Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Surgery

Pemphigus Vulgaris: A Case Report

M.H. Bahalou[1]*, B. Hemmaoui[1], A. Jahidi[1]

[1]Department of Surgery

DOI: https://doi.org/10.36347/sjmcr.2025.v13i07.005 | **Received:** 11.05.2025 | **Accepted:** 23.06.2025 | **Published:** 02.07.2025

***Corresponding author:** M.H. Bahalou
Department of Surgery

Abstract **Case Report**

Pemphigus is an intraepithelial autoimmune blistering dermatosis. It often begins as a mucosal-only phase, primarily affecting the oral mucosa. When skin involvement occurs, it is typically secondary and presents as flaccid blisters on otherwise healthy skin, quickly replaced by post-bullous erosions with an epidermal collar. Nikolsky's sign is positive in perilesional skin. Diagnosis is based on histological examination showing intraepithelial blistering with acantholytic cells and the presence of antibodies on direct immunofluorescence. Treatment relies on low-dose systemic corticosteroids combined with immunosuppressive treatment for isolated oral involvement. Cases with cutaneous involvement require high-dose systemic corticosteroids.

Keywords: Autoimmune disease, pemphigus vulgaris, blistering lesions.

INTRODUCTION

Pemphigus vulgaris, also known as chronic malignant pemphigus, is a rare blistering disorder marked by extreme skin fragility. It is a serious autoimmune intraepidermal disease caused by autoantibodies targeting specific desmosomal proteins in the squamous epithelium. This immune response leads to acantholysis, a loss of cohesion between keratinocytes, resulting in multiple blisters across the mucocutaneous surfaces. Lesions typically first appear in the oral mucosa before spreading to the skin.

A clinical scoring system based on lesion extent and treatment requirements distinguishes three main forms:

Severe forms: Extensive, progressive disease with long-standing oral lesions that impair feeding. Mild forms: Isolated, non-progressive cutaneous lesions without systemic involvement. Moderate forms: Limited but progressive disease, without major systemic effects.

CASE REPORT

We report the case of a 44-year-old man, former smoker (quit 3 years ago), with no history of atopy or immunodeficiency, presenting with high dysphagia and odynophagia lasting over three weeks. Clinical examination revealed oral cavity lesions on the soft palate (Figures 1, 2), anterior pillar, tonsillar lodge, inner cheeks, and on nasofibroscopy at the tongue base and laryngeal face of the epiglottis. The disease progressed without fever. There was no recent drug intake. Oral lesions began as isolated blisters that rapidly ruptured, forming confluent erosions with erythematous patches.

Despite antibiotic therapy, no improvement was noted. An initial biopsy suggested herpes-like lesions, but antiviral treatment failed, and new lesions appeared during treatment. A second biopsy revealed acantholysis with intraepithelial blistering (Figure 2). Cytological and histological analyses, confirmed by direct immunofluorescence, established the diagnosis of pemphigus vulgaris. The patient was started on corticosteroids and immunosuppressive treatment, with a favorable response.

