MEDISAINS
Jurnal Ilmiah Ilmu - Ilmu Kesehatan

Case Report

Management of pemphigus vulgaris: case report

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ARTICLE INFORMATION

Received: August 12, 2021
Revised: September 03, 2021
Available online: September 06, 2021

KEYWORDS

Pemphigus; Autoimmune Diseases; Rare Diseases; Skin Abnormalities

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A B S T R A C T

Background: Pemphigus Vulgaris is an autoimmune disease attacking the skin and mucous membranes. This disease is considered a rare disease. Epidemiological data for Pemphigus Vulgaris in Indonesia remains limited, correspondingly, nursing management in this disease. This study aims to describe nursing management in patients with Pemphigus Vulgaris.

Case presentation: a case of Pemphigus Vulgaris in a 28-year-old male was reported, with complaints of blisters all over his body appearing since 2019 and have not been completely recovered at the Banyumas General Hospital, Central Java, Indonesia.

Conclusion: A combination of wound care, nutrition, vitamins, and pharmacology can accelerate the recovery of patients with Pemphigus Vulgaris.

INTRODUCTION

The number of cases of Pemphigus Vulgaris worldwide has an incidence that varies from 0.5 to 3.2 cases per 100,000 population with a mortality rate of 5-15%. Mortality in patients with Pemphigus Vulgaris is three times higher compared to the general population.[1] Epidemiological data for Pemphigus Vulgaris in Indonesia are still limited. The prevalence of this disease in Indonesia is 1-4 cases per 100,000 people, with mortality reaching 75%.[2] There were five patients with Pemphigus Vulgaris cases at the Banyumas General Hospital in January 2020 - December 2020.[3]

Up to the present time, research and data discussing the management of Pemphigus Vulgaris patients remain limited. The management mechanism of nursing management in this disease has not been widely discussed. This study reports on the prevalence of Pemphigus Vulgaris in a male patient and describes the nursing management of patients with Pemphigus Vulgaris from various literature.

CASE PRESENTATION

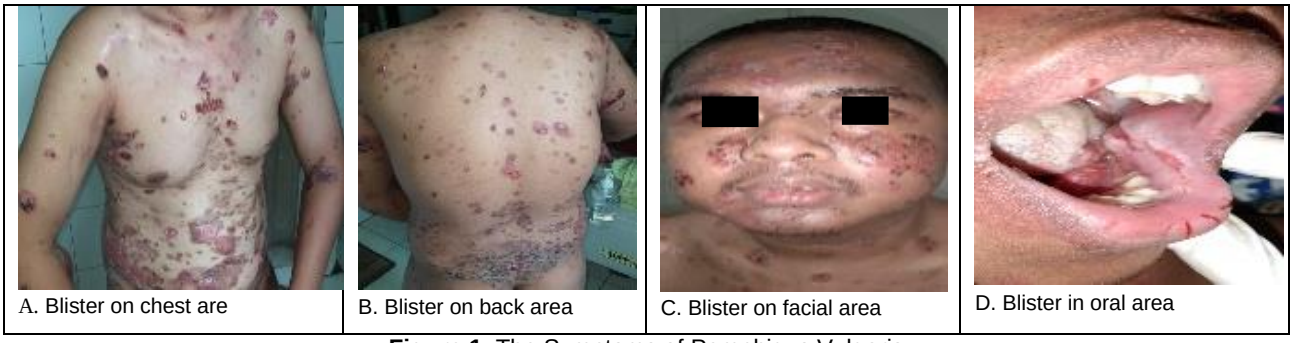
A 28-year-old male came to the Hospital Emergency Unit complaining of blisters all over his body and oral area. The patient said that the wound was burning, itchy and painful. Based on the anamnesis results, the patient said that these symptoms had appeared since 2019 and had been treated at several hospitals. Nevertheless, the wound had never fully recovered. The patient denied a history of drug and food allergies. The general condition of the patient was pretty fit. Compos mentis consciousness, blood pressure 120/70 mmHg, pulse 80 times/minute, temperature 37^0 C, respiration 20 times/minute.

