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Aggressive Refractory Pemphigus Vulgaris: A Rare Case Report

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ABSTRACT

Background An autoimmune condition called pemphigus vulgaris causes ulcers on the skin and mucous membranes as well as intraepithelial blistering. Between 50 and 70 years old is the typical onset age. In children, pemphigus rarely occurs. Therapeutic plasma exchange is thought to eradicate pathogenic autoantibodies, and this is necessary in resistant severe cases. It corresponds with the level of circulating autoantibodies. Presentation of case: - a case of a 15-year-old boy; he came with a complaint of fluid fill lesions all over the body with itching since from 15 days. The patient was apparently alright 15 days back when he noticed small fluid-filled lesions over the abdomen that gradually progressed to the chest, back, upper limbs, face, and scalp. The lesion were initially of small size to begins with and then gradually progressed to the coin sized lesion. The lesion were inicially associated with mild to moderate Grade-I itching. The fluid filled lesion would burst on iching and also spontaneously with little tendency to heal. He also complained of oral ulser, burning sensation, Photosensitivity and fever. The patient was visited to the native place physician, who gave symptomatic relief treatment. After that, he visited our hospital. We did a physical examination; we noted fluid fill vesicle and bulla over the trunk, chest, and back of size 3×4 cm (Figure 1). All necessary investigations were done such as history collection, Physical examination General Examination of the poor and Nutritional Status adequate. In the laboratory, the investigation physician revealed he had pemphigus Vulgaris. The immediate actions were taken: Urinary catheterization with intravenous (IV) fluids to rectify the electrolyte imbalance and the circulatory system, early intravenous antibiotic care of sepsis, and appropriate opioid pain relief were the first steps done.

Categories: Dermatology

Keywords: Pemphigus Vulgaris, blisters, vesicle and bulla.

Introduction

A collection of potentially fatal autoimmune bullous disorders known as pemphigus vulgaris are typified by flaccid blisters and erosions of the skin and mucosal membrane. The typical onset age ranges from 50 to 70 years old. Children rarely get Pemphigus Vulgaris.[1] The disease's gradual course, which includes enhanced body catabolism, loss of bodily fluids and proteins, and secondary bacterial infections that can cause sepsis and heart failure, determines how serious it is.[2]

Corticosteroids are still the cornerstone of treatment, whether or not adjuvant therapy is used. Adjuvant therapy consists of immunotherapy procedures and steroid-sparing medications. Dermatologists worldwide have ventured into using new modalities of adjuvant therapies, such as intravenous immunoglobulins and therapeutic plasma exchange, to address the growing number of patients with severe PV who show little or no response to conventional corticosteroid treatment, due to the range of extent, varied response to standard therapeutic protocols, and severe side effects.[3]

Presentation of case: -

A case of 15-year-old boy; he came with a complaint of fluid fill lesions all over the body with itching since from 15 days. The patient was apparently alright 15 days back when he noticed small fluid-filled lesions over the abdomen that gradually progressed to the chest, back, upper limbs, face, and scalp. The lesion

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The patient seemed gloomy and a little boring. Her vital signs were as follows: 38.5°C for temperature, 90/60 mmHg for blood pressure, 115 beats per minute for heart rate, and 24 breaths per minute for respiratory rate. Her mouth was pallid. His capillary refill duration was less than two seconds. His skeleton was in normal condition. His check of his lymph nodes turned up nothing. On cardiac examination, he had no murmur and tachycardia. His examination of his lungs showed no wheezing or rales. He was a little dazed and confused. Her cranial nerve examination came out normal. His pupils had normal pupillary light reactions, both direct and indirect. His motor assessment showed normal muscular tone and mass. He has complete bilateral strength. He had symmetrical, normal reflexes at his knees, ankles, triceps, and biceps. He had flexor plantar responses. His evaluation of his senses showed that his fingers and toes had typical sensations of mild touch, pin prick, location, and vibration.

Urinary catheterization with intravenous fluids to rectify the electrolyte and circulatory imbalance, early intravenous antibiotic therapy of sepsis, and appropriate opioid pain medication were the first steps taken promptly.

The Blood analysis- The following results were obtained from the patient's full blood count: White blood cells: 18,000 C/UL 18 x 109/L; neutrophils: 86%; lymphoid count: 10%; hemoglobin, which: 10 mg/dl 100g/L; hematocrit: 30.3% 0.303 proportion of 1; mean corpuscular volume: 79.5 fl; indicate corpuscular hemoglobin: 22.8 pg; platelets: 150 C/UL 150 x 109/L; total Bilirubin,: 4.3 µmol/L; lactate dehydrogenase: 408 U/L; erythrocyte sedimentation rate: 27 mm/hour; C-reactive protein: 48 mg/L; blood ureanitrogen: 2.8 mmol/L; creatinine that is: 50.8 µmol/L; potassium: 3.5 mmol/L; and sodium levels: 136.4 mmol/L.

