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Case Report
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# Mucosal Lichen Planus Pemphigoides: A Distinct Entity?

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## Abstract

We report a case of lichen planus pemphigoid with isolated mucosal involvement and a refractory course requiring cyclophosphamide. Our case highlights the nosological issues associated with this rare form of autoimmune blistering dermatosis and its therapeutic challenges.

**Keywords: Lichen planus pemphigoides; Lichen planus; Mucous Membrane Pemphigoid (MMP); Direct immunofluorescence; Cyclophosphamide**

## Introduction

Lichen Planus Pemphigoides (LPP is a rare autoimmune blistering disease which, owing to the advent of immunopathological techniques, is now recognized as a distinct entity within the pemphigoid group [1]. Clinically, blisters occur in association with Lichen Planus (LP) lesions and, immunohistologically, LPP exhibits both LP and Bullous Pemphigoid (BP) features [2]. LPP with exclusive mucosal involvement appears to be clinically and prognostically distinct from the previously described form. The aim of this paper is to present a new case of mucosal LPP and to clarify the nosological framework of this disease.

## Case Presentation

A 68-year-old male presented with one year of slowly progressive dysphagia and intraoral burning sensation. His past medical history was significant for systemic hypertension. He denied a history of any dental restorations or smoking. Intraoral examination revealed patterned white streaks (Wickham striae) in bilateral jugal mucosa, along with painful erosive lesions of the tongue displaying white borders and desquamative gingivitis (Figure 1). Oral LP with esophageal involvement was suspected. An esophagogastric endoscopy revealed several pharyngeal and esophageal erosions. Extraoral examination (skin, genitalia, eyes) was within normal limits. A mucosal biopsy taken from the tongue showed a lichenoid mucositis with hydropic degeneration of the basal cells and a band-like lymphocytic infiltrate in the superficial lamina propria (Figure 1). The diagnosis of erosive LP was made. The patient was started on oral prednisolone (1 mg/kg/day for a weight of 80 kilos) and topical corticosteroids (5 mg prednisolone dissolved in 20 cL water) as a mouthwash 4 times. After 5 months of steroid therapy, the disease remained active and the patient gradually developed ocular symptoms including xeropthalmia, photophobia and significant conjunctival hyperemia.

Anterior segment examination showed the presence of right eye symblepharon and left eye ankyloblepharon (Stage III and Stage IV respectively based on Foster's Classification System scoring) (Figure 2). A second oral biopsy and a conjunctival biopsy were performed. Both histologic tests demonstrated subepithelial clefting, with a mixed eosinophilic and neutrophilic infiltrate localized in the underlying lamina propria without any lichenoid features. Direct Immunofluorescence (DIF) demonstrated linear deposition of IgA (2+) and IgG (1+) on the Basement Membrane Zone (BMZ) (Figure 3). Indirect Immunofluorescence (IFI) of 1M NaCl-split skin detected positive reactivity with epidermal side for IgA (2+) and IgG (1+) antibodies. Enzyme-Linked Immunosorbent Assays (ELISAs) using the commercial kit (Euroimmun, Lubeck, Germany) were weakly positive for anti-BP180 NC16a-domain autoantibodies and negative for anti-PB230 autoantibodies. Based on the clinical, histopathological features and immunological findings, the diagnosis of LPP with isolated mucosal involvement was made. The patient was treated with dapsone (100 mg/day) and azathioprine (150 mg/day) in adjunction to short pulse oral corticosteroids. Topical therapy including artificial tears, corticosteroids and cyclosporine eye drops were added. Three months later, the patient underwent bilateral scar tissue excision with conjunctival human amniotic membrane graft. At one

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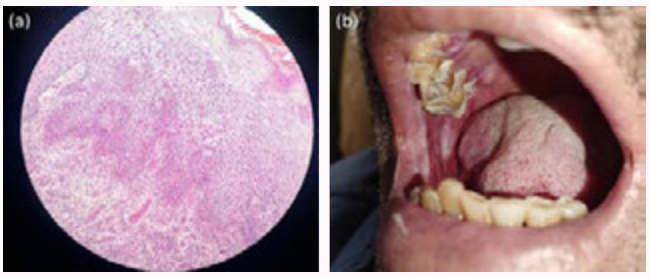


**Figure 1:** Clinical and histopathological features at initial presentation. (a) Multifocal areas of gingival and buccal mucosal involvement. Ulcerations and erythema of the right maxillary buccal gingiva surrounded by white striae. (b) Parakeratosis, acanthosis and interface dermatitis (Hematoxylin and eosin, original magnification x200).

