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*Case Note*

# A rare presentation of oral pemphigus vulgaris as multiple pustules

**Rimal J[1], Sumanth KN[2], Ongole R[3], George T[4], Chatterjee S[5]**
[1]Specialist Resident, [2]Associate Professor, [3]Reader, Department of Oral Medicine and Radiology, [4]Reader,[5] Post Graduate Student, Department of Oral Pathology and Microbiology, Manipal College of Dental Sciences, Mangalore

**Abstract**
Pemphigus vulgaris is an autoimmune blistering disease affecting the mucous membrane and skin. Ulcers, vesicles, bulla, erosions are the common manifestations of the disease. It is uncommon to find multiple pustular lesions in the oral cavity. Here, we report the first case of multiple pustules involving the lateral borders of tongue, buccal mucosa, hard palate, soft palate, vestibule and the gingiva of a 53 year old male. Histopathologic and Immunofluoroscence study was suggestive of pemphigus vulgaris. The condition improved with systemic corticosteroid along with adjuvant therapy.

**Key words:** pemphigus vulgaris, multiple pustules, immunofluorescence, corticosteroids

Pemphigus Vulgaris (PV) is a chronic mucocutaneous disease which usually manifests first in the oral cavity which later may spread to the skin or the other mucous membrane. As it is a life-threatening disease condition it is important that the dentist is able to recognize oral manifestations of PV and treat or refer appropriately. Apart from ulcers, vesicles, bulla, and erosions it can also be presented as pustular lesions. Here, we report a case of intraoral PV presenting as a multiple pustular lesions.

**Case Report**

A 53 years old male was referred from a private dental practitioner to the Department of Oral Medicine and Radiology, Manipal College of Dental Sciences, Mangalore. He complained of many ulcers in the mouth for the past 3 months. The lesion was insidious on onset and started on the lateral border of the tongue which subsequently involved the entire mouth. Progression of the lesions was rapid. There was associated discomfort while eating, speaking and swallowing which had significantly affected his quality of life. There was also a history of constipation and piles and was on medication (Ayurvedic). But there was no history of fever, other systemic drug intake, photosensitivity, skin lesions, arthralgia, abusive habits, genital lesions, malaise and weakness. Patient had visited a dentist 15 days before for the same problem and was put on Triamcinolone acetonide 0.1% cream thrice daily till the day of our examination . There was no relief of the symptoms; hence the patient was referred to the department for opinion.

On general examination, patient was moderately built, nourished for his age, conscious and co-operative. All the vital signs were within the normal limits. Visible areas of skin and nails were normal. On extra oral examination, skin over the face was normal with no abnormalities in the eyes. Bilateral submandibular group of lymph nodes were palpable which were soft, mobile and tender on palpation. Intraoral examination revealed poor oral hygiene with multiple missing teeth. There were multiple pustular lesions involving the bilateral lateral border of the tongue, buccal mucosa (Fig. 1 and 2), hard palate, soft palate(Fig. 3) , vestibule and the gingiva (Fig.4).Intra oral periapical radiograph was taken in order to rule out any periapical pathology in relation to 13, 14 region and was found to be negative. Each pustule was surrounded by an erythematous halo. The area was tender on palpation and the Nikolsky's sign was positive.

**Correspondence**
Dr. Jyotsna Rimal
Specialist Resident,
Department of Oral Medicine and Radiology,
Manipal College of Dental Sciences,
Mangalore-575001.
Email: josna21@yahoo.com

