

## Oral Pemphigus Vulgaris: A Case Report and Review

Bharathi U<sup>1</sup>, Naresh Lingaraju<sup>2</sup>, Srisha Basappa<sup>3</sup>, Mahesh M S<sup>4</sup>

(1. Post graduate, Department of oral medicine & radiology, Farooqia Dental College & Hospital, Mysore, Karnataka.

2. Professor and Head, Department of oral medicine & radiology, Farooqia Dental College & Hospital,

3. Professor, Department of oral medicine & radiology, Farooqia Dental College & Hospital.

4. Reader, Department of oral medicine & radiology, Farooqia Dental College & Hospital, Mysore Karnataka)

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**Abstract:** Pemphigus Vulgaris (PV) is chronic autoimmune mucocutaneous disease that initially manifests as intraoral lesions, which may later spread to skin. Dental professionals must be efficient to recognize the clinical features of pemphigus vulgaris to ensure early diagnosis and treatment, so that it determines the favorable prognosis and course of the disease. This paper reports a case of pemphigus vulgaris and review of literature.

**Key Words:** Pemphigus Vulgaris, Nikolsky's sign.

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### I. Introduction

Pemphigus Vulgaris is a chronic mucocutaneous disease which usually manifests first in the oral cavity, which later may spread to skin or other mucous membrane. As it is a life threatening disease, it is important that dentist is able to recognize oral manifestations of PV and treat and refer appropriately.<sup>1</sup> Pemphigus Vulgaris is an immunopathologic dermatologic disease that usually occurs in patients between the ages of 30 and 60. It is characterized by the development of flaccid, easily ruptured intraepithelial bullae on apparently normal skin and mucous membranes. The oral cavity is frequently affected in the course of the disease. Intraoral lesions may appear in as many as 50% of the patients without a simultaneous affection of the skin. Although any part of the oral mucosa may be affected, areas exposed to mechanical irritation are most commonly involved. The lesions tend to occur most frequently on the buccal and palatal mucosa and on the gingiva. The oral lesions begin as bleb like blisters or as diffuse gelatinous plaques. Rupture of the bullae occurs in an early stage and may be caused by slight rubbing or minimal mucosal trauma. The lesions are usually painful. Untreated generalized Pemphigus Vulgaris may be fatal. Therefore, by recognizing the oral lesions of Pemphigus Vulgaris, the clinician has a responsibility in the early diagnosis of the disease, which is of the utmost prognostic importance.<sup>2</sup>

### II. Case Report

A 31 year old female patient visited department of oral medicine and radiology of Farooqia Dental College & Hospital, Mysore with a complaint of ulceration and burning sensation on the lower lip, tongue and cheek mucosa since 2 years duration. Patient complained of difficulty in eating and swallowing. History of recurrence was present. No skin lesions were evident. On clinical examination, multiple ulcerations were seen on right buccal mucosa, lower lip on the mucosal side, ventral surface & lateral border of the tongue. Ulcerations were of varying sizes and irregular in shape having ill defined erythematous margins which had yellowish floor. Surrounding mucosa appears normal. Apart from confirming all the inspeactory findings, ulcers were tender on palpation. No discharge was elicited from the ulcers. Base of the ulcers were not indurated. There was no fixity to underlying structures. Nikolsky's sign was positive. Reason for arriving provisional diagnosis as pemphigus vulgaris is, history of frequent diffuse extensive ulceration which was preceded with rupture of the vesicles & presence of positive nikolsky's sign. Differential diagnosis includes; A) Recurrent Aphthous Stomatitis (lesion may be severe, but individual lesion heal and recur & in case of pemphigus the same lesion continue to expand peripherally over a period of weeks or months). B) Primary Herpetic Gingivostomatitis (occur on mucosa that is tightly bound to periosteum of hard palate gingiva and alveolar ridge). C) Bullous Pemphigoid (negative nikolsky's sign & oral lesions are rare). D) Erythema Multiforme (target lesions are present & negative nikolsky's sign). E) Mucous Membrane Pemphigoid (80% of the cases shows eye involvement & lateral shifting of the blisters is not possible). Incisional biopsy was done. Smear shows typical features of rounded up separated keratinocytes (called acantholytic cells within the blister just above the basal layer of the epidermis suprabasal clefting may be reported).