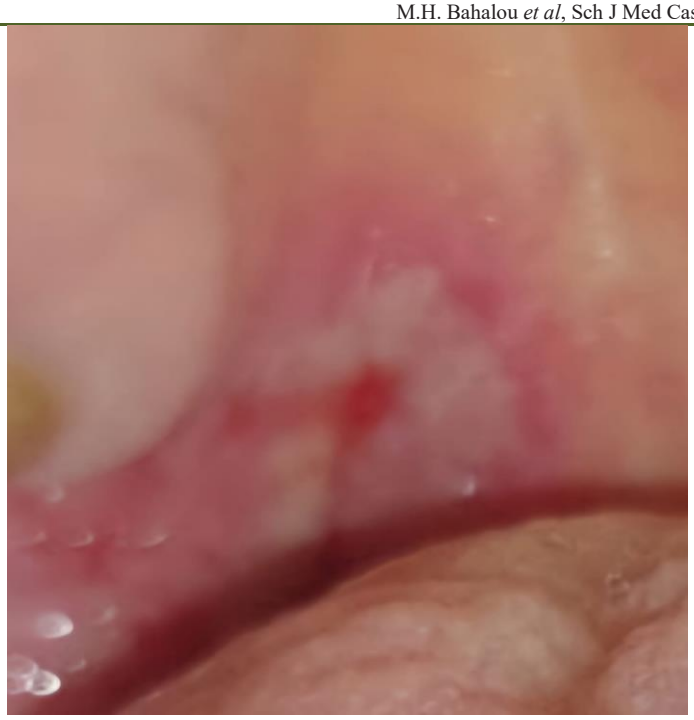

Figure 1: Pemphigus vulgaris lesion on the right side of the soft palate

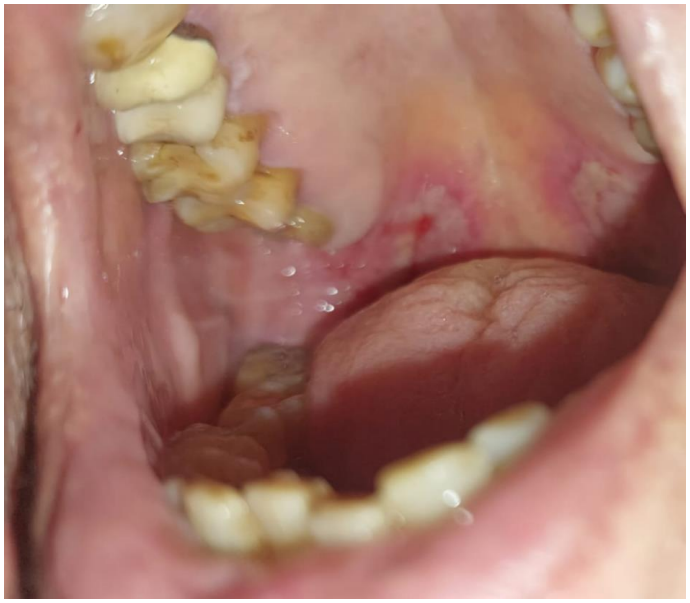

Figure 2: Extensive lesions on the soft palate

DISCUSSION

Pemphigus vulgaris is the most common autoimmune blistering disease, accounting for 85% of pemphigus cases. It primarily affects adults aged 40–60 but can occur in adolescents and rarely in neonates. Female predominance remains debatable [1;2].

It is a model autoimmune disease involving autoantibodies against desmosomal proteins, leading to intraepidermal blistering through keratinocyte detachment (acantholysis). In two-thirds of patients, initial symptoms are oral, preceding skin lesions by 2–3 months. Some cases remain confined to the oral mucosa, as in the case presented. Only 15% of cases never involve the oral mucosa. Most affected sites include the inner cheeks, palate (70–78%), and attached gingiva (20–25%). [1;3]

Oral lesions typically appear as irregular, painful erosions, sometimes with white debris, giving a "chewed" appearance. The palate may show small erosions punctuated by red dots (salivary duct openings). The gingiva may display erosive or desquamative gingivitis, mimicking other diseases. [4]

The lips may be covered in blackish hemorrhagic crusts, resembling erythema multiforme. The tongue and soft palate may show irregular erosions

with detached epithelial edges and whitish coating. These painful lesions bleed on contact, hindering nutrition and oral hygiene. [2;5]

In about 13% of cases, other mucosal sites are involved—pharynx, larynx, esophagus, nasal mucosa, genitals, or conjunctiva—as seen in our patient.[6]

Cutaneous blisters usually appear weeks to months after mucosal symptoms. They are well-demarcated, tense, on normal or erythematous skin, and rapidly evolve into large erosions. Nikolsky's sign (epidermal detachment under tangential pressure) is often positive. Pruritus is usually absent. [2;7]

General condition may decline: anorexia, vomiting, diarrhea, weight loss, and cachexia, though fever is often absent in early flares. Diagnosis involves the following investigations: [3;8]

Cytology – Tzanck smear: A smear from a fresh blister stained with M (ay-Grünwald-Giemsa reveals Tzanck acantholytic cells—characteristic but not specific.

