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Case Report

Mucous membrane pemphigoid of the oral cavity and Genitalia: A case report

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ABSTRACT

Mucous membrane pemphigoid (MMP) encompasses a diverse array of chronic autoimmune conditions characterized by the development of blisters predominantly affecting mucous membranes in various regions of the body. Commonly impacted areas include the mouth, mucosal linings of the eyes, throat, genitalia, and nasal passages. While MMP typically manifests in older women, with peak incidence occurring between 50 and 70 years of age, rare instances have been observed in children. Symptoms of MMP often entail recurrent blister formation, subsequent rupture, and potential scarring, with complications possibly affecting the eyes and throat. This article presents a unique case involving a 70-year-old female presenting with lesions localized to the oral and vaginal mucosa. A comprehensive discussion on histopathological examination findings and treatment modalities is provided. Notably, the absence of ocular involvement in this case distinguishes it from typical presentations of MMP.

1. Introduction

Mucous membrane pemphigoid (MMP) presents as a vesiculobullous disorder predominantly affecting older individuals. Often, the diagnosis of MMP is overlooked, particularly when the manifestations are confined to the oral cavity, with desquamative gingivitis being a primary symptom. Unfortunately, many older patients displaying symptoms akin to MMP receive a misdiagnosis of "desquamative gingivitis." Consequently, they may receive inappropriate treatment regimens aimed at managing chronic pain and achieving disease remission. This mismanagement not only compromises the patient's oral health but also interferes with their ability to consume adequate nutrition, as the discomfort associated with eating exacerbates the condition. Immunologic studies support the autoimmune nature of MMP and infer that it may be a variant of bullous pemphigoid.[1–15]

2. Case Report

A 70-year-old female patient comes with a chief complaint of pain, difficulty in mastication and peeling of mucosa on the palatal region in the last three months. The patient was all right three months back when she noticed fluid-filled blisters of the size of peanuts on the anterior and posterior parts of the hard palate. The blisters ruptured after 2-3 days, leaving painful, eroded areas. The pain was present until eroded regions were in the mouth. The pain was continuous, dull, and aggravated by eating and drinking. The patient complained of a salty taste in the mouth at the time of the rupture of the blisters. This process has occurred 3- 4 times in the last three months. The patient also complained of similar eruptions in the genital region, which have subsided after taking medications prescribed by the general physician. Patients with no known medical

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condition applied Betnovate-C ointment on the genital area for genital lesions. The patient has undergone multiple extractions in the past 20 years and is conscious, cooperative and well-oriented with time, place and person.

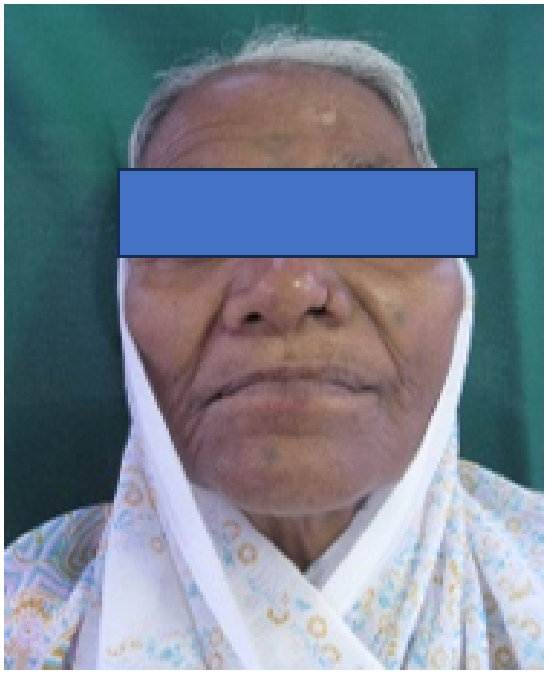

Figure 1:

2.1. *Extra-oral examination*

Bilaterally symmetrical face, non-palpable lymph nodes with bilateral synchronous movements of TMJ.

