

Pemphigus vulgaris in oral cavity: A case report

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يعتبر داء الفقاخ الشائع إصابة مناعية ذاتية تؤدي إلى تشكل نطفات حويصلية صغيرة داخل بشرة الجلد ومخاطية الفم. تبدأ الإصابة في الغالب في مخاطية الفم على شكل حويصلات وفقاغات مع تقرحات. يعتبر لأطباء الأسنان دورا كبيرا في الكشف المبكر عن هذه الإصابة وتشخيصها. تعرض هذه المقالة حالة لداء الفقاخ الشائع مع التأكيد بشكك خاص على وسائل التشخيص.

Pemphigus vulgaris is an autoimmune disease that produces intraepithelial blisters in the skin and the oral mucosa. The oral mucosa is often the first site to be affected by the disease. Initial lesions could appear in the oral cavity in the form of vesicles, bullae and ulcers. Dentists have a major role in the recognition and diagnosis of this disease. A case of oral pemphigus vulgaris is presented with special emphasis on diagnostic modalities.

INTRODUCTION

Pemphigus vulgaris (PV) is an autoimmune intraepidermal blistering disorder of the skin and mucous membranes.¹ Oral lesions have been reported as an initial manifestation of the disease in 50% of cases.² The fact that blisters on the oral mucosa are sometimes the first manifestation of the disease implies that the dental professionals must be sufficiently familiar with the clinical manifestations of PV to make a correct diagnosis and offer treatment. In this paper we describe a case of oral pemphigus vulgaris, with special emphasis on diagnostic modalities.

CASE REPORT

A 55-year-old male patient was referred to the Department of Oral Pathology, Government Dental College, Calicut, India with a three-month history of chronic oral ulcerations. The oral ulceration caused the patient considerable discomfort and significantly affected his normal oral function. This well built gentleman showed no lesions on the skin. He had

several superficial erosions on the buccal mucosa (Fig. 1), ventral surface of tongue (Fig. 2) and lip crusting was also present (Fig. 3) without any involvement of the gingiva or hard palate.

The patient stated that at least some of the erosions, particularly of the buccal mucosa started initially as vesicles. A vesiculobullous lesion was suspected based on the history of bullae formation, multiple chronic ulcerations and the apparent fragility of the oral mucosa experienced during examination. A provisional diagnosis of vesiculobullous lesion was made.

Routine blood investigations were carried out. The results of complete blood count, urine analysis and blood



Fig. 1. Erosion on buccal mucosa

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Fig. 2. Erosion covered by white slough on ventral surface of tongue.



Fig. 3. Lip crusting

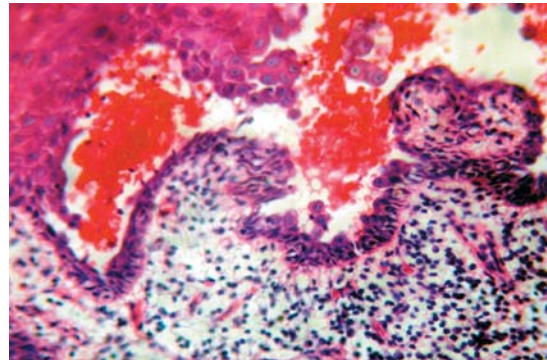


Fig. 4. Photomicrograph showing suprabasilar cleft with acantholytic cells.

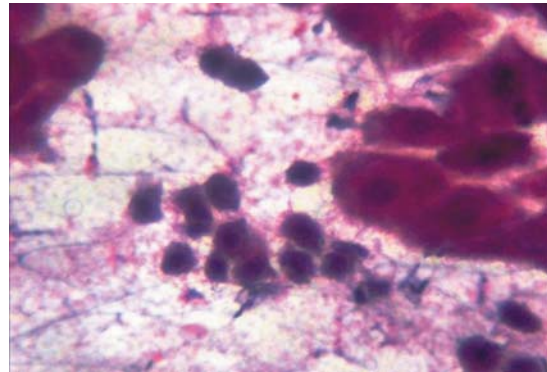


Fig. 5. Cytologic smear showing acantholytic cells.

glucose were within normal limits. Since the patient had a complaint of burning sensation, candida smear was advised. Surprisingly, candida smear revealed acantholytic cells in the absence of candida hyphae. A cytological smear was also carried out to confirm the presence of acantholytic cells. An incisional biopsy was performed on the lower lip and buccal mucosa and the specimens were submitted for conventional histology and direct immunofluorescence (DIF) studies. Indirect immunofluorescence was not done. Histopathological examination revealed suprabasilar clefting with acantholytic cells (Fig. 4). Exfoliative cytology (Fig. 5) showed acantholytic cells along with normal desquamated epithelial cells. Granular deposits of IgG and C3 in the intercellular spaces between the keratinocytes was

evident in the DIF technique (Fig. 6). A final diagnosis of oral pemphigus vulgaris was made.

