*International Journal of Health Sciences and Research*
*Vol.12; Issue: 10; October 2022*
*Website: www.ijhsr.org*
*Case report* *ISSN: 2249-9571*

# A Case of Bullous Pemphigoid - Potentially Fatal Condition

**Ragesh Gurumoorthi[1], Saranya Ramasamy[2], Sindhiya Kulandaisamy[3], Thrisha Sajith Kumar[4]**

[1,2,3,4]Department of Pharmacy Practice, Sri Ramachandra Faculty of Pharmacy, Sri Ramachandra Institute of Higher Education and Research (DU), Porur, Chennai

Corresponding Author: Ragesh Gurumoorthi

**DOI: https://doi.org/10.52403/ijhsr.20221019**

## ABSTRACT

Bullous pemphigoid is a rare autoimmune skin disorder characterized by enormous, fluid – filled blisters that appear on skin lower belly, upper thighs and armpits. It occurs when the body's immune system targets the tissue layer beneath the top layer of skin. The cause of this attack is not known. A 48 years old male patient received in the ward with the complaints of fluid filled lesions all over the body including oral cavity, history of rupture of lesions spontaneously to form raw areas, itching before the onset of lesions since 3 days. The incisional biopsy was done and the lesion was clinically confirmed to be a bullous pemphigoid. After the medical treatment with steroids, the patient was symptomatically better and was discharged with medications.

***Keywords:*** Autoimmune, blisters, lesions, erosions, paronychia

## INTRODUCTION

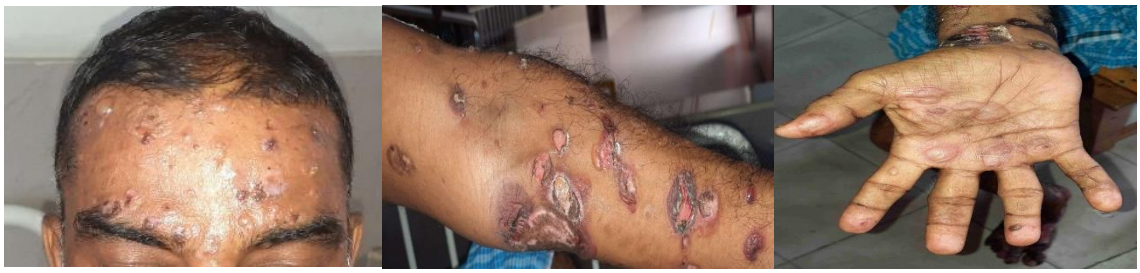
Pemphigoid is a blistering; painful disease most commonly found in the elderly and can seriously affect quality of life as well as life expectancy. BP is an autoimmune disorder caused by IgG antibodies that attack structural components of keratinocytic hemidesmosomal proteins, BP180 and BP230. [1] The antibodies and complement components trigger an inflammatory reaction at the dermo-epidermal junction, resulting in blisters formation. There is a tense blister appearance that may be widespread or localized on erythematous skin of limbs and trunks associated with BP. Oral and genital mucosa may exhibit bullae and/or erosions. In some instances, pruritus and erythema may occur prior to the emergence of bullae by weeks or months, and bullae may not be clinically apparent until weeks or months later. [2] Pemphigoid is primarily treated with systemic glucocorticoids. BP has shown efficacy with super potent topical corticosteroids, but they are unlikely to be practical for treating the whole body on a daily basis. [3] For cases of moderate to severe BP, rituximab and corticosteroids were suggested as first-line therapy; the regimen yielded a higher rate of complete remission when compared to treatment with rituximab alone. [4] The purpose of this case report is to highlight the different treatment options and urge referral to a dermatologist because of the potential severity of this condition.

## CASE REPORT

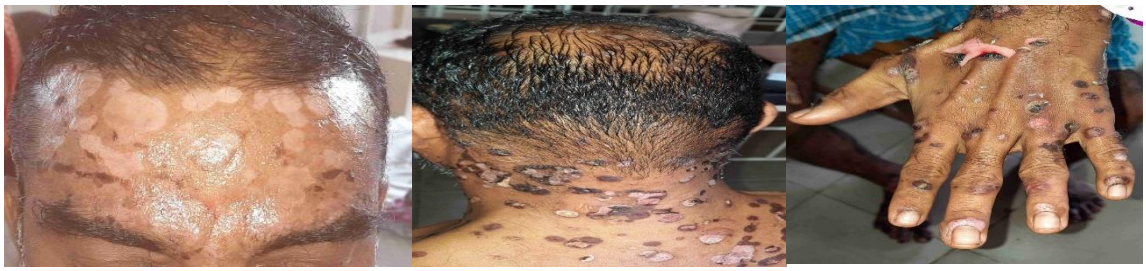
A 48 years old male patient was admitted with the chief complaints of fluid filled lesions all over the body for past 3 months, history of rupture of lesions spontaneously to form raw areas. He had history of itching before the onset of lesions, history of oral

