Gingival Pemphigus Vulgaris-challenges and solution: A case report

ABSTRACT

Pemphigus vulgaris (PV) is a potentially life threatening and rare mucocutaneous disease that usually manifests first in the oral cavity and may later spread to the skin or other mucous membrane. Lesions may occur anywhere on the mucosa but it is unusual for PV to present over the gingiva as a primary site of involvement. A 64 year old female patient reported with a chief complaint of reddish, painful gums with burning sensation since 8 years. The diagnosis of PV is based on clinical findings (Nikolsky's sign positive) and confirmed by histopathological analysis. Medications were prescribed as per indications and requirements. Oral hygiene instructions were given. No recurrence was observed at 1½ year of follow-up. Thus, this case serves to enhance our awareness of gingiva as a site at which systemic disease can manifests itself.

Keywords- Desquamative gingivitis, Mucocutaneous, Nikolsky's sign

Introduction

Pemphigus vulgaris (PV) is a potentially life threatening autoimmune disease that causes blisters and erosions of the skin and the mucous membrane. The term pemphigus was originally named by Wichman in 1791.[1] The word pemphigus originates from Greekword 'pemphix', which means blister or bubble.[2] It is a rare disease (0.1-0.5 cases/100,000 inhabitants/yr), with onset in the fifth or sixth decade of life.[3]

Between 50 to 90% of patients with PV present with oral lesions and 50% of them will have only oral symptoms. For this reason, dentists may be the first to diagnose the disease.[4]

In more advanced stages of PV, desquamative or erosive gingivitis can be observed.[3] Pemphigus is a rare case of desquamative gingivitis (DG).[5]

The gingiva is the least commonly affected site and desquamative gingivitis is a common manifestation of the disease.[6]

Diagnostic delays of greater than 6 months are common in patients with oral PV. This paper describes a rare and unique case of a patient presenting with burning sensation of gingiva since 8 years, but still no systemic manifestation, who was finally diagnosed as having PV.

Case Presentation:

A 64 year old female patient reported to the department of Periodontology of Rama dental college hospital and research centre, Kanpur with a chief complaint of reddish, painful gums with burning sensation since 8 years. The patient initially saw peeling of the gingiva while brushing and aggravation of burning sensation specially after taking hot and spicy food. She also complained of bleeding gums. Though, the patient had visited multiple dentists and received oral prophylaxis and oral hygiene instructions, but there was no improvement in her gingival signs and symptoms. On intraoral examination, both attached and marginal gingiva were intensely erythematous and erosive in appearance [Figure 1A]. These oral lesions

Access this article online	
Website: www.ujds.in	Quick Response Code
DOI: https://doi.org/10.21276/10.21276/ujds.2020.6.2.1	

[1]GUPTA I, [2]SINGH A, [3]GUPTA N, [4]GUPTA R

[1,3,4] Department of Periodontics & Oral Implantology, Rama Dental College-Hospital & Research Centre, Kanpur

[2]Consultant Periodontist, Noida

Address for Correspondence: Dr. Ira Gupta
Postal address:-3A/206 , FLAT: 1-A, Ram Kutir Appartments, Azad Nagar,
Kanpur.-208002
Email : driragupta@yahoo.co.in

Received : 04 May 2020, **Published :** 31 August 2020

How to cite this article: Gupta, I., Singh, A., Gupta, N., & Gupta, R. (2020). Gingival Pemphigus Vulgaris-Challenges and Solution : A Case Report. UNIVERSITY JOURNAL OF DENTAL SCIENCES, 6(2): 75-8.

occurred in repeated cycles of remissions and exacerbations. There was no involvement of buccal mucosa or tongue. Gingiva was the only site involved. Gingival recession was present with periodontal pocket of less than 5mm. Patient had poor oral hygiene with halitosis. Orthopantomogram showed generalized horizontal bone loss. She had no skin or other extraoral lesions and a review of her medical history included history of postmenopausal state and other related systemic diseases and family history was non-contributory. A positive Nikolsky's sign on the gingiva strongly suggested a vesiculobullous disorder [Figure-1B]. Based on the history, clinical signs and symptoms of intraoral lesions, a provisional diagnosis of desquamative gingivitis was given.

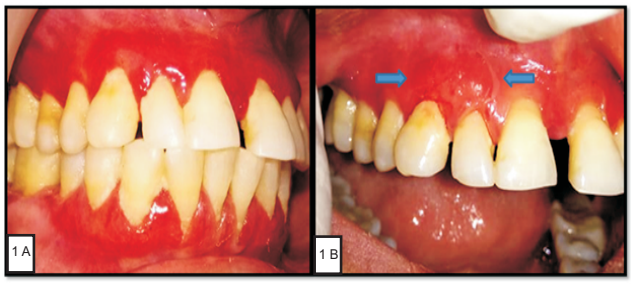

Figure 1: A) Preoperative photograph showing attached and marginal gingiva were intensely erythematous and erosive in appearance. Gingiva was the only site involved. B)Positive Nikolsky's Sign

Routine hemogram, urine and blood sugar examination of patient was done. With a due consent of the patient, a biopsy was taken from peri-lesional site of the involved gingiva with intact epithelium under local anesthesia (LA) [Figure 2A]. The sample was sent for histo-pathological examination to Department of Oral Pathology.