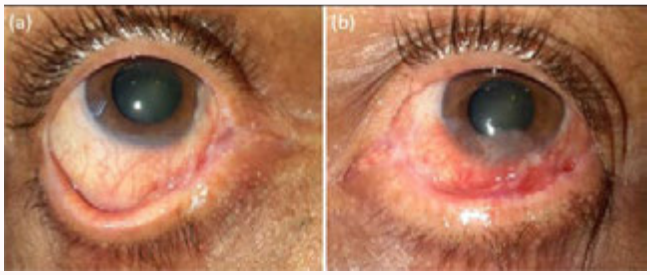


**Figure 2:** Right eye on the left (a) showing the presence of underlying symblepharon without restrictive ocular motility when trying to pull on the lower eyelid. Left eye on the right (b) showing a marked inferior fornix shortening with corneal scarring and restrictive motility when the patient was asked to look up while trying to pull on the lower eyelid.

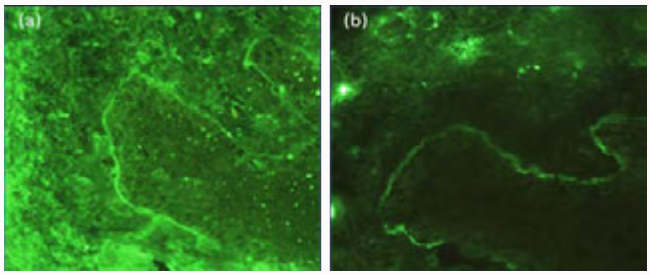


**Figure 3:** Direct immunofluorescence study showed linear deposition of (a) IgG and (b) IgA along the basement membrane zone (Hematoxylin and eosin, original magnification x100).

month follow up, ocular examination showed the presence of left eye symblepharon. Due to the rapid progression and the severity of the disease, intra-venous (i.v.) cyclophosphamide (600 mg/m$^2$) was initiated with ocular stabilization.

## Discussion

Oral lesions in autoimmune bullous diseases and oral LP may be difficult to distinguish both clinically and histopathologically. Our patient initially presented with oral lesions consistent with the diagnosis of erosive LP. Based on the lack of clinical improvement under treatment, in addition to the ocular involvement with rapid and severe onset, the differential diagnoses included LPP, Mucous Membrane Pemphigoid (MMP), epidermolysis bullosa acquisita, and linear IgA disease. Clinically, oral LPP is characterized by white striations (Wickham striae), erosions, desquamative gingivitis, ulcerations and sometimes bullae [3]. In the present case, the diagnosis of LPP was confirmed by histopathological examination which revealed both LP features (first biopsy) and MMP (second biopsy), as well as DIF findings consistent with pemphigoid. ELISAs for Bp180 and Bp230 IgG antibodies were weakly positive for anti-BP180 NC16a-domain autoantibodies and negative for anti-PB230 autoantibodies. This result was expected, as circulating antibodies to BMZ components were predominantly of the IgA class. There remains significant controversy as to whether LPP exhibiting exclusive mucosal involvement corresponds to MMP with LP-like clinical and histopathological features or if it is the result of the "epitope spreading" phenomenon from one immunologically unrelated disorder (oral LP) to another (BP/MMP) [4]. Recently, Schmidt et al. demonstrated that patients with LPP have a mixed Th1/Th2 cell response against BP180 [5]. These findings reinforce the first hypothesis. A subset of patients may indeed exhibit a distinct type of MMP that combines TH1 and TH2 cell response targeting hemidesmosomal components [4]. Regardless of the suggested pathophysiological mechanisms, the exclusive and treatment-resistant mucosal involvement renders the diagnosis of classic LPP less probable. LPP is usually a mild bullous dermatosis, characterized by almost constant cutaneous involvement and often a rapid, favorable progression that often obviates the need for intensive therapy. Taking all pertinent factors into consideration, we suggest that the various terms used in the literature, including MMP of the oral lichen type, oral LPP, and lichen planus mucous membrane pemphigoid, denote the same specific clinical condition [3,4,6].

## Conclusion

This case demonstrates a recalcitrant course of a rare and controversial entity of mucosal LPP. Our patient failed to respond to conventional therapy and eventually achieved remission with cyclophosphamide. A DIF test should be performed in cases of persistent oral erosions to differentiate oral LP from mucosal LPP.

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