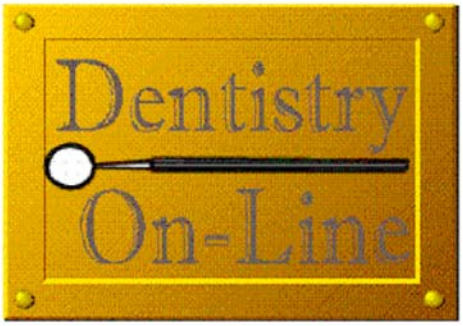

MUCOUS MEMBRANE PEMPHIGOID: CASE REPORT WITH REVIEW OF LITERATURE

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ABSTRACT

Mucous membrane pemphigoid (MMP) is a rare group of chronic autoimmune disorders. These disorders are characterized by vesiculobullous lesions that primarily affect various mucous membranes of the body. This is a case report of 43-year-old female patient who presented to our dental clinic with characteristic lesions of mucous membrane pemphigoid. A special emphasis on clinical and microscopic features and the importance of periodic revaluation of this case would be beneficial to the general dentists in its diagnosis and therapy.

Key Words: Mucous membrane pemphigoid, autoimmune disorder, vesiculobullous lesion, revaluation, therapy

INTRODUCTION

Mucous Membrane Pemphigoid (MMP) is a rare group of chronic autoimmune disorders characterized by blister producing lesions that primarily affects various mucous membranes of the body. It is the variant most likely to occur in the oral cavity and eyes although other membranes of the body may also be affected. MMP is now the preferred term for lesions only involving the mucosa and bullous pemphigoid when the condition primarily involves the skin[1]. Disease onset is usually between 40 and 70 years and oral lesions are seen as the initial manifestation of the disease in about two thirds of the cases. There is no racial or ethnic predilection although most studies have demonstrated a female-male ratio of approximately 2:1[2].

Patients with MMP commonly have gingival lesions resulting in sloughing during eating or tooth brushing followed by the involvement of the palatal and the buccal mucosae. Chronic oral soreness is common and can be worse with the intake of spicy food[2]. The clinical appearance is one of gingival erythema and loss of stippling, extending apically from the gingival margins to the alveolar mucosa. The desquamation may vary from mild,

insignificant patches to widespread erythema[3, 4]. The typical lesion is a small or large, clear –fluid blister, which breaks fairly rapidly in the mouth leading to pseudomembrane-covered, irregularly shaped erosions. These erosions have a yellowish slough and are surrounded by an inflammatory halo[1]. Oral scarring is rare. Some of the patients may also demonstrate a positive Nikolsky´s sign[4].Usually patients with oral involvement will lack major skin involvement, but the eyelids and genital mucosa are quite susceptible to the blistering phenomenon[4]. Patients with ocular involvement may present with pain or the sensation of grittiness in the eye and conjunctivitis. Erosions may be seen on the conjunctival surface. Involvement of the oropharynx may present with hoarseness or dysphagia .[1, 5].

The diagnosis of MMP is mainly based on history, clinical examination and biopsy of the lesions[1, 6]. When performing a biopsy, it is best to include a vesicle and a perilesional tissue not the erosion itself[1]. Cytology smears are not useful. Oral bullae demonstrate separation of the epithelium from the basement membrane with small amount of chronic inflammatory cells. Most lesions, however, are ruptured at the time of biopsy, hence, the subepithelial separation may only be found at the edge of an otherwise non specific inflammatory ulceration[1]. The

cause of autoimmune diseases is not known but genetic make up and environmental exposure may be factors. None of these diseases are contagious but certain medications may trigger the development of these disorders.

A case of MMP in a female patient with special emphasis on its clinical and microscopic features is presented along with a brief review of literature.

CASE REPORT

A 43 year old female patient presented in our dental clinic with a chief complaint of burning sensation of the gums for the past 8 months which would be aggravated on the intake of spicy food. In addition she also complained of small balloon like formation on the gums off and on which would crack down and was accompanied by pain. She denied the presence of any other systemic diseases or ongoing medical therapy at the time of arrival at our dental clinic .She also did not notice any changes in the eyes, skin or genital areas.

Clinical examination revealed generalized desquamation of the free and attached gingiva in the posterior areas. Traumatic

ulcerations due to deep bite were observed on the palatal aspect of anterior teeth. Applying gentle pressure on the normal mucosa induced a positive Nikolsky's sign. In the right posterior region an intact bullae was observed on the lingual aspect.

A mucosal biopsy was performed on an area adjacent to the bullae in the right mandibular area, so that intact tissue was submitted for laboratory examination. Tissue was submitted in 10% formalin for histopathological examination. It revealed parakeratinized stratified squamous epithelium with subepithelial clefting. Scattered mild, mixed chronic inflammatory cells were noted in the underlying connective tissue along with a few eosinophils. The basal cells were attached to the overlying epithelium and not to the basement membrane, suggestive of mucous membrane pemphigoid. The patient was referred to an ophthalmologist to rule out any conjunctival lesions. The ophthalmologist reported negative findings.

This disorder is extremely difficult to eliminate[2]. The goal of treatment is to suppress the blister formation and promote healing of the erosions[7]. Oral mucosal lesions are usually managed with topical steroids hence the patient was prescribed topical corticosteroids Kenacort (0.1%Triamcinolone acetonoide) 3 times a day followed by scaling and oral hygiene instructions.