Histology: Biopsy of a fresh intact blister fixed in formalin shows intraepidermal blisters due to acantholysis. Oral biopsies may be less clear and sometimes need repeating.

Direct Immunofluorescence: Perilesional frozen skin shows interkeratinocyte IgG (sometimes IgM/IgA) and C3 deposits in the Malpighian layer.

Indirect Immunofluorescence: Detects circulating anti-intercellular substance antibodies in over 80% of cases, with titers generally correlating with disease activity. Pemphigus vulgaris should be distinguished from [3;9]:

Herpes simplex: by its acute onset and cytological findings; Bullous pemphigoid: different on histology and immunology; Erythema multiforme: features target lesions and hemorrhagic crusts; Bullous/erosive lichen planus: can mimic pemphigus.

Before modern therapy, pemphigus vulgaris was fatal in 75% of cases, typically progressing to cachexia. Today, mortality is around 10%, thanks to earlier diagnosis, improved treatments, and better complication management.

Treatment should be tailored to the clinical form and requires a multidisciplinary approach, especially when ENT or ophthalmologic involvement is present. Diagnosis confirmation relies on: Clinical signs; Histology; Immunofluorescence (direct and indirect). [10]

A comprehensive pre-therapeutic assessment is vital to rule out comorbidities that could affect management. Individualized treatment and regular follow-up are essential in managing this potentially life-threatening disease.

REFERENCES

1. Amagai M. Autoimmune and infectious skin diseases that target desmogleins. Proc Jpn Acad Ser B Phys Biol Sci 2010;86(5):524- 37.
2. Su O, Onsun N, Meric Teker A, *et al*. Upper airway tract and upper gastrointestinal tract involvement in patients with pemphigus vulgaris. Eur J Dermatol 2010;20(6):792-6.
3. Espana A, Fernandez S, del Olmo J, *et al*. Ear, nose and throat manifestations in pemphigus vulgaris. Br J Dermatol 2007;156(4):733-7.
4. Jamora MJ, Jiao D, Bystryn JC. Antibodies to desmoglein 1 and 3, and the clinical phenotype of pemphigus vulgaris. J Am Acad Dermatol 2003;48(6):976-7.
5. Zagorodniuk I, Weltfriend S, Shtruminger L, *et al*. A comparison of anti-desmoglein antibodies and indirect immunofluorescence in the serodiagnosis of pemphigus vulgaris. Int J Dermatol 2005;44(7):541-4.
6. Kavala M, Altintaş S, Kocaturk E, *et al*. Ear, nose and throat involvement in patients with pemphigus vulgaris: Correlation with severity, phenotype and disease activity. J Eur Acad Dermatol Venereol 2011;25(11):1324-7.
7. Mahmoud A, Miziara ID, Costa KC, *et al*. Laryngeal involvement in pemphigus vulgaris: A proposed classification. J Laryngol Otol 2012;126(10):1041-4.
8. Fawzy MM, Hegazy RA, Abdel Fattah AF. Ear, nose, and throat involvement in Egyptian patients with pemphigus vulgaris: A step towards a better management. Int J Dermatol 2013;52(10):1268- 73.
9. Robati RM, Rahmati-Roodsari M, Dabir-Moghaddam P, *et al*. Mucosal manifestations of pemphigus vulgaris in ear, nose, and throat; before and after treatment. J Am Acad Dermatol 2011;67(6):e249-52.
10. Fernandez S, Espana A, Navedo M, Barona L. Study of oral, ear, nose and throat involvement in pemphigus vulgaris by endoscopic examination. Br J Dermatol 2012;167(5):1011-16.