



Pemphigus Vulgaris – A Case Study

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Abstract

Pemphigus is a rare autoimmune disease that causes blisters in the skin and oral cavity. Pemphigus vulgaris is one of the most common types of Pemphigus that affects the mucous membrane, which is found in the areas including mouth, eyes, throat, nose, and lungs. Eighty to ninety percent of patients with PV (Pemphigus Vulgaris) start with oral lesions. A 25-year-old female patient came up with a complaint of lesions over oral cavity which spread all over the body followed by the difficulty to open the mouth, increased red skin lesions, loss of appetite, loss of weight (10 kg in few months). Due to early diagnosis by the physician and the dentist, the treatment was started within a short period of time with steroids, immunosuppressant, antifungal, and antibiotics to control the disease.

Keywords

Pemphigus vulgaris, Autoimmune disease, Oral Cavity, Nikolsky's sign.

INTRODUCTION

Pemphigus is a life-threatening autoimmune disease which causes blisters and erosions of the skin and the mucous membrane ⁽¹⁾. Pemphigus has been classified into different types - Pemphigus vulgaris, Pemphigus foliaceus, Pemphigus vegetans, Pemphigus erythematosus, Paraneoplastic pemphigus and IgA pemphigus ⁽²⁾. Each type of Pemphigus is described based on the area where the blisters are formed. Pemphigus vulgaris is an uncommon immune system illness that causes tormenting rankling on the skin and the mucous layers ⁽¹⁾. Pemphigus vulgaris influences the mucous layers, which are found in the mouth, throat, nose, eyes, privates, and lungs ⁽³⁾. This illness generally begins with rankles in the mouth after which it starts to spread all over the skin. These blisters at times influence the layers of the genital areas ⁽⁴⁾. The diagnosis of PV is done through Tzack

smear and nikolsky's sign is positive ⁽⁵⁾. Treatment for Pemphigus vulgaris is fundamental, and generally involves in the utilization of corticosteroids to smoothen the condition of the patient. The main aim of treating the patient is to reduce the blisters and erosions on the mouth as well as in the oral cavity ⁽⁶⁾. The standard treatment for Pemphigus vulgaris is corticosteroids although it is very effective it has a substantial adverse effect ⁽⁷⁾. Once the symptoms start to reduce slowly then they start with immunosuppressant with a low dose of steroids.

CASE DISCUSSION

A 25-year-old female patient was admitted in the dermatology department with a complaint of lesions over oral cavity for 2 months, lesions and blisters (fluid filled) extended all over the body since 4 days and difficulty in open her mouth. The patient was

apparently normal 2 months back after which she developed lesions over tongue (whitish in colour) and gradually spreaded to entire oral cavity and lips. Lesions ruptured spontaneously on its own with erosion, crusting, and mousy odour. The patient underwent treatment at the local hospital for the same and she treated with T. acyclovir, T. candiforce, Inj. Taxim and candid mouth paint. The patient initially improved with the treatment, but it occurred after one week and started to spread all over the body. History of itching increased with red skin lesions, onset of blisters, throat pain, burning micturition, loss of appetite, weight loss (10kg in a few

months) was observed. No history of native medication/ photosensitivity, fever, abdomen pain, altered bowel habits, ocular pain/ discharge, loose stools and she was diagnosed for VZV infection four months back. On lab investigation, the patient was found to be positive for Nikolsky's sign. Skin biopsy test revealed the skin with hyperkeratosis and parakeratosis, epidermis shows mild spongiosis with acantholysis and suprabasal bullae with keratinocytes are attached to basement with pigment incontinence in the dermis. From these features it was confirmed that is a condition of bullous pemphigoid or Pemphigus vulgaris.