lesions for past 3 days. Multiple tense vesicles with bullae of size varying from 0.5 X 0.5 cm to 4X4 cm were seen all over the body- face, scalp, trunk, bilateral upper limb, lower limb. Multiple crusted erosions of size 8*6 cm with necrotic slough were seen over right foot. Purulent discharge over bilateral feet and multiple raw areas occurred. Palm showed new flaccid blisters. Multiple erosions occurred on face, hands & leg. Scalp showed tense vesicles along with crusted erosions [Figure 1]. Soft palate erosions were observed in oral cavity. Bilateral feet showed large erosions over dorse with serious discharge. Erosions over palate, buccal mucosa and single intact build over left anterior pillar were observed in oral cavity. Bulla spread sign seemed to be positive. Past medical history reveals that the patients are a known case of diabetes mellitus since 20 years and on oral hypoglycemic agent (Tab. Metformin and Tab. Glimipride) which were stopped and insulin was started for better glycemic control.

Complete blood count results revealed low lymphocyte counts, high eosinophil counts, low RBC counts, and low albumin levels. Using aseptic precautions, incisional biopsy was done on intact vesicle on left arm and another sample taken from perilesional skin on left arm and the results showed features consistent with bullous pemphigoid. Direct immunofluorescence (DIF) test showed linear IgG and C3 deposition along the dermoepidermal junction. Mild perivascular infiltrate of lymphocytes admixed with few eosinophils was seen in upper dermis. Bulla roof showed sub epidermal separation plane with occasional eosinophils. Paronychia and anonychia were seen over finger and toe nails. Peripheral smear examination showed eosinophilia, Ultrasonography (USG) interpretation showed fatty liver grade 1 and Nikolsky's sign was negative.



**Figures 1: Patient before treatment – Erythematous tense bullae on face, hand and palm showed tense vesicles along with crusted erosions**



**Figures 2: Patient after treatment – There is no new bullae and he was symptomatically better**

Patient was started on Inj. Decadron 4mg (Dexamethasone) IV once daily and was stepped to 8 mg intravenously since the new lesions were continued to develop. Tenovate ointment (Clobetasol propionate) was applied all over the skin to reduce the redness, swelling and irritation. T. Niacin was started to reduce the bullous in the skin, it improves the acne by its anti-inflammatory action and by reducing the sebum. Saline soaks & KMnO4 baths were done. Banana leaf and liquid paraffin dressing for dependent areas of pressure at rest was done. Empirically Inj. Augmentin

1.2 g bd was started, which was switched to Inj. Linezolid 600 mg bd after pus culture and sensitivity test. K. Pneumonia is organism isolated from the pus sample and showed sensitivity to Linezolid. T. Xyzal 5mg (Levocetirizine) is an antihistamine was started for treating the itching, redness, swelling, rashes and discomfort in the skin. Development of new blisters over armpits, thighs and fresh erosions where bullae have been deroofed. Ulcers over bilateral dorsa of feet was healed well. There were single erythematous papulae over right lateral binder of tongue. Mucopain gel was given to reduce the pain and discomfort associated with mouth ulcers and irritations. Scalp showed scaly plaques over temporal region. Complaints of multiple new intact tense vesicles on right arm (post aspect), right and left palms. Skin examination showed multiple fresh bulla (tense) on back and T. Linox 600 mg was added. It is a new class of antibiotic called oxazolidinones used to treat bacterial infections in the skin especially for bullous skin. There were no new blisters on body surface area and complaints of itching reduced. HbA1C level was 9.5 and endocrinologist opinion was obtained. T. Glynase 5 mg and injection insulin were prescribed according to CBG levels. Absolute eosinophil counts were raised along with reduced absolute lymphocyte counts. General physician opinion was obtained and T. Azoran 25 mg was started in view of lymphopenia with a plan to monitor the hemogram and continue the drug. Patient completed 15 days of steroid therapy and he was symptomatically better [Figure 2] and was discharged with medications for 7 days and asked to come for the follow-up. Patient instructed to use KMnO4 soaks in morning and saline soaks at night followed by T. Bact ointment. He counselled to avoid walking in barefoot and insisted to report immediately if any episodes of hypoglycemia/hyperglycaemia or if skin lesion worsens.

