

Mucous Membrane Pemphigoid: A Case Report

Isha Singla¹, Soheyl Sheikh², Shambulingappa Pallagatti², Deepak Gupta³

¹Senior Lecturer, Department of Oral Medicine and Radiology, M.M. College of Dental Sciences and Research, Mullana, Ambala, Haryana, India

²Professor, Department of Oral Medicine and Radiology, M.M. College of Dental Sciences and Research, Mullana, Ambala, Haryana, India

³Reader, Department of Oral Medicine and Radiology, M.M. College of Dental Sciences and Research, Mullana, Ambala, Haryana, India

ABSTRACT

Benign mucous membrane pemphigoid (MMP) is an autoimmune disease characterized by chronic vesiculo-bullous eruptions, predominantly on mucous membranes but occasionally on the skin. The diagnosis is hard to achieve due to similarity in clinical picture with other vesiculo-bullous lesions such as pemphigus or bullous pemphigoid, thereby, posing a diagnostic dilemma for the clinician. Histopathology can confirm the diagnosis; however, corticosteroids can be started to relieve the symptoms while confirmatory reports are not available as it is common treatment for most mucosal lesions. After

histopathology report, if diagnosis of MMP is established, anti-leprotic drug can be started along with corticosteroids. We report a case of MMP in a 43 year-old female who presented with difficulty in eating and pain in her mouth (buccal mucosa). Skin lesions were also present on legs and shoulder. Oral mucosal lesions were managed with anti-leprotic drug (dapsone), and topical and systemic corticosteroids. Such approaches not only result in immediate relief of symptoms and improved clinical outcomes but also provide a better quality of life for patients.

Keywords: Mucous Membrane Pemphigoid; Histopathology; Corticosteroids

INTRODUCTION

Bullous diseases are divided into two forms, either pemphigus with intraepithelial separation of cells or pemphigoid with subepithelial separation of cells. The pemphigoid family of diseases is part of a group of autoimmune subepidermal/subepithelial bullous disorders [1]. The pemphigoid group encompasses bullous pemphigoid, mucous membrane pemphigoid (MMP), linear IgA disease, chronic bullous dermatosis of childhood, and epidermolysis bullosa acquisita [2]. Benign MMP is an autoimmune disease characterized by chronic vesiculo-bullous eruptions, predominantly on mucous membranes but occasionally on skin [3]. It is also known as cicatricial pemphigoid as it has a chronic course and a tendency toward scar formation. It occurs worldwide in the people of all races [1]. MMP primarily affects middle-aged adults and females are more commonly affected than males [4]. It can affect any mucosal surface but the oral mucosa is the most common site and the nose, esophagus, larynx and genitals are less frequently

affected [5, 6]. Patients often present with the complaints of bleeding, pain, dysphagia and peeling of the mucosa but the most serious complications of the disease result from scarring [5]. Erosions and scarring of the mucosa might result in significant morbidity and treatment is usually challenging [6]. However, oral lesions, despite being extremely painful, generally are self-limiting and do not result in scar formation or organ dysfunction [4].

Diagnosis is based on history, clinical presentation, histopathology and immunofluorescence examinations. The management of a patient with MMP is complicated by the chronic nature of the disease [5].

Topical corticosteroids are the first line treatment for oral MMP, particularly for localized lesions. Extensive oral lesions or spread of the disease to other mucous membranes require prolonged administration of systemic corticosteroids [7]. Alternatively, steroid-sparing drugs such as dapsone, azathioprine, cyclophosphamide or intravenous immunoglobulins can also be given [7].

MMP has been reported in Indian literature quite

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Correspondence to: Dr Isha Singla

Address: Department of Oral Medicine and Radiology, M.M. College of Dental Sciences and Research, Mullana, Ambala, Haryana, India

E-mail: drishasingla@gmail.com

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Figure 1A/1B: Irregular ulcers on left & right side of buccal mucosa, covered with pseudo-membrane and having an erythematous floor



Figure 2: Extra-oral lesion on leg presenting as 0.5-1 cm in diameter ulcers covered with the scab and surrounding normal area

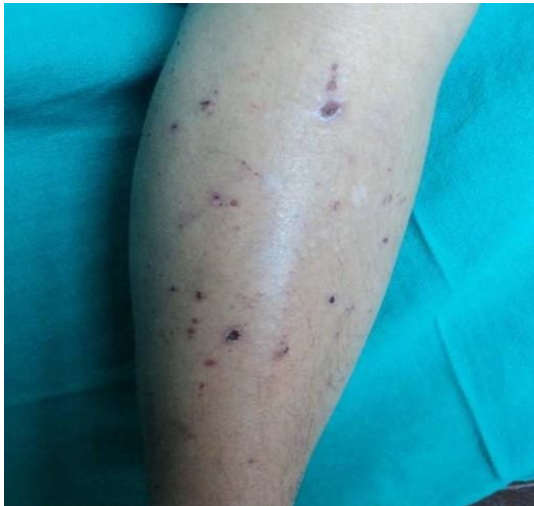
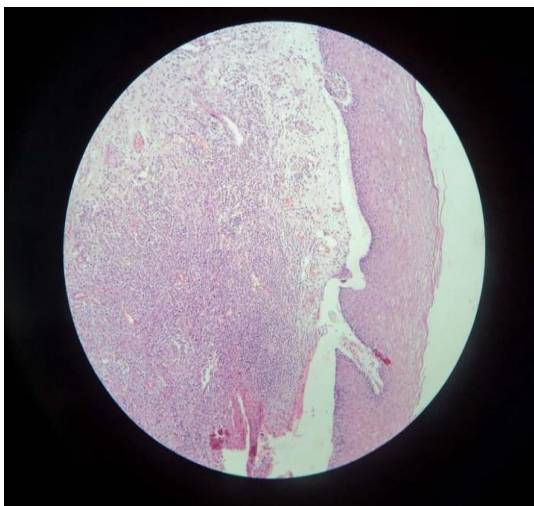


