

Pemphigus Vulgaris-A Case Report

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Abstract: Pemphigus is a term derived from the Greek word Pemphix (bubble or blister) for a group of potentially life threatening autoimmune mucocutaneous diseases characterized by epithelial blistering affecting cutaneous and/or mucosal surfaces. 1 Pemphigus is an autoimmune disease in which a patient's own circulating antibodies become altered so that they attack the points of adhesion of the epithelial cells, one to another, of the skin and mucous membranes. It most commonly develops in the fourth to sixth decades of life. We present an 18 year old patient with pemphigus vulgaris. The etiology was unknown.

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I. Introduction

Pemphigus vulgaris is a chronic autoimmune intraepithelial blistering disease. It is characterized by intraepithelial blister formation that results from breakdown of the cellular adhesion between epithelial cells. In 1964 autoantibodies against keratinocytes surfaces were described in patients with pemphigus. Clinical and experimental observations indicate that the circulating autoantibodies are pathogenic. 2 Pemphigus vulgaris almost always affects the mouth and it can be the initial site of presentation in 50% of cases, before skin and other mucosal sites become involved. Skin lesions present as flaccid fluid-filled blisters on sites exposed to trauma. The blisters break to form large denuded areas of skin which can prove fatal if extensive areas are involved.

The aim of this article is to distinguish the diagnosis of oral pemphigus vulgaris, which often results in the death, if untreated, from other similar oral lesions, and the importance of the roles of dentists in early diagnosis and treatment.

II. Case Report

An 18 yr old patient came to the department of oral medicine & radiology with a chief complaint of recurrent ulcerations on lips & inside the mouth since 3 years. Recurrent ulcers since 3 years. This is the 5th time & this time present since 1 week. Ulcers arise spontaneously, pain present while swallowing, the ulcers appear on the lip & then progresses inside the mouth, first blisters filled with fluid appear & swelling of the lips start. Ulcers also bleeds, salivation increased. Burning sensation present while chewing food. Taking treatment from an ENT specialist since 2006 relief occur after taking treatment & lesions heal within 15-20 days. Pain-VAS-100%. Burning sensation-100%. In extraoral examination erosion was present on the left ear. In intraoral examination swelling present on lips, blood stained crustation present on the right side of lower lip, diffuse Erythematous area was present on upper and lower labial mucosa. Multiple ulcers were present on the right and left buccal mucosa.

III. Investigation

Punch biopsy was done from a advancing edge of the bulla, where areas of characteristic suprabasilar acantholysis was observed. Cytologic smear was taken from early, freshly opened vesicle. This showed "Tzanck cells". Complete haemogram was done which was normal. Patient also showed a positive Nikolsky's sign.

IV. Discussion

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, the first signs of disease appear on the oral mucosa. While the lesion can be located anywhere within the oral cavity, they are

most often found in areas subjected to frictional trauma, such as the cheek mucosa, tongue, palate, and lower lip.³ It develops in 4th to 6th decades of life.

Genetic predisposition linked to HLA class II alleles seems to be of some significance, because it has been shown to occur with increased frequency in certain ethnic groups and within families.

Ashkenazi Jews and person of Mediterranean origin are especially at risk for pemphigus.

Classic lesion of pemphigus is a thin-walled bulla arising on otherwise normal skin or mucosa. Bulla rapidly breaks but continues to extend peripherally, eventually leaving large areas denuded of skin.⁴ A characteristic sign of the disease may be obtained by application of pressure to an intact bulla. In patient with PV, the bulla enlarges by extension to an apparently normal surface.

Another characteristic sign of the disease is that pressure to an apparently normal area results in the formation of a new lesion. This phenomenon is called as Nikolsky sign.⁴ Any mucosal and skin surface may be involved, and in severe cases, the conjunctival, pharyngeal, and laryngeal mucosa may all be involved, along with extensive skin lesions.

In 90%, of cases oral lesions develop and in 60% cases they occur first. Initial lesion most frequently occur on buccal mucosa because the epithelium demonstrate less intercellular substance and fewer intercellular junction making the area more susceptible to acantholysis. Palate and gingiva are other common site of involvement. Oral lesions may begin as classic bulla on noninflamed base with formation of shallow ulcer as bullae break rapidly. A thin layer of epithelium peels away in an irregular pattern, leaving a denuded base.⁵ The edges of the lesion continue to extend peripherally over a period of weeks until they involve large portion of the oral mucosa. It is common for the oral lesions to be present for months before the skin lesion appear. Major variants of pemphigus are Pemphigus Vulgaris, Pemphigus Vegetans, Pemphigus Foliaceus, Pemphigus Erythematosus, Paraneoplastic Pemphigus.

