

# Management of a Case of Mucous Membrane Pemphigoid – A Case Report

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## Abstract

Mucous membrane pemphigoid (MMP) is a rare, chronic, autoimmune, sub-epithelial blistering, and erosive disease that affects the mucosal surfaces of the mouth (gingiva, movable mucosa, tongue, and palate), eyes, nose, nasopharynx, hypopharynx, larynx, esophagus, genitals, and/or anus. MMP may cause tissue destruction, chronic mucosal inflammation, pain, and scarring. Gingival inflammation and scarring might cause gingival recession and loss of teeth while conjunctival scarring may contribute to blindness.

**Keywords:** Autoimmune, Bulla, Corticosteroids, Immunofluorescence, Mucosa, Vesicle.

## INTRODUCTION

Mucous membrane pemphigoid (MMP) is a sub-epithelial, blistering disease predominantly involving the mucosal surfaces and occasionally involving the skin (in up to 30% of patients).<sup>[1]</sup> MMP had recently been classified into subgroups defined on the basis of the autoantibodies present. *Oral pemphigoid or OMMP* (patients with oral lesions only), *anti-epiligrin pemphigoid (AECp)*: The diagnosis of AECp was based on chronic sub-epithelial blistering lesions of mucous membranes and skin, *Anti-bullous pemphigoid (BP)* *Ag mucosal pemphigoid* (oral mucosal and skin lesions): In direct immunofluorescence (DIF) findings were similar to BP (high frequency of circulating auto antibodies). A high frequent reactivity to BP antigens was observed in ocular pemphigoid (OCP) (*patients with ocular lesions*).<sup>[2]</sup>

MMP has an annual incidence of 0.5–3.2/100,000 population. MMP is predominantly a disease of women, with a mean age of onset between 51 and 62 years.<sup>[2,3]</sup> Children are rarely affected.<sup>[2]</sup> One of every 15,000–40,000 patients treated in an eye clinic might have OCP. The most common areas of involvement were the oral cavity (85%) and conjunctivae (64%). The oral mucosa is often the initial site of MMP lesions. Vesicles or bulla were the presenting features and there could be a positive Nikolsky sign.<sup>[3,4]</sup> The MMP was one of the main causes of desquamative gingivitis (DG) and

DG was the main oral feature of MMP. Eye involvement usually began as chronic conjunctivitis. Scarring following repeated fibrosis could lead to the fusion of the scleral and palpebral conjunctivae (symblepharon). The conjunctiva might contract and invert the eyelid margins (entropion), leading to inversion of eyelashes onto the corneal surface with subsequent irritation (trichiasis). The combination of entropion and trichiasis might lead to blindness.<sup>[5]</sup> Cutaneous involvement was uncommon (up to 25% of patients). Some patients might have lesions of other stratified squamous epithelia where scarring might cause serious complications. In rare cases, MMP was disseminated.<sup>[2]</sup>

Diagnosis of oral MMP as a part of DG-associated disorders requires a logical succession of clinical, laboratory, and histopathology procedures. Differentiating these disorders is important because though they share a common clinical appearance, often their prognosis is different and requires different management. It includes classical histopathological features including a sub-epithelial split.<sup>[1-3]</sup> The DIF findings

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showed deposits of IgG and C3 in a homogeneous linear manner in the basement membrane zone (BMZ).

## CASE REPORT

A 40-year-old female patient reported to the Department of Periodontics, Dr R Ahmed Dental College and Hospital, Kolkata, with the chief complaint of burning sensation, redness and gum bleeding, and occasional pain for past 2 years with no systemic features (Figure 1). The disease got aggravated on eating spicy food with no relevant medical, drug, and topical substance use history. The intraoral examination revealed a bright red glazed lesion on gingiva in relation to attached gingiva maxillary and mandibular region with vesicle formation on the left maxillary canine-premolar region. Positive Nikolsky sign was observed in mandibular anterior region (Figure 2). Buccal and palatal mucosa seems fine. Local factors were present. After scaling and root planning, lesional with perilesional tissues was obtained for histopathological and immunofluorescence examination (Figure 3). The patient was motivated to maintain oral hygiene and was asked to rinse with 0.2% of chlorhexidine mouthwash twice daily. Histopathological report showed mild degree of spongiosis,

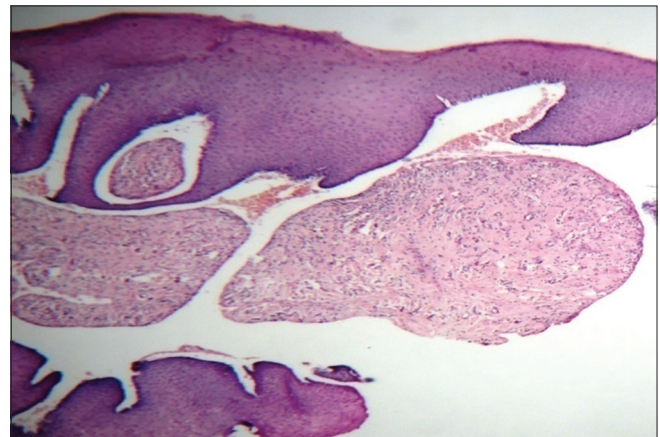
a sub-epithelial split with a chronic inflammatory infiltrate containing eosinophils, lymphocytes, and neutrophils in the lamina propria (Figure 4). The DIF findings showed deposits of IgG and C3 in a homogeneous linear manner in the BMZ (Figure 5).