The results of the wound assessment indicated that there were multiple erythematous macules, bullae, erosions all over the body with blackish-brown crusts, generalized distribution was in irregular shape, solid edges, and uneven surface. Sizes varied from 3x4 cm to 10x10 cm, and there was a solitary spherical erosion with a diameter of 3 cm covered with blackish-brown crusts (Fig. 1.A,B,C,D). The result of the Nikolsky signs physical examination on the

https://doi.org/10.30595/medisains.v19i2.11237

patient was positive. Based on the history and physical examination, and laboratory blood tests, the doctor diagnosed this patient with Pemphigus Vulgaris.

The patient was treated with daily wound care and administration of silver sulfadiazine ointment to prevent skin infection. Patients were provided with high calories and protein diet to help accelerate wound healing. Doctors also provided corticosteroid drug therapy, antibiotics, vitamin C, antihistamines, and adjuvant therapy to reduce the side effects of corticosteroid medication. After 21 days of treatment, the wound appeared to be healing, and the tissue damage began to decrease, as shown in Figure 2.A,B,C,D.



A. Blister on chest are | B. Blister on back area | C. Blister on facial area | D. Blister in oral area

Figure 1. The Symptoms of Pemphigus Vulgaris

A. Wound healing on chest area | B. Wound healing on back area | C. Wound healing on facial area | D. Wound healing in oral area

Figure 2. Wounds After 21 Days of Treatment

DISCUSSION

Pemphigus is an autoimmune disorder characterized by blisters on the skin and oral area. The wounds are formed due to acantholysis (separation of intra-cell cells) in the epidermis, which clinically appears as bullae. Acantholysis of pemphigus Vulgaris occurs in the sub-basal, particularly in the stratum spinosum or stratum granulosum and oral lesions. [4–7] Up to now, the exact cause of Pemphigus Vulgaris is unknown. However, the disorder is associated with impaired immune regulation that produces autoantibodies that attack cell junctions (desmosomes) responsible for adhesion to other cells in cell tissue. [8–10].

Risk factors that can increase or worsen pemphigus Vulgaris include genetic factors (human leukocyte antigen DRw6 and human leukocyte antigen RD4, which are associated with major histocompatibility complex II molecules), environmental factors such as ultraviolet radiation, autoimmune diseases (such as myasthenia gravis and thymoma) and the use of drugs such as D-Penicillamine and Captopril. [8,9,11,12] In this case, the exact cause of Pemphigus Vulgaris suffered by the patient is not yet known, and the symptoms first appeared in 2019 and have never fully recovered. The patient also had no history of drug or food allergies.

The clinical depiction of pemphigus Vulgaris is characterized by erosions/blisters on the skin and mucosal layers. Wounds can also affect the skin all over the body.[6,7] In about 60% of cases, the lesions/wounds first appear in the oral area; the remainder appears first on the scalp, face, neck, armpits, or genitals. Characteristics of the lesion are usually not itchy but painful. Bullae/blisters that rupture will form erosion and then crusting. This is a way for secondary infections that can increase mortality.[13,14] In this case, the symptoms of blisters in the mouth and all over the body feel burning, itchy, painful, they have been treated in many hospitals, but the wound has never recovered completely.

The diagnosis of pemphigus Vulgaris can be established if positive results are found on clinical examination, histological examination, and immunologic tests.[15] On the physical

examination, there is a Nikolsky sign, this sign can be seen by rubbing the hands from the normal area to the lesion, positive results appeared when there is peeling of the skin layer that indicates the shedding of the superficial layer of the basal layer of the epidermis. In addition, there is an Asboe-Hansen sign in the form of a bulla that widens when the middle part is pressed.[13] Histological depiction of pemphigus Vulgaris is a suprabasal bulla with acantholysis. Sometimes it is seen that keratinocytes are released into the bulla, and the superficial parts of the epidermis are seen intact.[16]

