The Journal of Dental Panacea 2022;4(4):183–186

Content available at: https://www.ipinnovative.com/open-access-journals

The Journal of Dental Panacea

Journal homepage: https://www.jdentalpanacea.org/

Case Report

# Pemphigus vulgaris – A case report

**Varsha Patel G.B[1,*], Sujatha G.P[1], Sangeeth Siddabasappa[1], Ashok L[1]**

[1] *Dept. of Oral Medicine and Radiology, Bapuji Dental College and Hospital, Davangere, Karnataka, India*

ARTICLE INFO

*Article history:*
Received 21-07-2022
Accepted 11-11-2022
Available online 24-11-2022

*Keywords:*
Autoimmune disorders
Oral mucosa
Pemphigus vulgaris

ABSTRACT

An important group of lesions in the oral cavity are autoimmune mucocutaneous diseases. The diagnosis of pemphigus vulgaris is made using clinical, histologic, and immunologic criteria, with older patients being the most frequent victims. Pemphigus vulgaris is an autoimmune blistering disorder that typically affects the oral mucosa. Autoantibodies against the desmoglein component of keratinocytes are the hallmark of this disease. Clinically, it appears as vesicles, bullae, or desquamative gingivitis, and histological analysis reveals that acantholysis is present. The diagnosis is typically confirmed by immunofluorescent evidence of IgG antibodies against desmoglein. Pemphigus vulgaris must be considered in the differential diagnosis of oral vesiculo-bullous lesions due to its clinical similarities to other oral illnesses. The physician can provide appropriate care by using diagnostic and therapeutic methods with sufficient understanding. This paper presents a case report of pemphigus vulgaris affecting oral cavity. This paper presents a case report of pemphigus vulgaris affecting oral cavity.



## 1. Introduction

Skin and mucous membrane blistering are hallmarks of autoimmune bullous disorders of the upper aerodigestive tract. The term "pemphigus" refers to a category of autoimmune skin and mucous membrane blistering illnesses. Pemphigus vulgaris (PV), a bullous disease affecting the skin and mucous membranes, is a member of this category and may be fatal if not treated with the proper immunosuppressive medications.[1]

PV has an incidence of 0.1–0.5 cases per 100,000 people per year1 and equally affects both sexes. The average age of onset is between 50 and 60.[2]

Pemphigus vulgaris, pemphigus vegetans, pemphigus foliaceus, pemphigus erythematosus, paraneoplastic pemphigus (PNP), and drug-related pemphigus are the main variations of the disease. Over 80% of cases of pemphigus are Pemphigus Vulgaris, making it the most prevalent type.[3]

IgG autoantibodies that attach to keratinocyte cell surface molecules are linked to blisters. PV antibodies attach to desmosome-free regions of the keratinocyte cell membrane as well as to desmosomes in keratinocytes.[4] Anti-inflammatory drugs, drugs that decrease the production of antibodies, and procedures that improve antibody elimination are only a few of the therapy options available for this disorder.[5] The purpose of this case study is to describe a typical instance of this ailment and to draw attention to the various therapeutic choices available.

## 2. Case Report

A 59-year-old female patient reported with the chief complaint of ulcers in the mouth and difficulty in swallowing food since 1 year. Patient first noticed difficulty for swallowing solid food and liquid which progressively

* *Corresponding author*.
*E-mail address*: ashoklingappa1@gmail.com (Varsha Patel G.B).

https://doi.org/10.18231/j.jdp.2022.039

increased in severity. The patient had noticed ulcers of the mouth which bleed on brushing. The patient did not report any skin lesions or involvement of other mucosal sites. A review of medical and family history was noncontributory.