As the increased overbite was aggravating the condition on the palatal aspect, the patient was referred to an orthodontist in our clinics for correction of the anterior deep bite.

The bite-opening appliance was a fixed anterior biteplane. A framework of 0.9mm wire supported the acrylic biteplane. The supporting wire was soldered to bands cemented on the first permanent molars. Wax spacers were used during acrylisation to relieve the palatal mucosa. Occlusal rests were fabricated on the premolars to prevent the bite plane from bobbing up and down and contacting the palatal mucosa. The rests were cemented using restorative composite. A clearance of 4 mm was seen between the posterior teeth on bite plane placement.

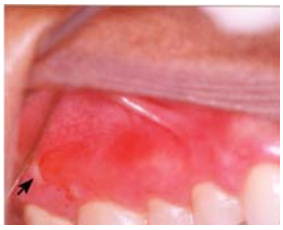

Figure 1. Clinical slide showing desquamation after the rupture of bulla.

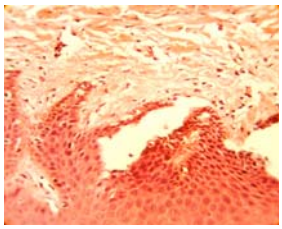

Figure 2. Histopathological slide showing sub-epithelial clefting.

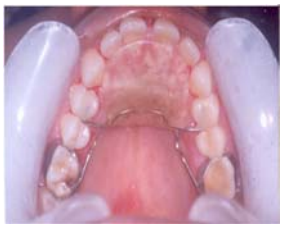

Figure 3. Correction of deep bite with anterior biteplane.

The patient was reviewed every 2 weeks, for the first one month, the lesions had subsided with topical steroids within 4 weeks of starting the treatment. The patient was asked to stop the topical application and reinforcement of oral hygiene instructions was given. Since the lesions can recur, the patient is under observation and the correction of increased overbite is also being reviewed.

DISCUSSION

MMP consists of a group of subepithelial blistering diseases primarily involving the mucosal surfaces. Mucous membrane pemphigoid has been known by many different names within the medical literature including benign mucous membrane pemphigoid, cicatricial (scarring) pemphigoid, and ocular cicatricial pemphigoid. A consensus group of researchers in 2002[1] determined that mucous membrane pemphigoid was the best designation for this group of disorders. The term "benign" mucous membrane pemphigoid was deemed inappropriate because of the potential for serious complications in some cases specially the lesions were very severe. The term "cicatricial" pemphigoid excluded affected individuals who do not develop scarring. Site-specific terms such as "ocular" cicatricial pemphigoid excluded individuals with multiple site involvement[1]. Since in this patient the characterestic lesions were restricted only to the mucosa it was diagnosed as MMP[2, 3, 8].

The disease is autoimmune in nature. Pathogenesis of MMP probably includes an autoantibody-induced, complement-mediated sequestration of leukocytes with resultant cytokine and leukocyte enzyme release and detachment of the basal cells from the basement membrane zone but there may also be complement-mediated cell lysis. MMP is characterized by junctional separation at the level of the basement membrane, which gives rise to a sub-basilar split[4], which was observed in the present histopathologic section. Pemphigus is similar in its clinical presentation to MMP but it produces acantholysis with cleavage of the spinous cell layer. In pemphigus the vesicle is intraepithelial whereas in MMP due to the separation of connective tissue from the epithelium the vesicle is subepithelial[1, 9]. In contrast to lichen planus the inflammatory infiltrate is non-specific in nature comprising of lymphocytes, plasma cells neutrophils with few eosinophils. Patients with MMP rarely have circulating autoantibodies to BMZ components hence indirect immunofluoresence is usually not indicated as a diagnostic procedure[6, 9].

Treatment of MMP is based on the severity of symptoms and the site involved[1, 2]. Patients with mild localized lesions may often

benefit from topical steroids such as beclomethasone dipropionate, betamethasone, clobetasol propionate, fluocinomide. Since our patient had lesions which were restricted to the oral mucosa only topical steroids were preferred as the first line of treatment [1, 7]. Patients with more extensive lesions can be prescribed systemic steroids like prednisolone [1]. Other treatment regimens, which were effective in certain resistant cases, were immunosuppressive agents such as azathioprine, cyclophosphamide, cyclosporine and dapsone. Sulphononamides and tetracyclines can also be implemented[10].

In conclusion, there is no cure for this disease due to its autoimmune nature. Lesions occur intermittently and may affect different parts of the oral mucosa at different times. The patient should be followed carefully by an ophthalmologist whether or not conjunctival involvement is seen at the time of oral lesion diagnosis. Dentists should be in close contact with an ophthalmologist and dermatologist in cases with possible conjunctival or skin involvement [2, 11].

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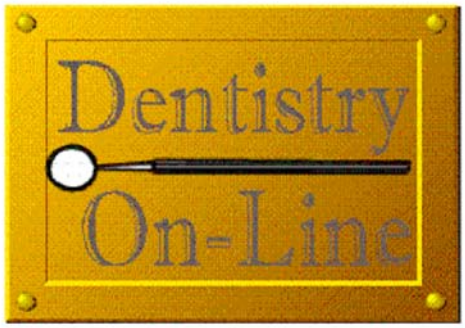

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