Journal of Pharmaceutical Research International

33(43A): 324-328, 2021; Article no.JPRI.72943
ISSN: 2456-9119
(Past name: British Journal of Pharmaceutical Research, Past ISSN: 2231-2919, NLM ID: 101631759)

Pemphigus Vulgaris- A Case Report

Mayur B. Wanjari[1*], Deeplata Mendhe[1], Pratibha Wankhede[1] and Sagar Alwadkar[1]

[1]Department of Community Health Nursing, Smt. Radhikabai Meghe Memorial College of Nursing, Datta Meghe Institute of Medical Sciences, Sawangi (M) Wardha, Maharashtra, India.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/JPRI/2021/v33i43A32493
Editor(s):
(1) Dr. Prabakaran Nagarajan, The Ohio State University, USA.
Reviewers:
(1) Tulika Rai, Banaras Hindu University, India.
(2) Priyanka Shukla, ERA University, India.
Complete Peer review History: https://www.sdiarticle4.com/review-history/72943

Case Study

Received 02 August 2021
Accepted 25 August 2021
Published 06 September 2021

ABSTRACT

Introduction: Pemphigus is a rare and life-threatening autoimmune disease characterized by blisters and erosion of the skin and mucous membranes throughout the entire body. It mostly affects the mouth, eye, nose, throat as well as genitals. Pemphigus vulgaris is most common type of pemphigus. The epithelial lesions are caused by autoantibodies reacting with desmosomal glycoproteins found on the keratinocyte's cell surface. The binding of immunoglobulin G autoantibodies to desmoglein 3, a transmembrane glycoprotein adhesion molecule found on desmosomes, is the underlying process that causes intraepithelial lesions.
Case Presentation: A 40-year-old male patient came to the hospital with a complaint of a mouth ulcer and a genital lesion that had been since 15 years. He is having a history of oral ulcer in oral mucosa and involvement of genital area in glans penis with a history of pain, bleeding, difficulty in swallowing.
Intervention: The patient was admitted to the hospital on 29/07/2021 and taken Inj. Rituximab 1gm in 500Ml Normal Saline in infusion pumps in over 6 Hours.
Conclusion: In this case report, we mainly focus on expert dermatological management and excellent nursing care in managing the rare complicated case nicely. Early diagnosis and treatment of pemphigus Vulgaris help determine the course of the disease of the patient and is done by a dermatologist.

**Corresponding author: E-mail: Wanjari605@gmail.com;*

Keywords: Pemphigus; pemphigus Vulgaris; blisters; erosions; autoantibodies; desmosomal glycoproteins; keratinocyte.

1. INTRODUCTION

Wichman coined the name "Pemphigus" in 1791 to describe a chronic blistering disorder derived from the Greek word "Pemphix," which meaning "bubble or blister." [1] Pemphigus Vulgaris is a severe and rare inflammatory autoimmune bullous disease of the skin and mucus membrane. A male to female ratio of 1:2 is the highest in the 5th and 6th decades of life, with the highest prevalence of cases per 100,000 people each year.[2] Some weird cases in children and the elderly have been reported.[3] The more common variants of pemphigus are Pemphigus Vulgaris, Pemphigus Vegetans, Pemphigus Foliaceous, Pemphigus Erythematosus, Paraneoplastic Pemphigus [PNP]and Drug-related Pemphigus but from that Pemphigus Vulgaris is most common of pemphigus and it accounts for over 80% of cases. [4] When just mucosal involvement is observed, it can be difficult to diagnose, as the majority of cases are affected by the oral mucosa.

2. PATIENT AND OBSERVATION

2.1 Patient Information

A 40-year-old male patient was a resident of Wardha, Maharashtra, and come to the outpatient department of dermatology with a chief complaint of ulcer in the mucosal membrane of mouth with difficulty in swallowing of solid food as well as lesion over the genital area in glans penis with pain, bleeding, and fluid-filled lesion.

The patient was apparently alright 15 years ago when he started to notice the development of a single ulcer in the mouth which was pea-sized, gradually progressive in number. The lesion did not increase in size and was not also with pain or itching, discharge. The lesion would appear on and off and would heal spontaneously patient visited a dermatologist and found relief on the treatment.