PRE-TREATMENT PHOTOGRAPHS



Figure 1: Lesions on left lateral border of the tongue

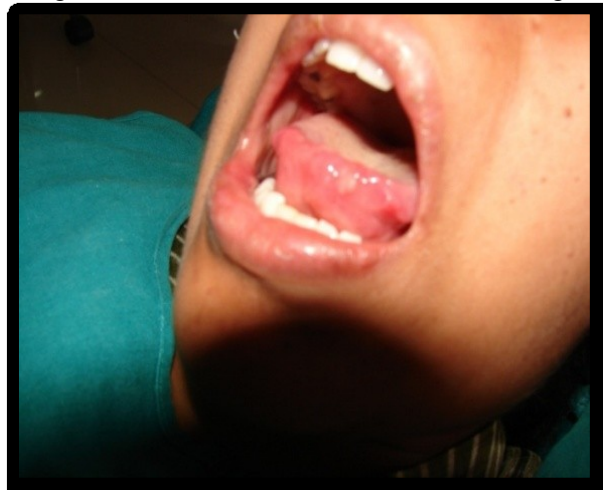


Figure 2: Lesions on the ventral surface of the tongue



Figure 3: Lesions on the lower labial mucosa

POST –TREATMENT PHOTOGRAPHS



Figure 1: Healed lesions of left lateral border of the tongue



Figure 2: Healed lesions of ventral surface of the tongue



Figure 3: Healed lesions of lower labial mucosa

HISTOPATHOLOGY IMAGES

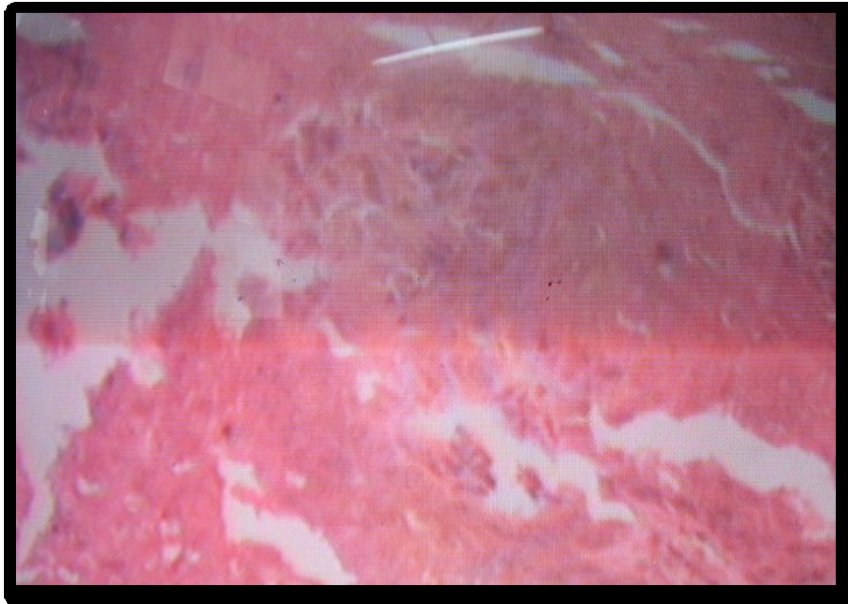


FIGURE 1

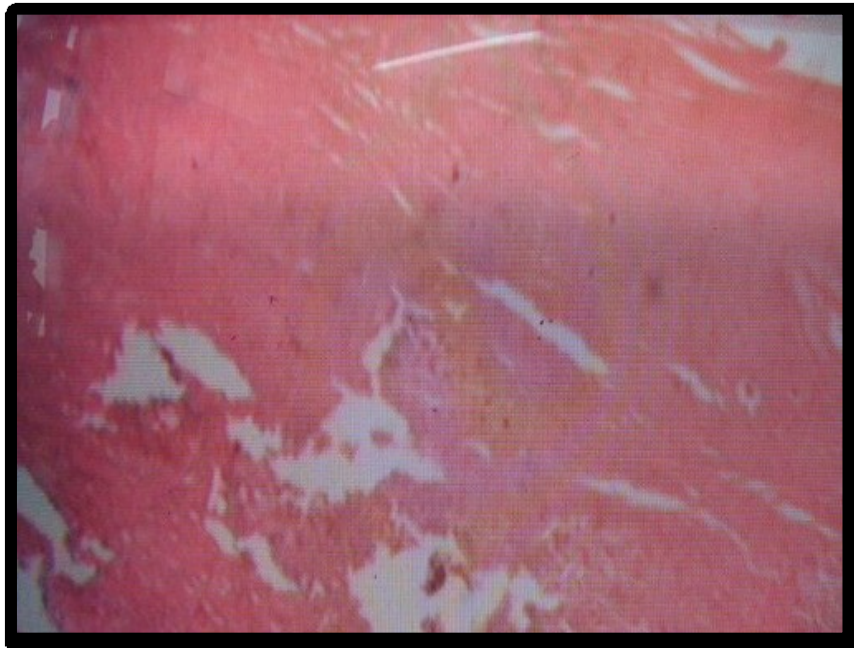


FIGURE 2

### III. Discussion

‘Pemphix’ in greek means “bubbles or blisters” and vulgaris in latin means “common”. Though pemphigus is a rare disease, pemphigus vulgaris is the commonest of all, comprising of the disease entity. Pemphigus vulgaris is a chronic autoimmune intraepithelial blistering disease. It always affects the mouth and it can be the initial site of presentation in 50% of cases before skin and other mucosal sites (oesophagus, pharynx, larynx, nasal and genital) become involved.<sup>3</sup> the reported incidence is 0.1-0.5 cases per 100,000 individuals worldwide per year. Its gender distribution is generally reported to be equal but few studies have shown it to be slightly predominant in women<sup>4,5</sup>. PV is characterized by intraepithelial blister formation that results from breakdown of cellular adhesion between epithelial cells. In most cases (70- 90%), the initial symptoms of disease appear on the oral mucosa. The oral lesions may be painful and may interfere with eating or other oral functions. The most common sites of oral involvement include the buccal mucosa, soft palate, labial mucosa, and gingiva, although any oral site may be affected.<sup>6</sup> The etiology of Pemphigus Vulgaris is still unknown although the disease has raised much concern. The pemphigus-group diseases are characterized by the production of autoantibodies against intercellular substances and, therefore, classified as autoimmune diseases.

The presence of a viral infection may also be involved in autoantibody production. When the disease is initiated by exogenous substances, such as medication, it is called induced pemphigus. Tsankov et al. Described the development of intra-oral pemphigus after exposure to the pesticide phosphamide in aerosol.<sup>7</sup> In PV, autoantibodies are produced against desmosomes<sup>8,9</sup>. The main antigen in PV is desmoglein (dsg) 3, a protein constituent of the desmosomes. Most patients with PV have circulating IgG autoantibodies against dsg3. These antibodies bind to the dsg3 on the epithelial cell membrane and may evoke acantholysis<sup>9,10</sup>. Most patients with oral lesions could be initially misdiagnosed, usually as aphthous stomatitis, gingivostomatitis, erythema multiforme, erosive lichen planus, or oral candidiasis, and may be improperly treated for months or years. Other differential diagnoses include dermatitis herpetiformis and cicatricial pemphigoid.