

# Benign mucous membrane pemphigoid: Oral, ocular and laryngeal lesions

Shaik Begum Khalida, M. Manjunath, A. G. Annaji, Lakhani Himanshu

## ABSTRACT

**Introduction:** Benign mucous membrane pemphigoid is a disorder that is autoimmune in nature and is characterized by subepithelial bulla formation. It may involve multiple mucosal sites that include oral cavity, conjunctiva, larynx, esophagus, genitourinary tract and anus. An early detection of this disease and its treatment may decrease its complications. **Case Report:** An Indian 54-year-old female reported with ulcers in the mouth since few months. On examination multiple vesicles were noted on the hard palate, labial mucosa and also ulcerative lesions on the entire oral mucosa except the gingiva. Evidence of symblepharon and ulcers in the larynx showed an extensive involvement of extraoral mucosae. Microscopic examination showed a subepithelial split suggestive of benign mucous membrane pemphigoid. **Conclusion:** Thus we hereby

report an advanced case of benign mucous membrane pemphigoid. The choice of treatment depends on the site of involvement, its severity and progression. Thus oral physicians should therefore thoroughly examine the other mucosal sites in obtaining a proper diagnosis.

**Keywords:** Mucous membrane pemphigoid (MMP), Oral, Ocular, Subepithelial bulla

### How to cite this article

Khalida SB, Manjunath M, Annaji AG, Himanshu L. Benign mucous membrane pemphigoid: Oral, ocular and laryngeal lesions. J Case Rep Images Dent 2016;2:19–22.

Article ID: 100007Z07SK2016

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doi:10.5348/Z07-2016-7-CR-5

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Received: 27 January 2016  
Accepted: 26 February 2016  
Published: 08 April 2016

## INTRODUCTION

Benign mucous membrane pemphigoid, also known as cicatricial pemphigoid, is a chronic vesiculobullous disorder that is autoimmune in nature. It predominantly involves the oral mucosa and conjunctiva, but rarely the skin. Most of the patients may present with only oral lesions, but its extension into the esophagus and pharynx can result in symptoms of sore throat and dysphagia [1, 2]. The etiopathogenesis is unclear, but is considered as an autoimmune in nature, with participation of basement membrane directed antibodies, thus leading to subepithelial blistering. Recent studies have shown an evidence of increase in eosinophils and also the proportion of type I and type III collagen in these patients [3]. Clinical presentation may vary; most commonly

involves the elderly women. Generally, oral lesions occur in more than 90% and ocular lesions in 60–70% of cases [4–6].

## CASE REPORT

A 54-year-old Indian female reported to the outpatient Department of Oral Medicine and Radiology with a chief complaint of ulcers and burning sensation in the mouth since six months. She gave a history of vesicles formation and difficulty in swallowing food since one month. She also gave history of difficulty in completely opening both the eyes since six months and dry cough since 3–4 months. On general physical examination, reduced eye opening noted in both the eyes, left being more affected than the right eye. On examination, there was a complete adhesion of the lower palpebral conjunctiva of the eyelid to the bulbar conjunctiva of the eyeball (symblepharon) (Figure 1). On intraoral examination, a solitary intact vesicle measuring about 5 mm in diameter was present on the lower labial mucosa (Figure 2A), multiple intact vesicles 8–10 in number, size ranging from 4–6 mm in diameter were noted on the hard and soft palate with multiple erosive areas (Figure 2B). Extensive shallow ulcerative areas were noted on the left lateral border of the tongue, ventral surface of the tongue, right lateral surface of the tongue, right buccal mucosa and upper labial mucosa but sparing the gingiva. Based on patient's history and clinical presentation, a provisional diagnosis of benign mucous membrane pemphigoid was considered.

Later on, the patient was referred to a general physician regarding her symptoms of dry cough. To rule out tuberculosis influence, Mantoux test was advised but the results were negative. To evaluate the cause for dysphagia, she underwent upper gastrointestinal endoscopy that revealed ulcerative lesions on the left false vocal cord and right aryepiglottic fold with edema of the surrounding structures (Figure 3A–B). Based on the history and clinical presentation we can probably consider a list of vesiculobullous diseases as our differential diagnosis. After a thorough clinical examination, a smear was taken from the fluid obtained by rupturing the intact vesicle on the hard palate which on microscopic examination revealed the presence of acantholytic cells suggestive of vesiculobullous diseases and therefore a biopsy was advised to arrive at a definitive diagnosis. An incisional biopsy was performed from the intact vesicle of the lower labial mucosa to obtain two tissue specimens, one for histopathologic evaluation and other for direct immunofluorescence test. Microscopic examination with H & E stained section showed Hyperplastic parakeratinized stratified squamous epithelium of irregular thickness with Subepithelial Split (Figure 4). Direct immunofluorescence of labial mucosal biopsy showed intercellular positivity for IgG and IgM in the lining epithelium and acantholytic cells. Based on the history, clinical examination and the laboratory



Figure 1: Complete adhesion of the lower palpebral conjunctiva of the eyelid to the bulbar conjunctiva of the eyeball (symblepharon).



