MUCOSAL LICHEN PLANUS SIMULTANEOUSLY INVOLVING ORAL MUCOSA, CONJUNCTIVA AND LARYNX

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INTRODUCTION

Lichen planus is an inflammatory mucocutaneous condition that usually exhibits a distinctive morphology. The prevalence of lichen planus is unknown, but it is estimated to occur in less than 1 percent of the population. Estimates of the prevalence vary among different populations, but the condition does not show a racial predilection. It most commonly affects middle-aged adults of either gender, with a slight female predominance.1,2 Mucous membrane lesions occur in about two-thirds of all cases and are found on the buccal mucosa, tongue, lips, gums, palate, tonsils and pharynx and also throughout the gastro-intestinal tract from the stomach to the rectum and anal mucosa. Genito-urinary involvement is also no exception. Oral lichen planus classically presents as a white, lacy, reticular pattern on the buccal mucosa. Papular, atrophic or erosive lesions may also occur. Patients with long-term oral mucosal lesions have an increased risk of developing squamous cell carcinoma within the lesions.1-4

Involvement of eye in lichen planus may be limited to eyelid or may develop an ocular surface disease with cicatricial conjunctivitis, keratouveitis, kerato-conjunctivitis sicca, punctate epithelial erosions, and corneal ulceration.5,6 Laryngeal disease, like other mucosal involvements, although possible but has been very rarely reported in literature.7 Clinical diagnosis of lichen planus is confirmed by histopathology that reveals characteristic findings of a band-like infiltrate of lymphocytes at the epidermal-dermal junction, vacuolar degeneration of the basal cell layer and pigmentary incontinence.

While most cases of lichen planus are idiopathic, some are linked to medication use or hepatitis C virus infection and lymphocytes, especially T-cells seem to play a major role in pathogenesis.1,2,8 The principle of treatment is directed towards the immunopathologic processes and corticosteroids (topical or systemic) are the agents of choice.7,8 Occasionally, when the disease is not responding adequately, a combination of azathioprine may be helpful.1,2 Topical cyclosporin may be required to stabilize ocular disease in severe cases.5 This case report describes an extensive multifocall occurrence of the disease in head and neck region.

CASE REPORT

A young male, aged 18 years, presented in the outdoor department of dermatology at Military Hospital Rawalpindi with complaints of hoarseness of voice, oral ulcerative lesions and symblephron. Mucosal biopsies (buccal mucosa and supraglottic area) revealed the diagnosis of lichen planus and he was managed accordingly. Although oral lichen planus is frequently reported but simultaneous involvement of oral, ocular and laryngeal mucosa has not been reported earlier.

As a case of mucosal pemphigoid but the biopsies done from lesional mucosae (buccal and supraglottic) did not reveal any subepidermal split and it was highly consistent with diagnosis of lichen planus (Figure 3). Moreover no deposition of immune deposits was detected on direct immunofluorescence (DIF). He was finally diagnosed as a case of mucosal lichen planus and was treated with tablet prednisolone 30 mg daily, tablet azathioprine 100 mg daily along with symptomatic topical treatments for oral mucosa and eyes. His ocular symptoms responded adequately and oral lesions substantially regressed but hoarseness did not improve. He was re-assessed by otolaryngologist but no surgical intervention was done keeping in view the diffused involvement and fragile nature of laryngeal mucosa. He was advised to continue medication and monthly follow-up by otolaryngologist and dermatologist.

**DISCUSSION**

About two-thirds of all cases of lichen planus have mucous membrane involvement. While approximately 50% of patients with skin lesions have oral lesions, these may be the only manifestation of the disease in about 25% of cases. Both antigen-specific and non-specific mechanisms may be involved in the pathogenesis of mucosal lichen planus. Antigen-specific mechanisms include antigen presentation by basal keratinocytes and antigen-specific keratinocyte killing by CD8+ cytotoxic T-cells. Non-specific mechanisms include mast cell degranulation and matrix metalloproteinase activation in mucosal lesions. These mechanisms may combine to cause T-cell accumulation in the superficial lamina propria, basement membrane disruption, intra-epithelial T-cell migration, and keratinocyte apoptosis. Oral mucosa is frequently involved mostly but cases of conjunctival and laryngeal disease are sparse in medical literature.

This case was unique in the sense as it presented mainly as voice hoarseness and there was simultaneous involvement of three different mucosae. To the best of our knowledge, no such case has yet been reported in literature. Mucosal lichen planus can closely mimic other autoimmune diseases such as mucous membrane pemphigoid, pemphigus vulgaris, systemic lupus erythematosis. Paraneoplastic lichen planus is another possible cause where cicatrising conjunctivitis may be associated with inflammatory skin and mucous membrane disease. Oral mucosal, conjunctival and laryngeal involvement in this case initially suggested cicatrical pemphigoid but characteristic histology and lack of immunofluorescent findings of pemphigoid led to the diagnosis is of this unusual case.

It is therefore suggested that, in patients presenting with simultaneous oral, conjunctival and laryngeal mucosal lesions, lichen planus must also be suspected in addition to mucosal pemphigoid and a thorough histological analysis, including immunofluorescence staining, should be performed in order to make a definite diagnosis.

**REFERENCES**