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Bullous Pemphigoid- A Rare Case report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Introduction: The most severe autoimmune subepidermal blistering condition of the skin and mucous membranes is bullous pemphigoid (BP). In Europe, it is estimated to affect 1 in every 4,000 people. Currently incidence range between 2-22/1,000,000 worldwide. It primarily affects the elderly and is diagnosed using clinical, histologic, and immunologic criteria. Clinically, it appears as diffuse eczematous, pruritic, urticaria-like lesions with the later emergence of tense bullae or blistering lesions filled with clear fluid.
Case Presentation: Here, we report a case of a 50- years old female patient with a complaint of itchy lesions with wounds all over the body present with an 8-month of history. A subepidermal blister with eosinophils and neutrophils infiltration was discovered on histopathological evaluation. Salt-split indirect immunofluorescence revealed linear deposition of IgG at the dermo-epidermal junction. On further investigation, using diagnostic and Interventional aids a final diagnosis of Bullous pemphigoid.

Keywords: Bullous pemphigoid (BP); subepidermal blister; eosinophils; neutrophils infiltration.

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1. INTRODUCTION

Bullous Pemphigoid (BP) is an uncommon skin condition characterized by massive, fluid-filled blisters. They appear on flexible skin areas including the lower abdomen, upper thighs, and armpits. Clinical, histologic, and immunologic criteria are used to diagnose it in the elderly. Anti-inflammatory drugs, pharmaceuticals that inhibit antibody production and treatments that enhance antibody elimination are among the therapy options available for this illness. The purpose of this case report is to describe a perfect illustration of this ailment, raise knowledge of different treatment choices, and urge for referral to a dermatologist due to its potential severity [1].

2. EPIDEMIOLOGY AND ETIOLOGY

According to published literature, Bullous Pemphigoid (BP) is the most common Autoimmune Sub-epidermal Blistering disorder of the skin and mucous membranes. In Europe, it is estimated to affect 1 in every 4,000 people. Currently incidence range between 2-22/1,000,000 worldwide [2].

3. CASE PRESENTATION

Patient information: A 50-year-old female patient, who was alright 8 months back when she developed a fluid-filled pea-sized lesion on her arm progressing gradually to the whole body and others compliant like weight loss, itching, burning sensation, fever, joint pain, raw area at trauma prone area is present. Her medical history revealed that he was a known type II diabetes mellitus patient and not taking any medication for that. Salt-split indirect immunofluorescence revealed linear deposition of IgG at the dermo-epidermal junction. After all, examinations were completed, the patient was diagnosed with Bullous Pemphigoid. The Patient was taking Tablet Prednisone 60 mg continuously on daily basis and she responded to the treatment well with a decrease in the number of bullae as well as reduced erythema and pruritus.

Clinical findings: On physical examination multiple vesicles and bullace on erythematous base with multiple raw areas on bilateral upper limb, lower limb, trunk, back and face nikolsky sing was negative but bulla sign is positive.

Pathological Finding: For diagnosis of this case, the skin biopsy was done and the sample was taken from hand it revealed that white tissue piece measuring less than 0.5x0.5x0.2 cm as well as the section was done from given tissue piece which shows histopathological features suggestive that Bullous Pemphigoid.

Differential Diagnosis: Some types of acquired bullous epidermolysis and anti-P200 pemphigoid are the main differential diagnosis. Mucous membrane pemphigoid may resemble BP with mucosal involvement, even though mucosal involvement is uncommon in BP.

4. MANAGEMENT AND TREATMENT

Most of the countries in the world recommended a Systemic Corticosteroid (CS) (Tablet Prednisone: 0.5-1 mg/kg/day) is to be as the standard line of treatment. In European countries, they make standards for first-line treatment is to apply ointment of a Super Potent Topical Corticosteroid on the whole-body surface area and only limited damaged areas will be treated, with a maintenance or tapering plan in place if necessary. Immunosuppressive medications (Methotrexate, Mycophenolate Mofetil) are commonly used in individuals with resistant BP and multiple relapses. The patient how had allergic to immunosuppressive medication or in poor health conditions for that Doxycycline can be administered. Currently, novel treatments are available including Rituximab and Omalizumab, have been explored, but their indications are yet unknown.