Based on the history and the clinical features the tentative diagnosis of pemphigus vegetans (Hallopeau) was given with the differential diagnosis of pyostomatitis vegetans, and oral Crohn's disease. Blood reports for complete blood picture, ESR, platelet count were within the normal limits. Incisional biopsy report showed a nonkeratinized stratified squamous epithelium with suprabasal clefting. Focal areas of epithelium also showed increased intercellular oedema with eosinophilic infiltration. Clusters of acantholytic cells showing degenerative changes were seen within the cleft. The underlying connective tissue stroma composed of loosely arranged collagen fibres with mixed infiltrate of inflammatory cells seen just beneath the epithelium (Fig. 5,6), and was suggestive of Pemphigus Vulgaris. Direct immunofluorescence report revealed Intercellular deposit of IgG and weak intercellular deposits of C3 in the basal epithelium (Fig. 7) and negative for IgM, IgA and Fibrinogen. On indirect Immunofluorescence IgG deposition in intercellular stroma was 1: 10 and C3 deposition was weak with the ratio of 1: 80. Pus collected from the pustule was sent for culture and was negative for any microbial agent. The patient was referred to Department of Gastroenterology for consultation regarding the patient's gastrointestinal condition; colonoscopy was performed and was negative for Crohn's disease and ulcerative colitis. Based on all these features, a final diagnosis of Pemphigus vulgaris was made.

As an initial treatment, patient was put on Prednisolone 10mg thrice daily for 7 days, followed by twice daily for 4 days, then 5mg thrice daily for another 4 days. The patient was recalled after 15 days for review. On the 15 days follow up, old lesions had healed and new lesions occurred on the right lateral border of the tongue and floor of the mouth. Lesions on the other areas of the oral cavity were still persisting. Patient was advised following medications:

- Prednisolone 5mg thrice daily for 7 days, tapered to twice daily for another 7days, followed by once daily for7 days.
- Dapsone 100mg once daily for 6 months.
- Azathioprine 50mg once daily for 15 days.

After 15 days follow up, old lesions had healed and new lesions were present on the right buccal mucosa. At this visit, patient complained of skin rashes for the last 1 month. This was attributed to the side effect of Prednisolone. Prednisolone and Azathioprine was discontinued and only Dapsone was continued because of its steroid sparing action and thus reduces the steroid dependency. Two months follow up revealed a single new lesion on the right buccal mucosa. The skin rashes had subsided. There was improved oral functions and overall, the quality of life.

