Pemphigus Vulgaris in a Male Patient: A Case Study

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Abstract

Pemphigus vulgaris (PV) is a chronic, autoimmune, intraepidermal blistering disease of the skin and mucous membranes. The initial clinical manifestation is frequently the development of intraoral lesions, and later, the lesions involve the other mucous membranes and skin. The etiology of this disease still remains obscure although the presence of auto antibodies is consistent with an autoimmune disease. Because only oral lesions are present initially, the chances of misdiagnosing the disease as another condition are increased, leading to inappropriate therapy. In this article, we report a case of PV with only oral manifestations in a 40-year-old male.

Key words: Auto antibodies, intra epithelial vesicle, pemphigus, ulceration

Introduction

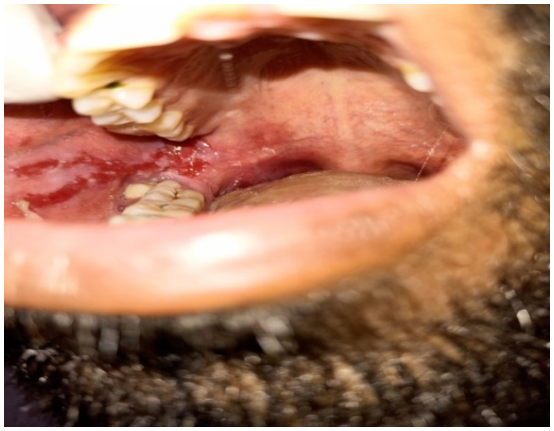
Pemphigus refers to a group of autoimmune, mucocutaneous blistering diseases, in which the keratinocyte antigens are the target of the auto antibodies, leading to acantholysis and blister formation. [1] The word pemphigus originates from Greek *pemphix*, which translates as blister or bubble. Pemphigus can be classified into five major groups: Pemphigus vulgaris (PV), pemphigus foliaceus, paraneoplastic pemphigus (PNP), drug-induced pemphigus and immunoglobulin A (IgA) pemphigus. Oral lesions have been associated with only PV and PNP.[2] Pemphigus is a chronic inflammatory autoimmune bullous disease. There are 0.5 to 3.2 cases reported each year per 100,000 populations, with the highest incidence in the 5th and 6th decade of life, with male to female ratio of 1:2.[3]

Pemphigus Vulgaris is a chronic mucocutaneous disease which usually manifests first in the oral cavity, which later may spread to skin or other mucous membrane. As it is a life threatening disease, it is important that dentist is able to recognize oral manifestations of PV and treat and refer appropriately. [4] It is characterized by the development of flaccid, easily ruptured intraepithelial bulla on apparently normal skin and mucous membranes. The oral cavity is frequently affected in the course of the disease. Intraoral lesions may appear in as many as 50% of the patients without a simultaneous affection of the skin. Although any part of the oral mucosa may be affected, areas exposed to mechanical irritation are most commonly involved. The lesions tend to occur most frequently on the buccal and palatal mucosa and on the gingiva. [4] The oral lesions begin as bleb like blisters or as diffuse gelatinous plaques. Rupture of the bullae occurs in an early stage and may be caused by slight rubbing or minimal mucosal trauma and the lesions are usually painful. Untreated generalized Pemphigus Vulgaris may be fatal. Therefore, by recognizing the oral lesions of Pemphigus Vulgaris, the clinician has a responsibility in the early diagnosis of the disease, which is of the utmost prognostic importance. [5] Here, we report a case of PV with only oral manifestations in a 40-year-old male.

Case report

A 40-year-old male patient resident of Kanpur reported with the chief complaint of ulcers in the mouth and inability in eating spicy food since 4 months. History revealed that patient first noticed a vesicle on right cheek region which progressively increases; later on lesion also appears on left cheek region and posterior part of the palate. The patient had noticed ulcers of mouth which bleed on brushing and increased salivation in the morning was reported. Patient did not report of any skin lesions on other parts of body. The patient had no history of tobacco or any other addiction. There was no history of long-term treatment for any chronic illness or continuous drug intake. No significant family history was found. There was no noticeable finding on general examination. Intra-oral examination revealed ulcerative lesions irregular in shape covered by pseudo membrane with erythematous surrounding, present bilaterally on buccal mucosa along the line of occlusion extending from retro molar trig one towards retro commissural area and involving faucal pillars bilaterally. [Figure 1&2] Nikolysky's sign showed a positive reaction over buccal mucosa and tissue tag was present. Generalized gingival inflammation along with maxillary right first premolar was carious. The clinical presentation of chronic multiple oral ulcers, positive Nikolysky sign ,

tissue tag and lesion extending gradually towards retro commisural area led to provisional diagnosis of oral Pemphigus Vulgaris Differential diagnosis includes mucous membrane pemphigoid,erosive lichen planus and bullous lichen planus.



