

CASE REPORT

Pemphigus Vulgaris: Diagnosis with Oral Lesions

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ABSTRACT

Pemphigus vulgaris (PV) consists of a group of autoimmune blistering diseases of the skin and mucous membrane. The blisters occur in the epithelium where the patient's immunoglobulin G (IgG) autoantibodies are produced in response to triggering factors that target two structured proteins of desmosomes identified as desmogleins (Dsg) 1 and 3. It is essential that dentists should know these pathologies in order to be able to diagnose them at an early stage of the disease and to direct patients for adequate treatment. This article presents a case report along with its treatment plan.

Keywords: Blister, Corticosteroid, Desmoglein, Pemphigus vulgaris.

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INTRODUCTION

Pemphigus vulgaris is a group of autoimmune blistering diseases of the skin and mucous membranes.¹ Pemphigus vulgaris in most patients manifest with oral lesions initially. They are most commonly seen on the oral mucosa, but the buccal mucosa is affected first, followed by the palatal, lingual, and labial mucosae. Majority of patients show oral lesions followed by the development of skin lesions. Consequently, if oral PV is recognized early, treatment can be done to stop the progression of the disease to skin involvement. Early onset of oral lesions of PV is difficult to identify since the oral lesions may be nonspecific, manifesting as superficial erosions or ulcerations and rarely presenting with the formation of intact bullae.²

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CASE REPORT

A 50-year-old patient reported to the dental college with the complaint of a blister and pain in the lower right back region since 2 months. The patient noticed slight bleeding and a blister associated with the lower right second molar on the gingiva 1 year back. He had visited a dentist where periodontal treatment, scaling and root planing were done. Initially, the bleeding subsided and the blister reduced in size but recurred after 6 to 7 months.

Intraoral examination revealed localized erosions involving marginal and attached gingiva of the mandibular right first and second molar. Nikolsky's sign was positive, and the epithelium could be easily peeled away (Fig. 1). Skin lesions were not present. Differential diagnosis of mucous membrane pemphigoid, erosive lichen planus, and PV was arrived at. A gingival biopsy was performed from the perilesional site of the involved gingiva and sent for histopathological examination.

Histopathological examination showed acantholysis in the suprabasal region with a "suprabasillar split" (Fig. 2). Intraepithelial vesicle showed Tzanck cells. Connective tissue stroma was loose collagenous with dense inflammatory cells, confirming PV. Patient was asked to undergo Dsg 1 and Dsg 3 tests.

Anti-Dsg 1 antibodies were 31.2 U/mL. Anti-Dsg 3 antibodies were 117.4 U/mL.



Fig. 1: Clinical photograph showing lesion on the gingiva in 46, 47 region

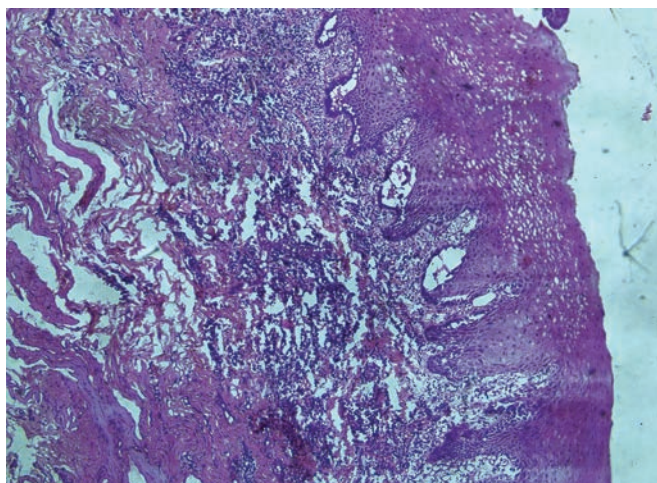


Fig. 2: Photomicrograph showing acantholysis in the suprabasal region with a suprabasilar split (hematoxylin and eosin stain, $\times 10$)



Fig. 3: Complete healing of lesion after 1 month

DISCUSSION

Pemphigus vulgaris is a chronic inflammatory autoimmune bullous disease. There are 0.5 to 3.2 cases reported each year per 100,000 population, with the highest incidence in the fifth and sixth decade of life, with female to male ratio of 2:1.³

Blisters occur in the epithelium where the patient's IgG autoantibodies are produced in response to triggering factors that target two structured proteins of desmosomes identified as Dsg 1 and Dsg 3. The autoantibody formation in pemphigus foliaceus that affects the cutaneous site is Dsg 1. Dsg 3 is predominantly expressed in the oral epithelium, while both Dsg 1 and Dsg 3 are expressed in the skin (although Dsg 1 is expressed more intensely in the superficial layer, Dsg 3 is found more abundantly in basal and suprabasal layers). Dsg 1 and Dsg 3 are components of desmosomal cadherin responsible for holding cells of the epithelium together. The loss of the adhesive function among the spinous cells due to anti-Dsg 3 antibodies results in bullae formation immediately in the suprabasal region in PV. There has been a new pemphigus which has been identified as antigen Dsg 4 and other non-Dsg antigens like human α -9-acetylcholine receptor, which regulates keratinocyte adhesion and keratinocyte annexin-like molecules binding acetylcholine termed pemphaxin and catenin which are also thought to play a role in its etiopathogenesis.⁴ In the present case, Dsg 1 and Dsg 3 tests were performed which were positive. It was observed that Dsg 1 was twice more than the normal range, while Dsg 3 was 16 times more than the normal range. Probably this can be the reason for oral lesions to first appear.

The etiology of PV is usually unknown but sometimes it may have a strong genetic basis as it has been reported more frequently in certain racial groups.

In PV, strong oral associations with certain human leukocyte antigen Class II alleles have been demonstrated. Other initiating factors reported include certain foods (garlic), infections, neoplasms, and drugs. The drugs commonly implicated are those in the thiol group, in particular captopril, penicillamine, and others, such as rifampicin.⁵

Desquamative gingivitis causes severe pain, thus interfering with appropriate oral hygiene leading to plaque accumulation. Dentists should be aware of these pathologies in order to diagnose them at an earlier stage and to direct patients for adequate treatment. Furthermore, intraoral examination should be included as a routine practice in dermatological services.¹

Treatment of oral lesions of PV include topical or intralesional corticosteroids or immunosuppressant drugs. Specific therapies for the underlying disease are available with local immunosuppressive treatment, but systemic immunosuppressive therapy, notably corticosteroids, is almost inevitably required in pemphigus.⁶ In the present case, a dermatologist was consulted and the patient was put on deflazacort 6 mg for 3 days along with 100 mg of dapsone. He was also advised to use triamcinolone gel and metronidazole gel. Two more cycles of this regime in the interval of 2 weeks were planned along with 150 mg of fluconazole and butenafine gel. A review after 4 weeks of the therapy showed disappearance of oral lesions (Fig. 3).

CONCLUSION

Pemphigus vulgaris is an autoimmune disease with dermatologic lesion. It is the role of a periodontist to identify early and diagnose the lesion and refer to a dermatologist with a detailed clinical, histological, and immunohistological examination.

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