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ABSTRACT

physiologically like acute severe burn.



Case Study on Pemphigus Vulgaris



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1. Introduction

The word Pemphigus derived from the Greek word pemphix (bubble or blister) [1] and vulgaris derived from the Latin word (common) [2]. Pemphigus is a life-threatening condition that causes erosions and blisters of the skin and mucous membranes [3]. Pemphigus vulgaris is a chronic, limited, intraepidermal bullous disease with a severe life-threatening outcome which was originally named by Wickman in 1971. Pemphigus is infrequent disease incidence ranging from rate ranging from 0.2 to 3.3 in 1,00,000 per year [4]. Men and women are equally affected with the most occurring age of 50-60 years. It is a chronic mucocutaneous disease usually occurred in the oral cavity, which later may spread to skin and other mucous membranes. Many of dentists need to recognize these oral manifestations and treat, refer appropriately because it is a fatal disease [5].

Pemphigus vulgaris mainly characterised by the occurrence of flaccid, ruptured intra epithelial bullae on apparently normal skin and mucus membranes. The oral cavity is mainly affected intraoral lesions appears in 50% of the patients without any involvement of skin. But any part of the oral mucosa and areas exposed to mechanical irritation are commonly affected. The lesions prone to manifest most commonly on buccal and palatal mucosa and on the gingiva. The lesions initially manifest as a bleb like blisters or as diffuse gelatinous plaques. Lesions are mostly painful if untreated pemphigus vulgaris can be fatal. Due to this early recognition and diagnosis of the disease is most prognostic importance [6].

2. Case Study

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A male patient of age 53 years was admitted in dermatology ward with the chief complaints of multiple fluid filled lesions for 3 months. Present complaints started 3 months back initially over the limbs which break opens to form lesions. History of present illness, burning over the lesion and itching. He had taken the treatment outside hospital. No history of similar complaints in the past. Past history of hypertension since one year, not a known diabetic.

Pemphigus vulgaris is a limited chronic blistering skin disease. It is a type II hypersensitivity reaction,

with the formation of antibodies against desmosomes which are the components of the skin that

function to keep certain layers of skin bound to each other. When desmosomes attacked, the layer of

skin will be separated and the clinical picture resembles as a blister. As time progress the condition

worsened without the treatment lesions increase in size and scattered throughout the body, behaving

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On dermatological examination multiple flaccid bullae of six 1×1 cm and 5×3 cm is present over the upper chest, upper extremities and lower extremities. Multiple erosions with crusting and post inflammatory hyperpigmentation is seen at chest, upper and lower extremities. On CVS examination S1 and S2 sounds are present. On the pathology examination of fluid from bullae for Tzanck smear cytology method the smear is positive for Tzanck cells (multiple typical acantholytic cells). Based on the above physical and pathological examination the patient is suffered with Pemphigus Vulgaris. The patient was treated with the following medications. Inj. Decadron 4 mg OD, Tab. Taxim 200 mg BD, Tab. Rantac 150 mg OD, Tab. Atarax 10 mg SOS, Tab. Fourtz BOD, Tab. Shelcal OD, Tab. Meganeuron plus OD, Fusigen cream BD, Tab. Telvas 20 mg OD. The treatment was tolerated well the bullae, lesions over the patient body resolved with evidence post inflammatory hyperpigmentation. The patient was discharged from hospital after 8 days and advised to follow up after one month.

3. Discussion

In Pemphigus Vulgaris, lesions initially comprise small asymptomatic blisters, although these lesions are very thin walled, they easily rupture giving rise to painful, erythematous and haemorrhagic erosions. In most cases the first signs of disease appear on the oral mucosa.

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Figure: Lesions in Pemphigus Vulgaris

The ulcerations can affect other membranes including the conjunctiva, nasal mucosa as well as skin where blisters are commonly visible [8]. Most of the skin is involved in the above case. The aetiology of this disease still idiopathic. These class of diseases are commonly characterised by the production of antibodies against the intercellular substances, hence they classified as autoimmune diseases [7]. Other causative factors include food (spinach, garlic), infections, unknown insect bites, neoplasms and some medications like captopril, penicillin's and fifampicin [9]. In Pemphigus Vulgaris immune system produces auto antibodies against desmosomes especially desmoglein 3 (Dsg3). Another desmosome component is desmoglein 1. The initial target which is affected is subcutaneous sites only. Dsg3 expressed in oral mucosa and Dsg1 expressed in skin [10]. The dermal and mucosal changes involve the loss of coherence among the layers of keratinocytes. This is identified in early stages of disease; the primary lesion is a thin-walled bullae containing clear in different sizes. Under pressure it releases contents into surrounding epidermis and further increase in their size. Healing is very slow but no scar will remain in this disease. In the oral mucosa, lesions filled with fluid are occurred without any inflammation. Whenever the epithelial wall of bullae ruptures it becomes painful [11].

In this case, we assessed multiple drugs for this Pemphigus Vulgaris and administering of new drug therapy to treat Pemphigus Vulgaris with more antibiotics. We administered the steroids as immunosuppressive agents such as inj. Decadron 4 mg because this disease autoimmune in origin and antibiotics like Tab. Taxim 200 mg were also prescribed to prevent occurrence of infections from lesions.

4. Conclusion

Pemphigus vulgaris is a rare cause of chronic ulceration of oral mucosa and skin. Although Pemphigus Vulgaris is a rare and fatal disease, today it can be successfully treated with the combination of immunosuppressive agents and effective adjuvants. Newer diagnostic tests and better monitoring of the disease can be achieved now with role of anti-Dsg antibody-keratinocyte binding in blister formation. In future antigen specific immunotherapy may be an alternative therapy to current conventional treatment methods. Early diagnosis and management of pemphigus vulgaris leads to a better prognosis, lower mortality and good quality of life.

5. Acknowledgement

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6. Conflict Of Interest

The authors declared no conflicts of interests with respect to authorship and publication of this case.

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