World Journal of Pharmaceutical Sciences
ISSN (Print): 2321-3310; ISSN (Online): 2321-3086
Available online at: http://www.wjpsonline.org/
Case Study

CASE REPORT ON PEMPHIGUS VULGARIS

Mintu Augustine*, Suhaila Ahammed, Greeshma Rajesh, Ajimi A Rahiman, Fathimath Dilsha

Department of Pharmacy Practice, National College of Pharmacy, Manasserry, Calicut, Kerala, India

Received: 21-07-2020 / Revised Accepted: 25-08-2020 / Published: 30-08-2020

ABSTRACT

Pemphigus Vulgaris is an autoimmune disease mainly affects skin and may also affect the mucosae of the mouth, nose, conjunctivae, genitals, esophagus, pharynx, and larynx. PV is found mainly in middle-aged and elderly patients. PV is a rare, potentially life threatening disease.Mucus membranes are mostly involved. Oral lesions are most frequently observed. Skin involvement usually occurs after mucosal involvement which is characterized by flaccid blisters and erosive areas. Corticosteroids are the primary drug used for the treatment of PV. Immunoadsorption techniques, intravenous immunoglobulin and rituximab have given boost to effective management of PV in severe cases. Here we report a case on pemphigus vulgaris.

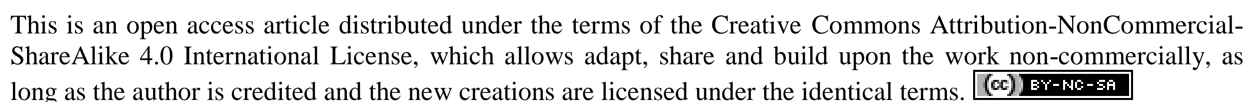
Key Words: Pemphigus Vulgaris, Acantholysis, Autoimmune, Mucocutaneous, Rituximab

INTRODUCTION

Pemphigus is a group of autoimmune, potentially life threatening mucocutaneous diseases characterized by epithelial blistering affecting cutaneous and/or mucosal surface [1]. Pemphigus can be grouped into 6types: pemphigus vulgaris, pemphigus vegetans, pemphigus erythematosus, pemphigus foliaceus, paraneoplastic pemphigus and IgA pemphigus. PV isthe most common variant, shows oral lesions as initialmanifestation in 50% of cases [2].Peak incidence of PV occurs between fourth and sixth decades of life, withmale to female ratio of 1:2[2]. According to the extent of cutaneous lesions, PV is further divided into mucosal dominant and mucocutaneous types [3]. Susceptibility of disease is strongly associated with some classII HLA antigens[4].Pemphigus is characterized by theproduction of pathogenic autoantibodies directed againstdiverse proteins of desmosomes. Binding of autoantibodies to desmosomal components results in disjunction of desmosomal areas, leads to acantholysis and formation of an intra-epidermal blister. Mucus membranes are mostly involved. Orallesions are most frequently observed. Skin involvement usually occurs after mucosal involvement which is characterized by flaccid blisters and erosive areas.
Nikolsky sign is commonly observed in perilesional area. Skin lesions are mainly localized

Address for Correspondence: Mintu Augustine, Department of Pharmacy Practice, National College of Pharmacy, Manasserry, Calicut, Kerala- 673602 India; Email: mintuaugustine8943@gmail.com

How to Cite this Article: Mintu Augustine, Suhaila Ahammed, Greeshma Rajesh, Ajimi A Rahiman, Fathimath Dilsha. Case report on Pemphigus Vulgaris. World J Pharm Sci 2020; 8(9): 156-158.


This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 International License, which allows adapt, share and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

© 2020World J Pharm Sci

in flexural areas, face, scalp, and extremities[4]. Though corticosteroids have remained primary therapy for pemphigus, immune adsorption techniques, intravenous immunoglobulin and rituximab have given boost to effective management of PV in severe cases.The incidence assessed by clinic-based survey conducted in Thrissur district, Kerala, was 4.4 per million populations per year [5].

