

Oral Lichen Sclerosus et Atrophicus – Literature Review and Two Clinical Cases

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Lichen sclerosus et atrophicus (LSA), also called Lichen sclerosus (LS) is a chronic, benign, depigmenting disease of the skin and mucous membranes, most frequently affecting genital mucosa and skin. Involvement of the oral mucosa without concurrent genital or skin lesions has very rarely been reported in the literature. Here we report on two Chinese women with LSA limited to the dorsum of the tongue, and describe the clinical manifestations and histopathological findings of these two patients.

Key words: *Lichen sclerosus et atrophicus, oral, tongue*

Lichen sclerosus et atrophicus (LSA) is a chronic inflammatory disease, predominantly affecting prepubertal girls and postmenopausal women on genital mucosa and skin. The etiology of LSA is not clear, but it has been linked with autoimmune mechanisms, infection, trauma, genetic susceptibility and hormone disorders¹. Clinically, it is characterised by white atrophic plaques in the anogenital region. The lesions are generally asymptomatic, but may cause discomfort with itching and pain, which may lead to erosions and purpura. Extragenital mucosa involvement is very unusual, and lesions limited to the oral mucosa are even less frequent. Oral mucosal lesions appear as well-demarcated whitish, flat lesions, where the size varies from small localized macula to lesions involving larger areas of the mucosa. Telangiectasia in relation to the lesion has been reported^{2,3}.

Skin and mucosal lesions show almost identical histologic features, characterised by a gradual epidermal atrophy and subepidermal homogenisation and hyalinisation of the collagen fibers with an underlying, often band-like, lymphatic inflammatory infiltrate. Hyperkeratosis may be absent in oral lesions but is always present in skin lesion. In view of the rarity of reported cases, two cases affecting only the dorsum of the tongue are presented. To date, only 26 patients with biopsy-proved isolated oral LSA have been reported in the literature. Among the cases, 10 had skin lesions and four had genital lesions. But till now, no cases of oral LSA were reported in China. The purpose of presenting these two case reports is to add new cases of oral LSA confined to the dorsum of the tongue to the limited number of already reported cases, especially in China for the first time, and to summarise the current knowledge of this relatively uncommon oral lesion.

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Case 1

A 54-year-old Chinese woman presented to the Department of Oral Medicine, for the white plaque on the dorsum of the tongue. The lesion had been found for 10 days and was asymptomatic. She had not taken any medication before that. No examination or therapy was carried out after that.

Medical history

The patient denied a history of trauma, injections, blisters, surgery or bleeding in this area. No history of cigarette and alcohol use was provided. No family history of similar appearing lesions was reported. In addition, no personal or family history of autoimmune disease was reported.

Physical examination

On oral examination, the patient had a well-demarcated atrophic, porcelain, white plaque on the anterior dorsum of the tongue sized 2 ~ 3 cm², with petechia in the centre of the plaque (Fig 1). The gingiva, lip and the rest of the oral mucosa surface were uninvolved. The patient was referred to a consultant dermatologist for further investigation but no skin and genital similar lesions were identified and vitiligo was excluded.

Accessory examination

The complete blood count was in the normal limit and the glucose count was 5.8 mmol/L, there was no testing of the thyroid function. An incisional biopsy was performed and the histological study revealed atrophic mucosal epithelium, liquefaction of the basal cell, diffused chronic inflammation with extensive fibrogenesis and hyaline degeneration in lamina propria and scattered telangiectatic blood vessels, which was in accordance with LSA (Figs 3 and 4).

Case 2

A 58-year-old Chinese woman without disease, antecedents of interest or known allergies or toxic habits (smoking, alcohol abuse) reported to the Department of Oral Medicine, with a white plaque on the dorsum of the tongue that had been present for two years and did not cause any discomfort. The patient did not have any examination or treatment before and after the lesion was found.

Medical history

The patient denied a history of trauma, injections, blisters, surgery or bleeding in this area. No family history of similar appearing lesions was reported. In addition, no personal or family history of autoimmune disease was reported.

Physical examination

On oral examination, the second patient had a white lesion, which measured 2.5 ~ 3.0 cm² on the left dorsum of the tongue, the lesion had an obscure border and was not as white as the first patient's (Fig 2). The gingiva, lip and the rest of the oral mucosa surface were uninvolved. The patient was referred to a consultant dermatologist for further investigation but no skin and genital similar lesions or vitiligo was identified.

The second patient underwent some accessory examinations, which showed a normal level of blood cell count, and the glucose level was 6.52 mmol/L, but later the endocrinologist excluded the diagnosis of diabetes. There was no testing of the thyroid function. An incisional biopsy was performed and the histological study revealed atrophic mucosal epithelium, diffuse chronic inflammation with local fibrogenesis and hyaline degeneration in lamina propria, which was consistent with the early stages of LSA (Figs 5 and 6).