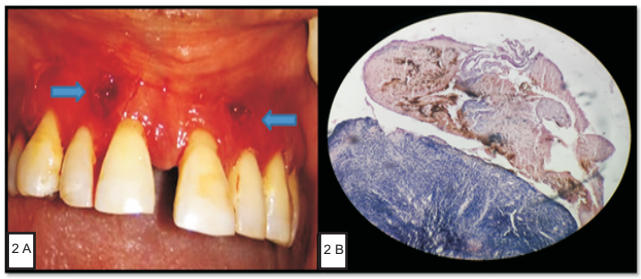

Figure 2: A) incisional biopsy done
B) histopathological picture showing the supra-basilar split (10X Magnification)

The cytological smear was prepared, which showed clusters of darkly stained, round acantholytic cells. Histopathology revealed an area of normal stratified squamous epithelium with a patchy distribution of denuded epithelium and an intra-epiththelial clefting with hyperchromatic acantholytic cells along with chronic inflammatory cell infiltrate in the sub-epithelial and perivascular regions [Figure 2B]. Based on histopathological findings, a final diagnosis of pemphigus vulgaris was made.

The treatment plan comprised of phase 1 periodontal therapy which included scaling and root planing. Oral prednisolone (omnacortil) 40 mg/day for a month with tapering dose, topical steroid oral paste (triamcinolone acetonide 0.1%) twice daily, topical antioxidant application of CNBC gel, Oxum spray along with Multi-vitamin and calcium supplements were prescribed. Soft tooth brush and other oral hygiene instructions were given. On the respective follow-ups, reduction in signs & symptoms were noticed; Pre-operative intense erythematous and erosive marginal and attached gingival lesion [Figure 3A] healed almost completely in 3-4 months. Patient came for regular check up at 6 month [Figure 3B] and she is under follow up since 1½ year [Figure 3C]. Till date, no recurrence & further involvement of other sites is observed.

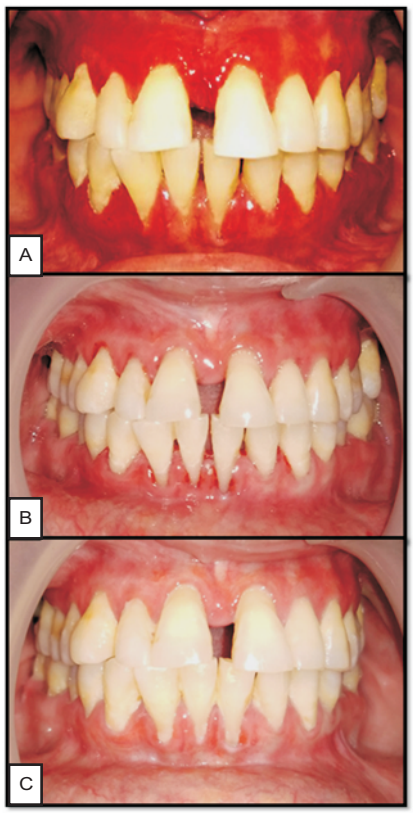

Figure 3: A) Preoperative photographs showing attached and marginal gingiva, intensely erythematous and erosive in appearance. B) 6th month follow-up. C) 1 ½ year follow-up.

Discussion:

Pemphigus is a rare autoimmune disorder with intra-epidermal bullous lesions, which affects the oral, genital or ocular mucosa and the epidermis.[7] Many a times, the mouth may be the only site of involvement for a year or so and this may lead to delayed diagnosis and inappropriate treatment of a potentially fatal disorder.[8] In 60% of cases, oral lesions precede skin lesions by as much as 6 months to 1 year.[9] The Oral lesions of pemphigus vulgaris often extend at sites of irritation or trauma and can range from small vesicles to large bullae and ulcerations.[8] The fragile blisters are easily broken, which leaves behind erosions surrounded by epidermal rings. Applying pressure on healthy skin causes either a bulla or erosion; this effect is known as Nikolsky's sign.[7] Pemphigus vulgaris is the most common form of pemphigus, accounting for over 80% of cases.[10]

The diagnosis of PV is based on three independent set of criteria: clinical features, histology, and immunological tests.[3]

