Pemphigus vulgaris : A Case Report

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ABSTRACT :

Pemphigus is a serious chronic skin and mucous membrane disease characterized by the appearance of vesicles and bullae, small or large fluid-filled blisters that develop in cycles. It is mediated by circulating auto antibodies directed against keratinocyte cell surfaces. Widespread ulceration following rupture of the blisters leads to painful debilitation, fluid loss, and electrolyte imbalance. Before the use of corticosteroids, death was not an uncommon outcome for patients with pemphigus vulgaris [1] . Dental professionals must be sufficiently familiar with the clinical manifestations of pemphigus vulgaris to ensure early diagnosis and treatment, since this in turn determines the prognosis and course of the disease. Here, we report a case of pemphigus vulgaris that was misdiagnosed in its earliest stage.

Keywords : Pemphigus, Oral ulceration, Topical prednisone, Systemic prednisone, Pemphigus vulgaris.

INTRODUCTION :

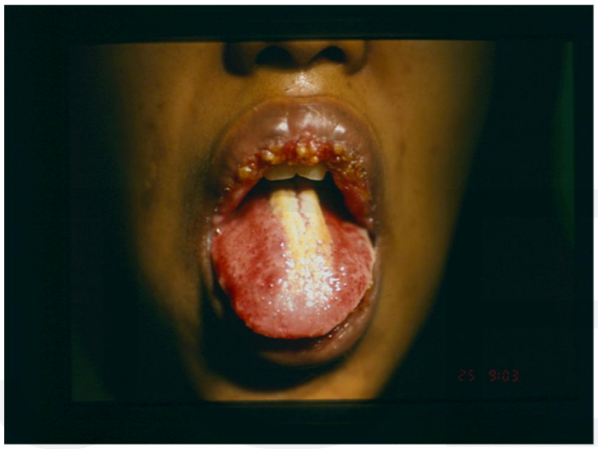
Pemphigus includes a group of autoimmune blistering diseases of the skin and mucous membrane, Characterized histologically by intradermal blisters and immunologically by the finding of in vivo bound and circulating immunoglobulin G (IgG) antibody directed against the cell surface of keratinocytes.[2]

The three primary subsets of pemphigus include pemphigus vulgaris (PV), pemphigus foliaceus, and paraneplastic pemphigus. Each type of pemphigus has distinct clinical and immuonopathologic features. Pemphigus vulgaris accounts for approximately 70% of pemphigus cases. [2]

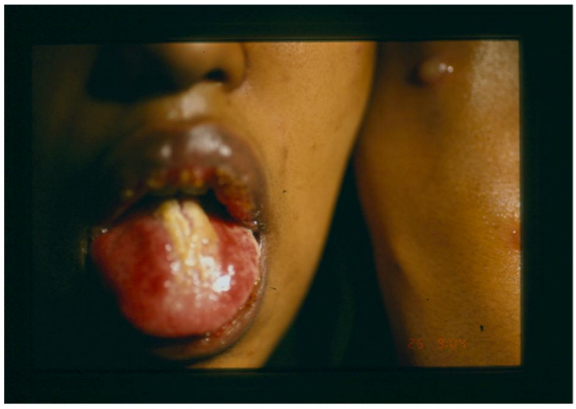
Pemphigus vulgaris (PV) is a rare immunobullous dermatosis with worldwide distribution. The core manifestation of the condition is mucosal erosions and easily ruptured bullae that emerge on an apparently normal skin and mucous membranes or on an erythematous base. The etiology of PV is still unknown, although the disease has attracted considerable interest. It is perhaps the most formidable dermatologic emergency which requires prompt treatment without which it may prove to be fatal. Though, new treatment modalities have decreased the mortality, nevertheless complications of the treatment are the main hazards presented by various clinical manifestations, and among them fever represents one of the most important presentations.

CASE REPORT :

A 21 years old arterian female patient presented with sever intra and extra oral ulcerative lesions involving oral mucosa, and lips(Figure1), the lesion started to appear 2months ago, and where getting worse. She gave a history of weight loss of about 10kg in last 2months due to painful oral ulceration and dysphagia. Patient had cutaneous lesions, a flaccid blister, on erythematous skin (Figure2). The affected skin was painful. She gave a history of receiving different courses of antibiotic, antiviral and antiphungal topical and systemic, medications were prescribed by dentist and none try or advise patient for taking a biopsy.



