International Journal of Pharmaceutical Research and Applications
Volume 6, Issue 4 July-Aug 2021, pp: 860-863 www.ijprajournal.com ISSN: 2249-7781

A Case Report On Pemphigus Vulgaris

Sunitha Mahadevan*[1] , Miss SowparnikaTreasa Sabu*[2]
*[1] PharmD Intern, Ezhuthachan College of Pharmaceutical Sciences
*[2] Assistant Professor, Department Of Pharmacy Practice Ezhuthachan College Of Pharmaceutical SciencesMarayamuttom,Neyyattinkara P.O, Thiruvananthapuram-695131

Date of Submission: 30-07-2021 Date of Acceptance: 08-08-2021

I. INTRODUCTION

Pemphigus vulgaris is a rare autoimmune disease causing painful blistering on the skin and mucous membranes and is the most common type of pemphigus[1]. Each type of pemphigus is characterised by blisters affecting the mucous membranes, which are found in areas including the : mouth, throat, nose, eyes, genitals and lungs[2]. This disease gradually starts with blisters in the mouth and then on the skin. These blisters can sometime affect the membranes of the genitals. Pemphigus vulgaris can be dangerous if left untreated leading to complications and typically involves the use of corticosteroids to suppress the immune system. The death rate from this disease had averaged 75 percent before corticosteroids were introduced in the 1950s[3]. This has improved with today's treatments. The symptoms include : painful blisters, oozing, crusting, or peeling at the blister site. The immune system produces proteins called antibodies, attacking harmful foreign substances like bacteria and viruses[4,5,6]. Pemphigus vulgaris occurs when the immune system mistakenly produces antibodies against proteins in healthy skin and mucous membranes[7,8,9]. The antibodies break down the bonds between the cells, and fluid collects between the layers of the skin. This leads to blisters and erosions on the skin. The exact cause of the pemphigus isn't known. Very rarely, certain medications can cause pemphigus vulgaris. These drugs include : penicillamine, which is a chelating agent that removes certain materials from the blood ans ACE inhibitors, which are a type of blood pressure medication[10,11,12,13]. Pemphigus vulgaris can affect people of all races, genders, and ages[14,15,16]. However, the condition is more common in the following groups: people of Mediterranean descent, eastern European Jews, people who live in the rainforests in Brazil, middle-aged and older adults[17]. A high dose of corticosteroids is the first line treatment for the condition. Common corticosteroids include prednisone or prednisolone and a high dose is usually needed to control the condition at first[18].

II. CASE REPORT

A 42 year old male patient was admitted in dermatology department with chief complaints of fluid filled lesions over body since 3 weeks, scaly erosion over scalp and oral erosion for past 2 weeks. The patient has no any concurrent co-morbidities.

On examination, the patient was found to be conscious with disphoric mood. presence of multiple flaccid vesicle and bullae with erosion and pus discharge was observed. The history of present illness started 2 weeks before. Lesions first erupted in the abdominal area then gradually spread all over the body upto scalp, sparing bilateral limb below knees and bilateral upper limb below elbows. No lesions were found in the genitalia. The patient has no recent history of travel or contact with allergen.

CLINICAL EXAMINATION

CVS: Normal S1 and S2 heart sounds.
Chest : Clear AEBE. Abdomen : soft, non-tender.Low grade fever present.
Dermatological examination: Hyper pigmented plaques with crusting are present over anterior and posterior aspect of trunk tiny erosions positive. Few hyper pigmented plaques with erosions are seen over both thighs. Few tiny crusted small plaques are present over chin, submandibular region. Hyper pigmented crusted small plaques seen over scalp. Tiny erosions over hard palate buccal mucosa present.

LAB INVESTIGATIONS

Skin biopsy was performed. Presence of postive Nikolsky and Ballar spread sign. Temperature:98.6f, Pulse rate:86, Respiratory rate:22, BP:130/80mmHg, SPo2:99%,

International Journal of Pharmaceutical Research and Applications
Volume 6, Issue 4 July-Aug 2021, pp: 860-863 www.ijprajournal.com ISSN: 2249-7781

Hb:15.5gm%, WBC:14600/cumm, Neutrophils;76%, Lymphocytes: 15%, Eosinophils:0%, Monocytes:9%, Basophils:0%, RBC:4.9million cells/cumm, Platelets:2.6lakhs, ESR:11mm/hr, Serum urea:24mg/dl, Serum creatinine: 1mg/dl, sodium:140 mmol/l, Potassium:3.9 mmol/L , Magnesium: 2.2mmol/l, Phosphorous:3.5mmol/L, Total bilirubin:1mmol/L, Direct bilirubin: 0.2mmol/L, SGOT:25IU/L, SGPT:67IU/L, ALP:77IU/L, Total protein: 6.8mg/dL, Serum albumin:4g/L and HbAIC:5%.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of Pemphigus Vulgaris can be done from other similar conditions by biopsy and direct immunofluorescence. Biopsies are best done on intact vesicles and bullae less than 24 hours old. The biopsy specimen should be taken from the advancing edge of the lesion, where the area of characteristic suprabasilar acantholysis can be observed by the pathologist. Supra basilar split seen in Pemphigus Vulgaris helps distinguish this condition from sub-epithelial blistering diseases such as mucous membrane pemphigoid, bullous lichen planus and chronic ulcerative stomatitis. Indirect immunoflorescence is helpful in further distinguishing pemphigus from pemphigoid and other chronic oral lesions and is useful in following the progress of patient for pemphigus.