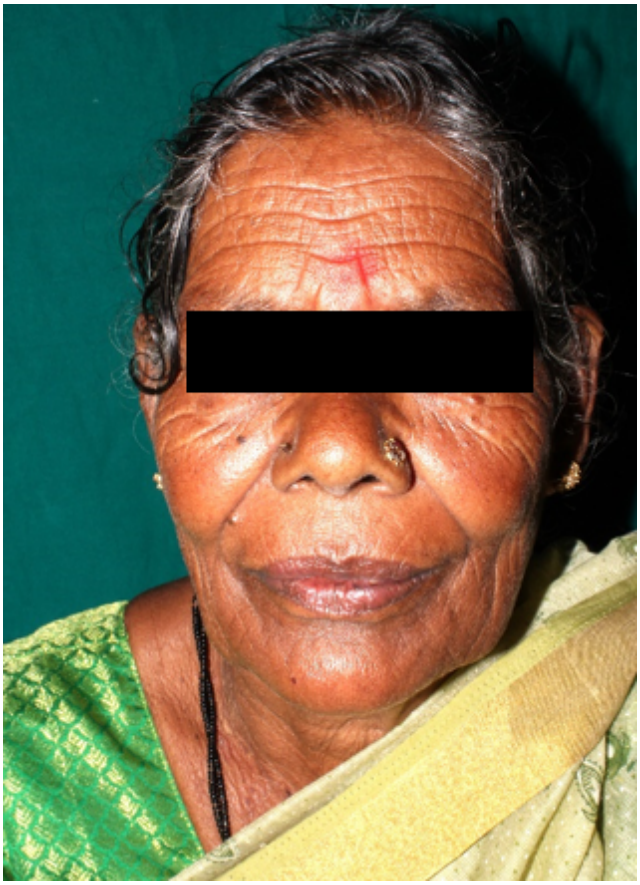

**Fig. 1:** Profile of the patient

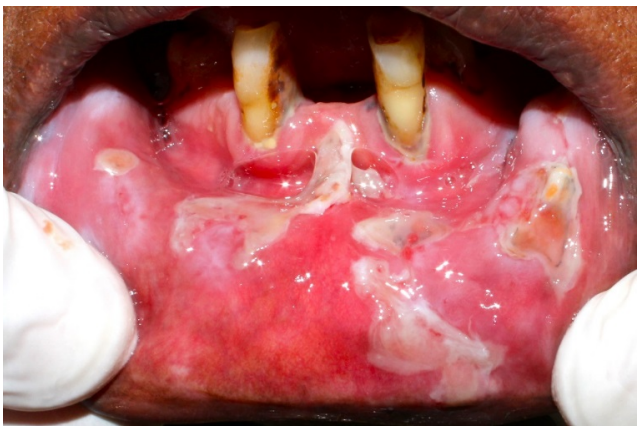

**Fig. 2:** Ulcerative lesions over the lower labial mucosa with irregular borders

On extra oral examination, [Figure 1] right and left submandibular lymph nodes were enlarged, palpable and

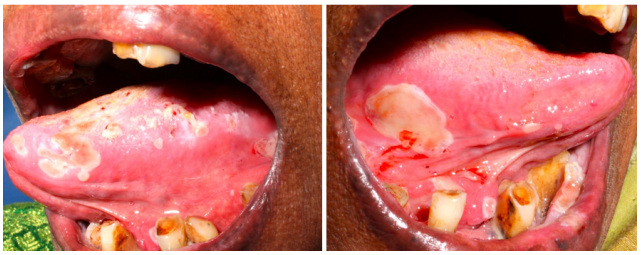

**Fig. 3:** Ulcerative lesions present on right and left lateral border of tongue

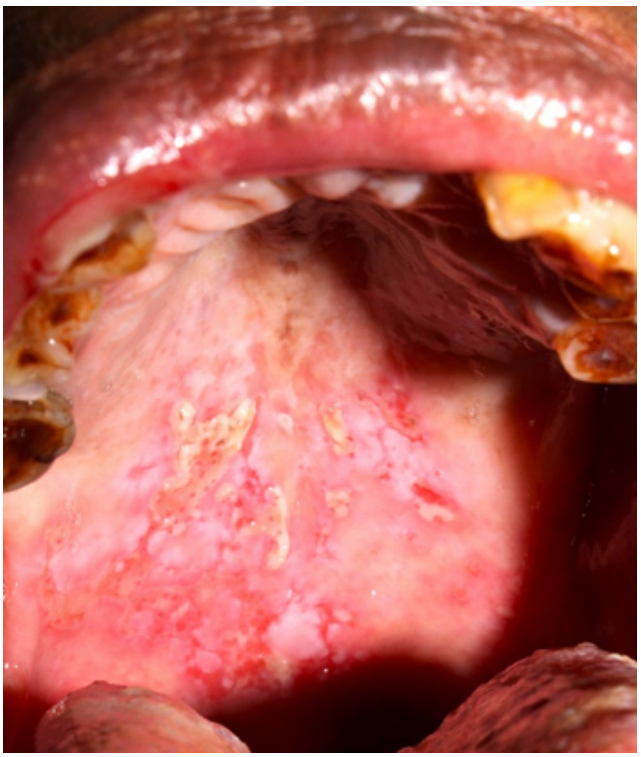

**Fig. 4:** Erosive lesions involving posterior hard plate, soft palate.

tender. Intra-oral examination revealed multiple ulcers present on upper and lower labial mucosa, right and left buccal mucosa, palate, tongue.[Figures 2, 3 and 4] Ulcers presented with the well-defined borders with yellowish slough with the areas of reddish erythema. Ulcers were tender on palpation and not attached to the underlying structures. Based on the clinical presentation, provisional diagnosis of pemphigus vulgaris was given.

