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Original Research Paper

Oral Medicine

MUCOUS MEMBRANE PEMPHIGOID WITH ORAL AND OCULAR LESIONS. A CASE REPORT.

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ABSTRACT Mucous membrane pemphigoid (MMP) is an autoimmune blistering disorder that is characterized by subepithelial bullae. Mucous membranes that may be involved include the oral cavity, conjunctiva, nasopharynx, larynx, esophagus, genitourinary tract. The most common site of involvement is oral mucosa and the disease may manifest as bullae, erosions, desquamative gingivitis, and occasional scarring. In this paper, we describe a case of oral and ocular mucous membrane pemphigoid.

KEYWORDS : subepithelial blistering diseases, mucous membrane pemphigoid, oral lesions, ocular involvement, symblepharon.

Introduction:

Mucous membrane pemphigoid (MMP) is a rare chronic progressive autoimmune disorder that predominantly involves mucous membranes with a tendency of scarring.[1, 2] Incidence of MMP varies from 1:12,000 to 1:40,000 in general population. It may affect any or all mucous membranes, with or without skin involvement, in decreasing frequency: oral cavity (90%), eye (65%), nose, nasopharnyx, anogenital region, skin (20-30%), larynx (8-9%), and oesophagus.[3] In severe cases, it may lead to blindness due to ocular involvement and may even be life threatening due to airway obstruction owing to scarring of the larynx. This paper describes clinical features, histopathological finding, treatment and follow up of a case of mucous membrane pemphigoid with oral and ocular involvement.

Description of case:

A 65year old female patient reported with a 3 years history of burning sensation in themouth and occurrence of blisters in her mouth which ruptured with clear discharge. She also gave 1 year history of burning sensation in the nose with occasional clear nasal discharge. Past medical history revealed that she had been treated by an ophthalmologist for burning sensation in the eyes 1year back. Ocular examination revealed fusion of palpebral and bulbar conjunctiva on the lateral aspect of right and left lower lids indicative of symblepharon (figure1 and 2). Intra oral examination revealed multiple shallow ulcers involving palate, right and left buccal mucosa surrounded by erythematous halo (figure 3). An intact bulla was present on left buccal mucosa which was taken for biopsy. A provisional diagnosis of MMP was made based on the history and clinical examination findings.

Histopathological sections revealed parakeratinized squamous epithelium with subepithelial split and dense inflammatory cell infiltration in the lamina propria along with fibrovascular stroma (figure 4). Based on this report a definitive diagnosis of benign mucous membrane pemphigoid was made. In view of the fact that the ocular and nasal lesions were healed, the patient was treated with topical 0.1% triamcinolone acetonide gel in ora-base for oral lesions. Complete relief from symptomsand healing of the ulcers and was noted on subsequent visits (figure 5). Patient was advised for ophthalmologic consultation where she was not advised any treatment as the healed ocular lesions were not interfering with the vision. However,the patient was instructed to report immediately if there was occurrence of ocular symptoms. A follow up for 8 months period did not show recurrence of oral lesions.

Discussion:

Vesiculobullous diseases, including mucous membrane pemphigoid, are relatively uncommon. Mucous membrane pemphigoid is a group of putative autoimmune, chronic inflammatory,subepithelial blistering diseases predominantly affecting mucous membranes, characterised bylinear deposition of IgG, IgA, or C3 along the epithelial basement membrane.[4] It was first described by Thost in 1911.[5] It predominantly affects elderly women with the mean age of onset between 51 to 62 years of age.6 Progressive scarring may potentially lead to serious complications owing to which the term "benign" mucous membrane pemphigoid was deemed. The term "cicatricial" pemphigoid should be used only for individuals who develop scarring.In severe cases, it may lead to blindness due to ocular involvement and may even be life threatening due to airway obstruction.Non specific chronic conjunctivitis is a frequent symptom that may progressively lead to scarring of the conjunctiva, symblepharon and ankylobepharon.[7] In the present case, the healed ocular lesions did not produce any symptoms or interfered with the vision. Diagnosisof MMP is mainly based on history, clinical examination, histopathology and immunoflorescent studyof the lesions.[4] In the present study, final diagnosis was based on the presence of multiple mucosal involvement, symblepharon and histopathological findings. Differentiating MMP from other blistering diseases is extremely essential as MMP may lead to serious complications owing to scarring and adhesions.