(Figure1) mouth erosions and ulcers



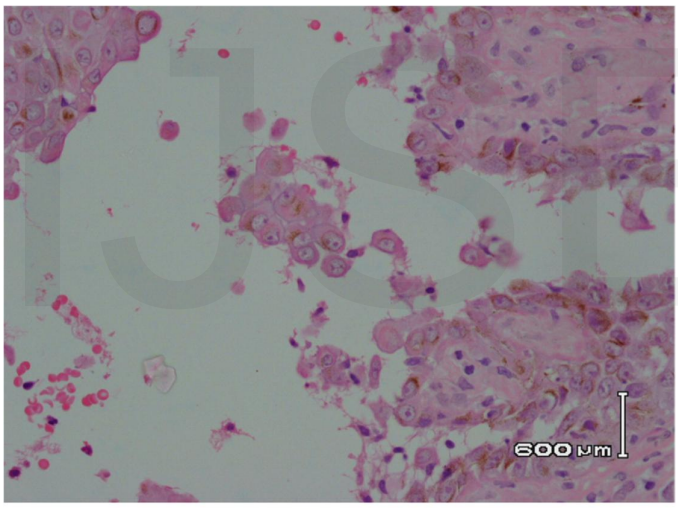
(Figure2) Intact and ruptured blisters on the skin of the hand

Differential diagnosis. According to the clinical presentation of the patient were: Pemphius vulgaris, Mucous membrane pemphigoid. A biopsy was taken to confirm the diagnosis.

Histopathological result. Showed suprabasal splitting of the epithelium and loss of intracellular attachments leading to intraepithelial vesicles formation with rounded acantholytic or Tzanck cells floating freely in the vesicular fluid(Figure 3,4,5).

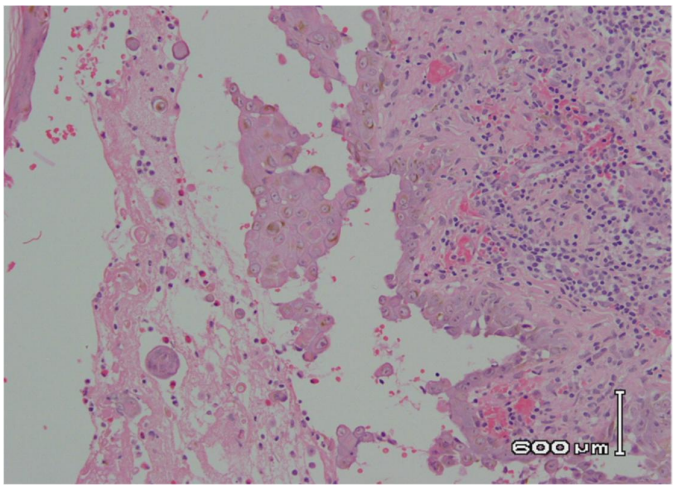
suprabasal splintting of the epithelium and loss of intracellular attachements

(Figure3)



loss of cell-cell adhesion, a process termed **acantholysis.**

(Figure 4)



(Figure 5)

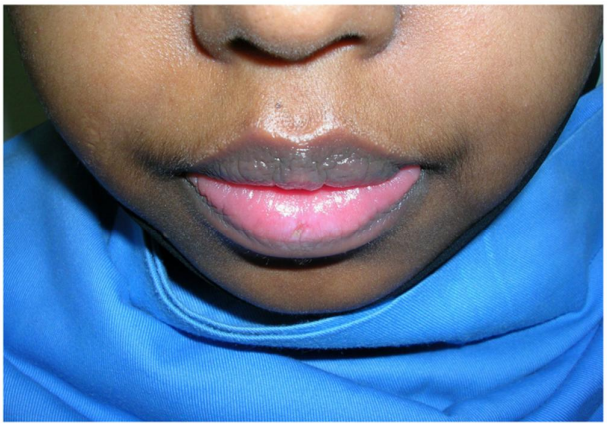
Loss of cell-cell adhesion, a process termed acantholysis and formation of rounded acantholytic or Tzanck cells floating freely in the vesicular fluid.

Histopathlogical result. Confirmed that the pemphigus vulagaris is the final diagnosis.

