower lip or buccal mucosa. They can assume a reticular aspect, difficult to differentiate from lichen planus (1,2,9,12).

Malignant or premalignant transformation may occur in LS patients. In an analysis of squamous cell carcinoma of the vulva, only 4.4% originated from LS lesions (1,4,12,13).

In this article, we describe a woman with lichen sclerosus affecting the skin, oral and genital areas.

CASE REPORT

A 70-year-old female patient, Caucasian, from Rio de Janeiro, presented to Dermatology Clinic of the Clementino Fraga Filho University Hospital, with complaints of asymptomatic spots on the chest that were increasing in number and size, and vulvar pruritus in evolution for 3 months. She had no other complaints. Dermatologic examination showed hypochromic, atrophic plaques, some with follicular keratosis in the inframammary and dorsal area, upper and lower limbs. In the vulvar region, there was an achromic, atrophic lesion with areas of purple and waxy shine, beside fusion of the labia minora and mild phimosis of the foreskin. Oral cavity examination revealed an asymptomatic leukoplakic lesion in the right oral mucosa, tongue dorsum and upper left gingiva (Figs. 1-4).

Incisional biopsies with histopathology were performed on the inframammary lesion area, genitals, right buccal mucosa, tongue and left gingiva. In the inframammary and genital lesion area, atrophic rectified epidermis with vacuolar alteration of basal cells and pigmentary incontinence was observed, associated with areas of collagen hyalinization, consistent with lichen sclerosus (Figs. 5-8).

Superficial squamous epithelium atrophy with collagen hyalinization in the lamina propria was

found in the upper left gingival; changes consistent with lichen sclerosus were also observed in the lesions of the right buccal mucosa and dorsum of the tongue.

Laboratory tests excluded other diseases; the patient was treated with topical tacrolimus showing discrete improvement, and has been regularly monitored at the General, Oral and Genital Dermatology Clinic.

DISCUSSION

The involvement of the oral mucosa alone or in conjunction with other forms of lichen sclerosus is extremely rare, and so far less than 30 well-documented cases of oral lesion were reported in the English-language literature. The most common area of involvement of the mouth is lower lip (11 reported cases), followed by buccal mucosa (9 cases), upper lip (7 cases) and tongue (5 cases). The palate, gingiva and tonsillar pillars may also be affected (4).

Oral lesions appear as white maculae or plaques of reticular pattern, and may have superficial ulceration. They are usually asymptomatic, but may cause itching, burning, pain and limitation in mouth opening (3,4).

The histology of LS includes varying degrees of epithelial hyperplasia and hyperkeratosis with vacuolar degeneration of the basal layer, hyalinization of subepithelial collagen, decrease or loss of elastic fibers in the upper dermis, and lymphocytic infiltrate. In more recent lesions, it tends to be located near the dermoepidermal junction, migrating to the deep dermis in later stages of the disease. Such changes occur both in oral LS lesions and in the skin, while hyperkeratosis tends to be more acute in skin lesions (3,4,8,12,14).

Differential diagnosis of oral lesions of LS includes localized or systemic scleroderma, lichen planus, vitiligo, oral leukoplakia, oral submucous fibrosis and lesions of graft *versus* host disease. Histopathology is mandatory for a definitive diagnosis (1,4,8,14) (Table 1).

Oral leukoplakia is characterized by a white plaque not associated with any specific disease (idiopathic keratosis). It is most often caused by chronic irritation of the oral mucosa, and the etiologic factors such as smoking and infective agents. The histopathologic findings are hyperkeratosis, changes in epithelial thickness with varying degrees of acanthosis and epithelial maturation disorder.

Scleroderma located in the oral cavity is rare and does not present the histopathologic features of LS. In systemic sclerosis, oral manifestations may



Figure 1. Atrophic hypochromic plaque in the inframammary region.



Figure 4. Leukoplakic lesion at the dorsum of the tongue.



Figure 2. Achromic, atrophic lesion, with areas of purpuric and waxy shine.