Therapeutic intervention- The patient received intravenous ceftazidime 1 g three times a day, oral azathioprine 150 mg/day (3 mg/kg/day) upon admission for seven days, and oral levofloxacin 500 mg once a day as part of their treatment. The administration of these antibiotics persisted until the cultures of pus and blood were collected. The bullous lesions of the skin had broken down, exposing painful, degraded patches of skin that resembled burn victims' wounds. Following an early evaluation by the plastic surgery team and dermatologists, the proper dressings were used to protect the skin wounds and ease the patient's discomfort. He did not exhibit any signs of improvement one week into the treatment, and he was experiencing daily temperature spikes. P. aeruginosa was found in blood and pus cultures taken from the erosions. He began receiving intravenous colistin (9 mIU/day) in three separate doses for ten days based on sensitivity reports. Three days following the start of the antibiotics, the prednisolone dose was increased to 1.5 mg/kg/day since the disease remained threatening. After that, the erosions continued for ten days, but there was a faint hint of re-epithelialization. The patient proceeded to get a few additional blisters and his Nikolsky sign remained positive. He also experienced genital edema. We were unable to consider administering intravenous corticosteroids to him because the disease was still active and he was in sepsis with a persistent fever.

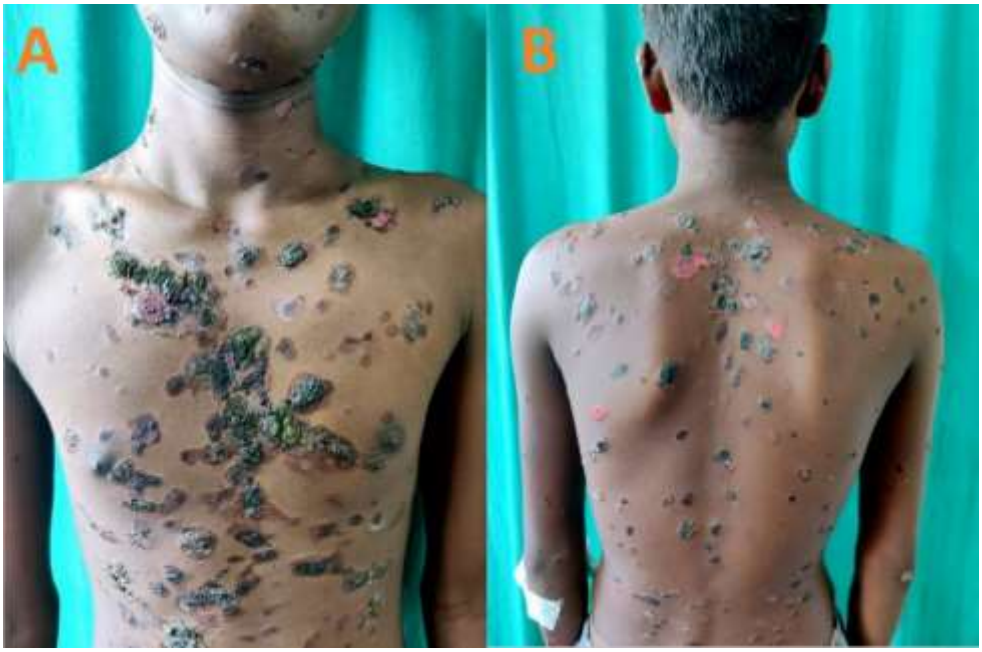

Figure 1: shows fluid fill vesicle and bulla over the trunk, chest, back.

Treatment was started with intravenous antibiotics, fluids, and ionotropic support. Because of poor GCS, tachycardia, and tachypnoea, Ultrasonography abdomen and pelvis were done which revealed an increased texture of the liver, minimal free fluid in the abdomen, and involuntary uterus. CT brain plain was done which revealed no obvious abnormality in brain parenchyma. MRI brain was done which showed no obvious abnormality in the brain parenchymal. Under all aseptic precautions, a lumbar puncture was performed and CSF fluid was sent for analysis. CSF Study show RBCS= 0-1 RBCs/HPF WBCS= 0-1 WBCS/HPF TLC- Approximately 7 cells/cumm DLC= Predominance of mononuclear cells (Lymphocytes). Repeat CSF done showed RBC - Nil/HPF WBC - 3-4 Cells/HPF DLC - Predominantly mononuclear cell (lymphocyte) TLC - approximately 320 cells/cumm. ECG and chest X-ray were done. An ophthalmologist's opinion was taken which showed no evidence of papilledema in both eyes. Neuropysiotherapist call was done and prom EXT for UE and le b/l x 10rep x 1 reps, ta, hamstring stretching, and positioning was given. A cardiorespiratory physiotherapist call was done and advised for manual chest vibration, passive end pressure, deep breathing exercise, neck isometrics and shoulder shrugging. The patient's condition started improving gradually. Patient vitals were monitored throughout the stay. Adequate hydration with IV and oral fluids was provided .in due course of the hospital stay patient's condition gradually improved and had no active complaints. Treatment was given in the hospital. Injectable meropenem 500 mg iv for 7 days, Injectable clindamycin 600 mg iv for 5 days, Injectable vancomycin 500mg iv for 3 days, Injectable linezolid 600 mg iv for 5 days, Injectable acyclovir 500mg iv for 14 days, Injectable doxy 100mg iv bd for 5 days, Injectable ciprofloxacin 400 mg iv for 7 days, Injectable colistin for 14 days, Injectable MPs 250mg iv od for 7 days, Injectable Emset 4mg iv /sos, Injectable lomoh 0.6ml/od for 31 days, Injectable mannitol 150mg iv for 14 days, Injectable pan 40mg iv for 7 days, Injectable Emset 4mg iv for 5 days, Injectable levipil 500mg iv for 5 days, tab folic acid 5mg for 5 days, tab sporolac 2 tabs for 5 days, NEB colistin IVM for 5 days, NEB mucomix for 5 days, for 5 days neb Doolin budecort, injection optineurin for 5 days, high protein diet, rt feed 100ml /2hrly, The patient's general condition improves. Hence being discharged. Treatment on discharge- tab Augmentin 625 mg bd for 5 days, tab levipil 500 mg bd for 1 month, tab folic acid 5 mg od for 1 month, Syp zincovit od for 1 month, tab ivabradine 2.5 mg od for 1 month, tab c2d3 od for 1 month, tab Ultracet bd for 7 days, cap rabesecd od for 10 days, advice - frequent body position change, limb physiotherapy, review in medicine OPD after 1 month.