Figure 2: (A) A solitary intact vesicle measuring about 5 mm in diameter was present on the lower labial mucosa, (B) Multiple intact vesicles 8–10 in number, size ranging from 4–6 mm in diameter were noted on the hard and soft palate with multiple erosive areas.

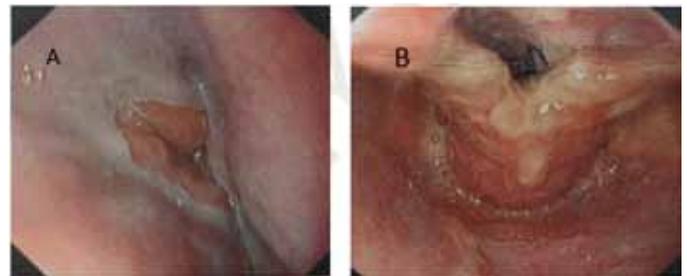


Figure 3: Upper gastrointestinal endoscopy showing (A) Ulcerative lesions on the left false vocal cord, and (B) Right aryepiglottic fold with edema of the surrounding structures.



Figure 4: Microscopic examination showing hyperplastic parakeratinized stratified squamous epithelium of irregular thickness with subepithelial split (white arrows) (H&E stain, x100).

findings a definitive diagnosis of mucous membrane pemphigoid was arrived. Based on the definitive diagnosis the treatment for oral lesions was initiated by topical application of 0.1% triamcinolone acetonide (kenacort oromucosal paste) twice daily and systemic corticosteroid (tab prednisolone 20 mg) thrice a day for 1 week. Following the remission of the lesions, the dosage was gradually tapered and stopped. Later on, the patient was referred to an ophthalmologist for the treatment of symblepharon.

## DISCUSSION

Benign Mucous membrane pemphigoid is a chronic autoimmune subepidermal bullous disease, which primarily involves mucous membranes, mainly oral cavity and eyes, but rarely the skin [7]. Most of the patients may present with only oral lesions, but its extension into the esophagus and pharynx can result in symptoms of sore throat and dysphagia [2]. More than 70% of cases may present with ocular involvement. Recent studies have shown that this disease is not a single entity but a complex disease [8, 9]. The etiopathogenesis of this disease is considered to be an autoimmune, with participation of basement membrane directed antibodies, thus causing a subepithelial blister. A dense inflammatory infiltrate and few areas of granulation tissue are noted in the substantia propria. Eosinophils and increased proportion of type I and type III collagen have also been shown [9]. Clinical presentation may vary according accordingly, primarily affects the elderly women. Generally, oral lesions have been reported in more than 90% and ocular lesions in 60–70% of cases [10]. Ocular complications include chronic scarring with progressive fibrosis, symblepharon formation between bulbar and palpebral conjunctiva [9, 10]. Oral lesions may manifest as scattered painful erosions or desquamative gingivitis with smooth erosions along the fixed gingiva. Buccal mucosal scarring may result in chewing and swallowing difficulties. Other mucosae such as the esophagus, larynx or genitalia may also be involved with possible adhesion and stricture formation [10]. Microscopic examination of an intact blister shows subepidermal separation. Direct immunofluorescence has shown to be positive in 50–60% of cases, displaying IgG, C3 and occasionally IgA deposits on the basement membrane zone.

The disease related complications can be decreased by its early detection and subsequent treatment. If a patient shows signs and symptoms of only one mucosal site involvement, then a thorough review of symptoms highlighting involvement of other areas should be considered. The treatment of choice for these patients mainly depends on the site, severity and progression of the disease. A multidisciplinary approach has been put forth by the consensus group for its treatment. The multidisciplinary group should include the dentists,

dermatologist, ophthalmologist, gastroenterologist and gynecologists. The patients were grouped into two categories: (1) low-risk groups and (2) high-risk groups for providing appropriate treatment. High-risk treatment included patients showing rapid progression with oral, ocular, esophageal, laryngeal and genital mucosae. This includes initial treatment with prednisone and cyclophosphamide. Alternative therapy includes dapsone, azathioprine, and intravenous immunoglobulin. Low-risk patients are those in which there is either oral or both oral and skin involvement. These patients are treated more conservatively as the incidence of scarring is less.

## CONCLUSION

Thus an early detection and treatment may thereby decrease their complications. In a patient who presents with involvement of only one site, a thorough review of symptoms highlighting any other area of involvement should be noted. The choice of treatment for these patients mainly depends on the site of involvement, severity and disease progression.

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## Author Contributions

Shaik Begum Khalida – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

M. Manjunath – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

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## Guarantor

The corresponding author is the guarantor of submission.

## Conflict of Interest

Authors declare no conflict of interest.

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