Acta Scientific Pharmacology

Volume 1 Issue 8 August 2020

Case Report

A Case Report On Potentially Fatal Condition-Bullous Pemphigoid

K Mounika* and E Alekhya Goud

Department of Pharmacy Practice, Santhiram College of Pharmacy, Nandyal, Kurnool, Andhra Pradesh, India

***Corresponding Author:** K Mounika, Department of Pharmacy Practice, Santhiram College of Pharmacy, Nandyal, Kurnool, Andhra Pradesh, India.

Received: June 27, 2020

Published: July 30, 2020

Abstract

Bullous pemphigoid is a rare autoimmune disease which involves blisters formation between skin layers. It occurs due to production of autoantibodies against hemidesmosomal autoantigens BPAG1 and BPAG2. It mainly affects skin but rarely mouth, eye and genitals are also affected. The formation of blisters on the skin takes place due to malfunction in the immune system. The symptoms include severe itching, or erythematous skin with urticated and infiltrated plaques that are annular pattern. It is usually diagnosed by clinical, histopathological and immunological tests. The first line treatment is corticosteroids and the other treatment options are Immunosuppressant's and certain antibiotics to decrease infection. A 64 years male patient admitted with C/O fluid filled lesions over hands and legs since 6 days. On cutaneous examination, patient has multiple tense dome shaped bullae of size ranging from 1 × 1 cms to 2 × 3 cms over flexor aspects of both upper limbs. Multiple wheals of size ranging from 1 × 1cms to 4 × 5 cms present over dorsum of both hands, upper back and lateral aspects of both arms. On Histological examination, infiltration of eosinophils and lymphocytes were seen in upper dermis and bulla. Patient showed negative to Nikolsky sign and bulla spread sign. Patient was treated with Inj. Decadron OD, Tab. Taxim BD, Inj. Avil OD, Tab.Rantac BD, Tab. Fourts B OD, Fucibet cream BD, Tab. Bepozic OD.

Keywords: Bullous Pemphigoid; Autoimmune; Hemidesmosomal; Blisters; Nikolsky sign

Introduction

Bullous pemphigoid is a rare autoimmune disease which involves blisters formation between dermis and epidermis skin layers [1]. It occurs due to production of autoantibodies (Ig G) against hemidesmosomal autoantigens collagen XVII (BP 180 or BPAG2) and plakin family protein BP 230 (BPAG1) [2]. Bullous pemphigoid mainly affects skin but rarely mouth, eye and genitals are also affected. Bullous pemphigoid affects the people who are below 70 years and the average estimated rate is 7-14 cases per million per year. The main cause of Bullous pemphigoid is unknown. The formation of blisters on the skin takes place due to malfunction in the immune system. Normally body produces antibodies against viruses or bacteria or any other harmful substances. But in this case our body produces antibody against particular tissues in the body with unknown reason in the skin basement membrane. As a result blisters will form in the skin due to inflammatory activity of antibodies [3]. Bullous pemphigoid occurs with no reason but some precipitating factors include the usage of medications such as sulfasalazine, furosemide, penicillin, etanercept and exposure to UV light radiation [4]. The clinical manifestations of early stage of Bullous pemphigoid include severe itching or Urticarial lesions on the skin. The later stage symptoms include erythematous skin with urticated and infiltrated plaques that are annular pattern [5]. In Bullous pemphigoid binding of auto antibodies to hemidesmosal components BPAG 1 and BPAG 2 leads to activation of complements and inflammatory pathways leads to mast cell degradation

and accumulation of eosinophils and lymphocytes that results in subepidermal blistering with the release of proteinases. Bullous pemphigoid is usually diagnosed by clinical, histopathological and immunological tests [6]. There is no cure for bullous pemphigoid, but symptoms can be reduced. The first line treatment of bullous pemphigoid is corticosteroids which reduces inflammation, heals blisters and reduces itching. The other treatment options are immunosuppressants and certain antibiotics like azithromycin and tetracycline to decrease infection [7].

Case Report

A 64 years male patient admitted in Dermatology, Venerology, and Leprosy (DVL) ward with chief complaints of fluid filled lesions over hands and legs since 6 days. Patient was apparently normal 6 days back and developed small bullae initially over left forearm which gradually spread over to right forearm and over to thighs over a duration of 4 days. Lesions slowly progressed in size. He has taken treatment from local dermatologist but lesions have not subsided. Patient had no similar complaints in the past. Patient was not known case of Diabetes milletus, Tuberculosis, Hypertension.