The lesion subsite at that time 7 years later, lesions started to reappears assist on the genital area with pea-sized, not associated with any complaints. A similar lesion appeared in the oral mouth patient visited a doctor and found relief on his treatment and stopped his total treatment after he found relief 3 months ago, the lesion reappeared over the genital, was associated with pain bleeding.

He took treatment found the same relief. But now the patient was facing the same problems and he comes to the hospital for further treatment dermatologist advice to do an investigation of Desmoglein I Antibody it was negative (6.35RU/mL) and Serum Anti Desmoglein 3 it was positive (43 U/ml). The doctor advises the patient to take Inj. Rituximab 1gm at Acharya Vinobha Bhave Rural Hospital, Sawangi (Meghe), Wardha, Maharashtra, India.

The patient is not having a history of illnesses like diabetic Mellitus, Hypertension, and Tuberculosis. No similar complaint in the family.

2.2 Diagnostic Assessment

On the general examination of the patient is conscious, cooperative, and well oriented to person, time, and place.

On cutaneous examination, Erosions are present over the glans penis with bleeding present.

Undergone through all investigation and physical examination final diagnosis is made Pemphigus Vulgaris.

2.3 Therapeutic Intervention

As said earlier, Pemphigus Vulgaris is a more life-threatening highly life-threatening autoimmune disease condition, if not diagnosed and treated earlier. The treatment given to our patient is as follows:

2.3.1 Pre-infusion medications

Inj. Methylprednisolone 100Mg with 100ML Normal saline Intravenous over 30Min, Inj. Avil 2 cubic centimeter (cc) Intravenous, Tab. Paracetamol 1gm.

2.3.2 Infusion Medication

Inj. Rituximab 1gm in 500Ml Normal saline over 6 hours.

3. DISCUSSION

Pemphigus is characterized by a thin-walled bulla that forms on seemingly normal skin or mucosa, breaks immediately, and spreads peripherally, eventually leaving vast denuded areas. [5] In patients with pemphigus, this disease also introduces a positive “Nikolsky's sign” - the ability to produce a peripheral extension of a blister and/or depletion of the epidermis by applying tangential pressure with a finger or thumb to the afflicted skin, peri-lesional skin, or normal skin[6].

The oral mucosa is the site of the disease's initial manifestation in the vast majority of cases (70–90%)[7]. In areas that are prone to frictional damage, such as the cheek, pharynx, larynx, esophagus, genital mucosa, and skin, where intact blisters are common, the sores can appear anywhere in the mouth[8].

The more common variants of pemphigus are Pemphigus Vulgaris, Pemphigus Vegetans, Pemphigus Foliaceous, Pemphigus Erythematosus, Paraneoplastic Pemphigus [PNP] and Drug-related Pemphigus.[9] Our case represents Pemphigus vulgaris. An antibody directed against different target cell surface antigens has caused a lesion to form in different layers of the epithelium in each form of this disease. [10,11] Pemphigus Vulgaris is the most common of pemphigus and it accounts for over 80% of cases [4,12].

Pressure applied to intact bullae results in a recognizable symptom of the disease that Bullae increase by extension to a normal surface in a patient with Pemphigus Vulgaris, and another symptom of the condition is pressure applied to a seemingly normal location, resulting in the development of a new lesion.[13] The phenomenon caused by the upper layer of skin peeling away from the basal layer is known as the Nikolysky sign and is also the positive sign in toxic epidermal necrolysis, scalded skin syndrome, and mucous membrane pemphigoid (both of which are acute illnesses) [14].

Early identification of the disease is a critical path to inpatient management since a smaller dose of medicine can be given for a shorter period of time to manage the disease. To ensure early diagnosis and treatment of pemphigus Vulgaris, dentists must be well trained in identifying clinical manifestations and providing guidance on disease progression and disease course. In early-stage take treatments that can prevent major involvement of the mucosal and cutaneous area, as well as lethal significance. To treat Pemphigus Vulgaris, local and systemic