Figure1: Patients Observation during admission

a, b- Painful blisters and sores in the mouth

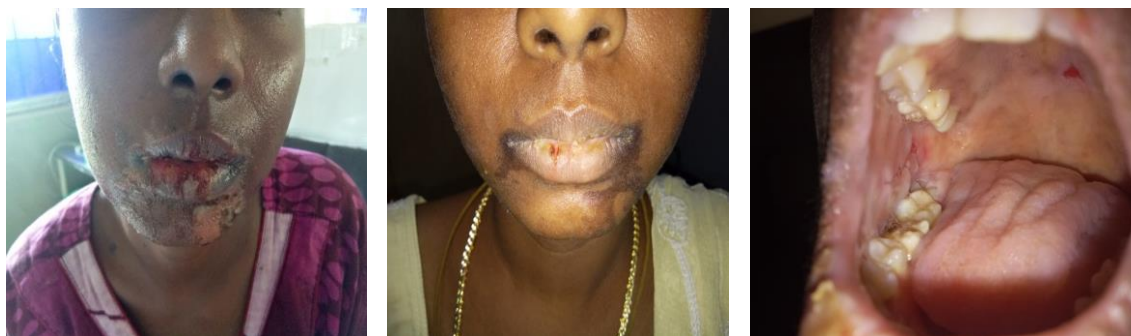


Treatment

Treatment of pemphigus vulgaris follows a standard therapy of corticosteroids with an immunosuppressant. In this patient, the drugs given by the physician are Steroids (dexamethasone 2CC iv) which is a standard treatment for Pemphigus vulgaris. Immunosuppressant (T. Cyclophosphamide 50 mg) was given for reducing the side effects of steroids and also a dose of steroids reduced gradually based on lab reports and patient recovery. Meanwhile blisters pus exudates were taken for microbial culture test and the report revealed that a growth of microorganisms such as streptococcus, staphylococcus, Klebsiella pneumonia, Enterococcus Faecalis, E-coli. The organisms have shown a susceptible to ciprofloxacin. Therefore, to suppress these organisms growth Inj. Cifran 200 mg was prescribed. Inj. Flagyl 500 mg and Inj. Fluconazole

200 mg were given to eradicate the fungal infections. Patient prescribed with Candid mouth paint and saline gargles to reduce the severe mouth ulcers. IV fluid, saline, Fusiwal ointment was given to reduce the erosion due to biopsy, Sitz bath is recommended for the patient because of blisters which causes irritation at perineal area and saline soaks was given to heal the sores and keeps the skin clean. Anti-viral (T. Acyclovir 400 mg) is given to the patient for prophylactic use because there are chances for viral infection and patients already had a past medical history of VZV infection three months back. Anthelmintics (T. Mebex 100mg) are also prescribed for treating worm infections. After 7 days of therapy, all the skin reactions were subsided. Patients discharged on their request and she advised to come for follow up of every 2 weeks once.

Figure 2: Patient recovery after 1 week of therapy
a-For the period of hospital Stay, b & c- During the discharge



CONCLUSION

PV is a rare and chronic autoimmune disease. In olden days, the mortality of PV leads to death because of the lack of proper treatment. Nowadays, mortality of PV is reduced due to an early prognosis of treatment. There is no cure for pemphigus vulgaris, but treatment help to keep the symptoms under control. Steroids are given to treat PV with immunosuppressant which leads to decrease the blister formation, promote healing of blisters and erosions, and also to determine the minimal dose of medication necessary to control the disease process. Even though their lesions are slowly improving but they are completely healing without a scar. Once the symptoms are under control, the steroid medication will gradually be reduced to the lowest possible dose that can still control the patient symptoms. This will help to reduce the risk of side effects. Therapy must be tailored for each patient, taking into account of preexisting and coexisting conditions. Clinical pharmacist must educate the patients regarding the oral hygiene, cleanliness and food practices etc... Early diagnosis leads to a better prognosis of this disease.

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ABBREVIATIONS

PV-Pemphigus Vulgaris

VZV- Varicella Zoster Virus

Conflict of Interest: Nil

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