Discussion:

The fact that pemphigus vulgaris rarely affects youngsters and often manifests between the ages of 50 and 70 adds to the case's significance. The majority of studies on pemphigus vulgaris that have been published in medical journals have focused on individuals between the ages of 40 and 80. It is possible to explain the girl's exceptional case of pemphigus vulgaris by pointing to the psychological impact of the terrible circumstances she has lived with in her community. Pemphigus vulgaris is a dangerous illness that can be fatal. Because of the potential for a subsequent infection, the loss of bodily fluids and the disruption of the epidermal barrier can be lethal. It takes an early and precise diagnosis to initiate treatment right away and avoid deadly consequences. Histopathological analysis is required to validate the clinical diagnosis.[4] The primary stay of care for pemphigus vulgaris is the use of systemic corticosteroids, either with or without suppression. Nonetheless, certain individuals continue to get intolerably high corticosteroid dosages. We are unable to reduce their high dosages of medications or provide treatment for those with severe illnesses.[5] Topical corticosteroids' place in PV is debatable.[6] An essential part of PV pathogenesis is the existence of circulating antibodies. Reducing the titers of circulating auto-antibodies with plasmapheresis has been associated with improved clinical outcomes.[7] This case report aims to illustrate the aggressive presentation of PV, particularly given that the disease started early and rarely manifests in children. Given the paucity of studies showing plasmapheresis's efficacy in causing partial or complete remission in pediatric patients, this case emphasizes the significance of plasmapheresis as a helpful intervention in PV patients who are not responding with standard therapy.

Conclusion

This case report aims to describe the aggressive presentation of pemphigus vulgaris, with a particular focus on the patient's early disease beginning. Given that there are few studies demonstrating the efficacy of plasmapheresis in causing partial or complete remission in younger patients, this case report emphasizes the significance of plasmapheresis as a helpful intervention in patients with pemphigus vulgaris who are not responding to conventional therapy. The disease rarely starts in growing up.

COMPETING INTERESTS :

As stated by the author, there are no conflicting interests.

FINANCIAL RESOURCE OF THE STUDY: Self

CONSENT :

Written consent from the patient had been obtained in accordance with university or international requirements.

References

1. Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K. Fitzpatrick’s dermatology in general medicine. 8. New York: McGraw-Hill; 2012.

2. Hertl M, Jedlickova H, Karpati S, et al. Pemphigus. S2 guideline for diagnosis and treatment – guided by the European Dermatology Forum (EDF) in cooperation with the European Academy of Dermatology and Venereology (EADV) J Eur Acad Dermatol Venereol. 2015;29(3):405–414. doi: 10.1111/jdv.12772. - DOI - PubMed

3. Chan W. Adjuvant therapies of pemphigus vulgaris: a review. XianggangPifu Ji Qingbing Xue. 2002;10(1):4–9.

4. Devaraj NK. A recurrent cutaneous eruption. BMJ Case Rep. 2019;12(2):bcr-2018-228355.

5. Bagchi S, Chatterjee R, Das S, Mahmud A. Dermatologic and oralmanifestations of pemphigus vulgaris: a case report with review. OralMaxillofac Pathol J. 2019;10:85–9.

6. Turner MS, Sutton D, Sauder DN. The use of plasmapheresis andimmunosuppression in the treatment of pemphigus vulgaris. J Am AcadDermatol. 2000;43(6):1058–64.

7. Devaraj NK, Aneesa AR, Abdul Hadi AM, Shaira N. Topical corticosteroids inclinical practice. Med J Malaysia. 2019;74(2):187–9.

8. Tan-Lim R, Bystryn JC. Effect of plasmapheresis therapy on circulating levelsof pemphigus antibodies. J Am Acad Dermatol. 1990;22(1):35–40.