The patient was commenced on systemic corticosteroids (Prednisolone) at an initial dose of 0.5 mg/kg/day. This initial dose failed to control the disease but after stepping up to 1mg/kg/day, there was marked improvement in two weeks. The patient was instructed to apply oral Cort E paste (0.1% triamcinolone acetonide and lignocaine). In addition, the patient was given nystatin mouth wash to prevent secondary fungal infection. Over the past 6 months, the prednisolone was reduced to 10mg / day. The patient is currently on the same daily low dose systemic corticosteroid therapy and to date, no other sites are involved.

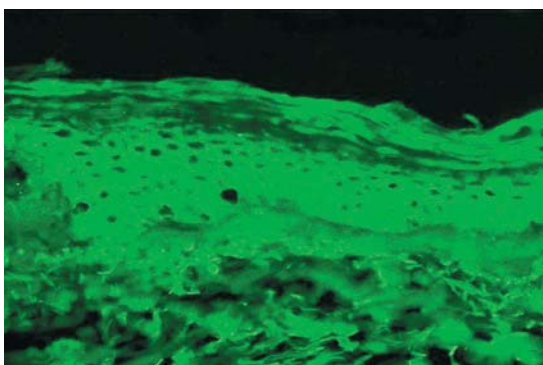


Fig. 6. Direct immunofluorescence demonstrating intercellular deposition of IgG

DISCUSSION

Pemphigus vulgaris affects the mucosa and the skin, resulting in superficial blisters and chronic ulceration.³ The etiology of PV is uncertain. PV is an uncommon condition affecting males and females in the 4th to 5th decade of life.² This is consistent with our case.

Diagnosis is usually based on three independent sets of criteria – clinical features, histology and immunological tests.⁴ The classic findings of oral PV are multiple chronic oral ulcers and a positive Nikolsky sign. Histologically, there is an intra epidermal blister associated with acantholytic cells.⁵ The acantholytic cells can also be confirmed using exfoliative cytology.⁵ In our case, these characteristic cells were evident even in candidal smear apart from cytologic smear and histopathologic examination.

Direct immunofluorescence (DIF) of oral mucosa biopsy specimens or Tzanck smears of PV is important for a definitive diagnosis, especially when the lesions are confined to the oral mucosa.⁶ DIF evaluation of fresh lesion specimens reveals IgG or IgM and complement fragments in intercellular spaces.⁷

The immunoperoxidase method proved to be a viable alternative to the use of DIF.⁸ It may be of particular value to the oral pathologist, who is more likely to be

dealing with oral PV. In addition, the interpretation does not require specialized microscopy and tissue sections and can be stored and retrieved for retrospective study.

Indirect immunofluorescence technique searches for circulating autoantibodies in the patient's serum and is usually performed after DIF studies reveal antibody deposits in the mucosa or skin.³ In PV, autoantibodies are produced against desmosomes,⁸ specifically desmoglein 3. Another important component of desmosomes is desmoglein 1, which is the target of autoantibody formation in pemphigus foliaceus that affects cutaneous sites.⁹

Specific enzyme linked immunosorbent assays (ELISA) are also available for detecting desmoglein 3 and desmoglein 1 autoantibodies.¹⁰ In future, the diagnosis and long-term follow up of patients with pemphigus would rely on detecting and quantifying antibodies against desmoglein proteins using ELISA.

CONCLUSION

Clarification and understanding of the spectrum of clinical manifestations of oral PV in correlation with histologic and cytologic findings are of great help in establishing a correct diagnosis. In addition to history and clinical examination, the diagnostic approach requires biopsy submitted for both routine histologic and DIF examination. Newer diagnostic tests and better monitoring of the disease process will help in early diagnosis and appropriate treatment of oral PV. Dental professionals can play a significant role in the early diagnosis and treatment of oral PV.

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