The classic histologic feature seen in pemphigus is acantholysis. In pemphigus vulgaris, intercellular edema develops within the epithelial layers, dissolution of the intercellular bridges occurs, and the intercellular spaces widen, leading to separations between the cells and the formation of blisters just above the basal cell layer (suprabasilar). The free-floating, rounded, acantholytic epithelial cells found within these vesicles can be detected by Tzank preparation, wherein, the base of a blister is scraped and examined microscopically for the acantholytic cells. The basal cells remain tightly attached to the basal lamina, but lose their attachments to one another, producing a so-called "tombstone" appearance. There are relatively fewer inflammatory cells in pemphigus vulgaris compared with other bullous diseases.[9]

A supra-basilar split seen in Pemphigus Vulgaris helps to distinguish this condition from sub-epithelial blistering diseases such as mucous membrane pemphigoid, bullous lichen planus and chronic ulcerative stomatitis.[10]

Though the ELISA testing is the confirmatory test but due to financial constraint of the patient it was not done. In present case, a classic signs of oral or gingival PV i.e., multiple erosions or desquamation and a positive Nikolsky sign along with an intraepithelial blister associated with acantholytic cells in biopsy was seen. This lead to a definitive diagnosis of PV.

Treatment was planned on the basis of clinical and histo-pathological findings which were suggestive of PV. Treatment was beneficial, as, no remission of lesion was seen even after one and a half year follow up.

Pemphigus Vulgaris is generally managed with local and systemic corticosteroid therapy. Treatment of PV is divided into 2 phases. First one is the loading phase, which is for controlling the disease and second one is the maintenance phase, which consists of consolidation and treatment tapering. Local treatment consists of a paste, an ointment or a mouthwash administered alone or in conjunction with systemic treatment. The initial dose of 0.5–2 mg/kg prednisone is recommended. Depending on the response, the dose is gradually decreased to the minimum therapeutic dose, taken once a day in the morning to minimize side effects. When steroids are used for longer periods of time, adjuvants such as Azathioprine or Cyclophosphamide are added to the regimen to reduce the complications of long term corticosteroid therapy.

PV as any autoimmune disease has no definitive cure. However, it has a good prognosis. Approximately, 50% of patients heal without recurrence and 20% of them will require maintenance treatment. A long-term follow-up is mandatory in these patients while they remain on therapy.[4]

Conclusion :

This case serves to enhance our awareness of gingiva as a site at which systemic disease can manifest itself. Timely recognition and therapy of such oral lesions is critical. Thus, periodontist plays a significant role in the early diagnosis and management of such lesions to prevent grave consequences. 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.

Acknowledgement :

We would like to thank Dr. Avinash Singh Bhadauria, MDS, Oral & Maxillofacial Surgeon and Dr. Vishal Mehrotra, Professor & Head of Department of Oral Medicine and Radiology along with the Department of Oral Pathology for helping us with the diagnosis and treatment planning of the case.

References :

1. Javali M.A and Zainab H. Pemphigus vulgaris presenting as gingival involvement. Indian Dermatol Online J. 2012;3:202-04.
2. Kumar S.J, Anand S.P. N, Gunasekaran N and Krishnan R. Oral pemphigus vulgaris: A case report with direct immunofluorescence study. J Oral Maxillofac Pathol. 2016;20:549-53.
3. Bascones-Martinez A, Munoz-Corcuera M, Bascones- llundain, C and Esparza- Gomez G. Oral Manifestations of Pemphigus Vulgaris: Clinical Presentation, Differential Diagnosis and Management. J Clin Exp Dermatol Res. 2010;1:112-15.
4. Gonzalez-Serrano J, Paredes V.M, Lopez-pintor R.M, Arriba L.D and Hernandez G. Case report-Successful treatment of oral pemphigus vulgaris in an insulin-dependent geriatric patient. Gerodontology. 2016;33(4):569-72.
5. Rath S.K and M. Reenesh M. Gingival pemphigus vulgaris preceding cutaneous lesion: A rare case report. J Indian Soc Periodontol. 2012;16(4):588–91.
6. Ohta M, Osawa S, Endo H, Kuyama K, Yamamoto HN and Ito T.Pemphigus Vulgaris Confined to the Gingiva: A Case Report. Int J Dentistry.2011:1-4.
7. Okoh M, Omoregie OF and Otakhoigbogie U. Pemphigus Vulgaris: A case report and literature review.Afr J Oral and Maxillofac Path. Med. 2015;1(2):1-7.
8. Shiezadeh F, Khadem S.S, Golshan A, Mahammadipour H.S, Soufizadeh R and Forouzanfar A.Pemphigus vulgaris that was misdiagnosed as diffused gingivitis, a case report and review.American J Oral Medicine and Radiology. 2015;2(4):195-98.
9. Weinberg M.A, Insler M.S and Campen R.B. Mucocutaneous features of autoimmune blistering diseases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod.1997;84:517-34.
10. Rai A, Arora M,Naikmasur V, Sattur A and Malhotra V.Oral Pemphigus Vulgaris: Case Report. Ethiop J Health Sci. 2015;25(4):367-72.