2.2. *Intraoral examination*

Three irregularly shaped erythematous areas of roughly 2.5 cm x 2.5 cm were seen extending.

1. Anteroposteriorly from the central part of 15 to the lateral aspect of maxillary tuberosity. Mediolaterally, 1cm from the midline of the palate till proper maxillary buccal vestibule.
2. Size: Roughly 1.5 cm x 1.5 cm extending anteroposteriorly from the distal end of 15 to the anterior part of maxillary tuberosity. Mediolaterally, roughly extending bilaterally 0.8cm (each side) from the midline of the palate.
3. Approximately 0.8 cm x 2.5 cm. Extending Anteroposteriorly distal end of 26 to the anterior part of maxillary tuberosity. Mediolaterally, roughly extending 1 cm from the midline of the palate to the buccal vestibule.

The smooth erythematous area is seen in the maxillary anterior region. Extent: anterioposteriorly: from the labial vestibular region covering the alveolar ridge and extending posteriorly till the incisive foramina. Mediolaterally: from the mesial side of 12 till the distal end of 23.

The genital mucosa showed generalised erythematous areas extending throughout.

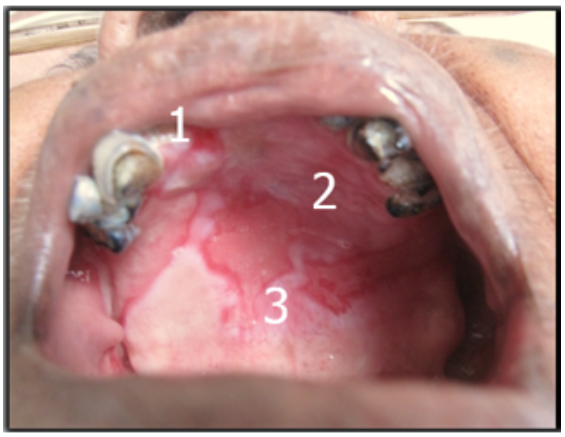

Figure 2:

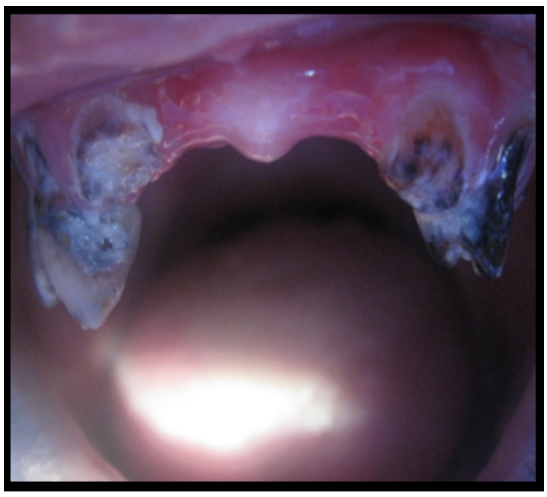

Figure 3:

2.2.1. *Clinical differential diagnosis*

1. Pemphigus
2. Mucous membrane pemphigoid
3. Bullous pemphigoid
4. Linear IgA
5. Recurrent Aphthous Ulcers
6. Erythema multiforme

2.3. *Investigations*

All haematologic inquiries were within normal range. An incisional biopsy was performed on the oral lesion and sent for histopathologic evaluation. The bit was 1cmx0.8cm in dimension, greyish-white in colour, and firm in consistency.

