PRESENTATION AND MANAGEMENT OF ORAL SYMPTOMS OF PEMPHIGUS VULGARIS: A CASE REPORT

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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)
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.

Management 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.

References

1. Shafer, Hine, Levy. Disease of skin. 6th ed. India: Elsevier; 2009. Shafer's Textbook of Oral Pathology; pp. 816-22.
2. Sandhya T, Avinash T, Daivat M.B, Sudhir B, Treville P “Pemphigus vulgaris” Contemp Clin Dent 2011 Apr-Jun; 2(2): 134-137.
3. Robinson NA, Yeo JF, Lee YS, Aw DC. “Oral pemphigus vulgaris: a case report and literature update.” Ann Acad Med Singapore. 2004 Jul; 33 (4 Suppl):63-8.
4. Nada M S, Anne N A, Raouf W A, Hussein S, Anne C J “Clinical and histological characterization of oral pemphigus lesions in patients with skin diseases: a cross sectional study from Sudan” BMC Oral Health. 2013 13: 66.
5. BystrynJC, Rudolph JL. “Pemphigus” Lancet. 2005 Jul 28 366 (9479):6173.
6. Pradeep A. R., S. Thorat M k, Arjun R “Pemphigus vulgaris associated with significant periodontal findings: A case report” International Journal of Medicine and Medical Sciences. Vol. 1 (8), pp. 297-301, August, 2009.
7. Fassmann A., Dvo.Akova N., IzakoviaovaHolla L., Vanuk J,1, Wotke J. “Manifestation of Pemphigus Vulgaris In The Orofacial Region. Case Report” Scripta Medica (Brno)-76 (1): 55-62, January 2003

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