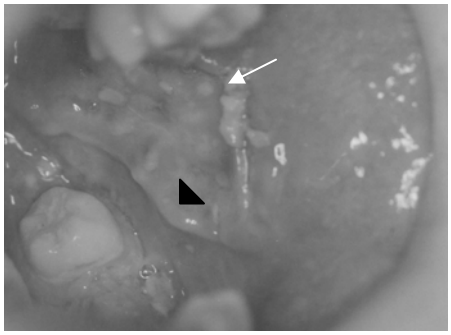

**Fig 1:** Multiple pustules on the left buccal mucosa (arrow) and the vestibule (arrowhead)

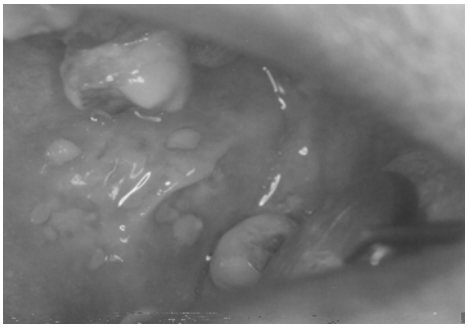

**Fig 2:** Similar pustules on the right buccal mucosa (arrows)

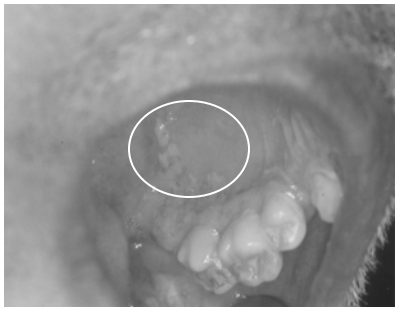

**Fig 3:** Palatal pustules (left side) with positive Nikolsky's sign

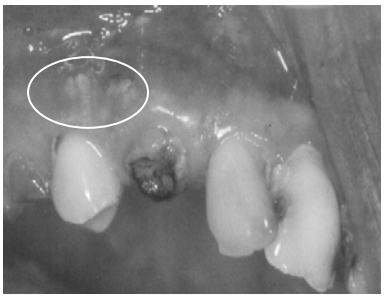

**Fig 4:** Involvement of the anterior gingiva on the right side

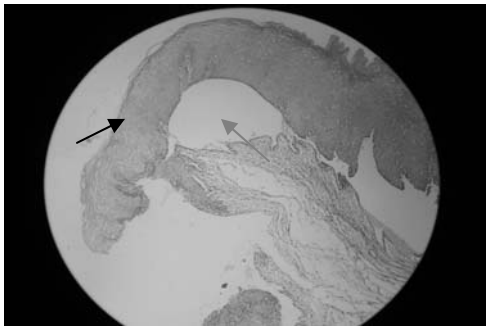

**Fig 5:** Low power photomicrograph of nonkeratinized stratified squamous epithelium (black arrow) with suprabasal clefting (grey arrow)( H&E original magnification X 10)

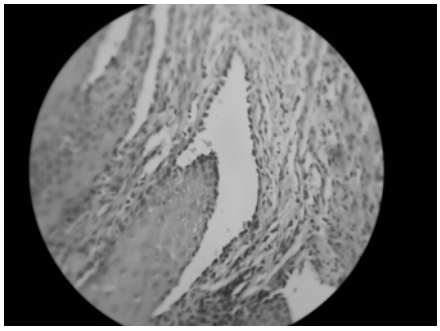

**Fig 6:** High power photomicrograph of clusters of acantholytic cells showing degenerative changes within the cleft. (H&E original magnification X 40)

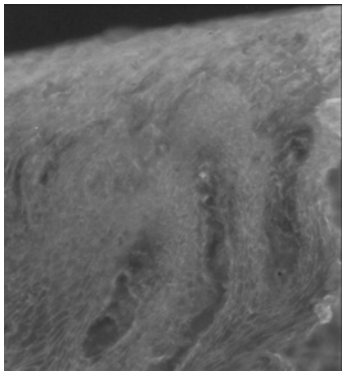

**Fig 7:** Intercellular deposition of IgG and weak intercellular deposits of C3 in the basal epithelium

**Discussion**

"Pemphix" in Greek means 'bubbles or blisters'[1] and "vulgaris" in Latin means 'common'[2]. Though pemphigus is a rare disease, pemphigus vulgaris is the commonest of all, comprising of 80% of the disease entity. The term pemphigus was originally named by Wichman in 1791[1]. It is mediated by circulating auto antibodies directed against keratinocyte cell surface. Mortality from pemphigus vulgaris before the development of effective therapies was as high as 90% and was often fatal mainly from dehydration or secondary systemic infection. Today, with treatment, it is approximately 5-15%[1]. Prognosis is worse in patients with extensive disease and in older patients. Oral lesions often represent the first clinical manifestation. It is thus important that the dentist is able to recognize oral manifestations of PV and treat or refer appropriately[3]. There is damage to desmosomes by antibodies directed against the extra cellular domains of cadherin-type epithelial cell adhesion molecule- the Desmoglein (Dsg) with immune deposits intra-epithelially, and loss of cell-cell contact (acantholysis), leading to intraepithelial vesiculation. The main antigen in pemphigus vulgaris is Dsg-3 but 50% of patients also have auto antibodies to Dsg-1. The proportion of Dsg-1 and Dsg-3 antibodies appears to be related to the clinical severity of pemphigus vulgaris those with only Dsg-3 antibodies have oral lesions predominantly. In PV mainly IgG antibodies are deposited intercellularly directed against the extra cellular domains particularly of Dsg3 and as oral epithelium expresses largely Dsg3 (skin expresses Dsg1 as well as Dsg3), oral lesions appear at an early stage[4]. Possible etiological agents are listed in the Table 1[7,4]

It is very important to differentiate Pemphigus Vulgaris and Pemphigus vegetans, though P. vegetans is a variety of
P. Vulgaris, lesions presenting as pustule has not been reported. Early lesions of P. vegetans may be similar to Pemphigus vulgaris and shows suprabasillar acantholysis. In later stages in Hallopeau type, pustules along with vegetative growth can be seen[6]. Histopathologically, intraepithelial abscess filled with eosinophillic infiltrate is diagnostic of P. vegetans along with papillomatosis, hyperkeratosis, and irregular downward extension of strands of epithelium[5].

