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Case Report

PEMPHIGUS VULGARIS: A CASE REPORT

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ABSTRACT

Pemphigus Vulgaris is a chronic mucocutaneous disease which usually manifests first in the oral cavity, which later may spread to skin or other mucous membrane. This is a case of 40yr patient who presented with complaints of hyperpigmented lesions over the upper limbs, lower limbs, trunk, back and abdomen and hyperpigmented crusted plaque over the scalp since 7 months. For diagnosis physical examination and immunoflouresense tests were performed. The treatment given was Tab. Methyl prednisolone (15mg-OD), Tab.Methotrexate(12mg/week), Tab.Fluconazole (150mg at night), LIVOGEN (ferrous fumarate-152mg+folic acid-1500mcg). Early the diagnosis and treatment less is the suffering to the patient.

INTRODUCTION

Pemphigus comprises a group of life threatening autoimmune mucocutaneous disease which presents at clinical examination as blisters, erosions, ulcerations of the skin and mucous membranes in which disease severity is based upon its progressive course accompanied by increased body catabolism and secondary bacterial and viral infections which may lead to sepsis and cardiac failure[1-2].“Pemphix‟ in greek means “bubbles or blisters” and vulgaris in latin means “common”. Although pemphigus is a bizarre disease, pemphigus vulgaris is the most common of all [3]. It is characterized by the presence of an autoantibody that is directed against the 130-kDa antigen in the intercellular cement substance (ICS) now characterized as desmoglein3. The binding of autoantibody to the epidermal keratinocytes causes a loss of intercellular adhesions resulting in a process called acantholysis [4]. The incidence rate of pemphigus vulgaris per 100million people is about 0.5 to 3.2 among which men and women are equally affected with the disease. It is commonly seen in the age group between 40-60 years [5]. Clinical lesions of pemphigus vulgaris consists of flaccid blisters on either normally or erythematous skin. These lesions are small and asymptomatic at first. They are thin walled and easily get ruptured giving rise to painful and hemorrhagic erosions. Any type of manual pressure to the skin of these patients may elicit the separation of the epidermis (Nikolsky’s sign) [6-7]. In addition to a full history and examination, biopsy and appropriate histopathological and immunological investigations of perilesional tissue are frequently indicated for the diagnosis [8]. Treatment of pemphigus vulgaris aims at acquiring disease remission which is possible through intensive therapy which is followed by maintenance therapy to stabilize the disease with administration of gradually decreasing doses of medication. For both intensive and maintenance therapy corticosteroids like prednisolone are used. Adjuvant drugs are the agents that support the effect of steroids, among which some act as “steroid -sparing agents”. Most commonly used Adjuvant drugs are Azathioprine, Cyclophosphamide, Methotrexate [9].

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Case Report

A 40yr old male patient was admitted into the hospital with the chief complaints of hyperpigmented lesions over the upper limbs, lower limbs, trunk, back and abdomen and hyperpigmented crusted plaque over the scalp since 7 month .Pain and burning sensation over mouth. Patient complained of difficulty in eating and swallowing. History of recurrence was present. On clinical examination, multiple and crusted ulcerations and bullae were seen on right buccal mucosa, lower lip on the mucosal side, ventral surface & lateral border of the tongue, hips and abdomen since 15 days which were associated with itching. Bullae are gradually increasing in number and size. Patient is also having oral candidiasis since 1 month.

Past Medical and Medication History: Patient has previous history of Diabetes mellitus since 5 years and oral candidiasis since 1 month. Patient was regularly taking ayurvedic medications for anemia.

Cutaneous Examination: Hyperpigmented, wealed lesions in the mouth, over the upper limbs, trunk, back, abdomen and crusted plaque over the scalp.