Figure 3: Histopathological picture showing epithelial connective tissue interface with subbasilar split



frequently [8]. However, we report a case of oral MMP that was managed successfully with topical steroid (triamcinolone acetonide 0.1%), systemic steroid and anti-leprotic drug (dapsone). This case is being presented because of difficulty in treatment approach of such patients. A different type of treatment plan was used which has been elaborated further.

CASE REPORT

A 43- year old female reported to the Department of Oral Medicine and Radiology with the chief complaint of difficulty in eating and pain in both left and right sides of mouth (buccal mucosa) for 2 months. Ulcers were associated with bleeding and surrounding area was slightly inflamed. The patient's past dental/medical history was unremarkable. Submandibular and submental lymph nodes were not palpable.

The oral examination revealed irregular ulcers measuring 2 cm X 2 cm and present on the left and right buccal mucosa, covered with pseudomembrane and had an erythematous floor. On the left side, ulcers extended 2.5 cm from the lip commissure to the posterior molar region and supero-inferiorly, ulcers extended 1cm above and 0.5 cm below the line of occlusion (Figure 1a). On right side, ulcers were 2 cm from the lip commissure to the posterior molar region and supero-inferiorly, ulcers extended 1.5 cm above and 1 cm below the occlusal line (Figure 1b). Skin lesions were present on legs and on left shoulder (Figure 2). The lesions on the skin were present as 0.5-1 cm in diameter ulcers covered with the scab. The surrounding area was normal. There were no signs observed or symptoms reported by the patient on the ocular or genital mucosa.

On palpation, oral lesions were tender and non-scrappable. Thin layer of epithelium peeled away in an irregular pattern leaving the denuded base.

The history and clinical findings of the patient helped to form a provisional diagnosis of pemphigus. A differential diagnosis of MMP and bullous pemphigoid was also considered due to similarity in clinical picture. An incisional biopsy was performed under local anaesthesia to establish a definitive diagnosis. Empirical treatment was started with topical steroids, hence the patient was prescribed topical corticosteroids (0.1% Triamcinolone acetonemide) 3 times a day. Histopathologically, the features were consistent with pemphigoid (Figure 3). Anti-leprotic drug (Dapsone) 150 mg/ day and systemic corticosteroid (6mg/day) were added to the treatment regimen. This was followed by scaling and oral hygiene instructions. The patient was re-evaluated every 2 weeks for three months. The skin and oral lesions had subsided within 16 weeks of starting the treatment. The patient was placed on maintenance dose of the topical corticosteroids and reinforcement of oral hygiene instructions were also given.

DISCUSSION

Cicatricial pemphigoid (CP) was first described by Cooper in 1857 [9]. In 1911, Thost separated CP from pemphigus [9]. The term MMP was first introduced by Lever in 1953 [9]. CP may remain localised to the oral cavity or it may be generalised. The most commonly involved sites are oral mucosa and ocular involvement but in the case reported above, oral mucosa and skin were the involved sites [10].

History, clinical examination, histopathology and immuno-fluorescence are helpful in diagnosis. The present case was diagnosed on the basis of classical clinical features and histopathology [8]. MMP can be difficult to treat and the results are often disappointing because the lesion can recur anytime despite compliance with treatment by the patient [6]. Early recognition and treatment can improve prognosis and avoid surgical intervention [8]. Therapeutic regimens for MMP are based primarily on clinical experience. Local therapy may be sufficient to control the disease, but systemic medications such as prednisolone, azathioprine, cyclophosphamide are indicated for severe ocular, laryngeal or esophageal involvement as well as for oral or cutaneous disease unresponsive to topical therapy [6].

Histopathologically, the MMP shows sub-epidermal vesicles with dense inflammatory infiltrate in the dermis or submucosa with lymphocytes, histocytes and plasma cell infiltrate [10]. In the present case, histopathological picture

revealed parakeratinized stratified squamous epithelium with underlying connective tissue. The epithelial connective tissue interface was predominantly flat with subbasilar split in most areas. Connective tissue showed intense inflammatory infiltrate of predominantly plasma cells. Numerous eosinophils and mast cells were also seen. These features are suggestive of benign MMP.

In the evaluation and treatment of MMP, the extent of disease, co-morbidities and the age of the patient are important considerations. Appropriate diagnosis and empirical treatment (with corticosteroids), before a definitive diagnosis is established, are important for managing this potentially debilitating disease. After confirmation of the diagnosis, other drugs can be added to the existing regime. Such approaches will not only result in improved clinical outcomes but also provide a better quality of life for the affected patients. Clinical awareness amongst health professionals and early recognition of the lesion should be emphasized.

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