## DISCUSSION

It is a type of blistering disorder frequently runs a chronic course of therapy. Initially it forms pruritic and urticaria like lesions with later forms a fluid filled bullae. Usually bullous occurs in lower leg, trunk, flexor extremities and axillary area. The blisters heal without scarring. [5,6] The clinical manifestations of bullous pemphigoid may resemble like a variety of other skin conditions hence, it is vital to confirm the condition with clinical, histological data and DIF findings. [7] As per the S2k guideline [8] the treatment proposed for bullous pemphigoid is based on the severity. For mild to moderate condition, only topical clobetasol propionate was recommended and for severe cases topical clobetasol propionate is recommended along with systemic treatment. But in case of elder patients, the practicability for using topical ointment in large skin is not feasible. This guideline also recommended the chlorhexidine, triclosan 1 % and octenidine is some topical antiseptics for bacterial superinfection and erosions. In this case, the patients affected with > 30 % body surface area and the patient categorised as severe. According to the guideline (S2k) initially this patient was treated with topical (Oint. Tenovate) and systemic corticosteroids (Inj. Decadron 8 mg) and symptomatic therapy was given to reduce the symptoms. Similarly, a case report from S. M. Biradar *et al*. reported that the treatment with systemic and topical corticosteroids forms the mainstay of treatment along with other adjuvant drugs. Also the use of prednisolone has proven its efficacy in the extensive disease state of bullous pemphigoid and improved the patient's quality of life. [9] In contrast, a case of a 65-year-old female with bullous pemphigoid formed resistant to corticosteroids therapy and suggested cyclophosphamide is the better alternative and showed immediate improvement in the symptoms. [10] An another case study reported that the reslizumab had showed a significant response in improvement of symptoms in bullous pemphigoid patient.

But recommended that further studies needed to confirm this result. Reslizumab is an anti-interleukin-5 mono-clonal antibody decrease the level of eosinophil in blood, bone marrow and tissue. Anti-IL-5 antibodies have shown efficacy and safety in patients with allergic and hypereosinophilic syndromes. The effects on patients with bullous pemphigoid, however, remain unclear. [11] Several studies have recommended and the use of steroid-sparing medicines and plasma exchanges, although more evidence is needed before their use can be considered as routine.

## CONCLUSION

The therapy can vary with the severity of disease that erratically affects the body tissues. Clinical examinations should be done regularly until the patient achieves complete clinical remission or is stopped from receiving treatment. By using systemic corticosteroids and symptomatic treatment, the main goals of therapy are to control the disease progression and to reduce the inflammation. The use of immunosuppressive agents may be helpful in preventing relapses. In order to monitor disease severity and remission, continuous follow-up is imperative. As compared to systemic steroids, topical steroids are more efficient and less likely to cause side effects. Correct identification of the condition, coupled with the appropriate maintenance therapy, helps in resolving the condition.

**Conflict of Interest:** None

## REFERENCES


1. Lo Schiavo A, Ruocco E, Brancaccio G, Caccavale S, Ruocco V, Wolf R. Bullous pemphigoid etiology, pathogenesis, and inducing factors: facts and controversies. Clin Dermatol 2013; 31(3):391–399.
2. S. M. Biradar1, S. Dhanavidya1, P. Kavya1, T. Keerthi1, N. Sunanda1, S. C. Marapur et al. Bullous pemphigoid a rare autoimmune disease: a case report. International Journal of Medicine and Medical Research 2018; 4 (2):20-23
3. Joly P, Roujeau JC, Benichou J. A comparison of oral and topical corticosteroids in patients with bullous pemphigoid. N Engl J Med 2002; *346 (5):321–7.*
4. Cho YT, Chu CY, Wang LF. First-line combination therapy with rituximab and corticosteroids provides a high complete remission rate in moderate-to-severe bullous pemphigoid. Br J Dermatol 2015; 173 *(1):302–4.*
5. Lo Schiavo A, Ruocco E, Brancaccio G, Caccavale S, Ruocco V, Wolf R. Bullous pemphigoid etiology, pathogenesis, and inducing factors: facts and controversies. Clin Dermatol 2013; 31:391–399.
6. Fuertes de Vega I, Iranzo-Fernandez P, Mascaro-Galy JM. Bullous pemphigoid clinical practice guidelines. Actas Dermosifiliogr 2014; 105:328–346.
7. Vaillant L, Bernard P, Joly P. Evaluation of clinical criteria for diagnosis of bullous pemphigoid. Arch Dermatol 1998; 134:1075–1080.
8. Enno Schmidt, Michael Sticherling, Miklos Sardy, Rudiger Eming, Matthias Goebeler, Michael Hertl, et al. S2k guidelines for the treatment of pemphigus vulgaris/foliaceus and bullous pemphigoid. J Dtsch Dermatol Ges 2015; 13(8):833-44.
9. Biradar SM, Dhanavidya S, Kavya P, Keerthi T, Sunanda N, Marapur SC. et al. Bullous pemphigoid a rare autoimmune disease: a case report. International Journal of Medicine and Medical Research 2019; 4(2), 20–23.
10. Sah M, Rozenman D, Cohen E, Ziv M. Bullous pemphigoid resistant to corticosteroids. Harefuah 2016; 155(10):593-595.
11. Rhyou HI, Han SH, Nam YH. Successful induction treatment of bullous pemphigoid using reslizumab: a case report. Allergy Asthma Clin Immunol 2021; 17(1), 117-119.


How to cite this article: Ragesh Gurumoorthi, Saranya Ramasamy, Sindhiya Kulandaisamy et al. A case of bullous pemphigoid – potentially fatal condition. *Int J Health Sci Res.* 2022; 12(10):150-153.
DOI: *https://doi.org/10.52403/ijhsr.20221019*

******