Case Report

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Phemphigus vulgaris an autoimmune acantholysis: a case report

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ABSTRACT

Pemphigus vulgaris is a rare chronic autoimmune skin disease characterized by a flaccid blister filled with clear fluid that arises on the skin or an erythematous base. The keratinocytes separated by the desmoglein present on the desmosomes which clinically manifests as fluid-filled blisters on the skin. We present pemphigus vulgaris with an unknown cause but good recovery after the management with monoclonal antibodies and corticosteroids in a 30-year-old male patient who came to the hospital with complaints of blisters on the epithelial layers of skin on the face and oral mucosa.

Keywords: Pemphigus vulgaris, Autoimmune, Blisters, Desmoglein desmosome

INTRODUCTION

The term "pemphigus" derived from the Greek word "pemphix" means bubble or blister and the term "vulgaris" derived from Latin word means common. This term "Pemphigus vulgaris" named by Wickman in 1791.

It is a rare chronic autoimmune intraepithelial bullous disease that usually affects the skin and mucous membrane. It is a rare disease an incident rate ranging from 0.5 to 3.2 per 1,00,000 per year. It mostly occurs almost in middle-aged or older people. It is a chronic mucocutaneous disease that usually starts first in the oral cavity then spread to the skin or mucous membrane. The primary lesion is a soft flaccid blister filled with clear fluid that arises on the skin. The major variants of pemphigus are pemphigus vulgaris, pemphigus vegetans, pemphigus foliaceous, pemphigus erythematosus, Paraneoplastic pemphigus (PNP) and drug-related pemphigus. Pemphigus vulgaris is the most common form of pemphigus, accounting for over 80% of cases. Lesions

may occur anywhere on the oral mucosa, but the buccal mucosa is the most commonly affected site followed by involvement of the palatal, lingual and labial mucosa. The lesions are painful. This case report describes the case of a patient complaining of oral lesions and skin lesions, who was diagnosed as Pemphigus vulgaris.

CASE REPORT

A 30-year-old male patient came to the hospital with the chief complaints of ulcer in mouth and tongue and had difficulty in swallowing solid food for the past 2 weeks and fluid-filled lesions over the right cheek. He also mentioned that the lesions over the cheek and in mouth (hard palate, on the uvula and lips) shown in figure 1,2,3 happened to bled frequently and was painful. History of present illness is itching, burning sensation present over the lesions. The review of medical and family history was noncontributory. The patient did not have any social habits. Skin biopsy shows pemphigus vulgaris. Oral mucosa was very feeble and peeling off with gentle scratching. The patient was then admitted to the ward for rituximab

infusion. The vital signs of the patient were normal, and he was conscious, oriented. All baseline and relevant investigations were done (haematological, renal, and liver function test). The patient was then shifted to Medical intensive care unit (MICU) for rituximab infusion. Premedications are given injection dexamethasone 8 mg, injection piriton 10 mg, injection emeset 8 mg, injection para 1 gm. Injection rituximab 1000 mg Intravenous (IV) in 500 ml Normal saline (NS) (total infusion time 5-6 hours) was given and the patient was then shifted to the ward. Discharge drugs after rituximab infusion are tablet prednisolone 30 mg once a day (OD), tablet pan 40 mg OD Before breakfast (B/F), tablet calcium 500 mg OD, kenocort oral gel twice daily (BD), zytee gel three times daily (TDS), tablet bact ointment BD, tablet cotrimoxazole 0-1-0 for 2 weeks.



Figure 1: Tongue lesions.



Figure 2: Buccal lesions.



Figure 3: Skin lesions on right cheek.

DISCUSSION

Pemphigus is defined as a group of life-threatening skin and mucous membrane disorders which is characterized by blisters of the skin due to acantholysis (loss of keratinocyte to keratocyte adhesion). This acantholysis is triggered by autoantibodies to intercellular adhesion molecules and it is induced by certain food (garlic), infections, neoplasms, and some drugs like captopril, penicillin, and rifampicin.^{5,6}

In pemphigus vulgaris, lesions were small asymptomatic blisters, which are very thin-walled and easily ruptured which is very painful and form haemorrhagic lesions. 70-90% of cases the first signs of disease appear on the oral mucosa. They are most commonly found in areas are check, mucosa, tongue, palate, and lower lip.

The mechanism involved is binding of Immunoglobulin G (IgG) autoantibodies to desmoglein 3 which is a transmembrane glycoprotein adhesion molecule attached to the desmosome. Some theory shows evidence that autoantibodies which directly block the adhesion of desmoglein. ^{7,8} A process of separation of cells takes place in lower layers of stratum spinosum called acantholysis and leads to the formation of the suprabasal bulla, this bulla increases its size in the epithelium containing fluids results in loss of large area of mucosa and skin. ⁹ 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. ¹⁰

CONCLUSION

The goal of the treatment is to reduce inflammation and autoantibody production. The most commonly used medications are corticosteroids (non-target specific treatment), later specific treatment targets have been validated. Rituximab was approved as first-line therapy for severe pemphigus vulgaris by Food drug administration (FDA) in June 2018. It is used along with short-term corticosteroids. It has shown better therapeutic outcomes. Untreated, pemphigus vulgaris is often fatal because of the susceptibility to infection and fluid and electrolyte disturbances. If the disease is early diagnosed, it is easy to prevent the progression of the disease but if it is delayed leads to an increased incidence of mortality.

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