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### RESEARCH ARTICLE

#### ORAL PEMPHIGUS: CASE REPORT & TREATMENT STRATEGIES

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#### Manuscript Info

#### Abstract

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#### Introduction:-

Pemphigus is a potentially fatal skin disease characterized by the appearance of vesicles and bullae. Pemphigus comes from the word *pemphix* which in Greek means a bubble or a blister. Originally coined by Wichman in 1971, Pemphigus includes a group of autoimmune blistering diseases of skin and mucous membrane. Intradermal blisters are its pathognomonic clinical feature and histologically characterized by IgG antibodies directed against the cell surface of keratinocytes. There are three primary subtypes of the disease that is, Pemphigus vulgaris, Pemphigus foliaceus, and Paraneoplastic pemphigus. Out of these most commonly seen variant is pemphigus vulgaris.

This disease has a very strong immunogenetic predisposition. Blisters are associated with the binding of antibodies to keratinocytes, desmosomes & desmosome free areas resulting in loss of cell adhesion. Disease activity can be linked to antibody titer. It affects all races with an equal gender predisposition. Mean age of onset is 50 to 60 years; however it may affect the younger and older individuals as well. The bullous lesions contain thin, watery fluid shortly after appearance but may soon become purulent or sanguineous. On rupturing a raw eroded surface can be seen. The loss of epithelium caused by rubbing apparently unaffected skin is known as NIKOLSKY'S SIGN. It occurs due to prevesicular edema that weakens the dermal-epidermal junction. Mucosal lesions often precede the lesions on the skin and other parts of the body. Patients may have irregularly shaped gingival, buccal or palatine erosions<sup>1</sup>. Affected individuals are unable to eat or drink due to the associated pain. It is characterized microscopically by suprabasilar split, acantholysis which causes clumps of cells to be suspended free within the vesicular space known as *Tzanck cells*<sup>1</sup>. Direct Immunofluorescent testing (DIF) seems to be the mainstay in diagnosing this lesion.

#### Case Report:-

A 34 year old patient reported to the department of Oral Medicine in AB Shetty Memorial Institute of Dental Sciences with the chief complaint of severe pain and burning sensation in the mouth with a resultant inability to eat and drink since the last three days. The ulcers first presented three months back on the tongue and the buccal mucosa and appeared like a white plaque, when the patient attempted rubbing it off it lead to bleeding and fluid discharge. Following this he took admission in a local hospital and was treated symptomatically. Remission occurred but within two weeks recurrence of ulcers was seen. Following this the patient also took homeopathic medication but the condition worsened and led to the present state. The patient had no history of tobacco consumption or any other addiction. No history of long-term treatment for any chronic illness, continuous drug intake or any significant family history. There was no significant finding on general examination. On intraoral examination multiple erosive lesions

were found with erythematous halo surrounding them, affecting bilateral buccal mucosa, tongue, floor of the mouth & lower lip with sloughing. Tenderness was present on palpation & Nikolsky's sign was positive. The clinical presentation of recurrent oral ulcers and positive Nikolsky's sign in this case led to provisional diagnosis of vesiculo-bullous lesion affecting the oral cavity. Differential diagnosis included aphthous stomatitis, chemical burns, pemphigus vulgaris, dermatitis herpetiformis, erythema multiforme bullosum, epidermolysis bullosa, erosive lichen planus, bullous pemphigoid & Para neoplastic pemphigus.

A smear was prepared and studied which on H & E staining showed epithelial & inflammatory cells. In a few areas clumps of rounded epithelial cells with enlarged & hyperchromatic nuclei were visible suggestive of Tzanck cells. Incisional biopsy was taken from a representative site, the microscopic features demonstrated stratified squamous parakeratinized epithelium with certain areas showing suprabasilar cleft and acantholytic cells. Underlying connective tissue showed dense chronic inflammatory cell infiltrate comprising of plasma & lymphocytes. The diagnosis of Pemphigus was given based on the histologic findings and to further confirm the diagnosis DIF test was performed. DIF demonstrated in vivo deposits of IgG (IgG1, IgG4) on the surface of keratinocytes, thereby confirming the diagnosis.

