CASE REPORT

A case report of Mucous membrane pemphigoid in a 17 year old female

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Received : 11-01-2024
Revised : 15-01-2024
Accepted : 22-01-2024

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Abstract:

Mucous membrane Pemphigoid is a heterogenous group of autoimmune subepithelial blistering disorder involving mucous membrane. The disease has got a strong female predominance with the peak of age involvement being the 5th decade. Typically the mucosal lesions occur on the oral mucous membrane and conjunctiva. Here we present a case of mucous membrane pemphigoid occurring in a female patient during her adolescent period.

Key words: Mucous membrane pemphigoid, autoimmune, subepithelial blistering.

INTRODUCTION

Autoimmune blistering disorders are a class of rare skin disorders that happen when the immune system attacks skin and mucous membrane. It includes a number of subepithelial blistering disorders such as pemphigus, pemphigoid, IgA mediated bullous dermatoses and epidermolysis bullosa acquisita.[1] Mucous membrane pemphigoid is an autoimmune blistering disease that predominantly affects mucous membrane, including the mouth and oropharynx, conjunctiva, nares and the genitalia. Skin involvement is restricted to the regions of head, neck, and upper torso. Patients with cutaneous involvement present with tense blisters and erosions where the blisters heal with scarring and pigmentation. Sequelae of mucosal involvement include decreased vision, blindness and supraglottic stenosis with hoarseness or airway obstruction.[2] Elderly females are commonly affected with mean age onset of 50-80 years. Intraoral features of MMP include desquamative gingivitis, vesicles, erosions covered by pseudomem branes and ulcers.[3] MMP related autoantigens include BP180, BP230, laminin 332, type VII collagen among which autoantibodies BP180 and LM 332 have been most often reported.[4]

CASE REPORT

A 17-year-old female patient presented with 2 year history of blister formations and ulcerations in the oral cavity. The patient experienced pain and a burning sensation when consuming hot and spicy foods. Although a physician prescribed a topical antiseptic gel for the ulcers, it provided no relief upon application. There was no relevant personal history, and no systemic disease was reported.

An extraoral examination revealed no lesions in the eyes, skin, or genital areas. Intraoral examination showed generalized gingival erythema on both the labial and palatal surfaces of the maxillary and mandibular gingiva. Additionally, the maxillary gingiva exhibited a loss of stippling from the attached gingiva to the mucogingival junction on both the labial and palatal aspects.Single mucosal erosions seen in labial mucosa in relation to 24,25 region. [fig:1,fig: 2].The entire oral mucosa was tender on palpation. The provisional diagnosis given was Bullous pemphigus.

An incisional biopsy was performed on the labial mucosa in relation to 23 region and palatal mucosa in relation to 24,25 regions and sent for histopathological examination. The laboratory received 2 bits of formalin fixed tissue of size 8x5 mm and 5x6mm, which were firm in consistency.

Microscopically, keratinized stratified squamous epithelium of variable thickness with subepithelial split was observed. The underlying connective tissue stroma showed moderately dense inflammatory cell infiltrate predominantly lymphocytes and capillary vessels [fig:3,fig:4] The clinical and histopathological features favor a confirmatory diagnosis of MMP.

