Case Report on Pemphigus Vulgaris

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Abstract: Pemphigus vulgaris (PV) is a chronic inflammatory autoimmune bullous disease. PV almost always affects the mouth and it can be the initial site of presentation in 50% of cases, before skin and other mucosal sites become involved. This is a case of 60-year-old female came with the chief complaints of pain and erosion in the mouth since 1 week. On intra-oral examination solitary, erythematous, pseudomembranous plaque present over the right buccal mucosa. As the patient was diagnosed with pemphigus vulgaris, the goal of therapy was to reduce inflammatory response & autoantibody production. The patient was treated with Methylprednisolone, antifungal & antibiotic along with local anesthetic gel for symptomatic relief. PV is a serious disease, and if left untreated, it could lead to patient's death.

Keywords: pemphigus vulgaris (PV), autoimmune, blistering, oral lesions, mucous membrane

1. Introduction

Pemphigus vulgaris is an autoimmune, intraepithelial, blistering disease affecting the skin and mucous membranes. Pemphigus Vulgaris is the most common form of pemphigus. It is mediated by circulating autoantibodies directed against keratinocyte cell surfaces. A potentially life-threatening disease, it has a mortality rate of approximately 5-15%. The primary lesion of pemphigus vulgaris is a flaccid blister filled with clear fluid that arises on healthy skin or on an erythematous base. 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. In many patients, oral lesions are followed by the development of skin lesions.

2. Case report

A 60-year-old female came with the chief complaints of pain and erosion in the mouth since 1 week. History revealed that the patient first noticed erythematous raised lesions which is gradually enlarged and become eroded following after which she started with pain and burning sensation over the eroded area mainly at the right side of Buccal mucosa. Patient had a past history of similar complaint 5 years back and on medication for the same. No significant family history was found. The patient had a history of tobacco addiction positive. On intra-oral examination solitary, erythematous, pseudomembranous plaque present over the right buccal mucosa. A few pus cells and gram-positive cocci (Staphylococcus aureus) in clusters was seen in the silt skin gram stain. The patient was diagnosed to have PV and was prescribed with Methylprednisolone (16mg OD per oral), Tolpa-D (15mg bidper oral) along with local anesthetic gel application for symptomatic relief. She was also given an antifungal (fluconazole 50 mg Bid per oral), Cefuroxime (500 mg bid per oral), Hydroxyzine (25mg OD per oral) Levocetrizine (5mg OD per oral) and Candid mouth paint.

3. Outcome & Follow Up

Goal of treating Pemphigus Vulgaris focus on healing the blisters and prevent the relapse of symptoms. A strict oral hygiene of the patient was maintained and monitored. A gradual reduction of symptoms with healing of lesions, erythema and inflammation in relation to ulcers was observed.

4. Discussion

Pemphigus vulgaris is an autoimmune disorder that causes severe blistering of the skin and of the mucous membranes lining the mouth, nose, throat and genitals. Autoimmune process and immunosuppressive therapy of pemphigus would predispose the patients to infections. S. aureus infection in patients with pemphigus was high compared to other bacteria. The bacteria could simply be an opportunistic infection, because pemphigus patients are treated with immunosuppressive therapy for a long time. Early recognition of concurrent pemphigus and bacterial infection, especially S. aureus, is extremely important because of the possible fatal consequences of the disease. Secondary infections are a common complication of pemphigus vulgaris.

5. Conclusion

Pemphigus vulgaris is a rare chronic autoimmune disorder affecting cutaneous-mucosa that is often misdiagnosed and late, even then oral lesions appear. If not treated promptly, the disease has a high morbidity rate, with a fatal rate of 5% to 10% in most of cases. The diagnosis is confirmed through physical and pathological examination. The primary aim of treatment is to decrease blister formation, prevent infections and promote healing of blisters and erosions. The therapeutic regimen, along with corticosteroid therapy and adjuvant treatments, helps patient to relive from symptoms. PV is a serious disease, and if left untreated, it could lead to patient's death.

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