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Review Article

# A pen sketch for oral pemphigus vulgaris: A review

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ABSTRACT

Pemphigus is a rare chronic mucocutaneous autoimmune bullous dermatosis. Based on clinical features and pathophysiology the various subtypes include pemphigus Vulgaris (PV), pemphigus foliaceus (PF), IgA pemphigus, and paraneoplastic pemphigus (PNP). Autoantibodies against desmogleins 1 and 3 cause pemphigus Vulgaris which results in acantholysis, or the loss of cell-to-cell adhesion ultimately causing potentially lethal bullae and erosion formation. 80 to 90% of patients develop oral lesions that are manifested before mucocutaneous lesions in more than half of patients.Dental professionals are pivotal and can thus diagnose the disease and prevent skin involvement through proper therapy. Treatment should include systemic corticosteroids and immunosuppressive drugs. Intravenous pulse therapy is instituted in severe cases of pemphigus. This article is an attempt to present clinical manifestations, pathophysiology, and newer medical treatment modalities of pemphigus.



## 1. Introduction

Pemphigus was named by wickman in 1791. The term Pemphigus has a greek origin from the word Pemphix standing for bubble or blister and vulgaris has latin origin which means common. Pemphigus is a rare disease with an incidence rate ranging from 0.5 to 3.2 per 100,000 per year with a slight prelidiction in women. Pemphigus primarily manifests in the $5^{th}$ and $6^{th}$ decade of life. Usually, presentation on the oral mucous membrane is as painful erosions secondary to vesicle formation and is seen as only a sign for a span of around 5 months prior to the development of skin lesion.[1]

## 2. Pathogenesis

The immunoglobulin (Ig) antibodies are present on the keratinocytes' cell surface proteins. These antibodies are being developed against the desmogleins and transmembrane glycoproteins which are associated with desmosomes. These desmosomes result in cell-to-cell adhesion within the epidermal layer.[2]

## 3. Clinical Features

A hallmark of PV is erosion and painful blisters that are predominately seen in the oral mucous membrane.[3] In Pemphigus Vulgaris oral lesions develops in 80-90 percent of patients and oral lesions are manifested before mucocutaneous lesions in more than half of patients.[4] Some patients may feel uncomfortable chewing and eating resulting in weight loss, poor nutrition, and fatigue.

The oral lesion is the first to show and the last to go.[5]

Figure 1 Erosive Lesion on right buccal mucosa & ulcers at the commisure.

Two major subgroups of PV are the mucosal-dominant type and the mucocutaneous type. Mucocutaneous produces

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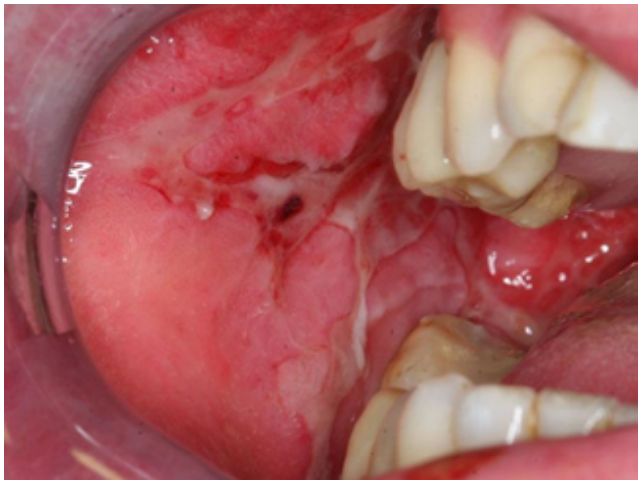

**Fig. 1:** Erosive lesion on rightbuccal mucosa

cutaneous blisters and erosion along with mucosal involvement while the mucosal-dominant causes mucosal erosions mainly.[6]

Desquamative gingivitis causes severe pain and interferes with appropriate oral hygiene, leading to plaque accumulation.