ASSESSMENT

Based on objective and subjective parameters the patient is suffered with Pemphigus Vulgaris.

Table I : Therapeutic options for the treatment of pemphigus vulgaris

Therapy	Method
First-line therapy	Corticosteroids (Prednisolone) Start with 1 mg/kg per day in severe cases 0.5–1 mg/kg per day in milder cases Doses may be increased by 50–100% every 5–7 days if blistering continues After installation of the remission period, the doses are gradually decreased (5–10 mg prednisolone/2 weeks down to 20 mg daily, then by 2–5 mg every 2–4 weeks down to 10 mg daily) Add an adjuvant immunosuppressant: Azathioprine 2–3 mg kg - 1 per day Mycophenolate mofetil 2–3 g per day Rituximab (rheumatoid arthritis protocol, 291 g infusions, 2 weeks apart)
Second-line therapy	If first-line treatment does not work, switch to alternate corticosteroid-sparing agent (azathioprine, mycophenolate mofetil or rituximab)
Third-line therapy	Cyclophosphamide Immunoadsorption Intravenous immunoglobulin Methotrexate Plasmapheresis or plasma exchange

Adapted from British Association of Dermatologists guidelines for the management of pemphigus vulgaris 2017

TREATMENT PROVIDED

Initially the patient was admitted and started empiral treatment with stat medications: Inj. TT(Tetanus Toxoid) 0.5ml IM, Inj Dexa(Dexamethasone) 4 mg iv, T. Atarax(Hydroxyzine) 10mg and Inj.Avil (Pheniramine maleate)1 amp iv.

Later pharmacological treatment continues with the following medications:

Inj.Taxim(cefotaxim) 1g iv BD, C.Cloxacillin 500mg Q6h, Inj. Betnesol (Betamethasone)1 CC iv BD, Inj.Pantop(Pantoprazole) 40 mg iv BD, T. Calcium+Vit D3 OD, Fucidin(Fusidic acid)cream LA BD, Candid(Clotrimazole) mouth paint BD, T.Zincovit(Multivitamin)OD, T.Deslor (Desloratidine) 5mg BD , T. Glyciphage (Metformin) 500mg BD , T.Allegra 180mg BD, Clonate (Clobetasol)lotion scalp LA ,Inj. Piptaz(Piperacillin+ Tazobactum)4.5 g iv TDS, T. Atarax (Hydroxyzine) 10mg, T.Nexito (Alprazolam) plus HS.

Others :

Potassium permanganate bath , Cetrimide 5% shampoo scalp application (1:2 diltuion), Dermadew aloe vera cream ans Sterile vaseline gauze was provided.

III. DISCUSSION

Pemphigus is defined as a group of autoimmune disorder of skin and mucous membrane characterized by acantholysis. The process of acantholysis is caused by autoantibodies to intercellular adhesion molecules . In majority of cases (70–90%), the first sign of the disease ie, blisters appears on the oral mucosa. While the lesions can be found anywhere within the oral

International Journal of Pharmaceutical Research and Applications
Volume 6, Issue 4 July-Aug 2021, pp: 860-863 www.ijprajournal.com ISSN: 2249-7781

cavity, they are most commonly found in areas subjected to frictional trauma ie, cheek mucosa, pharynx, larynx, esophagus, genital mucosa as well as the skin where intact blisters are commonly seen. The combination of systemic corticosteroids (prednisone/prednisolone, 1.0–1.5mg/kg/day) and potentially corticosteroid-sparing immunosuppressive drugs, azathioprine and mycophenolate mofetil, was regarded as standard first-line therapy for pemphigus vulgaris. To reduce the corticosteroids dose ,additional treatment with antibiotics, antivirals, and antifungals are prescribed accordingly. In severe cases plasmapheresis may be required. This procedure is intended to remove the antibodies attacking the skin from the blood. The plasma of the blood is removed by a device and replaced with donor plasma hence is very expensive. If the blisters are severe, numbing lozenges for mouth blisters, soothing lotions, wet dressings may be considered.

