A Case Report on Idiopathic Severe Bullous Pemphigoid


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ABSTRACT

Bullous Pemphigoid (BP) is an autoimmune disorder which exploits the immune system and affecting sub-epidermal region of skin causing mild itching to infection and blistering of sub-epidermal region. Clinical presentations are pruritis, urticarial, papular lesions. These later evolve to bullae in weeks to months and are typically present in the axillae, on the flexor surface of the forearms, medial thighs, trunk, and abdomen. The treatment for bullous pemphigoid is systemic corticosteroids, topical steroids in combination with nicotinamide plus tetracycline, minocycline or doxycycline have shown success in multiple cases. Immunosuppressive therapy is used when steroids do not control the disease or if patients have contraindications for systemic corticosteroid treatments. Patient was brought to emergency department with chief complaints itching, redness, bullous lesion all over the body for 2 months. Oral complaints including solitary erosion seen on right side of buccal mucosa, pharyngeal erythema. Laboratory investigations were carried out. Patient was diagnosed based on physical examination. Patient was provided with adequate treatment and counseling.

Keywords: Bullous Pemphigoid, corticosteroid, Autoimmune disorder, sub-epidermal region.

INTRODUCTION:

Bullous Pemphigoid (BP) is an autoimmune disorder which exploits the immune system and affecting sub-epidermal region of skin causing mild itching to infection and blistering of sub-epidermal region. Bullous Pemphigoid is a rare dermatological disorder which can be seen in all age groups but mainly affecting elderly patients.¹

Etiology:

Many cases of bullous pemphigoid are due to autoantibodies against proteins arranged at the dermal-epidermal junction. Drug-induced bullous pemphigoid occurs up to 3 months after medication initiation and is usually noted during a younger to older subset of patients. Drugs which trigger this reaction are diuretics such as furosemide and spironolactone, NSAIDs, amoxicillin, PD-1/PD-L1 inhibitors, gliptins, and TNF-alpha inhibitors.²

Pathophysiology:

There are 2 main components to the pathophysiology of BP: immunologic and inflammatory. The immunologic elements comprise autoantibodies against 2 parts of the basal keratinocyte hemidesmosomal proteins BP antigen 230 (BPAG1) and BP antigen 180 (BPAG2 or type XVII collagen)³

These antigens play an essential part in the adhesion complexes that promote epithelial-stromal adhesion. When autoantibodies bind to their target antigen, the inflammatory component follows which then activates complement and mast cells. This causes neutrophils and eosinophils to release a variety of inflammatory cells resulting in the release of proteolytic enzymes that damage the dermal-epidermal junction.⁴

Clinical Presentation:

In the prodromal phase, patients may experience moderate-to-severe pruritus alone or associated with urticarial, papular lesions.⁵ This later evolves to bullae in weeks to months and are typically present in the axillae, on the flexor surface of the forearms, medial thighs, trunk, and abdomen. Approximately 20% of patients will have neither bullae nor erosions at time of presentation.⁶

Constitutional symptoms are uncommon, except in widespread, severe disease. The bullous phase characteristically presents with vesicles and bullae on a background of normal or erythematous skin. The blisters are tense, up to 1 to 4 centimeters in diameter, and sometimes hemorrhagic. They typically contain clear fluid and should persist for several days before leaving erosions and crusts.
Unlike pemphigus vulgaris, the Nikolsky sign is negative in typical cases of bullous pemphigoid. 

**Standard Treatment:**

The treatment for bullous pemphigoid is systemic corticosteroids, but treatment ultimately depends on comorbidities and extent of disease. For localized disease, less than 20% body surface area in an elderly patient, super potent topical steroids such as clobetasol may be used. 

Topical steroids in combination with nicotinamide plus tetracycline, minocycline or doxycycline have shown success in multiple cases. 