The patients were both treated with 0.1% tacrolimus ointment. A periodical follow-up was strongly recommended.

Discussion

LSA is a chronic inflammatory disease predominantly affecting females on genital mucosa and skin. According to the epidemiology, LSA is more common in women than men: some studies give a female to male ratio of 10:1, others 6:1¹. Its aetiology is uncertain, but there is an increased incidence of autoimmune antibodies in LSA and an association with autoimmune diseases, such as vitiligo, thyroid disease and alopecia areata⁴; pernicious anaemia and diabetes mellitus have also reportedly been associated with LSA⁵. In our cases, vitiligo and anaemia were excluded, but for glucose levels, the first patient was normal and the second was a little higher than normal level, but diabetes was excluded afterwards. The thyroid function was not performed because the relevant signs and symptoms were negative.

According to the reports in the literature, the prevalence of LSA affecting the oral mucosa without skin and genital mucosa is exceedingly rare. To date, no more than 26 patients with biopsy-proved isolated oral LSA have been reported in the literature. Of these 26 cases, 17 occurred in women and 9 in men. The range of age was from 10 to 60 years. Among the cases, 10 had skin lesions and four had genital lesions^{2,3,6-8,9-15}. This suggests that oral LSA may appear without accompanying skin or genital lesions. The oral lesions of LSA are usually asymptomatic and may have been



Fig 1 Oral manifestation of the first patient.



Fig 2 Oral manifestation of the second patient.

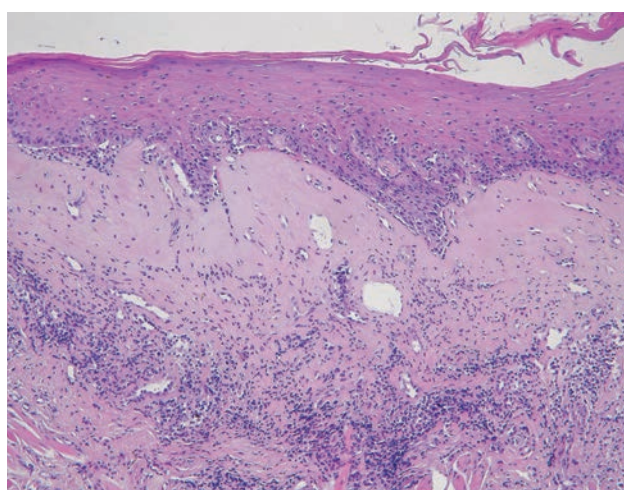


Fig 3 Superficial lamina propria showing homogenisation of the collagen fibres, HE ~ 10.

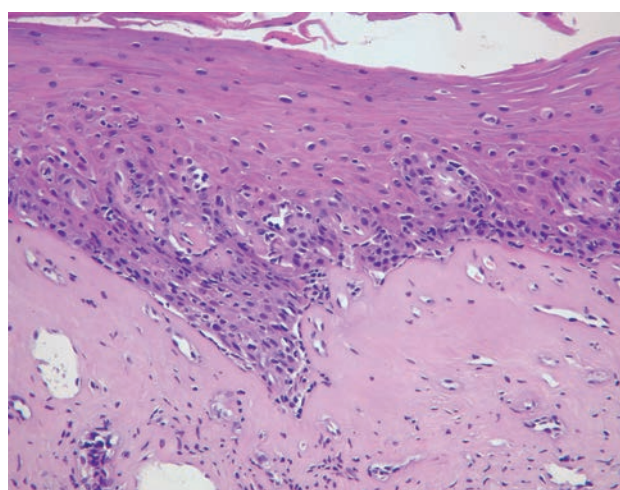


Fig 4 Higher power magnification showing homogeneous collagen fibres, HE ~ 40.

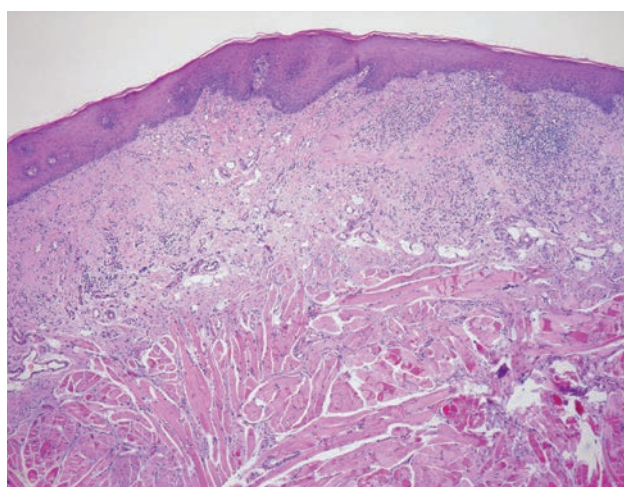


Fig 5 Superficial lamina propria showing homogenisation of the collagen fibres, HE ~ 10.