Table 1. Clinical finding

Investigation	Patient Value
Blood Investigation	
Hemoglobin	12.5 gm/dL
Total RBC Count	4.95 cells/mcL
Total WBC Count	6000 per microliter of blood
Haematocrit	39.2%
Mean Corpuscular Hemoglobin Concentration (MCHC)	31.8g/dl
Mean Corpuscular Volume	79.1fl
Mean Corpuscular Hemoglobin	25.1 picograms
Total Platelet Count	2.14 per microliter of blood
Monocytes	03
Granulocytes	65
Lymphocytes	30
Red Cell Distribution Width (RDW)	18.5
Eosinophils	02
Basophils	00
Urine Examination	
Urine Albumin	Absent
Urine Sugar	Absent
Epithelial Cell	Absent
Pus Cell	Present 2-4 Cells/HPF
Desmoglein I Antibody	Negative- 6.35RU/mL
Sr. Anti Desmoglein 3	Positive- 43 U/ml

corticosteroid therapy is commonly used. Treatment is divided into two stages: disease control and maintenance, which include consolidation and treatment tapering. In conjunction with systematic treatment can be used local treatment comprises a past, ointment or mouthwash. In chronic lesions treat corticosteroid intralesional injections have been used[14].

4. CONCLUSION

Pemphigus Vulgaris is a potentially threatening condition that can be treated with the proper level of care. Dermatologists and dentists are familiar with pemphigus vulgaris. If it is not treated early, the morbidity rate can be as high as 5-10%.[15] In this case report, a patient treated with Inj. Rituximab 1gm and showed progressive recovery.

This case report is a contribution to the knowledge of the rare fatal autoimmune disease of the skin.

5. RECOMMEDATION

The physician is treated with Tab. Bentnesol Forte, Tab. Azoran 50Mg, Tab. Pan 40Mg, Tab. Shelcal 500Mg, Tab. Dailyshine 60K IU, Lotion Momate F Cream for 15 days.

On discharge, the physician has advised the patient to come after 15 days to take a second dose of Inj. Rituximab 1gm.

CONSENT

While preparing case reports for publication patient informed consent has been taken from the patient.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Banerjee I, Bhowmik B, Maji A, Sinha R. Pemphigus vulgaris–A report of three cases and review of literature. Journal of family medicine and primary care. 2018;7(5):1109.
2. Shamim T, Varghese VI, Shameena PM, Sudha S. Pemphigus vulgaris in oral cavity: Clinical analysis of 71 cases.
3. Williams DM. Vesiculobullous mucocutaneous disease: pemphigus vulgaris. Journal of Oral Pathology & Medicine. 1989;18(10):544-53.
4. De Almeida PA. Pemphigus vulgaris: the manifestations and long-term management of 55 patients with oral lesions. British Journal of Dermatology. 1999;140(1):84-9.
5. Kavusi S, Daneshpazhooh 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):580-4.
6. 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):368-75.
7. Dagistan S, Goregen M, Miloglu O, Çakur B. Oral pemphigus vulgaris: a case report with review of the literature. Journal of oral science. 2008;50(3):359-62.
8. Huntley AC. pemphigus vulgaris and Vegetating and Verrucous lesions: Case Report. Dermatol Online J. 2004;9.
9. Mahoney MG, Wang Z, Rothenberger K, Koch PJ, Amagai M, Stanley JR. Explanations for the clinical and microscopic localization of lesions in pemphigus foliaceus and vulgaris. The Journal of clinical investigation. 1999;103(4):461-8.
10. Jensen PJ, Baird J, Morioka S, Lessin S, Lazarus GS. Epidermal plasminogen activator is abnormal in cutaneous lesions. Journal of investigative dermatology. 1988;90(6):777-82.
11. Stanley JR. Cell adhesion molecules as targets of autoantibodies in pemphigus and pemphigoid, bullous diseases due to defective epidermal cell adhesion. Advances in immunology. 1993; 53:291-325.
12. Urbano FL. Nikolsky's sign in autoimmune skin disorders. Hospital Physician. 2001;37:23-4.
13. Fellner MJ, Sapadin AN. Current therapy of pemphigus vulgaris. The Mount Sinai Journal of Medicine, New York. 2001;68(4-5):268-78.

14. Lever WF, Schaumburg-Lever G. Treatment of pemphigus vulgaris: results obtained in 84 patients between 1961 and 1982. Archives of dermatology. 1984; 120(1):44-7.

15. Harman KE, Albert S, Black MM. Guidelines for the management of pemphigus vulgaris. British Journal of Dermatology. 2003;149(5):926-37.

Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle4.com/review-history/72943