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Case Report

PRESENTATION AND MANAGEMENT OF ORAL SYMPTOMS OF PEMPHIGUS VULGARIS: A CASE REPORT
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ABSTRACT
Pemphigus Vulgaris is a rare autoimmune disease causing blistering of the skin due to the production of desmosomal antibodies. One of the most important clinical manifestations of pemphigus Vulgaris is the formation desquamated oral lesion on the gingiva. Such lesions are among the first to appear in patients who develop the disease and thus, knowledge of the presentation of the condition is of great importance as the dentist can be the first healthcare professional the patient suffering from the disease might seek. The clinician should not only assist with the early diagnosis but also provide symptomatic relief to the patient from oral discomfort as described in this case report.

INTRODUCTION
Pemphigus Vulgaris (PV) is an autoimmune Disease characterized by epithelial blistering affecting cutaneous and/or mucosal surfaces. The term is derived from the Greek word "Pemphix" meaning bubble or blister. The presentation of blisters on the skin and mucous membrane is due to the production of IgG antibodies against keratinocytes. The condition affects individuals of all races and ethnicity with both genders being affected equally. Clinically, the disease manifests as vesicles or bullae on the skin, filled with watery fluid. These vesicles, when rupture leave eroded surface on the skin. The oral lesions are characterized by blisters that rapidly rupture, resulting in painful erosions. The soft palate, buccal mucosa, and lips are commonly affected; however, the condition can involve any area of the oral cavity. Oral lesions are the preliminary symptoms of the disease in more than half of the cases. The current case report is of a 45 year old male presenting with lesions of unknown origin on the skin and gingiva.

CASE REPORT

Presentation
A 45 year old male patient was referred to the department of periodontics, VS Dental College and Hospital. The patient was referred by the department of dermatology, Kempegowda Institute of Medical Sciences, Bangalore. The patient presented with desquamative gingival lesions with burning sensation of mouth and ulcerative skin lesions over the skin. Medical history revealed that the patient was diagnosed with PV for which he was admitted in the hospital and was on medication for the same. Patient was under systemic and topical corticosteroid, immunosuppressant along with antiseptic mouth wash. Patient was referred due to complain of burning sensation on the gingiva. Nikolsky sign was positive. On extra oral examination, there were irregular lesion on the neck and cheek. Lesions were also present on other areas of the body like shoulder, arms and abdomen (Figure 1,2,3,4).

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Intraoral examination showed poor oral hygiene with desquamative lesions on the gingiva. Gingiva was friable and erythematous. There were no ulcerative lesions on the oral mucosa (FIG 5).

**Management**

The patient was admitted to Kempegowda Institute of Medical Sciences, Bangalore where he was diagnosed with PV and was on medication for the same.

The medication included topical and systemic corticosteroids and immunosuppressant. Management of oral lesion was directed towards providing symptomatic relief. Patient was prescribed topical corticosteroid (Tess Buccal Paste™). Oral prophylaxis was done with minimum damage to already friable soft tissue. After oral prophylaxis, patient was placed on chemical plaque control regime (Listerine Mouthwash™) for seven days. Patient was re-evaluated after 7 days. Patient reported subjective relief from symptoms including marked reduction of burning sensation of the gingiva. (Gig 7,8)

**DISCUSSION**

Pemphigus is a group of diseases that are characterized by the production of autoantibodies against intercellular substance and, therefore, classified as autoimmune disease. Viral infections have also been attributed for the production of antibodies hence, contributing to the etiology of the condition. Though the mechanism how antibodies directed towards desmosomes, which is the cell-cell junction, is fairly understood, the etiology the disease is still unknown. Desmosomes are responsible for holding the cells together.
Desmoglein-3 (Dsg 3) and Desmoliein-1 (Dsg 1) are proteins present in the desmosomes and are encoded by DSG3 and DSG1 genes, respectively. While both DSG3 and DSG1 are present in the skin, DSG3 is predominantly expressed in the oral epithelium. In Pemphigus Vulgaris, antibodies are produced against the Dsg 3 protein of desmosomes thus presenting the oral and extraoral symptoms.

Clinically, the condition presents itself with ulcerative lesions on the skin and mucous membrane. The primary lesion is a thin walled bulla which can be several centimetres in diameter. The bulla may containing clear fluid which on application of pressure, releases its content through the surrounding epidermis and further increases in size. However, conditions such as lichen planus and mucous membrane pemphigoid have similar presentation, hence, the diagnosis of pemphigus vulgaris should be based on clinical, histological and immunological tests.

Histologically, the conditions presents with acantholysis of the spinous layer. Tzanck smear to detect acantholytic cells is used as a screening procedure and rapid test for diagnosing oral PV. Immunohistochemistry of tissue samples from oral mucosa form PV patients show IgG in the intercellular junction of the keratinocytes.

Management of cases of pemphigus vulgaris is mainly directed towards prevention of production and autoantibodies and providing symptomatic relief to the patient. In our case report, the patient reported marked reduction of burning sensation of the gingiva following oral prophylaxis and administration of mouthwash.

Patient of PV are prescribed corticosteroids and immunosuppressant which reduce the production of autoantibodies against desmosomal proteins and prevent epithelial and mucosal ulceration.

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