On cutaneous examination, patient has multiple tense dome shaped bullae of size ranging from 1 × 1 cms to 2 × 3 cms present discretely bilaterally symmetrical over flexor aspects of both upper limbs. Similar type of lesions present directly over both anterior and medial thighs, posterior part of thigh and calf. Multiple wheals of size ranging from 1 × 1 cms to 4 × 5 cms present over dorsum of both hands, upper back and lateral aspects of both arms. Single crusted plaques present over flexor aspects of left forearm. On Histological examination, infiltration of eosinophils and lymphocytes were seen in upper dermis and bulla. Patient showed negative to Nikolsky sign and bulla spread sign.

Patient was treated with Inj.Decadron (Dexamethasone) 1cc IM once a day, Tab.Taxim (Cefotaxime) 200 mg twice a day, Inj. Avil (Pheneramine maleate) 2cc IM once a day, Tab. Rantac (Rabeprazole) 150 mg twice a day, Tab.Fourts B (Multivitamin) once a day, Fucibet cream (Betamethasone and fusidic acid) twice a day, Tab. Bepozic (Bepostatine) 10 mg once a day.

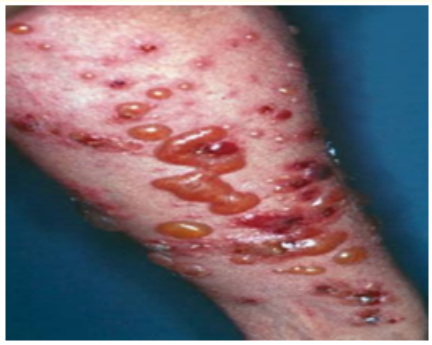

Figure 1: Showing bullae on forearm.

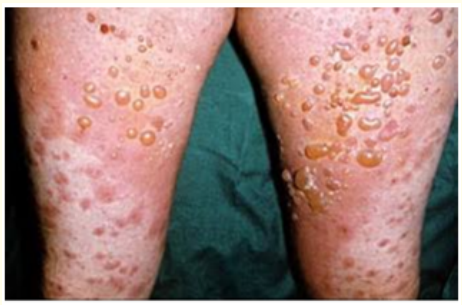

Figure 2: Showing bullae on both thighs.

Discussion

Bullous pemphigoid is a rare autoimmune disease which involves blisters formation between dermis and epidermis skin layers. The word Pemphigoid is derived from Greek words Pemphix (blister, bulla) and edois (form). It occurs due to production of autoantibodies BP 180, BP 230 against hemidesmosomal autoantigens collagen XVII (BP 180 or BPAG2) and plakin family protein BP 230 (BPAG1). Bullous pemphigoid is either acute or chronic which comes under Type-II hypersensitivity reaction. Bullous pemphigoid mainly affects skin but rarely mouth, eye and genitals are also affected. Elders are mostly affected to this disease compared to children and adults.

Bullous pemphigoid affects the people who are below 70 years and the average estimated rate is 7 - 14 cases per million per year [8].

The main cause of Bullous pemphigoid is unknown. The formation of blisters on the skin takes place due to malfunction in the immune system. Normally body produces antibodies against viruses or bacteria or any other harmful substances. But in this case our body produces antibody against particular tissues in the body with unknown reason in the skin basement membrane. As a result blisters will form in the skin due to inflammatory activity of antibodies. Bullous pemphigoid occurs with no reason but some precipitating factors include the usage of medications such as sulfasalazine, furosemide, penicillin, etanercept and exposure to UV light radiation [9].

The clinical manifestations of early stage of Bullous pemphigoid include severe itching or Urticarial lesions on the skin. The later stage symptoms include erythematous skin with urticated and infiltrated plaques that is annular pattern. The appearance of large blisters is the primary feature of Bullous pemphigoid. Blisters are tense which are clear and may contain blood. The patients with Bullous pemphigoid have reddish or darker skin around the blisters. Arms, upper thighs, abdomen and groin are commonly affected areas of Bullous pemphigoid. Sometimes blisters develop in mouth in only 10-30% of cases. If blisters develop on mucous membrane of eyes or mouth it is called as mucous membrane pemphigoid which is rare [10].

The pathogenesis of Bullous pemphigoid involves binding of autoantibodies to two hemidesmosomal components BPAG1 and BPAG2. This leads to activation of complements and inflammatory pathways leads to mast cell degradation and accumulation of eosinophils and lymphocytes that results in subepidermal blistering with release of proteinases such as metalloproteinase, elastin's etc [11].