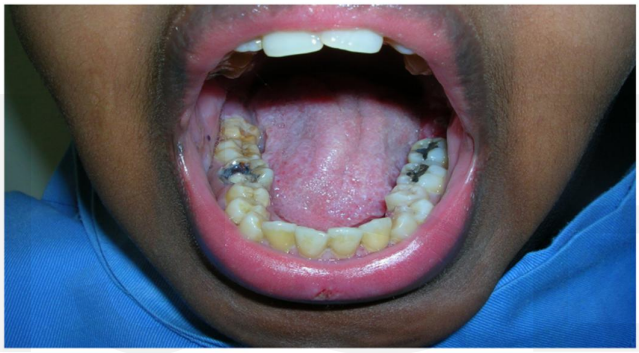
Management of this case was by using systemic and topical prednisone.

Follow up appointments :

Patient at follow-up appt. Healing of mouth and skin ulcerations



Patient at follow-up appt. Healing of lips ulceration



Patient at follow-up appt. Healing of intraoral and lips ulcerations

DISSCUSION :

Pemphigus vulgaris is a rare chronic autoimmune , intraepithelial, blistering disease affecting the skin and mucous membranes. It is mediated by circulating auto antibodies directed against keratinocyte cell surfaces. Blisters in PV are associated with the binding IGg autoantibodies to keratinocyte cell surface molecules. These intercellular or PV antibodies bind to keratinocyte desomsomes and to desmosome free areas of the keratinocyte cell membrane. The binding of autoantibodies results in a loss of cell-cell adhesion.[2]

Clinical Features. Pemphigus vulgaris affected all races, with an equal gender distribution among males and females. The mean age of onset is approximately 50-60years[2].

Oral manifestations. PV *involves oral mucosa in 50-70% of patients*. This may *limit oral intake secondary* to *dysphagia.* Almost all patients have mucosal lesions. Mucosal lesions may be the sole sign for an average of 5 months before skin lesions develop, or they may be the sole manifestation of the disease. Patients with mucosal lesions may present to dentists, oral surgeons, or gynecologists. Blistering and erosions secondary to the rupture of blisters may be *painful* and limit the patient's daily activities. Patients with PV typically heal without scarring unless the disease is complicated by severe secondary infection. *Nikolsky's sign*: In patients with active blistering, firm sliding pressure with a finger separates normal-appearing epidermis, producing erosion. This sign is not specific for PV and is found in other active blistering diseases. *Asboe-Hansen sign*: Lateral pressure on the edge of a blister may spread the blister into clinically unaffected skin.

Histologic Features. Pemphigus vulgaris appears as intraepithelial clefting with keratinocyte acantholysis (Figure 5). Loss of desmosomal attachments and retraction of tonofilaments result in free floating, or acantholytic, *Tzanck cells.* Bullae are suprabasal, and the basal layer remains attached to the basement membrane[1].

Frequency. PV is uncommon, and has been reported to occur worldwide. PV incidence varies from 0.5-3.2 cases per 100,000. PV incidence is increased in patients of Ashkenazi Jewish descent and those of Mediterranean origin.

The course of PV is variable one the disease terminating in death or recovery within a few days or weeks, or being prolonged over a period of months or even years.

Mortality/Morbidity. PV is a potentially life-threatening autoimmune mucocutaneous disease with a mortality rate of approximately 5-15%.

Complications. secondary to the use of high-dose corticosteroids contribute to the mortality rate. Morbidity and mortality are related to the extent of 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.

Management. Pemphigus vulgaris is generally managed with topical, oral and intralesional corticosteroids[3]. The current therapeutic regimen of pemphigus vulgaris is largely based on systemic immunosuppressant such as systemic corticosteroids along with other adjutants like methotrexate, cyclophosphamide, mycophenolate mofetil and intravenous immunoglobulin[4]. Here the case was managed using by using topical and systemic prednisone.

CONCLUSION :

A good history is very important for making a diagnosis. Examination and investigations may help to confirm or refute the diagnosis made from the history.

The history will also tell you about the illness as well as the disease. The illness is the subjective component and describes the patient's experience of the disease.

Try to follow the sequence history, examination, investigation when you see a patient. A common mistake is to rush into investigations before considering the history or examination.

Always remember to treat the patient and not the investigation. And remember that although we talk about "the patient," you should consider "the person".

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