Cutaneous lesions of PV typically reflect flaccid blisters and crusted erosions on an erythematous base. Due to the intraepidermal acantholysis because of anti-desmoglein antibodies blisters seen in PV are having flaccid nature. Positive Nikolsky sign is present in PV lesions, which is defined as a well-described clinical sign which manifests as dislodgement of the intact superficial epidermis by a shearing force, indicating a plane of cleavage in the epidermis. These lesions are excruciatingly painful.[7] Though cutaneous lesions can either be localized or diffuse and can affect any surface, the palms and soles are typically spared in patients with PV. This paucity of palmoplantar involvement can be helpful in distinguishing PV from other vesiculobullous dermatoses, such as PNP or erythema multiforme.[6] (Figure 3 Crusted vesiculobullous lesions of the elbow)

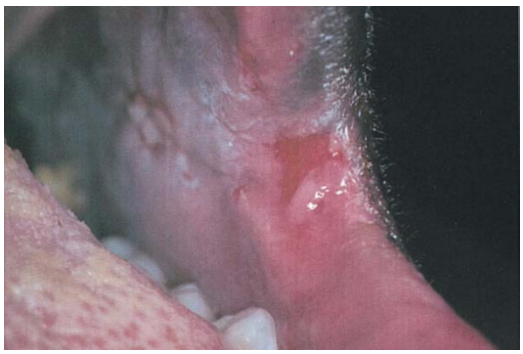

**Fig. 2:** Ulcers at the commissure

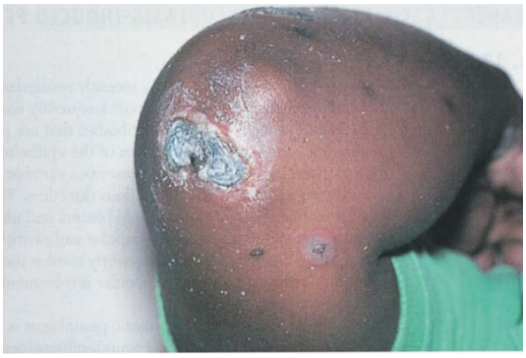

**Fig. 3:** Crusted vesiculobullous lesions of the elbow

## 4. Laboratory Diagnosis[4]

Establishing diagnosis requires compatible clinical features along with evidence of pathological features of involved mucosa.

The cytological examination which includes Tzanck smear can be helpful in the rapid demonstration of acantholytic keratinocytes of the spinous layer.

### *4.1. Histopathological examination*

A recent blister (< 24 hours of appearance) should be included in the biopsy. In case of rupture of the lesion, the biopsy should include a perilesional normal area. Histopathological examination indicates the level of epidermal cleavage that is suprabasal split helps to distinguish it from sub-epithelial blistering diseases for example mucous membrane pemphigoid, bullous lichen planus, and chronic ulcerative stomatitis.

### *4.2. Direct immunofluorescence examination*

Identification of IgG and C3 autoantibodies directed against the cell surface of keratinocytes is considered a "gold standard" for the differential diagnosis of PV.

To detect pemphigus autoantibodies the other methods are direct (DIF) and indirect immunofluorescence (IIF), immunoprecipitation, immunoblotting, and enzyme-linked immunosorbent assay (ELISA).

## 5. Treatment

In order to prevent serious involvement of other mucosa and cutaneous sites and fatal complications the treatment should be started promptly. Early diagnosis is a key aspect of patient management with the use of a lower dose of medication for a shorter duration. Dental professionals should be aware of the clinical manifestation of pemphigus Vulgaris to warrant early diagnosis and treatment, as this in turn determines the prognosis and course of the disease.[1]

### 5.1. Current therapies

Prednisolone is typically given as the first line of treatment for PV along with immunosuppressive drugs like azathioprine (AZA) and mycophenolate mofetil (MMF), or monoclonal antibodies against CD20, and cyclophosphamide. The most common treatment for Pemphigus Vulgaris is corticosteroid therapy, both locally and systemically.

Future Therapeutic Approaches for the treatment of PV Chimeric antigen receptor (CAR)-T-cell therapy and Anti-Neonatal Fc Receptor (FcRn) are promising therapy.[2]

## 6. Conclusion

In conclusion, in patients with PV who have lesions confined to the oral cavity, close follow-up is essential, and referral to specialists should be undertaken promptly in the event of the appearance of extraoral symptoms. Before the advent of corticosteroid therapy, pemphigus was fatal, still, the disorder is serious with a mortality rate of 5% to 10% which is now primarily due to treatment-related complications.

## 7. Source of Funding

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## 8. Conflict of Interest

None.

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