

CASE REPORT

Oral Pemphigus Vulgaris: Diagnosis To Management – A Case Report

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ABSTRACT

Background: Pemphigus vulgaris is a rare autoimmune disease affecting the mucous membrane and skin. The lesion can be systemically threatening causing blisters and erosions. In pemphigus vulgaris, the immune system makes autoantibodies that attack the skin and mucous membrane, leading to damage causing blisters and erosions. Timely recognition and proper treatment of oral lesions are necessary to prevent the involvement of skin lesions.

Case Details: This case report describes the case of a female patient aged 26 years reporting to the Department of Periodontology and Implantology, Jaipur Dental College, Jaipur, with chief complaint of gingival growth in between 42 and 43 with burning sensation in the same area. The paper highlights the investigations, differential and definitive diagnosis with treatment protocol rendered to the patient.

Results: Based on investigations, history, and clinical examination, the diagnosis of the lesion was of pemphigus vulgaris and appropriate treatment was rendered.

Conclusion: Dental professionals must be familiar with the clinical manifestations of pemphigus vulgaris to ensure early diagnosis and treatment planning.

Keywords: Autoantibodies, Nikolsky's sign, Pemphigus vulgaris, Tzanck cells.

How to cite this article: Marjara SR, Kudva PB, Bhat G, Kudva HP. Oral Pemphigus Vulgaris: Diagnosis to Management – A Case Report. *Int J Oral Care Res* 2018;6(1):S149-152.

Source of support: Nil

Conflicts of interest: None

INTRODUCTION

Pemphigus vulgaris is a rare autoimmune disease affecting the mucous membrane and skin. It affects

around 3 cases/100,000 population. The lesion can be systemically threatening causing blisters and erosions. Pemphigus vulgaris may cause blistering of the skin and the mucous membranes lining the mouth, nose, throat, and genital area. Oral lesions generally precede skin lesions in many patients.^[1,2] Different variants of pemphigus are pemphigus vulgaris, pemphigus foliaceus, pemphigus vegetans, pemphigus erythematosus, paraneoplastic pemphigus, and drug-related pemphigus. The most common form of pemphigus is pemphigus vulgaris and it accounts for over 80% of cases.^[3] In pemphigus vulgaris, the immune system makes autoantibodies that attack the skin and mucous membrane, leading to damage causing blisters and erosions.

CASE REPORT

A female patient aged 26 years reported to the Department of Periodontology and Implantology, Jaipur Dental College, Jaipur, with a chief complaint of gingival growth in between 42 and 43 [Figure 1] with burning sensation in the same area which aggravated while having food for 6 months. No history of systemic involvement and family history reported. The patient had fair oral hygiene with moderate deposits in lower anterior and cleaned her teeth once a day with toothpaste and brush. The patient was moderately built with no signs of anemia. Extraorally, blisters were reported 2 months back leaving behind a scar in the form of pigmented areas seen on the forehead of the patient [Figure 2]. Similar lesions were also seen on forearms [Figure 3] and upper chest of the patient. Intraorally, gingival overgrowth was seen in between tooth 42 and 43 which was sessile, pale pink in color, fibrotic in consistency, with smooth surface, and wider at the base measuring 5 mm × 5 mm × 8 mm and this resulted in spacing between the teeth and extended lingually [Figure 4]. Nikolsky's sign was checked by applying lateral pressure with thumb pad on skin over a bony prominence and was mildly positive. The clinical presentation in this case led to provisional diagnosis of vesiculobullous lesion. Differential diagnosis included pemphigus vulgaris, mucous membrane pemphigoid, bullous lichen planus, and pyogenic granuloma. Hematological and biochemical investigations were carried out, in which complete hemogram and fasting blood glucose test were done which showed normal values. Excisional biopsy

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Figure 1: Gingival overgrowth between 42 and 43



Figure 4: Gingival overgrowth which resulted in spacing between 42 and 43 and extended lingually



Figure 2: Scar in the form of pigmented areas on the forehead



Figure 5: Excisional biopsy with electrocautery



Figure 3: Scar in the form of pigmented areas on the forearms

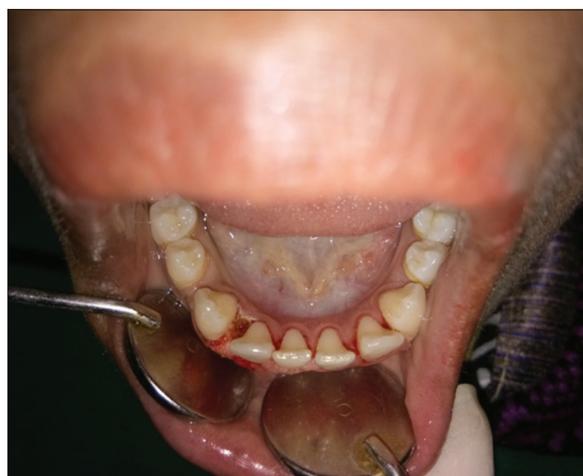


Figure 6: Excisional biopsy of gingival overgrowth performed

was done with scalpel and electrocautery unit [Figures 5 and 6] under local anesthesia. Single bit soft tissue specimen measuring 5 mm × 5 mm × 8 mm was sent for histopathological investigation. Histopathological investigation was done under photomicrograph ×10 and ×40 (hematoxylin and eosin). Hematoxylin and eosin stained section showed parakeratinized stratified squamous

epithelium with few rete ridges. The epithelium showed intraepithelial cleft formation [Figure 7], in which there was the presence of numerous acantholytic cells chiefly of squamous cells which gave the appearance of Tzanck cells [Figure 8]. The connective tissue stroma showed collagen fiber bundles and fibroblast. There was the presence of numerous chronic inflammatory cells chiefly

