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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
A 54yr old female Indian 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 symblepheron 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.
**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 oesophagus and pharynx can result in symptoms of sore throat and dysphagia [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].

**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 6 months. She gave a history of vesicles formation and difficulty in swallowing food since 1 month. She also gave history of difficulty in completely opening both the eyes since 6 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 (SYMBLEPHERON)(Figure 1). On intraoral examination, a solitary intact vesicle measuring about 5mm in diameter was present on the lower labial mucosa (Figure 2A), multiple intact vesicles 8-10 in number, size ranging from 4mm-6mm 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 GI 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 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 (kenocort oromucosal paste) twice daily and systemic corticosteroid (Tab Prednisolone 20mg) thrice a day for 1 week. Following the remission of the lesions, the dosage was gradually tapered and stopped. Later on, patient was referred to an ophthalmologist for the treatment of symblepheron

**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 oesophagus 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 [9]. The etiopathogenesis of this disease is considered to be an
autoimmune, with participation of basement membrane directed antibodies, thus
cause 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, symblepheron 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 gynaecologists. 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.

CONFLICT OF INTEREST
None

AUTHOR’S CONTRIBUTIONS
Shaik begum khalida
Group 1 - Conception and design, Acquisition of data, Analysis and interpretation of data
Group 2 - Drafting the article, Critical revision of the article
Group 3 - Final approval of the version to be published

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REFERENCES


**FIGURE LEGENDS**

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

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

Figure 3: upper GI endoscopy showing ulcerative lesions on the left false vocal cord (A) and right aryepiglottic fold with edema of the surrounding structures (B).

Figure 4: Microscopic examination with H & E stained section showed Hyperplastic parakeratinized stratified squamous epithelium of irregular thickness with subepithelial split (White arrows).

**FIGURES**

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Figure 3: upper GI endoscopy showing ulcerative lesions on the left false vocal cord (A) and right aryepiglottic fold with edema of the surrounding structures (B).
Figure 4: Microscopic examination with H & E stained section showed Hyperplastic parakeratinized stratified squamous epithelium of irregular thickness with subepithelial split (White arrows).