

# Pemphigus vulgaris presenting as gingival involvement

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## ABSTRACT

Pemphigus vulgaris (PV) is an autoimmune blistering disease affecting the mucous membrane and skin. Typically, oral lesions appear before skin lesions, and in a majority of the cases only oral lesions are present. The dentist may then be the first to recognize and diagnose this disease. It is unusual for PV to present over the gingiva as a primary site of involvement. Diagnosis is based on clinical presentation and confirmed by histopathological study. Early diagnosis and management can prevent the uneven life-threatening effects of this potentially chronic mucocutaneous disorder. The case serves to enhance our awareness of the gingiva as a site at which systemic disease can manifest itself.

**Key words:** Desquamation, gingivitis, mucocutaneous, pemphigus

## INTRODUCTION

Pemphigus vulgaris (PV) is a chronic autoimmune mucocutaneous disease that usually manifests first in the oral cavity and may later spread to the skin or other mucous membranes.<sup>[1]</sup> Apart from ulcers, vesicles, bullae and pustular lesions, it can present solely as mucosal erosions. Herein we report a case of intraoral PV presenting as intense erythema and erosions involving the gingiva.

## CASE REPORT

A 52-year-old female presented to the Department of Periodontics, Al-Badar Dental College and Hospital, Gulbarga with complaints of bright red gingivae and discomfort in her normal oral function. The patient initially saw peeling of gingivae while brushing. There was no involvement of oral mucosa, palate or tongue. The site most severely affected was her upper front buccal gingivae. The considerable pain and discomfort that the patient felt hindered her from carrying out effective oral hygiene measures and this in turn exacerbated the gingival symptoms. She had essential hypertension, diabetes mellitus and hyperlipidemia and her daily medications included insulin, antihypertensive drugs and statins. On intraoral examination, there were multiple large irregular erosions and

areas of intense erythema involving particularly the gingivae of both upper and lower arch, buccally and palatally/lingually [Figures 1 and 2]. Periodontal pockets deeper than 4 mm were noted. Orthopantomogram showed a combination of horizontal and vertical bone loss. The Nikolky's sign (loss of epithelium occasioned by rubbing apparently unaffected skin) that is a feature of PV was positive. The patient had no cutaneous involvement and other mucosal sites such as conjunctiva, nasal passages and oesophagus were free of lesions.

Based on the history, multiple erosions and the apparent fragility of the gingivae experienced during examination, a vesiculobullous disorder was suspected. Pemphigus and pemphigoid were considered in the differential diagnosis. As mucous membrane pemphigoid is usually seen in the elderly, the former was thought to be more likely. The other common conditions that can present with similar manifestations are oral lichen planus, drug hypersensitivity or idiopathic. A biopsy was drawn from perilesional site of the involved gingivae. Histopathological examination revealed suprabasal blister formation associated with extensive acantholysis of keratinocytes, suggestive of PV [Figure 3].

Since the patient only had isolated gingival involvement, she was started on systemic

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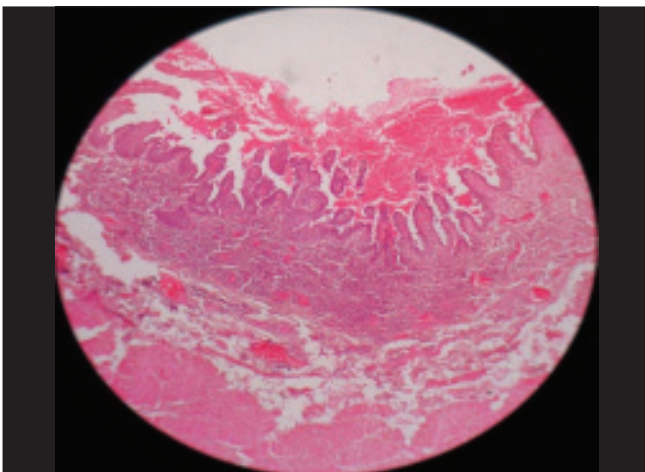
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**Figure 1:** Intensely erythematous gingivae with multiple erosions