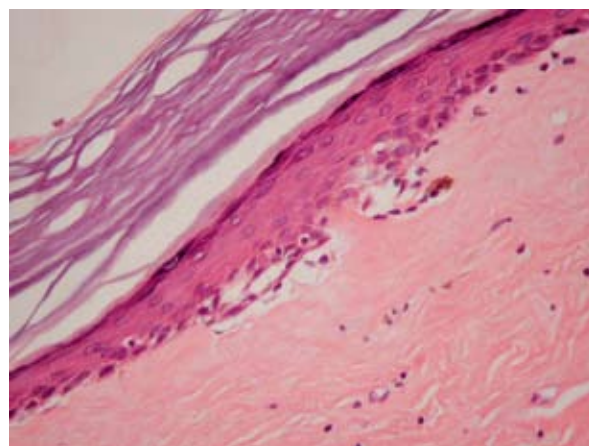


Figure 5. Histopathology – lesion of the inframammary region: thinned epidermis with hyperkeratosis, vacuolar degeneration of the basal layer and collagen hyalinization. (H&E, X100)



Figure 3. Leukoplakic lesion in the right buccal mucosa.

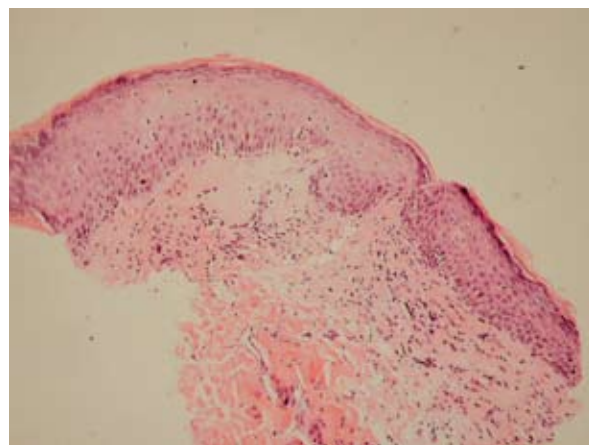


Figure 6. Histopathology – lesion of the vulva: area of collagen hyalinization. (H&E, X40)

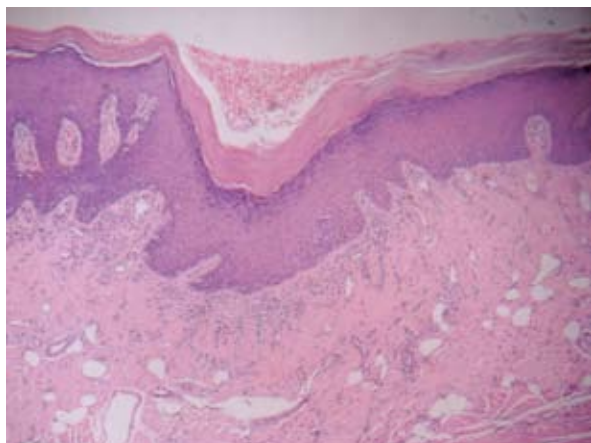


Figure 7. Histopathology – lesion at the dorsum of the tongue: intense mononuclear inflammatory infiltrate in the lamina propria, dilated capillaries, pigmentary incontinence and collagen hyalinization. (H&E, X40)

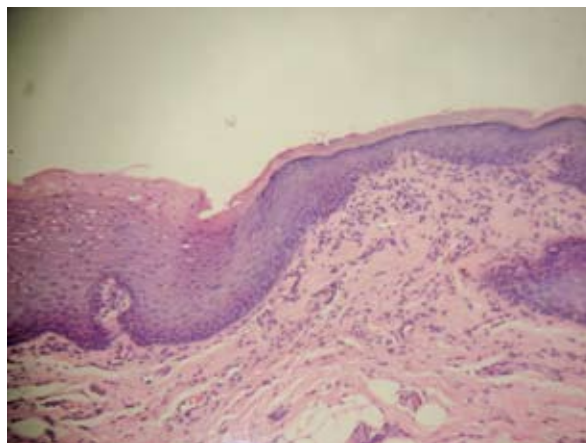


Figure 8. Histopathology – lesion of the buccal mucosa: clear transition of the epithelium and mild collagen hyalinization. (H&E, X100)