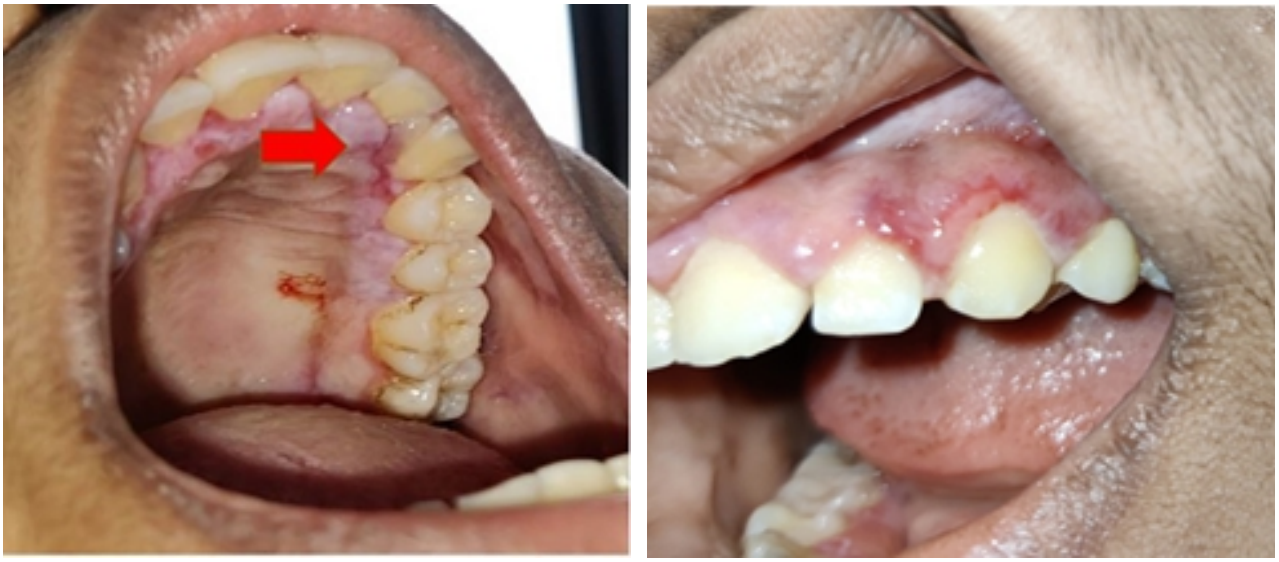


Figure 1&2 showing mucosal erosions in the palatal and labial mucosa

DISCUSSION

Mucous membrane pemphigoid (MMP) is a chronic, autoimmune vesiculobullous disease mainly affects mucosal tissues and, rarely seen in the skin. Mucous membrane pemphigoid was first reported by Wichmanns and Thost[5]. In the past, MMP was referred to as "benign mucous membrane pemphigoid," "cicatricial pemphigoid" and "ocular or oral–gingival pemphigoid."[6] In the literature on MMP, involvement of the skin is rare, but the conjunctiva may sometimes be affected, depending on the severity of the disease, and women are affected more than men. In our case also there were no skin lesions accompanying oral lesions. It is usually seen in adults over 40 years of age and occasionally seen in children. In our case the patient was a young female aged 17 years. There was no predisposing factors such as emotional stress, smoking, nutritional deficiency or systemic disease in that patient.[7]

The aetiology of MMP remains unclear. Severe mucosal inflammatory injury, medications (clonidine, D-penicillamine, indomethacin) viruses, UV light, and occasional occurrence with other autoimmune diseases, and genetic predispositions such as HLADQB1 are some of the suspected aetiological factors.[8,9]

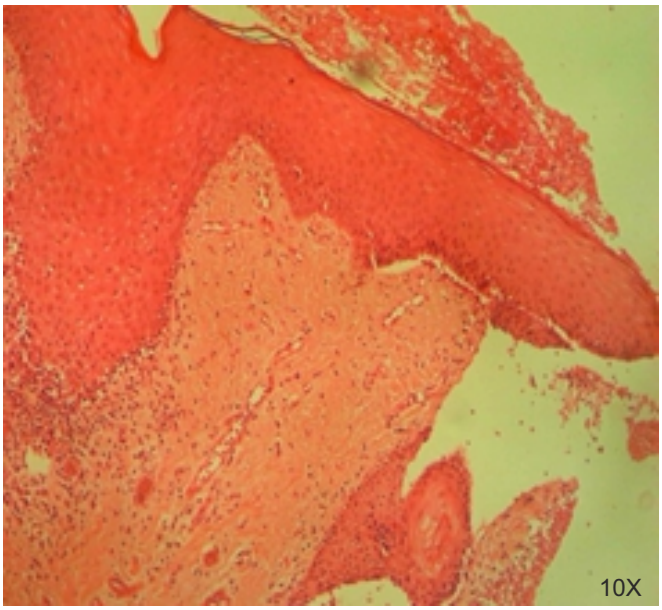
The most frequently presenting feature in MMP is desquamation of gingiva but other sites such as conjunctiva, nasal, esophageal, laryngeal, vaginal mucosa as well as skin may be involved.[10] There is no evidence in cited literature supporting the association of MMP and post-menopausal condition or habit of smoking.[11]