**Figure 2:** Erosive lesions on the gingiva palatally



**Figure 3:** Photomicrograph showing suprabasilar split and acantholytic cells

corticosteroids (Prednisolone 0.5 mg/kg/day). This initial dose was stepped up to 1mg/kg/day, which gave marked improvement in two weeks. Vitamins and calcium supplements were prescribed concominantly. The patient was instructed to apply twice daily topical steroid oral paste (triamcinolone acetonide 0.1%) along with local anesthetic gel. The patient was instructed to use a soft brush for brushing the teeth. In addition, an antifungal mouthwash was given to prevent secondary fungal infection. Sometime after commencement of corticosteroid therapy, scaling and root planing was performed.

Over the past 1 year, the prednisolone has been tapered to 10mg/kg/day. Currently, the patient is on this daily low-dose systemic corticosteroid therapy (Prednisolone), topical steroid oral paste (triamcinolone acetonide 0.1%) and supplementary medications. Till date, no other sites are involved.

## DISCUSSION

The term pemphigus was originally named by Wichman in 1791.<sup>[2]</sup> Pemphigus is a group of potentially life-threatening autoimmune mucocutaneous diseases characterized by epithelial blistering affecting cutaneous and/or mucosal surfaces, the term being derived from the Greek word *pemphix* (bubble or blister). Although 'vulgaris' means common in Latin, the worldwide incidence of PV is low and has been reported to be 0.1–0.5 per 100,000 persons per year.<sup>[1]</sup> PV is the most common variant of pemphigus, comprising of 80% of the disease entity. PV frequently involves the mouth<sup>[3]</sup> and has a fairly strong genetic background; ethnic groups such as Ashkenazi Jews and people of Mediterranean and Indian origin are particularly susceptible and there is a link to HLA class II alleles.<sup>[4]</sup> Clinically, PV appears to occur in males and females in an equal ratio,<sup>[5]</sup> and is most frequently reported in patients between the fourth and sixth decades of life.<sup>[6,7]</sup> It is mediated by circulating autoantibodies directed against the keratinocyte cell surface. Mortality from PV before the development of effective therapies was as high as 90%, mainly due to dehydration and secondary systemic infection.

Oral lesions are common and early manifestations of PV, seen typically in adults (rarely in childhood).<sup>[8,9]</sup> They typically run a chronic course, causing blisters, erosions and ulcers. However, the prevalence of oral involvement varies. One recent multicenter study in several countries showed that Bulgarian patients less frequently had oral mucous membrane lesions (66%) compared with Italian (83%) and Israeli (92%) patients.<sup>[10]</sup> Initially vesiculobullous, the oral lesions readily rupture and as the older ones rupture and ulcerate, new bullae develop. They are seen primarily in the buccal mucosa, palate and lips.<sup>[11]</sup> Gingival lesions are less common and usually comprise severe desquamative or erosive gingivitis, characterized by red erosions or deep ulcerative craters.<sup>[12]</sup>

In the present case, gingiva was the only site involved; the gingiva was intensely erythematous and erosive involving both attached and marginal gingiva. However, the patient's oral hygiene was very poor and this in turn contributed further to gingival inflammation, leading to generalized periodontitis.

Diagnosis is based on clinical presentation and histopathological examination.<sup>[13]</sup> The classic signs of oral or gingival PV are multiple erosions or desquamation and a positive Nikolsky sign which both of which were present in our case. Histologically, there is an intraepithelial blister associated with acantholytic cells,<sup>[7]</sup> features which were evident in this patient.

Systemic corticosteroids are the treatment of choice in patients with PV;<sup>[14]</sup> topical steroid therapy alone is insufficient for sustained control of the disease because of the systemic autoimmune nature of PV. In the present case, systemic and topical corticosteroid treatment and adjuvant therapy of antifungal mouthwash, use of soft brush and vitamin supplementation were instituted.

## CONCLUSION

PV is a chronic autoimmune mucocutaneous disease that often primarily involves the oral cavity. As it is a life-threatening disease condition, it is important for the clinician to be able to recognize oral manifestations of PV at an early stage and treat or refer appropriately. Dental professionals can thus play an important and significant role in the early diagnosis and management of PV.

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