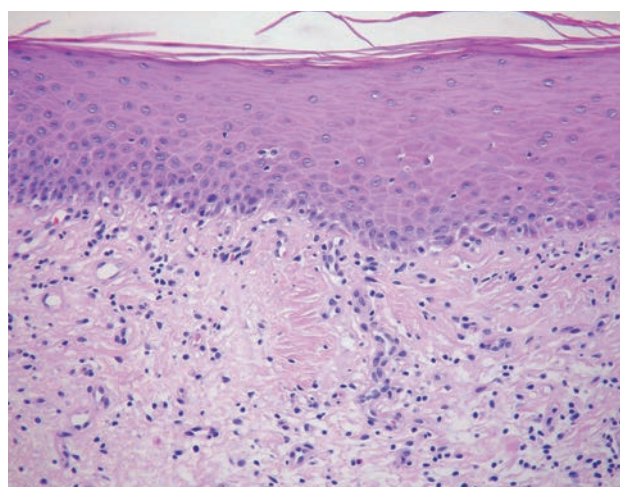


Fig 6 Higher power magnification showing homogeneous collagen fibres, HE ~ 40.

overlooked in the past. We purport that many oral mucosa LSA lesions are clinically and histopathologically misdiagnosed⁶. In 1957, Ravitis first described microscopic features of these lesions confined to the oral mucosa. The histological features observed for oral LSA are quite similar to those for skin, with the exception of hyperkeratosis.

LSA can occur on every site of oral mucosa, including buccal mucosa, tongue, lip and gingival mucosa. The main clinical characteristic of oral LSA is porcelain white plaque with a well-demarcated border. Extragenital LSA is usually asymptomatic. When the gingiva was involved, it may cause attachment loss and absence of the teeth¹³.

Knowledge of such lesions is important in order to establish a differential diagnosis with other white oral lesions, such as oral lichen planus (OLP), leukokeratosis or vitiligo. OLP is common, mostly asymptomatic and mostly in the posterior buccal mucosa bilateral and occasionally on the dorsum and/or ventrum of the tongue. Oral white lesions are the typical features. Six clinical types of oral OLP have been reported: reticular, papular, plaque-like, erosive, ulcerative and bullous types. Leukokeratosis is closely associated with local stimulation, and the lesion will release or disappear when the stimulation is removed.

Histological confirmation of the diagnosis is essential. At present, the most striking histopathological features of oral LSA are focused on the hydropic degeneration of basal cells, subepithelial hyalinization of the connective tissue, a slightly diffuse, band-like mononuclear infiltration in the deeper connective tissue and atrophy of the epithelium. It is sometimes difficult to differentiate oral LSA from OLP, oral submucous fibrosis, oral scleroderma and oral vitiligo by means of histological features. However, the band of lymphocytes at the epithelium connective tissue junction that is usually found in OLP is not evident in LSA, and scantiness or loss of elastic fibers, usually present in LSA, is not found in scleroderma. The presence of the melanocytes in the basal epithelium in the LSA eliminates vitiligo and the absence of the obliteration or narrowing of the blood vessels and epithelial atypia eliminate submucous fibrosis. Thus, because of the microscopic findings present in oral LSA, it is possible to establish a correct diagnosis for this lesion^{3,6}.

The oral manifestation of the second patient was similar to that of OLP, but the histopathological findings were characteristic with LSA, including the liquefaction of the basal cell and local hyalinised changes in the lamina propria, which reminds the clinician to carry out correct differential diagnoses.

Treatment of oral LSA is usually unnecessary because of its asymptomatic nature, benign behaviour, few cosmetic concerns and no evidence of recurrence. Treatment strategies for oral LSA are derived from the therapeutic experience with extraoral mucosal presentations. The most common and effective is topical corticosteroids. Ultra potent topical corticosteroids have been the first-line treatment for LSA at any site, but the outcomes are variable^{3,14,16}. Among some of the available drugs are topical clobetasol 0.05% ointment, topical testosterone propionate 2% in petrolatum, and 1% pimecrolimus cream¹⁷.

Although no cases of malignant change associated with oral LSA have been reported given the limited number of patients reported in the literature, a regular long-term follow-up is necessary and indicated⁶.

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