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MUCOUS MEMBRANE PEMPHIGOID – A CASE REPORT

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Abstract

Benign mucous membrane pemphigoid (MMP) is an autoimmune disease characterized by chronic vesiculo-bullous eruptions, predominantly on mucous membranes but occasionally on skin. The diagnosis is hard to achieve due similarity in clinical picture with other vesiculobullous lesions like pemphigus, bullous pemphigoid, etc., 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 of mucosal lesions. After histopathology report, if diagnosis of MMP is established, anti-leprotic drug can be started along with corticosteroids for better relief. We report a case of mucous membrane pemphigoid in a 43 year-old female with difficulty in eating food and pain in left and right side of mouth (buccal mucosa). Skin lesions were also present on legs and shoulder. Oral mucosal lesions were managed with anti-leprotic drug (Dapsone), 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 the affected patients.

Keywords: mucous membrane pemphigoid, histopathology, corticosteroids

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Introduction

Bullous diseases are divided into two form- either pemphigus with intraepithelial separation of cells or pemphigoid with subepithelial separation of cells. The pemphigoid family of diseases is a part of the group of autoimmune subepidermal/subepithelial bullous disorders.[1]

The pemphigoid group encompasses bullous pemphigoid, mucous membrane pemphigoid, linear IgA disease, chronic bullous dermatosis of childhood, and epidermolysis bullosa acquista.[2] Benign mucous membrane pemphigoid 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. It has a chronic course and a tendency toward scar formation. It occurs worldwide in the people of all


Figure 1a & 1b. Irregular ulcers on right & left side of buccal mucosa, covered with pseudomembrane and having an erythmatous floor

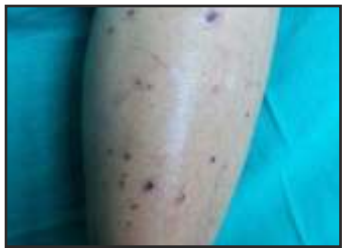

Figure 2: Extra-oral lesion on leg presenting as 0.5-1 cm in diameter ulcers covered with the scab and surrounding normal area

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

races.[1]

Mucous membrane pemphigoid primarily affects middle-aged adults and females are more commonly affected as compared to males.[4] It can affect any mucosal surface but the oral mucosa is the most common site[5] and the nose, oesophagus, larynx and genitals less frequently affected.[6] Patients often present with the complaint 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 of 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, histopathological and immuno-fluoresence examinations. The management of a patient with mucous membrane pemphigoid is complicated by the chronic nature of the disease.[5]

Topical corticosteroids are the first line of 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 be given.[7]

It has been reported in Indian literature frquently.[8] However, we report a case of oral mucous membrane pemphigoid that was managed successfully with topical steroid (Triamcinolone acetonide 0.1%), systemic steroid and anti-leprotic drug (Dapsone). This case was reported 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 food and pain in left and right side of mouth (buccal mucosa) since 2 months. It was associated with bleeding and surrounding area was slightly inflamed. Her past dental/medical history was unremarkable. Extraorally, lymph nodes were not palpable.

The intraoral examination revealed irregular ulcers measuring 2 cm X 2 cm present on the left and right buccal mucosa, covered with pseudomembrane and had an erythmatous floor. On left side, it extended 2.5 cm from the angle of mouth to the posterior molar region and supero-inferiorly, it extended 1cm above and 0.5 cm below the line of occlusion (Figure 1a). On right side, it was 2 cm from the angle of mouth to the posterior molar region and supero-inferiorly it extended 1.5 cm above and 1 cm below the occlusal line (Figure 1b). On physical examination, skin lesions were also present on legs and 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.

The inspectory findings were confirmed. On palpation, oral lesions were tender and non-scrapable. Thin layer of epithelium peeled away in an irregular pattern leaving the denuded base. The history and clinical findings of the patient helped form a provisional diagnosis of pemphigus. A differential diagnosis of mucous membrane pemphigoid 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 Kenacort (0.1%Triamcinolone acetonomide) 3 times a day.

Histopathologically, the features were consistent with those of pemphigoid (Figure 3). So other drugs were added to already existing regime of topical corticosteroids. Anti-leprotic drug (Dapsone) 150 mg/ day and systemic corticosteroid (6mg/day) were given to the patient. This was followed by scaling and oral hygiene instructions. The patient

was reviewed every 2 weeks for three months. The skin and oral lesions had subsided within 16 weeks of starting the treatment. The patient was put on a maintenance dose of the topical corticosteroids and reinforcement of oral hygiene instructions were given.

Discussion

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

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]

Mucous membrane pemphigoid can be difficult to treat and the results are often disappointing.[6] It is difficult to treat and results can be disappointing because the lesion can recur anytime despite of proper following of regime by the patient. Early recognition and treatment can improve prognosis and avoid surgical intervention.[8] Therapeutic regimens in mucous membrane pemphigoid are based primarily on clinical experience. Local therapy may be sufficient to control the disease, but systemic medications are indicated for severe ocular, laryngeal or esophageal involvement as well as for oral or cutaneous disease unresponsive to topical therapy.[6]

Histopathologically, the mucous membrane pemphigoid shows a sub-epidermal vesicles with a 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 mucous membrane pemphigoid.

Conclusion

In the evaluation and treatment of mucous membrane pemphigoid, the extent of disease, co-morbidities and the age of the patient are important considerations. Appropriate diagnosis and starting empirical treatment (with corticosteroids), before a definitive diagnosis is established, are important for managing this potentially debilitating disease. This helped to manage the condition of the patient in the present case as well. After confirmation of the diagnosis, other drugs can be added to the existing regime. This approach was followed in the above mentioned case also. 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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