Pemphigus Vulgaris : A Case Report with an Overview

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Introduction

Pemphigus is a group of potentially life threatening autoimmune mucocutaneous disease characterized by epithelial blistering affecting cutaneous and/or mucosal surfaces. The term was derived from the Greek word Pemphix meaning bubble or blister.[1]

Pemphigus vulgaris (PV) is the most common form and frequently involves the oral mucosa. PV runs a chronic course, almost invariably causing blisters, erosions and ulcers on the oral mucosa and skin PV is caused by autoantibodies against epithelial intercellular components, especially cadherins and particularly desmogleins. The main antigen involved in PV is Desmoglein 3(Dsg 3). The oral lesions are most commonly noted on the buccal mucosa, soft palate and alveolar gingiva.

Case History

A 39 year old female patient reported with a complaint of ulceration in the right & left lower region of the mouth since 7 years. Patient was taking treatment for the lesions but since there were frequent exacerbations & remissions, so proper treatment could not be advocated because of inconclusive diagnosis. Patient was apparently asymptomatic when 3 months back he again noticed lesions & was having discomfort. On examination, ulcerated lesion with ill defined margin was seen extending from premolar region of the right buccal vestibule to retromolar area (Figure1). Also Exfoliative cytology was performed and the findings are the presence of Tzanck cells in clusters (Figure 2).

On histopathological examination, intraepithelial split was seen and presence of Tzanck cells with hyperchromatic nucleus was seen within the split. Chronic inflammatory cell infiltration was evident in edematous to fibrovascular connective tissue stroma (Figure 3).

Discussion

There are many variants of Pemphigus Pemphigus Vulgaris, Pemphigus Foliaceous, Pemphigus Paraneoplastic & Drug Induced Pemphigus. Pemphigus vulgaris is the most common form & frequently involves mouth. The main antigen in pemphigus vulgaris is Dsg3 where as in pemphigus foliaceous is Dsg 1. However 50% of PV patients also have autoantibodies to Dsg 1, and the proportion of Dsg 3 & Dsg 1 antibody appear to be related to clinical severity. Those PV which are predominantly oral have only Dsg3 antibodies. In Paraneoplastic Pemphigus, Desmoplakin 1, Desmoplakin 2 & BP 230 antigens are involved. In Drug Induced Pemphigus Dsg 3 antigen is mainly involved. Patients with active disease have circulating and tissue bound autoantibodies of both the immunoglobulin G1(IgG1) and immunoglobulin G4 (IgG4) subclasses. The binding of autoantibodies results in a loss of cell-cell adhesion causing acantholysis. The cause of Pemphigus vulgaris remains unknown. However, several relevant factors have been identified such as genetic factor, age, disease association, drugs.[2]

In Pemphigus vulgaris, lesions at first comprise small asymptomatic blisters, although these are very thin walled and easily rupture giving rise to painful and hemorrhagic erosions. In most cases (70% - 90%), the first signs of the disease appear on the oral mucosa, while the lesions can be located anywhere within the oral cavity. They are most often found in areas subjected to frictional trauma such as buccal mucosa, tongue, palate and lower lip. The ulceration may affect other mucosal membranes including the conjunctiva, nasal mucosa, pharynx, larynx and genital mucosa as well as the skin where intact blisters are more commonly seen. The diagnosis is generally based on oral manifestations while confirmation is provided by the histopathological findings, which show the presence of intraepithelial blisters, acantholysis and Tzanck cells.[3] The diagnosis of Pemphigus vulgaris should be confirmed by direct immunofluorescence examination of fresh perilesional tissue or tissue submitted in Michel's solution. With this procedure, antibodies (usually IgG or IgM) and complement components (usually C3) can be demonstrated in the intercellular spaces between the epithelial cells in almost all patients with the disease. Indirect immunofluorescence is also typically positive in 80% to 90% of cases, demonstrating the presence of circulating autoantibodies in the patient's serum.[4]

Most patients could be initially misdiagnosed, usually as aphthous stomatitis, gingivo-stomatitis, erythema multiforme, erosive lichen planus or oral candidiasis and may be improperly treated for months or even years. The initial aim of the treatment is to induce disease remission. This should be followed by dosage of maintenance treatment using the minimum drug dosages required for disease control in order to minimize their side effects. Corticosteroids are the primary drugs used in the treatment of pemphigus vulgaris. Mild localized lesions of oral mucous membrane pemphigus in patients with low titers of circulating autoantibodies may be controlled, atleast temporarily, with topical corticosteroid rinses or creams, including agents such as clobetasol proprionate. Intralesional triamcinolone may be used for resistant local lesions.[5]

Conclusion

In the present case, denuded blisters of various sizes ranging from the size 0.25x0.25 cm to 0.5cmx0.5cms were seen on the lower lip and buccal mucosa. Histopathology showed Parakeratinized stratified squamous epithelium with intra epithelial split also showing basal cells abutting on the underlying connective tissue. Tzanck cells were seen within the split arranged in the groups or single cells. In our case, sections were obtained from intraoral biopsy specimen submitted for histopathological examination. The principal histologic characteristics in present case were evaluated which correlated well with the histological features seen in PV.

In this case, we describe the management of the patient who has previously undergone treatment based on misdiagnosis and whose complaints were not relieved even after a long time. We also distinguish the diagnosis of oral pemphigus vulgaris, which often results in the patient's death, if untreated from other similar oral lesions, and the importance of the role of dentist in early diagnosis and treatment.

References

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Legends

Fig. 1. Ulcers with irregular borders and mild erythema around the periphery. The surface was covered with yellowish slough.
Fig. 2. Photomicrograph of smear shows the presence of Tzank cells with hyperchromatic nucleus. (H & E 10x)
Fig. 3. Photomicrograph showing intraepithelial split and basal cells abutting on the underlying connective tissue. Tzank cells were seen within the split. (H & E 10x)

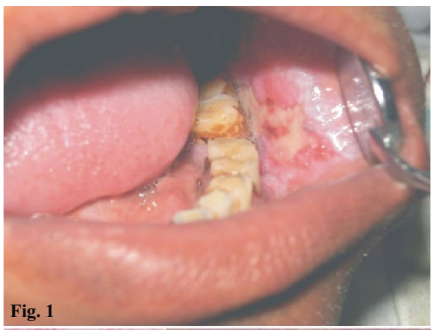
Fig. 1

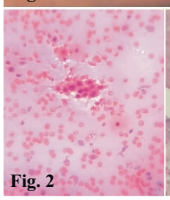
Fig. 2

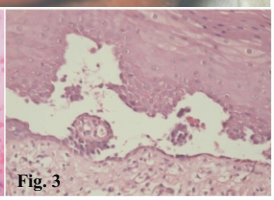
Fig. 3