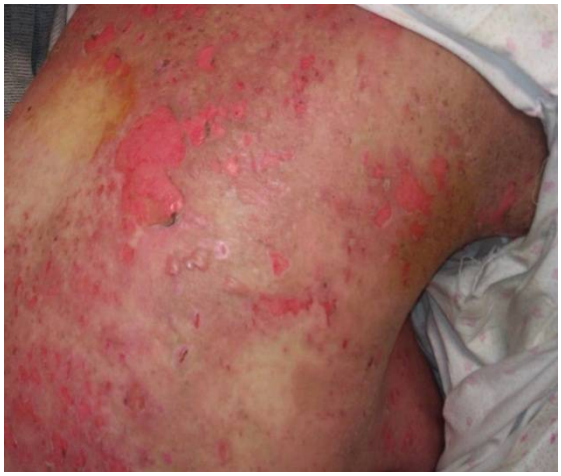

Fig 1 Lesions on back of the body

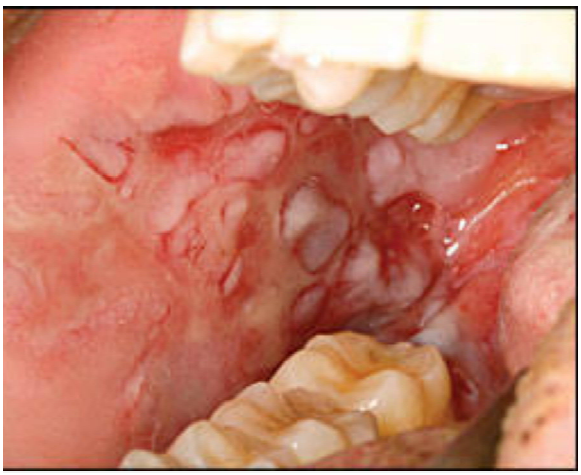

Fig 2 Lesions in the mouth

Laboratory Investigations: Apart from the above mentioned physical examinations the patient underwent routine test as well as definitive tests.

Routine Tests: Complete blood picture revealed decrease Hb levels- 8gm/dl as well as decrease RBC count -2.7 mill/cumm, a total WBC count of 7,200/cumm with a differential count of 40% lymphocytes, and Neutrophil 52%. The Erythrocyte sedimentation Rate(ESR) was 15mm at the end of 1 hour. Electrolytes revealed normal levels of sodium and calcium levels of 145mEq/L and 1.00mmol/L respectively. Patient was perfomed liver function test and was found to have normal levels- Total bilrubin , Direct Bilrubin, SGPT, Albumin level.

Definitive Tests: Direct Immunofluoresence was done to the patient to confirm pemphigus vulgaris. Direct Immunofluorescence revealed deposition of IgG antibodies.

Treatment: After performing cutaneous examination and definitive tests the patient was diagnosed with phempigus vulgaris and the treatment included Tab.Methyl prednisolone (15mg-OD) as a first-line treatment for phempigus vulgaris. As a second-line drug Tab.Methotrexate(12mg/week) was given, Antifungal Tab.Fluconazole(150mg at night) was given to treat oral candidiasis whereas candid mouth paint was given for symptomatic relief of candidiasis. Patient was being prescribed with LIVOGEN (ferrous fumarate-152mg+folic acid-1500mcg) to treat anemia. Patient was advised to maintain good oral hygiene and to avoid consumption of foods that can cause irritation to the skin**.**

Discharge Medications: The patient was discharged after 15 days when the lesions were seen to be resolving. The patient was advised to continue Methyl Prednisolone (10mg-OD) and Fluconazole (100mg at night), LIVOGEN (Ferrous fumarate 152mg+ Folic Acid 1500mcg) for 15 more days.

DISCUSSION

Pemphigus comprises of four related diseases with autoimmune etiopathogeneses: Pemphigus vulgaris, Pemphigus vegetans, Pemphigus erythematosus, and Pemphigus foliaceus. Among the four types pemphigus vulgaris is the most prevalent type [10]. Pemphigus Vulgaris is a chronic mucocutaneous disease which usually manifests first in the oral cavity, which later may spread to skin or other mucous membrane [2]. The pathophysiology responsible for PempigusVulgaris has been attributed to autoantibodies to DSG3 which is a member of the desmoglein family of transmembrane glycoproteins that are components of the desmosomes, which are the integral structures that mediate cell-to-cell adhesion [11].

Our patient presented with multiple and crusted ulcerations and bullae on right buccal mucosa, lower lip on the mucosal side, ventral surface & lateral border of the tongue, hips back and abdomen. As reported in literature our patient too presented with the two most common symptoms related to PV, that is, pain and burning sensation. For confirmation of diagnosis Direct Immunofluorescence was done which revealed deposition of IgG antibodies.

PV is generally treated with oral, and topical corticosteroids [12]. The present treatment regime in PV is based on systemic immunosuppressant like corticosteroids along with adjuvants like methotrexate, cyclophosphamide [13]. Our patient was treated with Tab.Methyl prednisolone (15mg-OD), Tab.Methotrexate(12mg/week), Antifungal Tab.Fluconazole(150mg at night) and LIVOGEN (ferrous fumarate-152mg+folic acid-1500mcg). After 15 days of treatment with these agents patient was discharges hen the lesion were seen to be resolving. However long term follow up is needed to identify the possible remission of the disease. Various authors have reported that the oral lesions can disappear after 2 months to one year [14,15], although it remains unclear whether the PV completely remits [16], and there are no well-defined criteria for the cure/remission of this disease [17].

CONCLUSION

Pemphigus comprises a group of life threatening autoimmune mucocutaneous disease which presents at clinical examination as blisters, erosions, ulcerations of the skin and mucous membranes. Detailed clinical and immunohistochemical tests are necessary to confirm the diagnosis. The early diagnosis and treatment less is the suffering of the patient.

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