Immunological examination plays an important role, including examining serum antibodies and skin lesions; antibody examination of lesions is more specific and sensitive than serum antibodies. Antibody testing also helps assess the success of therapy. Serological examination of Enzyme-linked immunosorbent assay (ELISA) test to determine the presence of antibodies that attack desmoglein 1 and desmoglein 3 can also support the diagnosis of pemphigus Vulgaris.[17] In this case, the diagnosis of pemphigus Vulgaris was established based on the clinical picture seen in the patient, namely blisters in the mouth and sores scattered throughout the body. Nikolsky's physical examination results also indicated positive (+) results. In this case, no histologic, immunologic, or serological tests were performed.

In this case, the patient's wounds were treated every day; the patient was also given silver sulfadiazine ointment. The use of silver sulfadiazine ointment is suitable to help prevent and treat infection in open wounds; this medication works by stopping the growth of bacteria in open wounds.[18,19] The provision of a high-calorie diet, protein, and vitamin C was also carried out to help accelerate wound healing and increase the patient's immune system. The patient was also given immunosuppressant in the form of steroids. The use of corticosteroids was started with a high initial dose with a total injection dose of methylprednisolone 125 mg administered daily until clinical symptoms improved. The corticosteroid dose would be reduced slowly to a maintenance dose of 40-50mg per day to avoid relapse. The administration of corticosteroid drugs was aimed to reduce inflammation. In this case, the treatment was combined with antibiotics ceftriaxone injection 1 gram per 12 hours and oral doxycycline 100 mg per 12 hours in order to prevent secondary infection.[4,20]

Corticosteroids have immunosuppressive solid and anti-inflammatory effects that can cause high mortality and morbidity in patients. Therefore, to overcome the side effects of corticosteroids, it can be combined with adjuvant therapy (adjuvant corticosteroid-sparing) in the form of azathioprine, cyclosporine, methotrexate, and mycophenolate mofetil.[4] In this case, the patient was also given methotrexate (MTX). The dose given is 2x12.5 mg/week orally. Cetirizine therapy 1x10 mg was also given to patients to treat allergy symptoms and skin rashes.[18]

In this case, a combination of the management of wound care, nutrition, and pharmacology in the form of corticosteroids, antibiotics, antihistamines, and adjuvant therapy (methotrexate) proved to be effective in relieving the patient's recovery. Although the treatment period in the hospital took a longer time, which took 21 days, there was a very significant improvement in the patient's wounds. The wounds started to heal. It was seen that the condition of the wounds throughout the patient's body had recovered, and tissue damage had declined as well.

CONCLUSIONS AND RECOMMENDATION

Management of Pemphigus Vulgaris patients focuses on daily wound care, administration of silver sulfadiazine ointment to prevent skin infections, and providing a high-calorie and protein diet to support wound healing and pharmacological therapy as pharmacological therapy corticosteroids, antibiotics, vitamins and antihistamines, and therapy adjuvants. In this case, it was declared cured after 21 days of treatment, with signs of healing wound and decreased tissue damage. Standard operating procedures for managing patients with Pemphigus Vulgaris need to be improved so that patients' length of stay in the hospital can be shortened.

REFERENCES

1. Zeina B, Sakka N, Mansoor S. Pemphigus Vulgaris. *Medscape*. 2020. https://emedicine.medscape.com/article/1064187-overview.
2. Kurniawan F, Arisanty R. Karakteristik Klinik histopatologik dan Profil Imunofluoresensi Kasus Pemfigoid bulosa di Departemen Patologi Anatomik Rumah Sakit Cipto Mangunkusumo Tahun 2011-2018. *Maj Patol Indones (The Indones J Pathol*. 2020;29(3):139-144.
3. RSUD Banyumas. *Medical Record*. Banyumas; 2021.
4. Porro AM, Hans Filho G, Santi CG. Consensus on the treatment of autoimmune bullous dermatoses: pemphigus vulgaris and pemphigus foliaceus - Brazilian Society of Dermatology. *An Bras Dermatol*. 2019;94(2 Suppl 1):20-32. doi:10.1590/abd1806-4841.2019940206
5. Cholera M, Chainani-Wu N. Management of Pemphigus Vulgaris. *Adv Ther*. 2016;33(6):910-958. doi:10.1007/s12325-016-0343-4
6. Moraes da Silva A de F, Bernabé DG, Miyahara GI, Biasoli ER, Callestini R, Tjioe KC. Pemphigus Vulgaris: How to Perform an Oral Biopsy Properly? *J Craniofac Surg*. 2016;27(8). https://journals.lww.com/jcraniofacialsurgery/Fulltext/2016/11000/Pemphigus_Vulgaris__How_to_Perform_an_Oral_Biopsy.98.aspx.