A systemic prednisolone at a dose of 0.5-1.0mg/kg per day is recommended. This dose of systemic corticosteroids controls the disease within approximately fortnight and should be slowly tapered over six to nine months or longer. This treatment regimen is limited by patient age, comorbidities and side effects. 

Immunosuppressive therapy is used when steroids do not control the disease or if patients have contraindications for systemic corticosteroid treatments. Alternative agents include azathioprine, mycophenolate mofetil, methotrexate, chlorambucil, and cyclophosphamide. If all other treatments fail, IVIG, anti-CD20 (rituximab) or omalizumab can be used for treatment-resistant cases. 

**CASE REPORT:**

A 72 year old male patient who is hypertensive was brought to the emergency department of the hospital with chief complaint of itching, redness, bullous lesion all over the body for 2 months.

Oral complaints including solitary erosion seen on right side of buccal mucosa, pharyngeal erythema.

These lesions started two months back. Firstly, these lesions were seen on forehead which gradually developed into blisters and are progressive in nature. Further progressing to face, upper limb, lower limb, trunk, this eroded in 3 days and crusted in a week.

**INVESTIGATIONS:**

Renal function tests were reported normal. 

Liver function tests were reported normal.

**HAEMATOLOGICAL TEST:**

- **White Blood Cells:** 13000 cells/cumm (Normal range: 4000-11000 cells/cumm)
- **Lymphocytes:** 15.7% (Normal range: 20%-40%)
- **Haemoglobin value:** 10.3 g/dL (Normal range: 11-17 g/dL)
- **Mean Corpuscular Volume:** 78.3 fl (81-101 fl)
- **Mean Corpuscular haemoglobin:** 25.4 picogram(pg) (Normal value: 27-32 pg)
- **Hematocrit:** 31.7% (Normal value: 40.7-50.3%)

**COMPLETE URINE EXAMINATION:**

All parameters are normal except for the traces of albumins were seen in urine examination.

**DIFFERENTIAL DIAGNOSIS:**

Physical examination like pruritus, papular lesion, blister all over the body (forearms, face, upper limbs, lower limbs, abdomen). Nikolsky sign is negative. Based on physical examination of blisters, it had been diagnosed as Bullous Pemphigoid (BP).

**TREATMENT:**

Upon admission, the patient was provided with intravenous dexamethasone 4 mg twice a day, intravenous pheniramine 2.25 mg twice a day, intravenous pantoprazole 40 mg once a day, tablet doxycycline 100 mg once a day, tablet linezolid 600 mg twice a day, tablet nicotinamide 250 mg twice a day, tablet calcium with vitamin D3 500 mg once a day, tablet ferrous Ascorbate + folic acid once a day, tablet fluconazole 150 mg once a day, syrup Lactulose 15 ml once a day, KMnO4 compression thrice a day, Mupirocin Ointment, Clobetasol (0.005% w/w)+ Fusidic acid (2% w/w). The patient was moved to isolation ward as a precautionary measure.

**OUTCOME AND FOLLOW-UP:**

The patient was kept under observation for 8 days. The patient had shown signs of improvement upon treatment. The patient should be closely monitored and counselled regarding the disease and prognosis of disease

**DISCUSSION:**

Bullous pemphigoid is chronic blistering of skin, it starts with mild itching and prognosis may lead to blistering and infections. BP is an autoimmune condition.

This patient came with the complaints of itching, redness, solitary erosion seen on right side of buccal mucosa, bullous lesion all over the body for 2 months. And was provided with corticosteroids, anti-histamine, antibiotics, antifungal, multivitamins systemic therapy,

Tropically the patient was provided with potassium permanganate compression which is gold standard in management of Bullous pemphigoid along with potassium permanganate topical antibiotic, topical glucocorticoids was provided.

During the discharge the patient and attenders were counselled in layman’s language regarding the disease conditions, prognosis of disease, application of topical medications, adherence to the medications, non-pharmacological remedies and was advised to review back to the dermatologist as directed.

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**REFERENCES:**