Many diseases, including bullous pemphigoid, pemphigus vulgaris and bullous lichen planus, should be considered in the differential diagnosis of MMP. The lesions of bullous pemphigoid and MMP closely resemble each other, the main difference being in their clinical presentation. The clinical course in patients with bullous pemphigoid is limited but the course in patients with MMP is usually protracted and progressive.[10]

The vesicular lesions of MMP are also similar to those of pemphigus vulgaris, particularly when the lesion is confined to the oral cavity. The lesions of pemphigus vulgaris appears first in the mouth, but soon spread widely on the skin. Acantholysis and intra-epithelial clefting are seen, and immunofluorescence reveals immunoglobulin G auto-antibody bound around the surface of the prickle cells in the epithelium, but in MMP, the G-auto antibody is bound along the basement membrane.[12]

On clinical examination bullous-type oral lichen planus may be confused with MMP. However, the histopathological subepithelial separation without degeneration of the basal layer of the epithelium seen in MMP is indicative in making the differential diagnosis. In bullous-type oral lichen planus, juxta epithelial inflammatory cell infiltration can be seen histopathologically. In addition, the presence of the characteristic clinical white striations at the periphery of the ulcerated areas in lichen planus helps to differentiate between the two conditions.[13]

Once the diagnosis has been established by microscopy and direct immunoflorescence, we can use topical



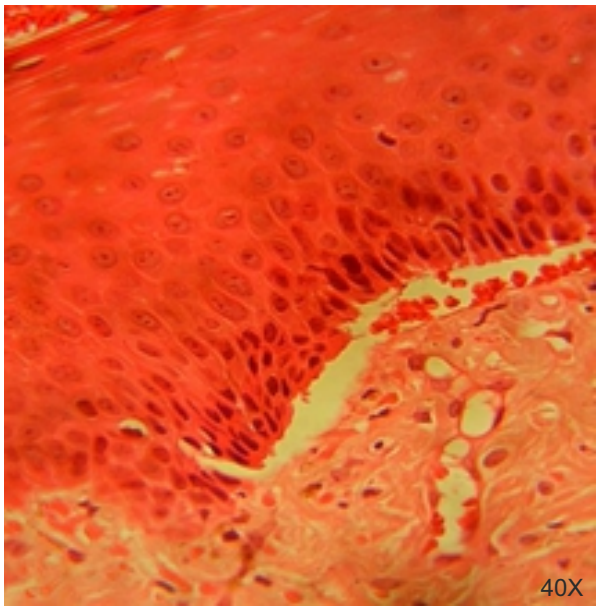


Figure 3 & 4 showing keratinized epithelium of variable thickness with subepithelial split

corticosteroids, either alone or in association with systemic corticosteroids for treatment.[14] If topical corticosteroids are unsuccessful, Immunosuppressive drugs such as methotrexate, azathioprine, levamisole, cyclophosphamide, and mycophenolate mofetil are used along with systemic corticosteroids.[15] Promising and dramatic responses have been achieved with Dapsone. Patient should be subjected to regular blood profile testing as prolonged dapsone use may induce hemolytic anemia.[16] Patients with less severe disease has also been treated with a tetracycline derivative or a combination of tetracycline and niacinamide.[17] Before choosing which medication to use, the oral hygiene of the patient should be improved. Intravenous immuno globulins, plasmapheresis, and Low level laser therapy (LLLT) constitute the recent development in the management of MMP.[18]

Conflict of interest: None

Source of support: Nil

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How to cite this article: Thomas N, Venugopal L, Steaphen P, George E. A case report of Mucous membrane pemphigoid in a 17 year old female. J Oral Biomed Sci 2024; 3:15-8.