IV. CONCLUSION

Pemphigus vulgaris is a rare autoimmune disorder that comprise of blistering and erosion of the skin and mucous membranes. It mainly occurs in middle-aged or older people (>40 years of age). The primary lesion of pemphigus vulgaris is a soft blister filled with clear fluid that appears on the skin. The blisters develops in the mouth, followed by skin blisters persisting for some time period. The blisters might be painfull, making it hard for eating food and rupture of blisters on the skin may limit the person's daily activities. The complications of the infection can be seriousas it may cause loss of body fluids and protein. The exact cause of pemphigus vulgaris is unknown, but the blisters in pemphigus vulgaris are associated with the binding of antibodies to the skin cells. Treatment is aimed at reducing symptoms and preventing complications, and may include the use of corticosteroids, immunosuppressive drugs, and more recently immunotherapy. Pemphigus vulgaris cannot be cured and require long term treatment to keep it in remission.

CONFLICT OF INTEREST

The author declares there is no conflict of interest.

REFERENCES

[1]. Shamim T, Varghese VZ, Shameena PM, Suddha S. Pemphigus Vulgaris in oral cavity. Clinical analysesof 71 cases. Med Oral Pathol Buccal. 2008;13:2622–2626. [Google Scholar]

[2]. Williams DM. Vesiculobullous Mucocutaneous Disease: Pemphigus Vulgaris. J Oral Pathol Med. 1989;18:544. [PubMed] [Google Scholar]

[3]. Scully C, Paes De Almeida O, Porter SR, Crilkes JJH. Pemphigus Vulgaris, manifestations and long term management of patient with oral lesions. British Journal of Dermatology. 1999;140(1):84–89.[PubMed] [Google Scholar]

[4]. Kauvsi S, Danesh pazhooh M, Farahani F, Abedini R, Lajevardi V, Chams davatchi C. Outcome of Pemphigus Vulgaris. Journal of the European Academy of Dermatology and Venereology. 2008;22(5):580–584. [PubMed] [Google Scholar]

[5]. Endo H, Rees TD, Hallmon WW. Disease progression from mucosal to mucocutaneous involvement in a patient with desquamatous gingivitis area with Pemphigus Vulgaris. Journal of Periodontology. 2008;99(2):368–375. [PubMed] [Google Scholar]

[6]. Michael H, Carrian S. Pathogenesis, Clinical Manifestation and diagnosis of Pemphigus. 2013. Jul 7, url: http//www.uptodate.com/store.

[7]. Dagistan S, Goregen M, Milogluo, Lakur B. Oral Pemphigus Vulgaris: A Case Report with review of literature. J Oral Sci. 2008;80:359–362. [PubMed] [Google Scholar]

[8]. Huntley AC. Pemphigus Vulgaris and vegetating and verrucous lesions. Case Report, Dermatol Online J. 2004:9. [Google Scholar]

[9]. Mahoney MG, Wang Z, Rothenberger K. Explanation for the Clinical and Microscopic localization of lesion in Pemphigus foliaceous and vulgaris. J Clin Invest. 1999;103:461–468.[PMC free article] [PubMed] [Google Scholar]

[10]. Jensen PJ, Barad J, Morioka S. Epidermal Plasminogen activator is abnormal in cutaneous lesion. J Invert Dermatol. 1988;90:777. [PubMed] [Google Scholar]

[11]. Stanley Cell adhesion molecule as target of autoantibodies in pemphigus and pemphigoid bullous due to defective cell

adhesion. Adv immunol. 1993;53:291–393. [PubMed] [Google Scholar]

[12]. Urbano FL. Nikolsky's Sign in Autoimmune Skin Disorders. Hospital Physician. 2001:23–24.[Google Scholar]

[13]. Kancoar AJ, Deo Pemphigus in India. Indian J Dermatol Venereai leprol. 2011;77:439–449.[PubMed] [Google Scholar]

[14]. Harman KE, Albert S, Black MM British Association of Dermatologists, author. Guideline for the Management of pemphigus Vulgaris. Br J Dermatol. 2003;149:926–937. [PubMed] [Google Scholar]

[15]. Anhalt GJ. Paraneoplastic Pemphigus. Journal of Investigative Dermatology Symposium Proceedings. 2004;9:29–33. doi: 10.1111/j.1087-0024.2004.00832.x. [PubMed] [CrossRef] [Google Scholar]

[16]. Fellner MJ, Sapadin AN. Current therapy of pemphigus vulgaris. Mt Sinai J Med. 2001;68(4–5):268–278. [PubMed] [Google Scholar]

[17]. Toth GG, Jonkman MF. Therapy of pemphigus. Clin Dermatol. 2001;19(6):761–767. [PubMed] [Google Scholar]

[18]. Lever WF, Schaumburg-Lever G. Treatment of pemphigus vulgaris. Results obtained in 84 patients between 1961 and 1982. Arch Dermatol. 1984;120(1):44–47. [PubMed] [Google Scholar]