Bullous pemphigoid is diagnosed based on clinical, histopathological and immunological criteria [12]:

- **Clinical:** Physician should ask the patient about medical history. Then he should ask the patient regarding time and type of onset of disease and further he should evaluate the symptoms of patient. To detect potential triggering factors physician should collect the information about previous drug intake, infections, neurological diseases and malignancies.
- **Histological:** It is the first step to confirm the disease. Biopsy should be performed by collecting the edge of active lesion and normal unaffected skin to find out level of blister. Subepidermal blistering and dermal infiltrate of eosinophils and lymphocytes are the findings of biopsy.
- **Immunological:** Immunological tests include Direct Immunofluorescence (DIF) and Enzyme Linked Immuno Sorbent Assay (ELISA). These tests are performed to detect the tissue bound autoantibodies. The levels of autoantibodies have been found to directly correlate with disease activity. While these tests are used for diagnosis if clinical presentation and biopsy are not sufficient.

There is no cure for bullous pemphigoid, but symptoms can be reduced. The first line treatment of bullous pemphigoid is corticosteroids which reduces inflammation and heals blisters and reduces itching. The other treatment options are immunosuppressants and certain antibiotics like azithromycin and tetracycline to decrease infection [13]. The Physician will choose the treatment depending on how much skin is involved. Drugs helps to control itching and blisters. The patients can remit within few months or years based on the severity of disease. The most commonly used medications are systemic and topical steroids like Prednisolone 0.5 mg once a day or Dexamethasone 0.75 mg once a day in the morning controls the disease within few weeks. Some clinical trials showed that clobetasol propionate cream is more beneficial when compared to prednisolone and dexamethasone. Dapsone, Methotrexate, high doses of intravenous Immunoglobins like rituximab, Omalizumab are also beneficial in the treatment of Bullous pemphigoid. Immunosuppressive agents like Azathioprine 0.5 to 2.5 mh/kg [14]. A combination of Tetracycline with Nicotinamide/Mycophenolate is beneficial. Some studies shows that Methotrexate was most effective in treating moderate to severe disease because it has less side effects [15].

Conclusion

Bullous pemphigoid is a rare autoimmune disease occurs in elderly individuals more than 60 years of age. Physician has to made appropriate diagnosis by using different laboratory tests in order to confirm the disease. Normally Bullous pemphigoid remits within few weeks but it has poor prognosis especially in older patients so, it should treat with appropriate care before turning into fatal condition.

Bibliography

1. Zillikens D., *et al*. "An autoimmune blistering disorders of the elderly". *Journal of Geriatric Dermatology* 4 (2016): 35-41.

2. Michael Kasperkewicz., *et al*. "Pemphigoid diseases: Pathogenesis, diagnosis and treatment". *Autoimmunity* 45.1 (2012): 55-70.

3. Lo Schiavo A., *et al*. "Bullous pemphigoid etiology, pathogenesis and inducing factors: facts and controversies". *Clinical Dermatology* 31 (2013): 391-399.

4. Jorge Parellada., *et al*. "A case on Bullous pemphigoid: A prevalent and potentially fatal condition". *Cureus* 10.4 (2018): 1-7.

5. Fuertes de Vega I., *et al*. "Bullous pemphigoid clinical practice guidelines". *Actas Dermosifiliogrficas* 105 (2014): 328-446.

6. Schmidt E and Zillikens D. "Modern diagnosis of Autoimmune blistering skin diseases". *Autoimmune Review* 10 (2010): 84-89.

7. Cortes B., *et al*. "Mortality of Bullous pemphigoid in Switzerland: a prospective study". *British Journal of Dermatology* 165 (2011): 368-374.

8. Joly P., *et al*. "Incidence and mortality of Bullous pemphigoid in France". *Journal Investigation on Dermatology* 132 (2012): 1998-2004.

9. Ujiie H., *et al*. "What's new in Bullous pemphigoid". *Journal of Dermatology* 37 (2010): 3194-204.

10. Cozzani E., *et al*. "A typical presentations of Bullos pemphigoid: Clinical and immunopathological aspects". *Autoimmune Review* 14 (2015): 438-445.

11. Michael Hertl. Autoimmune diseases of the skin: Pathogenesis, diagnosis and management, second revised and enlarged edition (2005): 84-95.

12. Viallant L., *et al*. "Evaluation of clinical criteria for diagnosis of Bullous pemphigoid". *Archives of Dermatology* 134 (1998): 1075-1080.

13. Mehta V and Balachandran C. "Localized flexural Bullous pemphigoid". *Indian Journal of Dermatology* 53 (2008): 157-158.

14. Ludwig RJ., *et al*. "Emerging treatments of Bullous pemphigoid diseases". *Trends in Molecular Medicine* 19 (2013): 501-512.

15. Yeh SW., *et al*. "Blistering disorders: Diagnosis and treatment". *Dermatology Therapy* 16 (2003): 214-223.