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A CASE STUDY ON PEMPHIGUS VULGARIS

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ABSTRACT

Pemphigus vulgaris is a rare autoimmune disease that causes painful blistering on the skin and mucous membranes. Pemphigus vulgaris is the most common type of a group of autoimmune disorders called pemphigus. Pemphigus vulgaris occurs when the immune system mistakenly makes antibodies against proteins in healthy skin and mucous membranes. This disease usually starts with blisters in the mouth and then on the skin. The blisters sometimes affect the membranes of the genitals.

INTRODUCTION

Pemphigus vulgaris is a rare chronic blistering skin disease and the most common form of pemphigus[1]. Pemphigus is derived from the Greek word pemphix meaning bubble or blister[1]. Pemphigus describes a group of chronic bullous diseases, originally named by Wichman in 1791[2]. It is classified as a type II hypersensitivity reaction in antibodies are formed against desmosomes[2]. As desmosomes are attacked, the layers of skin separate and the clinical picture resemble a blister[2]. These blisters are due to acantholysis, or breaking apart of intercellular connections through an autoantibody-mediated response[3]. Pemphigus vulgaris accounts for approximately 70% of pemphigus cases[3].Pemphigus vulgaris isn't contagious and cannot be transmitted from one person to another[3]. It also doesn't appear to be transmitted from parent to child[4]. It is mediated by circulating autoantibodies directed against keratinocyte cell surfaces[4].

A potentially life-threatening disease, it has a mortality rate of approximately 5-15%[5]. The primary lesion of pemphigus vulgaris is a flaccid blister filled with clear fluid that arises on healthy skin or on an erythematous base[5]. Pemphigus vulgaris incidence varies from 0.5-3.2 cases per 100,000 population[5]. Pemphigus vulgaris incidence is increased in patients of Ashkenazi Jewish descent and those of Mediterranean origin[6]. The term pemphigus refers to a group of autoimmune blistering diseases of the skin and mucous membranes characterized histologically by intraepidermal blister and immunopathologically by the finding of in vivo bound and circulating immunoglobulin G (IgG) antibody directed against the cell surface of keratinocytes[5].These intercellular or pemphigus vulgaris antibodies bind to keratinocyte desmosomes and to desmosome-free areas of the keratinocyte cell membrane[7]. The binding of autoantibodies results in a loss

of cell-to-cell adhesion, a process termed acantholysis[7].Over time the condition inevitably progresses without treatment: lesions increase in size and distribution throughout the body, behaving physiologically like a severe burn[7]. The immune system produces proteins called antibodies[8]. Antibodies normally attack harmful foreign substances like bacteria and viruses. Pemphigus vulgaris occurs when the immune system mistakenly makes antibodies against proteins in healthy skin and mucous membranes[8]. The antibodies break down the bonds between the cells, and fluid collects between the layers of the skin[8]. This leads to blisters and erosions on the skin[8]. The antibody alone is capable of causing blistering without complement or inflammatory cells[9]. Pemphigus vulgaris is a potentially life-threatening autoimmune mucocutaneous disease with a mortality rate of approximately 5-15%[9]. Mortality in patients with pemphigus vulgaris is 3 times higher than the general population. Complications secondary to the use of high-dose corticosteroids contribute to the mortality rate[10].

CASE REPORT

A 65 year old male patient was admitted under general medicine for the reason of generalised tiredness and fever. The patient has history of Type II DM, lef leg cellulitis, BP, Pemphigus vulgaris. Patient also had history of admission in hospital for the reason that bystander administered Inj.Rituximab for the treatment of Pemphigus vulgaris in sitting position and experienced depression like illness after that incident. This patient had ulceration over tongue, buccal mucosa and over right iliac crest.The patient was on T.Wysolone 30 mg for past 3 months.

CVS:

Normal S1S2, respiratory system was clear, drowsy, sick looking. Had ulceration over tongue, buccal mucosa.

DIAGNOSIS:

Temp-96.6 F, Pulse-112 bts/min, BP-90/60 mmHg, Hb-6.7, WBC-5900, ESR-130, PCV-20.8, RBS-203, Hba1c-6.87, urea-129, SGOT-8, SGPT-7, Albumin-1.5, Sodium-144,Potassium-3.2, Calcium-9.2.

Based on subjective and objective evidence case was diagnosed as Septic shock, Pemphigus vulgaris.

In this case the patient is treated with following medication:

The patient was treated with Inj.Tigecycline (Tigecycline) Tetracycline antibiotics 50mg BD, T.Forcan (Fluconazole) anti fungal 50 mg OD, C.Becosules (B-Complex) vitamin supplement 1-0-0,T.Celin 500 mg (Vitamin-C) vitamin supplement 1-0-0, T.Wysolone (Prednisolone) corticosteroid 20mg 1-0-0, Megaheal ointment (Amorphous hydrogel) for wound dressing,T.Benalgis(Benfotiamine) dietary supplement of Thiamine 1-0-0, Inj.Meropenem (Meropenem) broad spectrum antibiotic 1g Q8h, Syp.Potchlor(Potassium chloride) potassium supplement 5ml TDS, T.Pantop (Pantoprazole) gastric irritation, Candid mouth paint for LA (Clotrimazole) antifungal.

DISCUSSIONS

Pemphigus is characterized by a thin-walled bulla that forms on seemingly normal skin or mucosa, breaks immediately, and spreads peripherally, eventually leaving vast denuded areas. The oral mucosa is the site of the disease's initial manifestation in the vast majority of cases (70–90%). In areas that are prone to frictional damage, such as the cheek, pharynx, larynx, esophagus, genital mucosa, and skin, where intact blisters are common, the sores can appear anywhere in the mouth. The more common variants of pemphigus are Pemphigus Vulgaris, Pemphigus Vegetans, Pemphigus Foliaceous, Pemphigus Erythematosus, Paraneoplastic Pemphigus [PNP] and Drug-related Pemphigus. Our case represents Pemphigus vulgaris. An antibody directed against different target cell surface antigens has caused a lesion to form in different layers of the epithelium in each form of this disease. Pemphigus Vulgaris is the most common of pemphigus and it accounts for over 80% of cases. The principal dermal and mucosal changes involve the loss of coherence among layers of keratinocytes. This is manifested, in the early stages of diseases; the primary lesion is ath in walled bulla, several centimeters in size, containing clear fluid. Under pressure it releases its

contents through the surrounding epidermis and further increases in size. Healing is very slow, but no scars will remain. On the oral mucosa, bullae filled with fluid are also present but no inflammation develops. When the epithelial wall of the bulla ruptures it will become painful lesions.

CONCLUSION

Pemphigus vulgaris is a rare cause of chronic ulceration of mucosa. The severity and natural history of pemphigus vulgaris are variable, but before the advent of steroids, most patients with pemphigus vulgaris died. Treatment with systemic steroids has reduced the mortality rate. Most deaths occur during the first few years of disease, and, if the patient survives 5 years, the prognosis is good. Early disease probably is easier to control than widespread disease, and mortality rates may be higher if therapy is delayed.

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