

Case Report

Oral Pemphigus Vulgaris - A Case Report

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ABSTRACT

Desquamative gingivitis (DG) is characterized clinically by erythema with epithelial desquamation, ulceration, and/or the presence of vesiculobullous lesions of the free and attached gingiva. It is important to be aware of this rare clinical entity so as to distinguish DG from plaque induced gingivitis. Correct diagnosis is critical since proper treatment and follow-up will depend on which disease is involved. Diseases manifesting as DG pose diagnostic problems because the lesions resemble clinically and histologically. So immunohistochemistry is increasingly used along with histology to aid in accurate diagnosis. Hence this case report emphasized on investigation, diagnosis and management of a case of generalized desquamative gingivitis.

INTRODUCTION

Chronic desquamative gingivitis (DG) was first described by Tomes and Tomes¹ in 1894. In 1930, Prinz² and Merrit³ first proposed the term of chronic diffuse DG and first attempted to define the disease process. The term DG describes the clinical appearance of gingiva that are red, glazed, often edematous with loss of stippling and that have areas of superficial epithelial desquamation and/or ulceration. DG is not a disease entity but a clinical manifestation of several different disorders.⁴⁻⁶ As opposed to plaque induced gingivitis, DG is more common in middle-aged to elderly females, is painful, affects the buccal/labial gingiva predominantly, frequently spares the marginal gingiva but can involve

the whole thickness of the attached gingiva and its clinical appearance is not significantly altered by traditional oral hygiene measures or conventional periodontal therapy alone.^{7,8}

The majority of cases of DG are now known to be due to mucocutaneous conditions, in particular lichen planus, pemphigoid and pemphigus.⁹⁻¹² Other causes include allergic reactions to toothpastes/mouth rinses (plasma cell gingivitis),¹³⁻¹⁵ Crohn's disease,¹⁶ psoriasis,¹⁷ linear IgA disease¹⁸ and chronic ulcerative stomatitis.¹⁹

Although a definitive diagnosis of the specific disease or disorder causing DG is required to provide proper treatment, it is almost impossible to do so based solely on clinical manifestations. Therefore, histopathological

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examination and direct immunofluorescence(DIF) testing are often required to establish the final diagnosis. Hence this case report describes the diagnosis and management of a case of generalized DG.

CASE REPORT

A 40 year old female reported to the OPD of department of Periodontics ACPM Dental college and hospital, Dhule with a chief complain of bleeding and soreness of gums since four to six months with periods of exacerbation and quiescence. Her personal history revealed that she cleaned her teeth with finger and manjan due to sore and bleeding gums.

On extra-oral examination, dry patchy skin lesions were noticed on the back and facial region. Intra-oral examination revealed erythematous and generalized desquamative lesions, contour was scalloped with rolled marginal gingiva and blunt interdental papillae, and consistency of gingiva was soft and edematous gingiva, with loss of stippling, bleeding on probing and suppuration. The teeth were not mobile but were associated with presence of plaque and calculus.

Patient was subjected to complete hemogram and all the parameters were found to be within normal range. Orthopantomogram revealed generalized horizontal bone loss. Additional investigation was carried out including direct immunofluorescence test. Patient was subjected to Phase-1 therapy including the planned sessions of scaling and root planing. Patient was referred to a dermatologist regarding treatment for skin lesions. Topical metronidazole and chlorhexidine gel, chlorhexidine mouthwash rinses were prescribed to the patient. Patient was instructed to maintain good oral hygiene and was evaluated every 3 weeks till 6 months. Incisional biopsy was planned for the patient.

The specific ELISA test revealed intermediate presence of desmoglein III antibody (7.6 U/ml). On histopathologic examination, squamous epithelium with suprabasal cleavage and few acantholytic cells were noticed. Focus of dense chronic inflammation was seen. On co-relation of clinical and histologic findings, the features were suggestive of “Pemphigus vulgaris”.

DISCUSSION

Since DG is a common clinical presentation of a variety of diseases, definitive diagnosis and treatment are problematic.^{11,20,21,22} Routine histological examination sometimes cannot differentiate between the DG-causing diseases.^{11,23,24} Thus, immunohistology, particularly immunofluorescence, is increasingly being used with routine histology to more accurately diagnose DG diseases.^{11,23,24} The immune-reactants involved in the immunopathology of DG diseases are diagnostically and can be identified and analyzed by immunofluorescent or immunohistochemical methods.^{11,25,26} While immunohistochemical staining methods are very sensitive, they do not approach the resolution offered by fluorescent techniques, and endogenous enzyme activity often produces problems with heavy background staining.²⁷

Pemphigus vulgaris (PV) is a chronic autoimmune intraepithelial blistering disease. PV almost always affects the mouth and it can be the initial site of presentation in 50% of cases before skin other mucosal sites (oesophagus, pharynx, larynx, nasal, genital) get involved. Skin lesions presents as flaccid fluid-filled blisters on sites exposed to trauma. PV is an uncommon condition affecting males and females in 4th to 5th decade of life.²⁸ In PV autoantibodies are produced against desmosomes (adhesion proteins) specifically desmoglein 3 (Dsg 3). Dsg3 is predominantly expressed in oral

epithelium while both Dsg1 and Dsg3 are expressed in skin. Dsg1 and Dsg3 are components of desmosomal cadherin responsible for holding the cells of the epithelium together. The loss of adhesive function among the spinous cells due to anti-Dsg3 antibodies result in bulla formation immediately suprabasal in pemphigus vulgaris. Specific enzyme linked immunosorbant assay (ELISA) are now available for detecting Dsg1 and Dsg3 autoantibodies.²⁹

In the present case reduction in the clinical signs and symptoms were noticed following phase I therapy and the use of corticosteroids. However regular recall visits were carried out to perform professional scaling in order to reduce plaque associated inflammation.



Fig. I: Pre-operative
Fig. I (A): Frontal view



Fig. I (B): Right lateral view



Fig. I (C): Left lateral view



Fig. I (D): Maxillary palatal view



Fig. I (E): Mandibular lingual view



Fig. II: 1 month following phase I therapy
Fig. II (A): Frontal view



Fig. II (B): Left lateral view



Fig. II (C): Right lateral view



Fig. III: Skin lesion



Fig. IV: Orthopantomogram

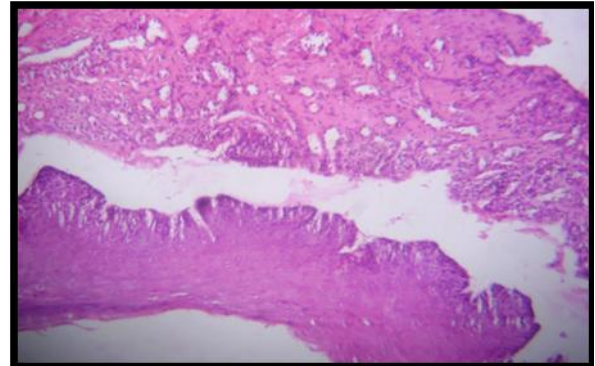


Fig. V: Histopathologic section

CONCLUSION

PV is a rare cause of chronic ulceration of oral mucosa. Newer diagnostic tests and better monitoring of the disease process can be achieved now with a clearer understanding of the role of anti-Dsg antibody-keratinocyte binding in blister formation. Corticosteroids dramatically improve mortality and are considered first choice therapy in PV. A regular follow up, decrease of adverse events, proper modification of the treatment and elimination of triggering factors are inevitable for long-term remission.

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