Pemphigus Vulgaris- A Case Report

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ABSTRACT: Pemphigus vulgaris is a chronic autoimmune mucocutaneous disease that initially manifests in the form of intra oral lesions, which spread to other mucous membrane and skin. The etiology of pemphigus vulgaris is still unknown. The pemphigus group of disease is characterized by the production of autoantibodies against intercellular substances and is thus classified as autoimmune disease. Most patients are initially misdiagnosed and improperly treated for many months or even years. Here we report a case of pemphigus vulgaris in a 50 year old male patient who reported to department of oral medicine and radiology with a complain of chronic oral ulcer.

KEY WORDS: Pemphigus Vulgaris , Oral Cavity, Autoimmune Disease.

INTRODUCTION:

Pemphigus refers to a group of rare chronic mucocutaneous diseases characterized by painful lesions caused by intraepidermal acantholytic structures in the skin and mucous membrane. The exact nature of the disease remains unknown. Pemphigus vulgaris (PV) is characterized by intra-epithelial bulla formation that result from breakdown of the cellular adhesion between epithelial cells.

Literature reveal similar prevalence between sexes. In 1964 autoantibodies against keratinocyte surfaces were described in patients with pemphigus. The disease has two known types – pemphigus vulgaris and pemphigus foliaceus. Although there has been reports of children with this condition. It is usually seen between 5 – 7 years of age. The mucosal lesions of PV are located most frequently in the oral and pharyngeal mucosa; however the conjunctiva, larynx, nasal mucosa, vulva, vagina, cervix may be involved. The bullous lesions are painful, slow to heal and show a tendency to become invasive. Although any part of the oral cavity may be affected, the soft palate, buccal mucosa and lips are the most common sites.

However, it is generally accepted that disease such as lichen planus and pemphigoid lesions sometimes show similar clinical appearance. In the literature, there have also been reports several other disorders manifesting DG including psoriasis, dermatitis, herpeticiformis, chronic ulcerative stomatitis, erythema multiforme, epidermolysis bullosa and Kindler syndrome. Similar appearances may be seen in reaction to dental materials, mouth washes and medications, and in lupus erythematosus, Crohn's disease, sarcoidosis and leukemia.

Oral lesions are a hallmark of PV and occur in almost all cases and present the preliminary in more than half of patients. Clinically the oral lesions are characterized by blisters that rapidly rupture, resulting in painful erosions.

Definitive diagnosis cannot be achieved with clinical examination alone, as several other vesiculobullous and ulcerative lesions have a similar appearance. Incisional biopsies are required.

Clinical and experimental observations indicate that circulating autoantibodies are pathogenic. An immunogenetic predisposition is well established. Blisters in PV are associated with the binding of IgG autoantibodies to keratinocyte cell surface molecules. These intercellular or PV antibodies bind to keratinocyte desmosomes and to desmosomes free areas of the keratinocyte cell membrane.

In the absence of systemic treatment, the oral lesions are almost invariably followed by the involvement of skin, on and occasionally other epithelial surfaces such as oesophagus. Unless there are localized oral lesion in which case treatment with tropical corticosteroids suffices for a time, systemic corticosteroids (eg prednisolone) are essential sometimes administered intravenously. Once the condition is under control, the dosage of prednisolone can be reduced.

Adjuncts alternatives include azathioprine, cyclosporine, cyclophosphamide, dapsone, levimasole,
Unfortunately the high doses and prolonged administrations of corticosteroids that are often needed to control the disease result in several side effects, many of which are serious and life threatening.

**CASE REPORT:**

A 50 year old male patient visited to the Department of Oral Medicine and Radiology with a chief complain of painful ulcerative area on the right buccal mucosa since last 10 months. The patient reported that the ulcer cause discomfort and affected his normal oral function. Patient reported that he was under treatment for the same in some local hospital but showed no improvement.

Personal and family history was uneventful. On intraoral examination, ulcer was seen on the buccal mucosa, with irregular border. It measured approximately 1.5X2 cm. It was about 1.5 cm deep. The overlying mucosa was punched out and ulcerated. On extra oral examination skin lesions were seen on the side of neck and under arms. It was approximately 0.5X0.5 cm. The overlying mucosa rubbed off on rubbing. On the basis of history and clinical examination a provisional diagnosis of pemphigus vulgaris was given. A differential diagnosis of chronic ulcer and pemphigoid was given. A biopsy was taken from the right buccal mucosa from the periphery of the lesion involving normal and ulcerated mucosa and was sent for histopathological examination. Histological finding in the case were characterized by parakeratinised stratified squamous epithelium. Predominant subepithelial split was evident with few areas showing detachment of epithelium overlying connective tissue. Inflammatory component was evident within the stroma. On the basis of histological findings, final diagnosis of pemphigus vulgaris was given.