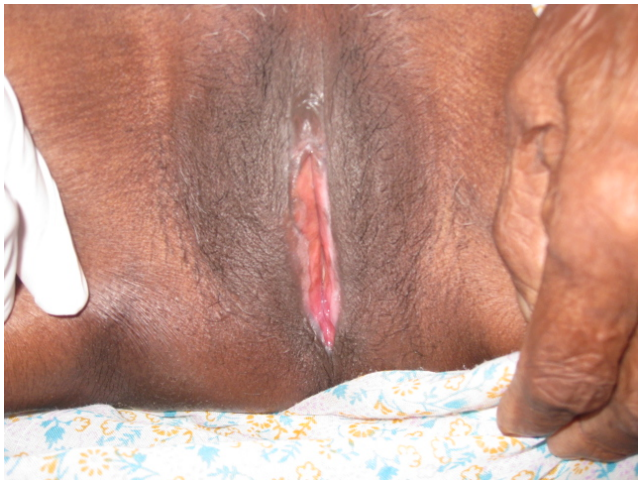

Figure 4:

The bit was divided into two halves longitudinally and sent for processing.

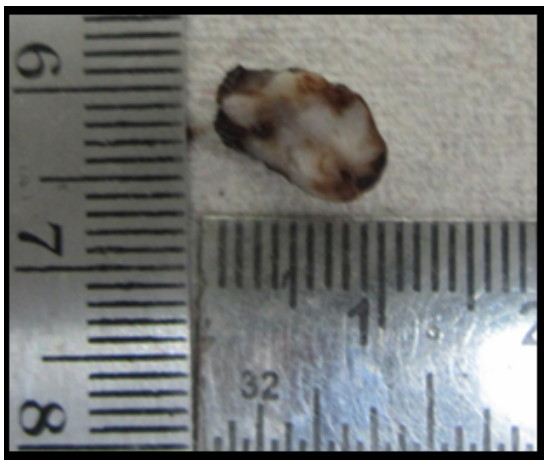

Figure 5:

2.4. *Histopathologic evaluation*

Microscopy reveals a sub-epithelial cleft with a variable inflammatory infiltrate rich in lymphocytes, neutrophils or both and plasma cells.

The bullae, or the cleft, is between the basal cell membrane and basal lamina.

Eosinophils are less frequent and often absent.

Older lesions reveal the appearance of granulation tissue and progressive scarring fibrosis. Ulcerated lesions were commonly seen. The infiltration typically contained plasma cells, histiocytes, and neutrophils. Marked dermal scarring was also seen.

Indirect immunofluorescence is positive in only 5 per cent of these patients, signifying a lack of circulating

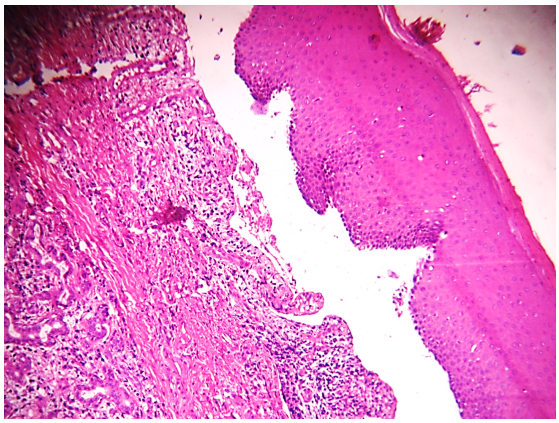

Figure 6: Marked dermal scarring is seen in addition to subepidermal vesiculation.

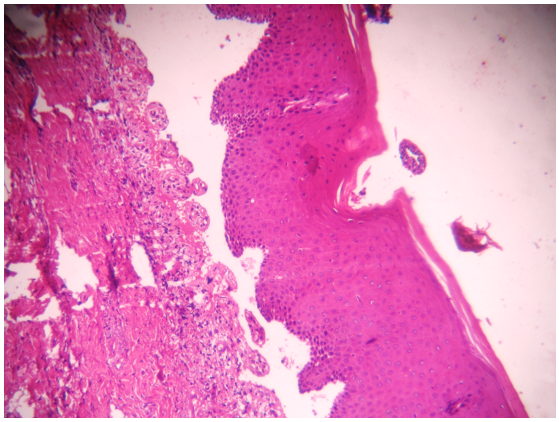

Figure 7: Characterestic Eosinophillic spongiosis is seen.