CASE REPORT

A 47 year old female patient was admitted to the Department of Dermatology with a two months history of multiple oral erosions. She also complained about pain on swallowing food two months back. She consulted a Doctor and was given Vitamin B complex tablets but showed no improvement and new lesions continued to appear. Later she noticed multiple blisters over abdomen and back. The lesions were flaccid and ruptured within one or two days. Similar lesions also appeared on the scalp two weeks back. She consulted a dermatologist and was advised further evaluation and skin biopsy. On examination multiple erosions of varying size with crusting present over scalp, chest, abdomen and back. Two flaccid vesicles were present on back. Nikolsky sign was negative while bullae spread seqn was found to be positive. Hemorrhagic crusting was present on lips, erosions of varying size were present in oral cavity. The patient was treated with tablet prednisolone 30mg which was tapered and then stopped. A diagnosis of pemphigus vulgaris was made after evaluating the skin biopsy. Skin biopsy was performed at multiple levels. Sections show mild hyperkeratosis and suprabasal bullae in epidermis containing neutrophils, eosinophils and acantholytic cells. Few dermal papilloma were seen covered by basal epithelial cells and the floor of the bullae. Papillary dermis shows edema, capillary congestion and inflammation including eosinophils, deep dermis and subcutaneous tissue necrosis. Features were consistent with pemphigus vulgaris. Immunofluorescence assay shows fishnet deposits of IgG 3 in the epidermis.

Patient was admitted in dermatology ward for first dose of rituximab infusion and was initially treated with tablet prednisolone 40mg, syrup potassium chloride, condy's bath and mupirocin ointment. Complete blood count, urine examination, liver function test, serum electrolytes, random blood sugar and c reactive protein, ECG and ECHO were done and was found fit for rituximab infusion. Injection rituximab 500mg/50ml infusion in 450ml normal saline was given under medicine ICU care. There were no infusion reactions, no breathing difficulty, fever, chills and rigors. Vitals were stable. Patient felt better and was discharged with medications. She continued hospital visits in order to take Rituximab infusion. Symptomatic relief was observed after completion of therapy compared to initial visit.

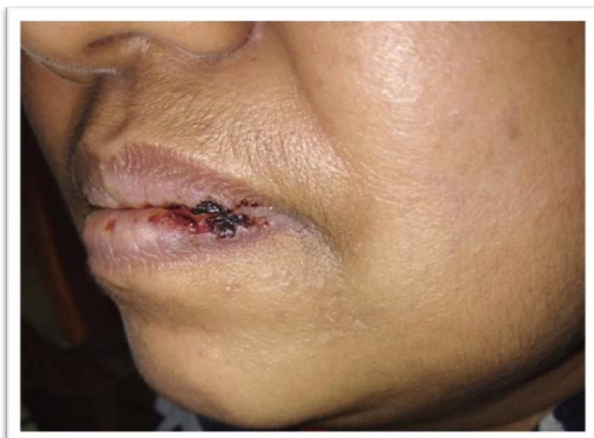


Fig no.1:Oral erosions during treatment.

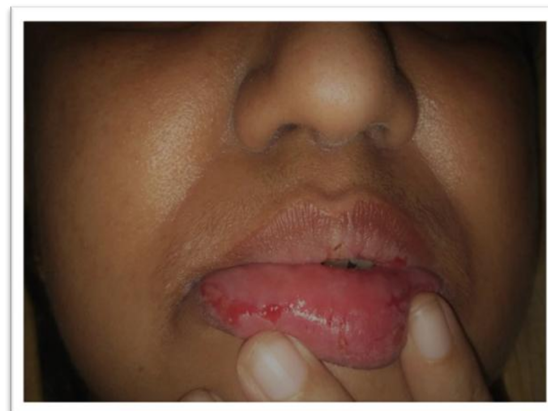


Fig no 2: Multiple oral erosions prior to treatment.