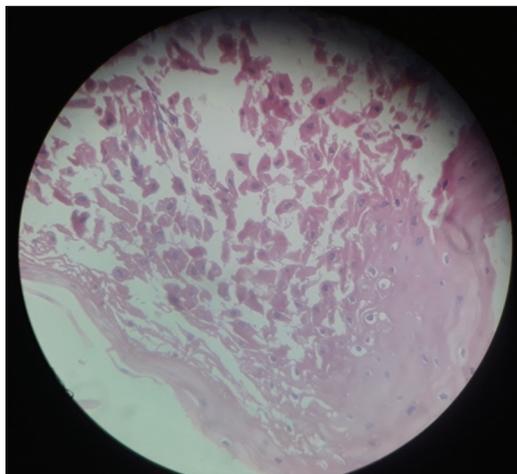


Figure 7: Photomicrograph $\times 10$ (hematoxylin and eosin) showing intraepithelial cleft formation

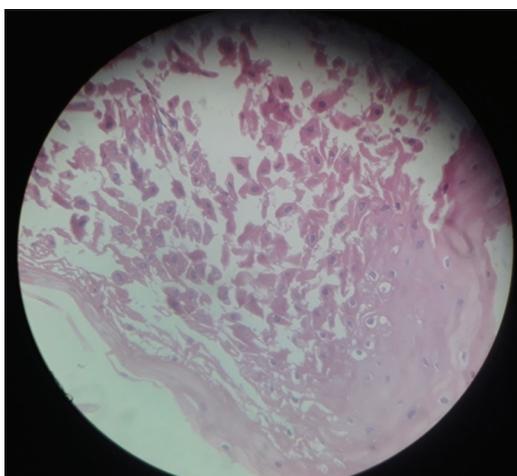


Figure 8: Photomicrograph $\times 40$ (hematoxylin and eosin) showing Tzanck cell



Figure 9: Post-operative photo showing healing after 1½ months

lymphocytes and plasma cells. There was also the presence of blood vessels lined by endothelial cells. Deeper connective tissue stroma also showed the presence of extravagated red blood cells and adipocytes. The patient was sent for further investigation, in which 5 ml of blood sample was collected from antecubital region to carry

out desmoglein ELISA test. Autoantibodies against desmogleins 1 and 3 were detected by ELISA. They were positive for both desmogleins 1 and 3. Based on the history, clinical examination, and investigations, a final diagnosis of pemphigus vulgaris was made and appropriate treatment was rendered. The patient was given an oral rinse containing turmeric (Oro-T mouthwash) and topical steroid ointment (0.1% triamcinolone acetonide ointment) for twice a day for 2 weeks. The patient was also given an antioxidant (oxitard) for once a day for 1 month. Follow-up was done at 3 and 6 months. There were no signs of recurrence [Figure 9].

DISCUSSION

Pemphigus vulgaris is defined as a group of life-threatening blistering disorder of skin and mucous membrane characterized by acantholysis. It may cause severe blistering of the skin and mucous membranes lining the mouth, nose, throat, and genital area. In pemphigus vulgaris, the immune system makes autoantibodies. Our immune system makes antibodies that are required to fight infection. Normally, these antibodies do not attack our own body systems. However, in an autoimmune disease, the immune system attacks our own body. It is seen that blisters occur in the epithelium of the patient where immunoglobulin G autoantibodies are produced in response to triggering factors, target the structured proteins of desmosomes identified as desmogleins 1 and 3. It has recently been found that a new pemphigus antigen desmoglein 4 and other non-desmoglein antigens like human α -9-acetylcholine receptor that regulates keratinocyte adhesion and keratinocyte annexin like molecules binding acetylcholine termed pemphaxin and catenin are also thought to play its role in etiopathogenesis.^[4,5] In general, pemphigus affects the patients in the fourth and fifth decades of life, with the frequency of females affected being more than the males.^[6] More than 50% of patients affected have been reported with initial lesions of oral mucosa followed by skin involvement. The average duration of these oral lesions is seen between 3 months and 1 year.^[7] In the current study, the diagnosis of the lesion was pemphigus vulgaris.

Histopathological examination and serological test (ELISA) were done as a part of investigation. Histopathological examination showed the appearance of Tzanck cells seen in pemphigus vulgaris. Tzanck cells are clumps of large hyperchromatic epithelial cells which are seen lying free within the vesicular space formed as a result of acantholysis in the spinous layer of the epithelium. The results seen are in agreement with various case reports done by Kurian *et al.*,^[8] Rai *et al.*^[9] like the presence of Tzanck cells typically found in pemphigus vulgaris.