Initially prednisolone 80 mg was given. Tab Pan D 20 mg was given for stomach acidity caused by steroids. For skin lesions gentian violet was given for topical application once in two days. Patient was recalled after 14 days. Prednisolone was decreased by 10 mg in the following week. At the end of 7th week prednisolone was decreased by 30 mg. After 6 months of treatment the cheek mucosa was healed without scarring.

**DISCUSSION:**

In PV acantholysis occurs deep in the stratum spinosum, creating a suprabasal cleft, whereas in pemphigus foliaceus the bulla formation occurs at a higher level. In PV the oral mucosa is the first site to be involved (upto 70% of cases) and it is the only site affected in over 50% of patients. Distal extension can occur in PV, affecting the pharynx, larynx and oesophagus producing dysphagia.

In pemphigus vulgaris, lesions at first comprise small asymptomatic blisters, although these are very thin –walled and easily rupture giving rise to painful and haemorrhagic erosions. In most cases (70 – 90%), the first sign of 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 mainly in the tongue, palate and lower lip. The ulcerations may affect other mucous membrane including the conjunctiva, nasal mucosa, pharynx, larynx, oesophagus and genital mucosa, as well as the skin where intact blisters are commonly seen. In the present case, the oral lesions were the first manifestation of the disease mainly cheek mucosa. The diagnosis is generally based on the oral manifestations, while confirmation is provided by the histological findings, which shows the presence of intraepithelial blisters, acantholysis and Tzanck cells. In our case, a biopsy of the intraoral lesion was obtained. Most patients could be initially misdiagnosed, usually as erythema multiforme, erosive lichen planus or oral candidiasis, and may be improperly treated for months or even years.

Other dermatological diseases associated with large bullae on the oral mucosa which are identified as differential diagnosis of pemphigus vulgaris, should be ruled out. One of them is pemphigoid, the bullous dermatitis of autoimmune origin that is relatively uncommon. It may accompany as a facultative paraneoplastic dermatosis, an underlying malignant disease. The oral mucosa is affected around in every fifth patient. The oral lesion does not precede dermatological symptoms. The bullae on the mucosa are smaller, their duration is short, and remaining erosions heal relatively fast without scars.

Oral signs are always missing in other bullous diseases such as pemphigus erythematosus, pemphigus foliaceus or pemphigus benignus familiaris chronicus. Hailey which is important for differential diagnosis.

The initial aim of treatment is to reduce disease remission. This should be followed by a period of maintenance treatment using the minimum drug doses required for disease control in order to minimize their side effects.

Systemic oral corticosteroids are the treatment of choice in the treatment of pemphigus vulgaris. Topical steroid therapy alone is insufficient for sustained control of the disease because of the systemic autoimmune characteristics of PV. There is no consensus regarding the initial steroid dosage needed to produce remission and its effect on the subsequent course of the disease, but it is generally agreed that low doses (below 60 mg/day) usually do not suffice to induce initial control. In the mild localized lesions of oral mucous membrane pemphigus in patients with low titres of circulating auto antibodies may be controlled at least temporarily, with topical corticosteroid.
rinses and cream, including clobetasol propionate. Intralesional triamcinolone may be used for resistant local lesions. PV is a chronic autoimmune mucocutaneous disease that primarily involves the oral cavity. Therefore, diagnosis of periodontal lesion is very important in facilitating early treatment of PV. Although there is no consensus regarding the initial steroid dosage needed to induce remission, it is suggested that administration of high dose corticosteroids may cause fatal complications.

CONCLUSION:

Pemphigus vulgaris is rare with reported incidence of 0.1-0.5 cases per 100,000 individuals worldwide per year. It is slightly predominant in women and primarily manifest in adults during the fifth and sixth decade of life. Juvenile cases have been reported, but are rare. In the majority of patients, pain, mucous membrane erosions are the presenting sign of PV and may be the only sign of PV for an average of 5 months before skin lesions develop.

Morbidity and mortality are related to the extent of the disease, the maximum dose of systemic steroids required to induce remission, and the presence of other diseases. Prognosis is worse in patients with extensive disease and in older patients.

REFERENCES:


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LIST OF PHOTOGRAPHS

Fig.1 Intra Pre-Operative oral photograph

Fig.2 Lesions on the skin

Fig.3 Post Treatment photograph