The treatment plan comprised oral prednisolone 60 mg/day along with multi-vitamin supplementation and analgesic in the form of a lidocaine gel for local application. After six days of steroid therapy, the patient was symptomatically better and clinically reduced sloughing and erythema was seen. The patient was discharged since he was relatively comfortable and could consume oral diet. Patient was reviewed after one week, there was complete resolution of sloughing and a few erythematous zones were noted. Due to the remission of symptoms the dose of prednisolone was reduced to 40mg. Again after ten days the patient was reviewed and was completely asymptomatic, therefore we reduced the dose of prednisolone to 20mg tapering it over a period of one week and then stopping the medication.

### **Discussion:-**

Generally in Pemphigus vulgaris small asymptomatic blisters are seen, they are really thin walled & tend to rupture, thereby causing pain and hemorrhagic erosions. Lesions can be found in any place in the oral cavity but the sites most vulnerable are the ones subjected to constant friction<sup>2</sup>. Due to the presenting features the disease tends to be misdiagnosed as aphthous stomatitis, gingivostomatitis, or erosive lichen planus. As was the case with our patient, who in his earlier visit to a doctor was misdiagnosed as a case of aphthous stomatitis. Therefore we will stress on the importance of an early correct diagnosis, which enables the use of lower dosage of medications over a shorter period of time.

Since our patient reported to us with an aggressive lesion, with acute symptoms, we conducted a smear test, biopsy and DIF simultaneously. Also, culture sensitivity of the lesional exudates along with a complete blood profile of the patient was done. Till the diagnosis was confirmed supportive therapy in the form IV fluids and local care was provided. The culture sensitivity was negative and all the blood profile was completely normally, however the smear, biopsy and DIF suggested the presence of pemphigus vulgaris.

Once the diagnosis has been confirmed the treatment should be started as soon as possible with systemic corticosteroids that remain the treatment of choice for this lesion<sup>3</sup>. Corticosteroids do cause rapid remission but also lead to various adverse effects like peptic ulcers, weight gain, hypertension, diabetes & osteoporosis<sup>3</sup>. Therefore when steroids have to be used for longer periods of time adjuvants such as Azathioprine or Cyclophosphamide are added to the regimen<sup>4</sup>. Even in our patient ant acids were prescribed to prevent gastritis.

Emerging therapies include plasmapheresis, intravenous immunoglobulins, immunoadsorption, cholinergic agonists, tumor necrosis factor-alpha antagonists (infliximab & etanercept) & cholinergic agonists<sup>3</sup>. Out of all these therapies plasmapheresis is the one that is being extensively studied and its use questioned for the treatment of pemphigus vulgaris. In a study by Matthew S. Turner, BS the use of plasmapheresis in conjunction with immunosuppression has been studied. They have suggested a series of five plasma exchanges within a period of eight days followed by administration of immunosuppressive drugs to prevent a rebound flare of the disease. They have concluded that this line on treatment should be employed for patients who do not respond to the conventional treatment or for patients with a severe form of the disease<sup>5</sup>.

**Conclusion:-**

In this article we illustrated the common oral presentation of pemphigus vulgaris, the importance of cinching the correct diagnosis and preventing it from being misdiagnosed as some of the other oral lesions with similar presentation. It should be considered as a differential diagnosis whenever a patient presents with a history of multiple, chronic, non-healing ulcerations that begin as a blister or bullae. Definitive diagnosis can be achieved by histological inspection and DIF. Once diagnosis is confirmed we stressed on the importance of administration of systemic corticosteroids to reach a disease free state & also the use of some new treatment modalities for a more severe form of the disease.

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