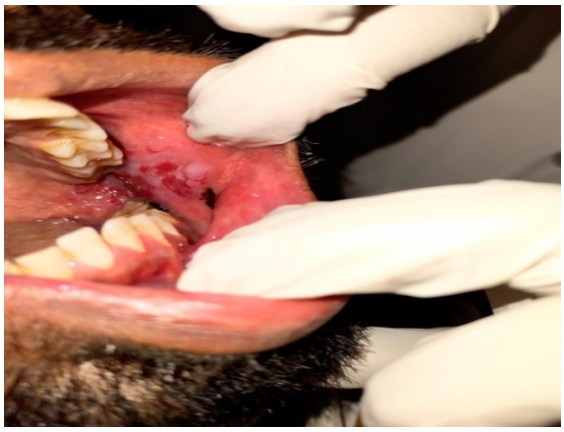
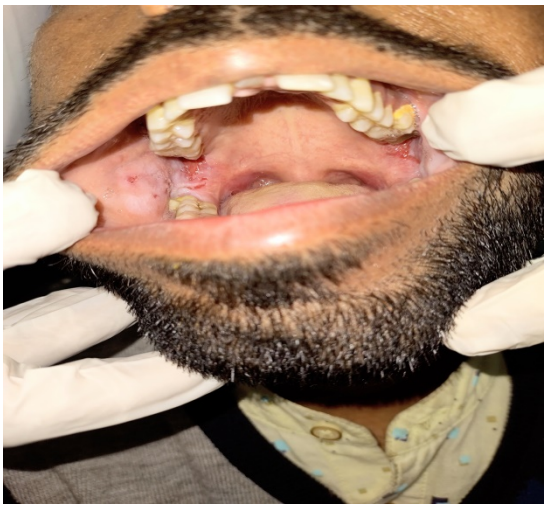


Figure 1&2 bilateral irregular ulcerative lesions present over buccal mucosa

The treatment plan comprised of prednisolone 30 mg/day for 7 days along with multi-vitamin. Triamcinolone acetonide 0.1% ointment 3-4 times per day for 15 days to be applied topically was also prescribed to the patient. Patient was advised to apply content of dexamethasone vial on the eroded areas with clean hand and cotton for seven days. On the first follow-up after 7 days, the patient had The patient responded positively with partial healing of lesion. [Figure 3,4] The dose of prednisolone was tapered to 20 mg/day for next 7 days along with topical application of Triamcinolone actinides 0.1% ointment. Patient further lost the follow up.



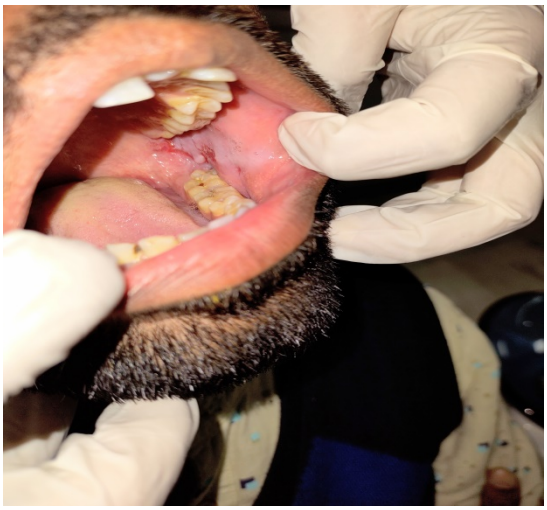


Figure: 3 & 4 bilateral partial healed lesion over buccal mucosa

Discussion

Pemphigus is defined as a group of life threatening blistering disorder of skin and mucous membrane characterized by acantholysis (loss of keratinocyte to keratonicyte adhesion).[6] In pemphigus vulgaris, lesions at first comprise small asymptomatic blisters, although these are very thin-walled, they easily rupture giving rise to painful and hemorrhagic erosions.70-90% cases the first signs of disease appear on the oral mucosa. While lesions can be located anywhere within the oral cavity, they are most commonly found in areas are check, mucosa, tongue, palate and lower lip. The ulcerations may affect other membranes including the conjunctiva, nasal mucosa, pharynx, larynx, esophagus and genital mucosa, as well as the skin where blisters are commonly seen.[7] Increased salivation and

problems with chewing and swallowing are the major subjective complaints.[8]

These groups of diseases are characterized by the production of antibodies against intercellular substances so, therefore, classified as autoimmune diseases.7 Other initiating factors reported included certain food (garlic), infections, neoplasm's and some drugs like caporal, penicillamine and rifampicin although the etiology for the present case is still unknown.[9]

The classical lesion of pemphigus is a thin walled bulla arising on otherwise normal skin or mucosa. A characteristic sign of the disease may be obtained by the application of pressure to intact bullae. In a patient with Pemphigus Vulgaris, bullae, enlarges by extension to an apparently normal surface. Another characteristic sign of the disease is the pressure to apparently normal area resulting in the formation of a new lesion. This phenomenon, called Nikolysky sign, results from the upper layer of the skin pulling away from the basal layer. [10]

An important aspect of patient management is early diagnosis when lower doses of medication can be used for shorter periods of time to control the disease. Dental professionals must be sufficiently familiar with clinical manifestation of pemphigus vulgaris to ensure early diagnosis and treatment, since this in turn determines the prognosis and course of the disease.[10] Pemphigus Vulgaris is generally managed with local and systemic corticosteroid therapy. Treatment is administered in 2 phases: a loading phase, to control the disease, and a maintenance phase, which is further divided into consolidation and treatment tapering.[10]

Conclusion

Pemphigus vulgaris is a rare cause of chronic ulceration of mucosa. The severity and natural history of pem¬phigus vulgaris are variable, but before the advent of steroids, most patients with pemphigus vulgaris died. Treatment with systemic steroids has reduced the mortality rate dramatically.

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