Table 1. Differential histopathologic diagnosis of oral lesions of lichen sclerosus (LS)

Oral lesion	Epithelium	Vacuolar degeneration of basal layer	Band-like inflammatory infiltrate	Decrease or loss of elastic fibers	Obliteration or narrowing of blood vessels
LS	Hyperkeratosis and atrophy	+ or -	-	+	-
Lichen planus	Hyperkeratosis and atrophy	+	+	-	-
Leukoplakia	Hyperkeratosis	-	-	-	-
Submucosal fibrosis	Atrophy	-	-	-	+
Scleroderma	Atrophy	-	-	-	+

Adapted from Buajeeb *et al.* (8).

occur such as xerostomia, microstomia, gum bleeding, tooth movement and expansion of the periodontal ligament space. Systemic sclerosis, unlike LS, can affect internal organs and present specific circulating autoantibodies. In histopathology, the changes in scleroderma occur especially in the deep dermis, as opposed to the superficial dermis in LS. The decrease or loss of elastic fibers, characteristic of LS, does not occur in scleroderma; besides, collagen synthesis is increased in scleroderma and reduced in LS.

The presence of basal melanocytes excludes the diagnosis of vitiligo. Differentiation from submucosal fibrosis is very difficult, but pyknotic nuclei in basal cells, epithelial atypia, obliteration or narrowing of blood vessels are not observed in LS (14).

Oral lichen planus clinically resembles LS, especially in the atrophic and reticulate form. Lichen planus is differentiated, in histopathology, by the presence of

a mononuclear inflammatory infiltrate in band at the dermoepidermal junction, and by not presenting epithelium atrophy or collagen sclerosis, as in LS (3,7).

Oral manifestation of acute graft *versus* host disease includes pain in the oral mucosa, desquamation and ulceration, cheilitis and lichenoid plaques. Small white lesions may prematurely affect the oral mucosa and tongue, disappearing in two weeks. Erythema and ulceration are more pronounced. The lesions of the chronic phase coincide with the skin lesions and appear as generalized erythema, lichenoid lesions and xerostomia. The clinical history of transplant is important in differentiating the two entities.

There is no consensus on topical or oral treatment for LS and in most cases it is not necessary, given that most lesions are asymptomatic and its benefit only lies in controlling the symptoms. There is no healing measure for the lesions (14).

The treatment is based on the use of topical steroids, with the option of calcineurin antagonists and vitamin D derivatives. In the presence of more severe symptoms, oral or intralesional steroids can be used. Small lesions can be surgically removed with resolution of symptoms; there are no reports of post-removal relapse. Colchicine can be used in inflammatory lesions due to its ability to inhibit microtubule polymerization, limit the chemotactic and phagocytic activity of inflammatory cells and modulate the synthesis of collagen. Its use has been reported in two cases with regression of skin lesions (4). A recently reported case of a 7-year-old girl, in which the author used a topical pimecrolimus application of 1% twice daily for four weeks, was able to avoid progression of the disease (15). Soria *et al.* report that topical rapamycin (1 mg/mL twice a day for 3 months) may be effective in some cases of refractory chronic erosive oral lichen planus. Its action is probably due to both its immunosuppressive and antitumor properties. There is negligible absorption into the blood and minimal side effects (16).

In genital LS, there are areas of hyperkeratosis, dysplasia and transformation into squamous cell carcinoma in 5% of patients. No cases of malignant transformation in oral lesions of LS have been reported so far (4).

We must emphasize the importance of oral cavity examination in all patients with genital and extra-genital LS, given that oral lesions may be asymptomatic.

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