CASE DISCUSSION

Pemphigus mainly affects skin and may also affect the mucosae of the mouth, nose, conjunctivae, genitals, esophagus, pharynx, and larynx. PV is found mainly in middle-aged and elderly patients [6]. PV is rare, those who affected with the disease condition presents a lifetime physical, emotional and monetary burden. Etiology of PV is multifactorial, with complex interactions of genetic and environmental factors contributing to the progression of disease and exacerbation[7]. Primary goal of treatment is to induce disease remission. Management of disease should be followed by a period of maintenance treatment using minimum doses of drugs required for disease control in order to minimize their side effects[6].

Corticosteroids are the primary drug used for the treatment of PV. Localized mild lesions of pemphigus in oral mucous membrane in patients with low titers of circulating auto antibodies may

be controlled, at least temporarily with topical corticosteroids rinses or creams[8, 9]. The minimal therapy includes prednisone with less than or equal to 10mg/day or a two months duration of minimal adjuvant therapy[10]. Conventional therapy includes high dose corticosteroids, immunosuppressants and immune globulins[11]. Morbidity and mortality rate depends on extend of disease progression, maximum dose of corticosteroids used to induce remission, presence of other diseases and in older patients.

In our case, the patient was presented with multiple oral lesions and erosion over abdomen and trunk. She also complained about pain on swallowing food. She was treated with prednisolone 40mg, condy's bath, saline compression over erosions, calcium supplements, potassium chloride syrup and rituximab infusion 500mg/50ml added to 50ml normal saline. Following therapy patient reported pain relief and was discharged with same medications. She continued hospital visit in order to take rituximab infusion. After the completion of therapy she feels much better.

CONCLUSION

Pemphigus Vulgaris is an autoimmune disease, mainly affects skin and mucous membrane. The disease diagnosed by skin biopsy and immunofluorescence assay. Corticosteroids are primarily used for the treatment. Rituximab can be used in severe cases.

Acknowledgement: The authors thank the Department of Dermatology.

Conflict of interest: The authors have no conflict of interest.

Abbreviations: PV-Pemphigus Vulgaris, IgA-Immunoglobulin A, HLA- Human Leukocyte Antigen, ECG- Electrocardiogram, ECHO-Echocardiogram.

REFERENCE

1. Scully C, ChallacombeSJ. Pemphigus Vulgaris: Update on Etiopathogenesis, Oral Manifestations, and Management. Crit Rev Oral Biol Med 2002; 13(5):408-397.
2. Shamim T et al. Pemphigus Vulgaris in Oral Cavity: Clinical analysis of 71 cases. Med Oral Patol Oral Cir Buccal 2008; 13(10):626-622.
3. Yoshida K et al.Cutaneous type pemphigus vulgaris: A rare clinical phenotype of pemphigus. J AM ACAD DERMATOL 2005; 52:845-839.
4. Joly P, Litrowski N. Pemphigus group(Vulgaris, vegetans, foliaceus, herpetiformis,brasiliensis).Clinics in Dermatology 2011;29:436-432.
5. Kanwar AJ, De D.Pemphigus in India.Indian journal of Dermatology, venereology and leprology 2011; 77(4):449-439.
6. Dagistan S et al. Oral Pemphigus Vulgaris: A Case report with review of the literature. Journal of Oral Science 2008; 50(3):362-359.
7. Gupta VK et al. A Globally available internet based patient survey of Pemphigus Vulgaris: Epidemiology and disease characteristics. Dermatol Clin 2011; 29:404-393.
8. Tsankov N et al. Contact pemphigus induced by dihydrodiphenyltrichloroethane.Eur J Dermatol 1998; 8:443-442.
9. Braun-Falco O et al.Blistering Diseases. Dermatology 2nd ed.Springer. Munich 2000:695-650.
10. Venugopal SS, Morrell DF. Diagnosis and clinical features of Pemphigus Vulgaris. Dermatol Clin 2011; 29:380-373.
11. Ahmed AR et al. Treatment of Pemphigus Vulgaris with Rituximab and Intravenous Immuneglobulin.N Engl J Med 2006; 355:1779-1772.