Incisional biopsy on left buccal mucosa was carried out and report revealed presence of epithelium and connective tissue. The epithelium is parakeratinized stratified squamous type showing acantholysis. A characteristic suprabasilar split with hemorrhagic spots and acantholytic cells known as Tzank cells are seen. Areas revealing basilar tombstones are present. Subepithelial connective tissue shows chronic inflammatory cell infiltration of lymphocytes and numerous

adipocytes. Few constricted blood vessels engorged with RBCs are seen. Deeper stroma shows both longitudinal and transverse sections of muscle bundles. Correlating with the given clinical features, the above 'histopathological features are suggestive of Pemphigus.

Treatment is usually targeted at controlling the severity and preventing relapses. Systemic corticosteroids remain the gold standard treatment for pemphigus. Patient was treated with topical Triamcinolone acetonide 0.1%, applied 3 times daily for oral lesions.

## 3. Discussion

Pemphigus is a category of potentially fatal blistering skin and mucous membrane disorders distinguished by acantholysis (loss of keratinocyte to keratonicyte adhesion).[3]

The Greek word "Pemphix," which means bubble, is the source of the term "pemphigus." A set of rare but potentially fatal autoimmune diseases known as pemphigus vulgaris are characterised by mucosal and cutaneous blisters. Epithelial blistering that develops from erythematous macules or urticarial bases is its defining feature.[6]

Wichman initially coined the term "pemphigus" in 1791 to refer to all blistering disorders collectively (Korman 1988). In 1953, Lever distinguished PV from bullous pemphigoid based on the histological image, clinical criteria, and progress of the disease.[7]

Desmoglein 3 (Dsg3) and desmoglein 1 (Dsg1) are desmosomal cadherins that Pemphigus targets, but Pemphigus foliaceus solely targets Dsg1. The distribution of blistering is influenced by the distinct expression patterns that Dsg3 and Dsg1 exhibit in mucosa and skin. Dsg1 is expressed significantly less and mostly in the superficial layers of the mucosa, but Dsg3 is expressed in much higher numbers throughout the entire epithelium. While Dsg1 is distributed across the entire epidermis and particularly strongly in the superficial layers, Dsg3 is extremely weakly expressed in skin and is only found in the basal and parabasal layers.[8]

People commonly develop pemphigus between the ages of 40 and 50, with femaless allegedly being affected more commonly than men. More than 50% of the patients affected have been observed to have oral mucosal symptoms at first, followed by skin involvement. It is discovered that oral lesions last, on average, between three months and a year.[9]

Pemphigus vulgaris oral symptoms can appear either beforehand or concurrently with cutaneous lesions.[10] Pressure or friction can cause blisters to form on normally looking skin or mucosa (Nikolsky sign). Clinically, the lesion presents as thin-roofed, short-lived blisters that are brittle and prone to rupture, leading to painful and hemorrhagic erosions. In our case, the disease first showed symptoms as oral lesions of the gingiva and palate.[11] The buccal mucosa is the most frequently affected area of the oral mucosa, followed by the palatal, lingual, and labial mucosa. Lesions can develop anywhere on the oral mucosa. The gingiva is least frequently affected, and desquamative gingivitis is the most typical symptom of the condition. In many patients, oral lesions are followed by the development of skin lesions.[3]

Early diagnosis, when lower dosages of medication can be administered for shorter periods of time to control the disease, is a crucial component of patient therapy. Topical and systemic corticosteroids are currently used in combination with immunosuppressants like azathioprine or mycophenolate to treat pemphigus vulgaris. Intravenous immunoglobulin or plasmapheresis, which work to lower levels of circulating autoantibodies, can also be added to the treatment. Short-term disease remission has been demonstrated to be effectively induced by anti-CD20 antibody rituximab therapy. After the lesions have disappeared, a maintenance regimen is required to keep the disease under control and lessen the negative effects of the medications. Depending on how much the lesion is involved, the length of medical treatment varies.[10]