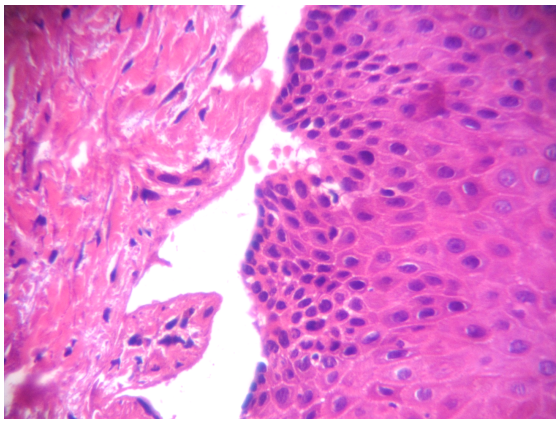

Figure 8:

autoantibodies. Hence, the final diagnosis was made based on clinical and histopathologic features.

Final Diagnosis: Mucous membrane pemphigoid

3. Discussion

In 1794, Wickmann documented the first instance of Mucous Membrane Pemphigoid (MMP) in a female patient. While the precise cause of MMP remains under investigation, several factors have been implicated, including severe mucosal inflammatory damage, certain medications (like clonidine, indomethacin, and D-penicillamine), viral infections, ultraviolet light exposure, and genetic predispositions such as the HLA-DQB1*03:01 allele.[16]

The association of HLA-DQB1*03:01 with MMP, initially observed in patients with ocular MMP,[15] later extended to all clinical variants. This allele is thought to play a role in T-lymphocyte recognition of antigens in the basement membrane zone.[17,18] The pathogenesis likely involves autoantibody-induced inflammation, complement activation, cytokine release, and leukocyte enzyme activity, leading to the detachment of basal cells from the basement membrane and the formation of vesicles beneath the epithelium.[19]

Specific antibodies targeting oral mucosal proteins, including the 168-KDa and α6 integrin, have been identified in oral MMP cases.[20] Additionally, a genotype of the interleukin four receptor A-1902 A/A has been linked to oral pemphigoid, with lower scarring rates observed in affected individuals.[21]

Diagnosis of MMP typically involves biopsy, with vesicular or perilesional tissue preferred over eroded areas. Histopathological examination often reveals a subepithelial split with an inflammatory infiltrate, including eosinophils. Direct immunofluorescence is essential for diagnosis,[22] showing linear deposition of IgG, C3, and occasionally IgA along the basement membrane zone.[23] Salt-split skin testing via indirect immunofluorescence can help detect circulating autoantibodies.

Financial constraints may limit additional diagnostic tests such as immunohistochemistry, but clinical evaluation can effectively differentiate MMP from other immunobullous disorders. For instance, the absence of skin involvement distinguishes MMP from bullous pemphigoid. In contrast, clinical features and the absence of specific triggers help exclude conditions like linear IgA disease and dermatitis herpetiformis.

In conclusion, while MMP diagnosis relies on a combination of clinical, histopathological, and immunological findings, thorough evaluation and exclusion of differential diagnoses are crucial for accurate management and treatment. Enzyme-linked immunosorbent assay systems for BP180 and laminin 332 are used to achieve target antigen.[24,25]

4. Conclusion

We had reported a patient with oral and genital lesions of MMP with different clinical presentations at the time of reporting. Dentists, ophthalmologists, and dermatologists need to monitor the patients constantly. MMP should be considered a differential diagnosis for any desquamative, vesiculobullous disease in an elderly female. The dentist's role in patient care is vital. The diagnosis is based on thorough history taking, clinical examination, and biopsy with histologic and immunofluorescence analysis. Amongst the numerous immunomodulatory drugs used in treating Oral mucous membranes, pemphigoid corticosteroids (topical and systemic) are used predominantly. Routine follow-up should be done to prevent exacerbations and remissions.

5. Source of Funding

None.

6. Conflict of Interest

None.

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