7. Kuriachan D, Suresh R, Janardhanan M, Savithri V. Oral Lesions: The Clue to Diagnosis of Pemphigus Vulgaris. *Case Rep Dent*. 2015;2015:593940. doi:10.1155/2015/593940
8. Hammers CM, Stanley JR. Mechanisms of Disease: Pemphigus and Bullous Pemphigoid. *Annu Rev Pathol*. 2016;11:175-197. doi:10.1146/annurev-pathol-012615-044313
9. Meyerle JH, Anhalt GJ. Pemphigus. BMJ Best Practice. https://bestpractice.bmj.com/topics/en-us/454. Published 2021.
10. Tamgadge S, Tamgadge A, Bhatt D, Bhalerao S, Pereira T. Pemphigus vulgaris. *Contemp Clin Dent*. 2011;2(2):134-137. doi:10.4103/0976-237X.83074
11. Ingold CJ, Khan MA. *Pemphigus Vulgaris*. Treasure Island (FL): StatPearls Publishing; 2021.
12. Hertl M, Sitaru C. Pathogenesis, clinical manifestations, and diagnosis of pemphigus. UpToDate, Inc. https://www.uptodate.com/contents/pathogenesis-clinical-manifestations-and-diagnosis-of-pemphigus#H460257. Published 2021.
13. James WD, Berger TG, Elston DM, Odom RB. *Andrews' Diseases of the Skin : Clinical Dermatology*. 11th ed. Philadelphia: Saunders Elsevier; 2011.
14. Bharathi U, Lingaraju N, Basappa S, MaheshM. S. Oral Pemphigus Vulgaris: A Case Report and Review. *IOSR J Dent Med Sci*. 2014;13:24-29.
15. Singh S. Evidence-based treatments for pemphigus vulgaris, pemphigus foliaceus, and bullous pemphigoid: a systematic review. *Indian J Dermatol Venereol Leprol*. 2011;77(4):456-469. doi:10.4103/0378-6323.82400
16. Payne AS, Stanley JR. Pemphigus. In: *Fitzpatrick's Dermatology in General Medicine, 8e*. New York: Mc Graw Hill; 2012.
17. Bystryn J-C, Rudolph JL. Pemphigus. *Lancet (London, England)*. 2005;366(9479):61-73. doi:10.1016/S0140-6736(05)66829-8
18. Abedini R, Mahmoudi H, Kordestani S, Habib FN, Abyaneh M, Rahemi H. Comparison of topical nanocolloidal silver formulation use with eosin 2% solution in management of hard-to-heal ulcers in patients with pemphigus vulgaris. *J Wound Care*. 2020;29(11):664-668. doi:10.12968/jowc.2020.29.11.664
19. Soheila K, Farzaneh NH, Hosein SM. A Novel Wound Rinsing Solution Based On Nano Colloidal Silver. *Nanomedicine J*. 2014;1(5):315-323. https://www.sid.ir/en/journal/ViewPaper.aspx?id=412079.
20. Rezeki S, Setyawati T. Pemphigus Vulgaris: Pentingnya Diagnosis Dini, Penatalaksanaan Yang Komprehensif Dan Adekuat (Laporan Kasus). *Indones J Dent*. 2009;16(1):1-7. doi:10.14693/jdi.v16i1.20