<sup>6</sup> During active stage of the lesion, when lateral pressure is applied on the blister or perilesional skin or normal appearing skin, it results in removal of upper layer of epidermis known as nikolsky's sign. Two types of nikolsky's sign are described – wet nikolsky's sign in which after separation of epidermis the base of skin is moist, glistening, and exudative, and the dry nikolsky's sign, if the base of eroded skin is relatively dry. Active disease is readily appreciated by presence of a wet nikolsky's sign, while a dry nikolsky's sign indicates re-epithelization beneath a remnant blister top. The same phenomenon can be appreciated in the mucous membrane also<sup>3</sup>. In high risk patients with multiple oral lesions, rapidly progressing spread of the disease to other mucosal membranes such as the eyes, genital, esophagus or nasopharyngeal zone.<sup>4</sup> pregnancy may precipitate or aggravate PV, as reported in better known autoimmune diseases such as systemic lupus erythematosus<sup>10</sup>. For the definitive diagnosis of PV, the following criteria must be fulfilled: 1) the presence of appropriate clinical lesion(s), 2) confirmation of acantholysis in biopsy specimens, and 3) confirmation of autoantibodies in tissue or serum, or both<sup>9</sup> although PV is an intraepithelial blistering disease, intact bulla formation of the gingiva is rare and the disease manifests with nonspecific symptoms. Therefore, the diagnosis of PV tends to be delayed. Clinical, histopathological, and immunological examinations should be undertaken to obtain a definitive diagnosis of PV.<sup>11</sup> Although there is no real cure for pemphigus vulgaris, the disease can usually be successfully controlled with immunosuppressive drugs, such as azathioprine and prednisone.<sup>12</sup> Topical application of corticosteroids is effective if small, isolated areas of the oral mucosa are involved. The acute phase of pemphigus is associated with changes in gastric mucosa and this condition is further aggravated by ingestion of corticosteroids. At present, administration of azathioprin (azamun or imuran), which is added to achieve a decrease in antibody production, permits a lower dose of corticosteroids. The combined use of these drugs has recently improved the prognosis of pemphigus; in some patients it may even be possible to discontinue corticosteroid therapy.<sup>13</sup> Drugs reported most significantly in association with PV include Penicillamine, Captopril, Cephalosporin, Pyrazolones, Nonsteroidal Anti-Inflammatory Drugs (NSAIDs), and other thiol-containing compounds. Rifampin, emotional stress, thermal burns, ultraviolet rays, and infections (Eg, coxsackievirus, herpesviridae family) have also been reported as triggers for Pemphigus Vulgaris.<sup>14</sup> In any event, in the absence of systemic treatment, oral lesions of PV are almost invariably followed by involvement of the skin or occasionally other epithelia such as the esophagus when systemic immunosuppression will almost invariably be required.<sup>15</sup> The prognosis depends on the age of the patient, the initial severity, the extent of lesions, the interval between symptom onset and start of treatment, and the drug dose required to control the disease.<sup>16</sup>

#### IV. Conclusion

The earlier the diagnosis of Pemphigus Vulgaris is made and treatment is started, the less the suffering of the patient and the more favorable the prognosis. Dental treatment of patients with pemphigus vulgaris should be done with extreme care. The dermatologist should be consulted when surgical procedures are indicated, because of the prednisone therapy.

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