The lab tests which can be done are- Biopsy- best for intact vesicle or bulla of less than 24 hours, and biopsy is done from advancing edge of the lesion to see acantholysis. Tzanck smear-it is done to demonstrate Tzanck cell which often are found lying freely within the vesicular space. Indirect immuno-fluorescent antibody test-antibodies against intercellular substance can be seen.⁶ The titers of antibody are directly related to the level of the clinical disease. Direct test- antibody will bind the immunoglobulin deposit in the intercellular substance and show positive fluorescence under fluorescence microscope.⁶ Differential diagnosis includes Recurrent aphthous stomatitis- these lesions are severe but heals and recure; but in pemphigus lesion, course extends over a period of weeks or months. Lesions of pemphigus are not round, symmetrical but are shallow and irregular and often have detached epithelium at the periphery. In Dermatitis herpetiformis mucosal involvement is rare, Tzanck test and Nikolsky's sign are negative. Skin changes are polymorphic. The target like lesion of the skin in erythema multiforme help to distinguish it. In viral infection the bulla are larger, Bullous pemphigoid are usually not intraoral, immunofluorescence can be done. Bullous drug induced exanthema- negative Tzanck test and Nikolsky's sign. In Benign mucous membrane pemphigoid- lateral shifting of the blisters usually not possible, skin involvement less frequent, eye involvement in 80% of cases, blisters are painless.⁷

In management of pemphigus Corticosteroids- topical or systemic prednisone is used to disease under control and once the disease is under control, dose of prednisone is reduced. Combination therapy is used in which high dose of corticosteroid are combined with immunosuppressive drug such as cyclosporine or azathioprine. Plasmapheresis is useful in patient's refractory to corticosteroid. Administration of 8-methoxypsoralen, followed by exposure of peripheral blood to ultraviolet radiation. Systemic corticosteroids remain treatment of choice for pemphigus as they are both effective & capable of inducing a rapid remission. However, adverse effects of corticosteroids are both time & dose dependent. Adjuvant therapies are, therefore, used to provide a steroid sparing effect. As these treatments typically have a slower onset of action (i.e., 4-6 weeks), they are most beneficial as maintenance therapies. Emerging therapies include, Intravenous Immunoglobulin, Plasmapheresis, Immunoabsorption, Extracorporeal Photochemotherapy, Rituximab, Tumour Necrosis Factor-alpha Antagonists (Infliximab & Etanercept), Cholinergic Agonists, Other Experimental Therapies - Desmoglein 3 Peptides & KC706, Gene therapy.

V. Figures



Figure 1: 18 yrs old patient



Figure2: Erosion was present on the left ear



Figure 3: Lips- blood stained crustation



Figure 4: clinical photographs of lips

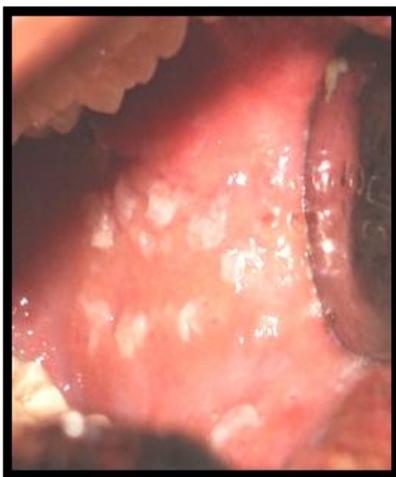


Figure 5: clinical photographs of buccal mucosa



Figure 6: clinical photographs of lips



Figure 7: clinical photographs of tongue

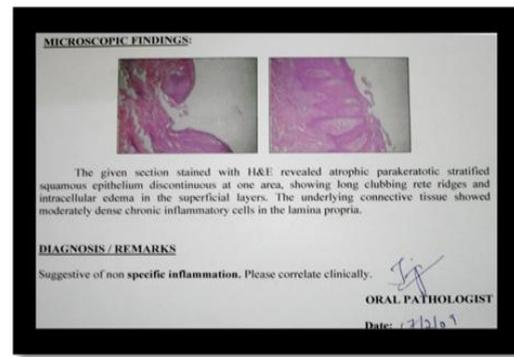


Figure 8: biopsy report

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