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Case Report

# Exceptional pemphigus case report: A scarcely seen presentation

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ABSTRACT

Pemphigus is a known but rare autoimmune mucocutaneous blistering condition. It has four variants; pemphigus vulgaris, pemphigus foliaceous, IgA pemphigus, and paraneoplastic pemphigus. There has been very few case reports about the occurrence of both p.vulgaris and p. foliaceus in the same patient. We report a case having concomitant P.vulgaris of oral mucosa and P. foliaceus of chest and back.



## 1. Introduction

Pemphigus is a seldom acquired autoimmune mucocutaneous blistering disease. It has four variants; pemphigus vulgaris, pemphigus foliaceous, IgA pemphigus, and paraneoplastic pemphigus. Each variant has distinct histological features and target antigens.[1] The two principal variety are pemphigus vulgaris (PV) and pemphigus foliaceus (PF).[2] Pemphigus Foliaceous depicts clinical blisters involving the face and trunk exempting oral mucosa involvement, and histopathology reveals sub-corneal acantholytic bullae. Pemphigus vulgaris clinically depicts flaccid blisters and persistent erosions antecedental on the intertriginous regions, almost always associated with oral mucosal lesions, and histopathology reveals suprabasal acantholytic bullae in the deep epidermis.[3]

Literature shows that Pemphigus vulgaris targets desmoglein 3 (Dsg3) and desmoglein 1 (Dsg1) hence affecting skin and mucosa both. Whereas Pemphigus Foliaceous targets only Dsg1 hence affecting only superficial skin and does not affect mucosa.[4,5]

There has been very few case reports about the occurrence of both vulgaris and foliaceus in the same patient.[6,7] We report a case having concomitant Pemphigus vulgaris of oral mucosa and Pemphigus foliaceus of chest and back

## 2. Case Report

A young male patient was referred to the ENT outpatient department with complaints of oral lesions along with ulcers for 3 months. The patient did not have any other immunocompromised conditions or comorbidities. He also had complaints of fluid-filled crusted lesions over the scalp, chest, and back for 1 month.

On examination there were multiple lesions over bilateral buccal mucosa, the lesions were hyperpigmented to violaceous, non-raised flaccid bullae with white coloured plaque, and surrounding mucosa having melanosis with crusting. On palpation, there was no induration, it was non-tender, and there were no signs of inflammation. His oropharynx and neck examination were normal.(Figure 1)

Biopsy taken from the lesions of buccal mucosa revealed histopathological features of pemphigus vulgaris with inflammatory cells. An intraoral biopsy of the flaccid bullous blistering lesion was derived and the tissue was

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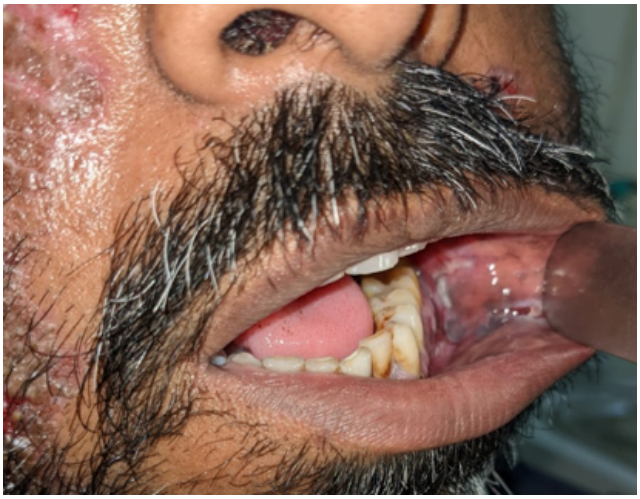

**Figure 1:**

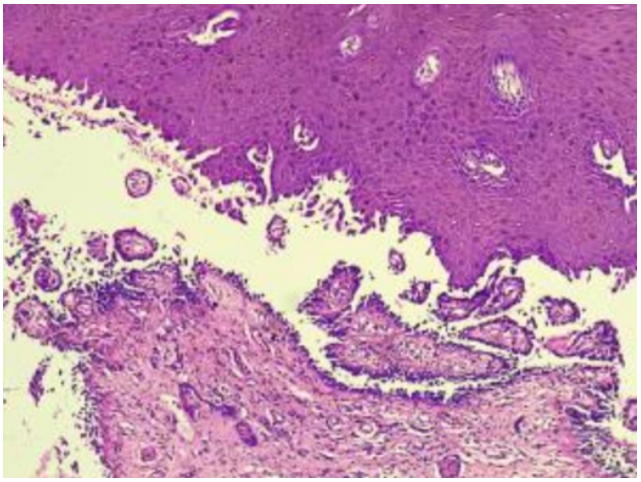

**Figure 2:**

stained and the histopathological examination revealed the main features of stratified squamous epithelium with intraepithelial bullae containing acantholytic cells and classical tombstone appearance.(Figure 2)

The patient was referred to the department of dermatology for lesions on the back and chest for which a biopsy and tzank smear were taken which showed features of pemphigus foliaceus.

The patient was treated primarily with topical antibiotics and multivitamins but there was no significant improvement.

Correlating the clinical and histopathological features, a final diagnosis of oral Pemphigus Vulgaris was made.

The patient was started on oral prednisolone at an initial dose of 40mg once daily for 5 days with a tapering dose of 20 mg for 5 days followed by 10 mg for 5 days and 5 mg for another 5 days.

Along with 20 days of oral prednisolone treatment the patient was also given topical triamcinolone application three times in a day for 10 days, followed by twice daily application for 5 days and once daily application for another 5 days. The total treatment was for a 20 days and the patient was followed up every week. The patient recovered completely and there was no recurrence of any lesions.

In addition, the patient was also given broad-spectrum antibiotics (tablet Doxycycline 100 mg) for a week l, supportive multivitamins, and probiotics to prevent any secondary infection. The patient was also asked to maintain good oral hygiene.

With the vigilant diagnosis and patient compliance to medications, there was complete recovery without any remissions.

## 3. Discussion

The quintessential lesion of pemphigus is a thin-walled bulla emerging on otherwise normal skin or mucosa, which swiftly breaks and continues to extend peripherally, ineluctably leaving large denuded areas. It also displays positive "Nikolsky's sign" – the capability to induce peripheral extension of a blister and/or removal of epidermis as a consequence of applying tangential pressure with a finger or thumb to the involved skin, peri-lesional skin, or normal skin in patients affected or suspected with pemphigus.[8,9] Pemphigus vulgaris affects both Dsg1 nad Dsg3 hence causing skin as well as oral lesions. However Pemphigus foliaceus affects only Dsg1 causing only cutaneous lesions. There have been few studies describing about the transitions and concomitant involvement of both the subtypes.[8]

A rare case study was reported by A.Komai in the British Journal of Dermatology in 2001 describing the clinical transition of P.foliaceus and P.vulgaris and assessing autoantibody profile by Elisa.[10] Whereas our study did not analyse any transition due to limited resources such as lack of immunofluorescence techniques for antibody testing and financial contraints of the patient.

The mainstay of treatment modalities remain limited to oral and topical tapering dose of steroids some patients have also known to benefit by biological agents like Mycophenolate Mofetil (MMF) and Azathioprine.[10]

## 4. Conclusion

This rare case report help in understanding the scarcely seen presentation of pemphigus wherein there is simultaneous occurrence of both the variants, Pemphigus vulgaris and Pemphigus foliaceus. It states that there should be more vigilant clinical examination in all patients with Pemphigus to rule out other subtypes hence leading to better treatment outcomes and good disease prognosis.

## 5. Source of Funding

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## 6. Conflict of Interest

None.

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