Corticosteroids are the main stay in treatment of MMP. The frequency and severity of the adverse effects associated with the use of systemic corticosteroids have led to the increased use of topical corticosteroids. In the present case the oral lesions were successfully treated with topical 0.1% triamcinolone acetonide gel in ora-base. In severe conditions, dapsone, cyclophosphamide, azathioprine, or methotrexate arethe systemic agents most often used.[8] Newer treatment regimens include antibiotics, nicotinamide, and intravenous immunoglobulins.[9] In general, MMP is one of the most difficult blistering diseases to control and it responds poorly to therapy.Intermittent exacerbations and waning of disease activity are commom, although some patients may experience long-term remissions. In the present case, 9 months follow up did not show any recurrence of the lesions.

Conclusion:

In the present case MMP involved oral, ocular and nasal mucosa. The oral lesions were treated successfully with topical triamcelone acetonide 0.1% and 8 months follow up for did not show recurrence of the oral lesions. We emphasize that any patient presenting with blistering disease of oral cavity should also be examined for other mucosal sites involvement. Early recognition of this disorder and treatment may decrease disease-related complications.

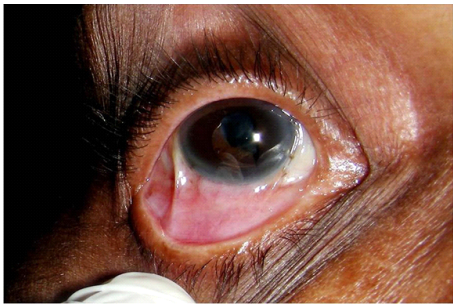

Figure 1: symblepharon of right eye

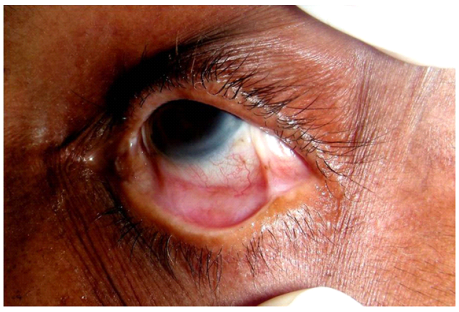

Figure 2: symblepharon of left eye

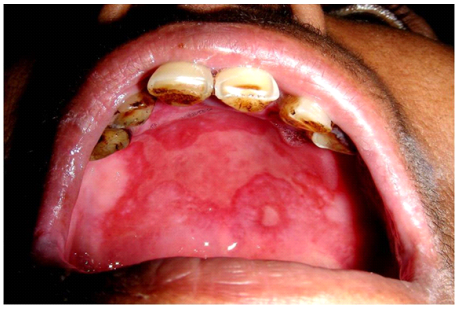

Figure 3: superficial ulceration in the palate

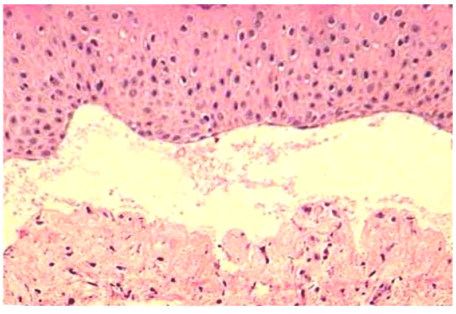

Figure 4: subepithelial split and inflammatory cell infiltration in the lamina propria

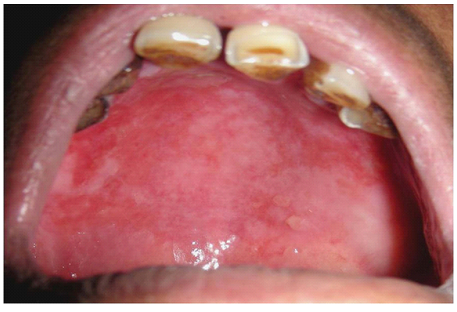

Figure 5: healed palatal lesion

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