Due to its chronic course and life threatening nature, a full history and examination, biopsy examination and appropriate histopathological and immunological investigations are frequently indicated. Biopsy of perilesional tissue, with histological and immunostaining examination, are essential to the diagnosis. ELISA has been proved to be helpful even for pemphigus Vulgaris. With appropriate dilution, ELISA detection of autoantibodies to Dsg3 and Dsg1 can provide useful information for assessing disease activity[4]. Management mainly comprises of corticosteroids with/without adjuvant[8] and drugs used are listed in Table 2.

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**Table1:** Possible etiological agents of the Pemphigus Vulgaris

| | |
|---|---|
| **Diet** | Members of Botanical family Allium<br>Garlic<br>Onion<br>Leeks |
| **Drugs** | Penicillamine<br>Captopril<br>Phenol drugs<br>Rifampicin<br>Diclofenac<br>Other ACE Inhibitors |
| **Viruses** | Herpes simplex<br>Human herpes virus 8 (HHV8) |
| **Other factors** | Possible contribution of high oestrogen levels (pregnant)<br>Increased exposure to pesticides |
| **Association with other disorders** | Rheumatoid arthritis<br>Lupus erythematosus<br>Myasthenia gravis<br>Pernicious anaemia |

**Table 2:** Drugs used in treatment of Pemphigus Vulgaris

| |
|---|
| **CORTICOSTEROIDS** |
| **LOCAL** |
| **SYSTEMIC** |
| Prednisolone |
| Triamcinolone acetonide |
| Betamethasone |
| Dexamethasone |
| **ADJUVANTS** |
| **ANTI-INFLAMMATORY** |
| Dapsone |
| Tetracycline |
| Minocycline |
| Gold Salts (aurothiomalate) |
| Retinoid |
| Thalidomide |
| **IMMUNOSUPPRESSORS** |
| Cyclophosphomide |
| Azathioprine |
| Methotrexate |
| Cyclosporin |
| Chlorambucil |
| **IMMUNOMODULATING THERAPY** |
| Plasmapheresis |
| Intravenous Gamma Globulins |

**Conclusion**

Pemphigus Vulgaris is a chronic mucocutaneous disease which usually manifests first in the oral cavity which later may spread to the skin or the other mucous membrane. Apart from ulcers, vesicles, bulla, and erosions it can also be presented as pustular lesions. Hence, dental professionals must be sufficiently familiarized with the intraoral clinical manifestations of pemphigus vulgaris to ensure early diagnosis and treatment, which determines the prognosis and course of the disease.

**References**


1. www.emedicine.com/DERM/topic319.htm accessed on 05/10/06
2. Neville BW, Damm DD, Allen CM, Bouquot IE. Oral And Maxillofacial Pathology, 1st edition 1995: Pg.559-561
3. Iamaroon A, Boonyawong P, Klanrit P, Prasongtunskul S, Thongprasom K. Characterization of oral pemphigus vulgaris in Thai patients. J Oral Sci. 2006 Mar; 48(1):43-6.
4. Black M, Mignogna MD & Scully C. 2005: *Number II* Pemphigus vulgaris. *Oral Diseases* 11 (3), 119-130
5. McCarthy P.L, Shklar G. Disease of Oral Mucosa 2nd edition 1980:Pg.276-287
6. Greenberg MS, Glick M. Burket's Oral Medicine, Diagnosis and Treatment, 10th edition 2003, Pg 71
7. Camacho-Alonso F, Lopez-Jornet P, Bermejo-Fenoll A. Pemphigus Vulgaris. A presentation of 14 cases and the review of the literature. Med Oral Patol Oral Cir Bucal 2005:10, 282-288
8. Lagha N B, Poulesquen V, Roujeau J. C, Alantar A, Maman L. Pemphigus Vulgaris: A Case-Based Update JCDA Oct 2005:71 (9) Pg. 667-674