Serological test (ELISA) for autoantibodies for desmogleins 3 and 1 was positive. In pemphigus vulgaris, the immune system makes autoantibodies that attack the skin and mucous membrane, leading to damage causing blisters to develop. Autoantibodies attack proteins called desmogleins. The desmoglein proteins are present on the cells in the outer layer of the epidermis. Desmogleins normally act as a glue to hold the cells of the epidermis together. When the autoantibodies attack the desmogleins protein, the cells in the skin and mucous membranes no longer hold together and separate. This causes blisters and erosions. In pemphigus vulgaris, the predominant antibodies produced are against desmoglein 3. In some patients, antibodies against desmoglein 1 can also be found. The results are in agreement with studies done by Gandhi *et al.*^[10] In the current study, the lesion healed well as seen post-operatively in the patient. Steroids are the general medication of choice for treating autoimmune disorders like pemphigus vulgaris. In general, steroids can be used topically, intralesional, and systemically. They are used because they have a strong anti-inflammatory and immunosuppressive effects. In the current study, topical steroid was given in the form of ointment (0.1% triamcinolone acetonide ointment) for twice a day for 2 weeks. The lesions healed well. The results are in agreement with the studies done by Dumas *et al.*^[11] In the current study, besides steroids, antioxidants were also used as these help to prevent oxidation-related tissue damage. These help the body by neutralizing and removing the free radicals from the bloodstream. These help in healing. The results are in agreement with studies done by Mohammadi *et al.*^[12] In addition, an oral rinse containing turmeric was also used as it helps to reduce the severity of the inflammatory response and prevent secondary infection. Moreover, it helped in maintaining oral hygiene.

CONCLUSION

Dental professionals must be familiar with the clinical manifestations of pemphigus vulgaris to ensure

early diagnosis and treatment which can help in better prognosis and course of the disease.

REFERENCES

1. Kavusi S, Daneshpazhooh M, Farahani F, Abedini R, Lajevardi V, Chams-Davatchi C. Outcome of pemphigus vulgaris. *J Eur Acad Dermatol Venereol* 2008;22:580-4.
2. Endo H, Rees TD, Hallmon WW. Disease progression from mucosal to mucocutaneous involvement in a patient with desquamating gingivitis area with pemphigus vulgaris. *J Periodontol* 2008;99:368-75.
3. Scully C, Paes De Almeida O, Porter SR, Gilkes JJ. Pemphigus vulgaris: The manifestations and long-term management of 55 patients with oral lesions. *Br J Dermatol* 1999;140:84-9.
4. Kljuic A, Bazzi H, Sundberg JP, Martinez-Mir A, O'Shaughnessy R, Mahoney MG, *et al.* Desmoglein 4 in hair follicle differentiation and epithelial adhesion: Evidence from inherited hypotrichosis and acquired pemphigus vulgaris. *Cell* 2003;113:249-60.
5. Nguyen VT, Ndoye A, Grando SA. Pemphigus vulgaris antibody identifies pemphaxin. A novel keratinocyte annexin-like molecule binding acetylcholine. *J Biol Chem* 2000;275:29466-76.
6. Iamaroon A, Boonyawong P, Klanrit P, Prasongtunskul S, Thongprasom K. Characterization of oral pemphigus vulgaris in Thai patients. *J Oral Sci* 2006;48:43-6.
7. Shamim T, Varghese VI, Shameena PM, Sudha S. Pemphigus vulgaris in oral cavity: Clinical analysis of 71 cases. *Med Oral Patol Oral Cir Bucal* 2008;13:E622-6.
8. Kurian D, Suresh R, Janardhananand M, Savithri V. Oral lesions: The clue to diagnosis of pemphigus vulgaris. *Case Rep Dent* 2015;2015:593940.
9. Rai A, Arora M, Naikmasur V, Sattur A, Malhotra V. Oral pemphigus vulgaris: Case report. *Ethiop J Health Sci* 2015;25:367-71.
10. Gandhi N, Jain S, Choudhary K, Kumar M. Usefulness of desmoglein 1 and 3 in serodiagnosis of pemphigus vulgaris and its correlation with disease activity-ELISA study. *J Orofac Sci* 2014;6:104-7.
11. Dumas V, Roujeau JC, Wolkenstein P, Revuz J, Cosnes A. The treatment of mild pemphigus vulgaris and pemphigus foliaceus with a topical corticosteroid. *Br J Dermatol* 1999;140:1127-9.
12. Mohammadi H, Djalali M, Daneshpazhooh M, Honarvar NM, Chams-Davatchi C, Sepandar F, *et al.* Effects of L-carnitine supplementation on biomarkers of oxidative stress, antioxidant capacity and lipid profile, in patients with pemphigus vulgaris: A randomized, double-blind, placebo-controlled trial. *Eur J Clin Nutr* 2017;72:99-104.