A lengthy clinical course and considerable morbidity and death are associated with pemphigus vulgaris. If pemphigus is not treated, mortality increases by 50% after two years and reaches almost 100% after five years. Significant skin involvement, septicemia, bronchopneumonia, electrolyte imbalance, and subsequent systemic infections were the main reasons of death.[10]

## 4. Conclusion

In this investigation, PV was the most common subtype of pemphigus. A common but potentially fatal autoimmune mucocutaneous condition, pemphigus vulgaris has a propensity to attack the oral mucosa. Although the exact cause of PV is unknown, a number of diverse elements may contribute to its pathogenicity. Most PV cases started off with mouth lesions. The clinician typically encounters the oral symptoms of pemphigus vulgaris first and occasionally as the only symptom. Timely diagnosis and prompt treatment are hence necessary for complete cure with reduced morbidity and mortality owing to the chronic and often fatal course of this condition.

## 5. Source of Funding

No financial support was received for the work within this manuscript.

## 6. Conflict of Interest

None declared.

## References

1. Pettini F, Ballini A, Capodiferro S. Management of oral pemphigus vulgaris: A case report and a clinical update. *Eur J Inflamm.*

2015;13(1):53–7. doi:0.1177/1721727X15576854.
2. Vinall C, Stevens L, Mcardle P. Pemphigus vulgaris: a multidisciplinary approach to management. *BMJ Case Rep*. 2013;p. bcr2013200479. doi:10.1136/bcr-2013-200479.
3. Arpita R, Monica A, Venkatesh N, Atul S, Varun M. Oral Pemphigus Vulgaris: Case Report. *Ethiop J Health Sci*. 2015;25(4):367–72.
4. Tarakji B. Pemphigus Vulgaris in Old Patient. *Case Rep Dent*. 2021;p. 3946161. doi:10.1155/2021/3946161.
5. Parellada J, Arencibia YO, Watson H, Parellada N, Saikaly LE, Saikaly SK, et al. A Case of Bullous Pemphigoid: A Prevalent and Potentially Fatal Condition. *Cureus*. 2018;10(4):e2533.
6. Hussain MH, Tanweer F, Sakagiannis G, Mair M, Mahmood S, Ashokkumar S, et al. Pemphigus Vulgaris and Bullous Pemphigoid of the Upper Aerodigestive Tract: A Review Article and Novel Approaches to Management. *ORL J Otorhinolaryngol Relat Spec*. 2021;83(6):395–403.
7. Challacombe SJ, Setterfield JF. Oral Vesicular and Bullous Lesions. In: Farah C, Balasubramaniam R, McCullough M, editors. ontemporary Oral Medicine. Springer, Cham; 2019. p. 1083–142. doi:10.1007/978-3-319-72303-7_13.
8. Perks AC, Farthing PM, Murphy R. A Case of Concomitant Pemphigus Foliaceus and Oral Pemphigus Vulgaris. *Head Neck Pathol*. 2018;12(4):592–7. doi:10.1007/s12105-017-0884-0.
9. Kuriachan D, Suresh R, Janardhanan M, Savithri V. Oral Lesions: The Clue to Diagnosis of Pemphigus Vulgaris. *Case Rep Dent*. 2015;p. 593940.
10. Ahankare P, Divyesh&patankar, & Sangeeta, Sridharan G. Pemphigus Vulgaris: Case Report and Review of Literature. *J Clin Diagn Res*. 2019;13(5):4–6.
11. Kumar SJ, Anand SN, Gunasekaran N, and RK. Oral pemphigus vulgaris: A case report with direct immunofluorescence study. *J Oral Maxillofac Pathol*. 2016;20(3):549. doi:10.4103/0973-029X.190979.

## Author biography

**Varsha Patel G.B,** Post Graduate Student https://orcid.org/0000-0001-7636-3682

**Sujatha G.P,** Professor https://orcid.org/0000-0002-6830-6263

**Sangeeth Siddabasappa,** Professor

**Ashok L,** Professor and HOD https://orcid.org/0000-0002-9363-8630

**Cite this article:** Varsha Patel G.B, Sujatha G.P, Siddabasappa S, Ashok L. Pemphigus vulgaris – A case report. *J Dent Panacea* 2022;4(4):183-186.