**Figure 3:** Study tools used



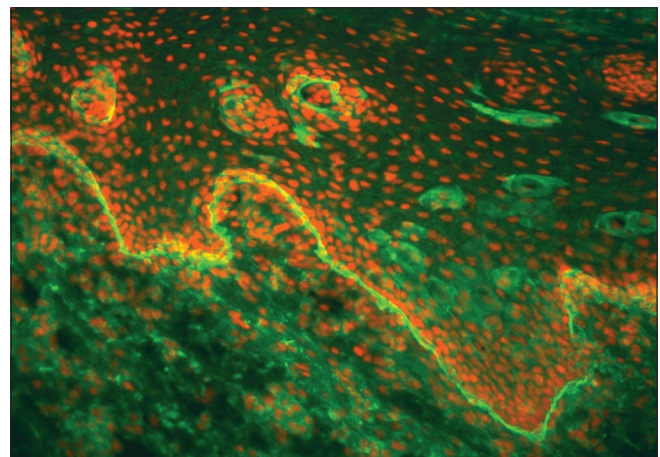
**Figure 1:** Vesicle formation on gingiva (Mucous membrane pemphigoid) presented as desquamative gingivitis.



**Figure 4:** Sub-epidermal split in H&E stained slide of Mucous membrane pemphigoid (40× magnification)



**Figure 2:** Nikolsky sign on gingiva



**Figure 5:** IgG deposits manifested in DIF presentation of Mucous membrane pemphigoid

Kenacort topical corticosteroid ointment 3 times daily was prescribed to the patient. The patient was also prescribed lycopene alongside. The patient was kept on regular follow-up every 2 weeks. The dose was eventually tapered to once daily in around 5 weeks. The patient was kept on follow-up for 6 months with no reoccurrence.

## DISCUSSION

DG is now recognized principally to be a manifestation of a number of disorders ranging from immune-mediated diseases (oral lichen planus [OLP], MMP, pemphigus vulgaris, erythema multiforme, erythema bullosa acquisita, dermatitis herpetiformis, contact stomatitis, lupus erythematosus, and linear IgA disease) to adverse reactions to a variety of chemicals or allergens (plasma cell gingivitis and CS) claimed MMP having the majority of presentation (90%) among their case series of 41 cases of DG.<sup>[7]</sup> A case series was published in 2008 involving 187 Caucasian subjects by Leao *et al.* who reported OLP as having the most common prevalence (70.5%) among the DG cases. MMP (14%) was reported to occupy the second position in their study report.<sup>[6,8]</sup>

Clinical presentations were grouped under three categories, that is, red, intermixed, and white lesions as per description. Red lesions predominated in the present case in accordance with clinical patterns observed.<sup>[6]</sup> Russo *et al.* 2010 observed that full mouth plaque score and full mouth bleeding score were higher in DG sites but not statistically significant<sup>[9]</sup> as observed in the present case report. Clinical presentation of pain and burning sensation was similar to observation reports of various workers.<sup>[6,10]</sup>

The clinical presentation of DG was polymorphous and the presence of plaque complicated the scenario.<sup>[7]</sup> In the absence of any ocular manifestations, diagnosis could not be provided to the current case of MMP. Histopathological assessment proved to be diagnostic in the present case with a sub-epithelial split with a chronic inflammatory infiltrate containing eosinophils, lymphocytes, and neutrophils in the lamina propria 1-3. DIF was considered to be gold standard for vesicular bullous lesions and was also considered as supportive for diseases like OLP and LE. Hence, it was added in the plethora of techniques for establishing diagnosis in addition to clinical and light microscopic examination.<sup>[7]</sup> The present case of MMP was found to have deposition of IgA, IgG, and C3. It was exactly analogous to the reports observed among 239 DG subjects.<sup>[7]</sup> Observations of the present study

were in harmony with the work of various researchers who had reported deposition of these substances to be as high as 70–80%.<sup>[11,12]</sup> Direct IF detection rate was observed as 70% among subjects with MMP.<sup>[13]</sup>

## CONCLUSION

DG is not a definitive diagnosis and was considered to be non-plaque-induced gingival disorder according to current periodontal diseases classification. Understanding a thorough diagnostic approach to clinical, histopathological, and immunofluorescence studies will yield proper diagnosis and hence better treatment protocol can be obtained.

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