5. DISCUSSION

Pemphigoid words come from the Greek words “Pemphix” which means bulla, blister and “eidos” which means form. With an annual frequency of 0.2 to 3 cases per 100,000 people, BP is the most prevalent of blistering illnesses. The disease has no gender bias, but it is more usually observed in people over the age of 75. Young folks and children may be impacted as well, but far less commonly [3,4].

BP is classified as an autoimmune disease when IgG antibodies are present against structural components of the Keratinolytic Hemidesmosome proteins BP180 i.e. Bullous Pemphigoid antigen 2 and BP230 i.e. Bullous Pemphigoid antigen 1. At the dermo-epidermal interface both antibody and complement, components deposit along the basement membrane, resulting in a severe inflammatory reaction with blistering blisters. Antibody levels

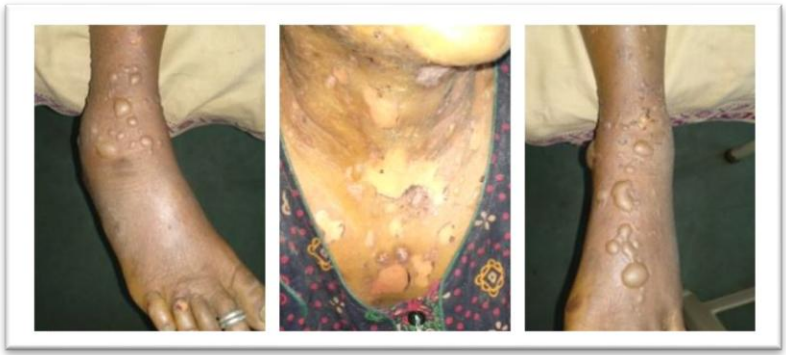

Fig. 1. Bullous Pemphigoid (BP)

Table 1. Treatment

Sr. No.	Name of Drug	Dose	Route	Frequency	Drug Action
1.	Inj. Insulin Mixtard	26 Unit	SC	BD	Helping move sugar from the blood into other body tissues
2.	Tab. Defcort	12 Mg	Oral	OD	Reducing the inflammation
3.	Tab. Cyclophosphamide	50 Mg	Oral	BD	Slowing or stopping cell growth
4.	Tab. Nicoglow	250 Mg	Oral	OD	Maintaining a general good health
5.	Tab. Orofer XT		Oral	OD	Elemental Iron
6.	Tab. Shelcal	500 Mg	Oral	BD	Maintaining healthy bones
7.	Tab. Pan	40 Mg	Oral	OD	Antacid
8.	Tab. Atarax	25 Mg	Oral	BD	Reduces activity in the central nervous system
9.	Tab. Dailyshine	60 K IU	Oral	Once a week	Fat-soluble vitamins synthesized in the human body from cholesterol

Table 2. Diagnostic Evaluation

Investigation	Patient Value
Blood Investigation	
Hemoglobin	11.3 gm/dL
Total RBC Count	4.21 cells/mcL
Total WBC Count	13100 per microliter of blood
Haematocrit	34.5%
Mean Corpuscular Hemoglobin Concentration	32.8 g/dL
Mean Corpuscular Volume	82.1fl
Mean Corpuscular Hemoglobin	26.9 picograms
Total Platelet Count	2.87 per microliter of blood
Monocytes	04
Granulocytes	65
Lymphocytes	30
Red Cell Distribution Width (RDW)	13.3
Eosinophils	01
Basophils	00
Urine Examination	
Urine Albumin	Present
Urine Sugar	Present
Epithelial Cell	Absent
Pus Cell	Present

will be proportional to disease activity. Tight sub-epidermal blisters are caused by a localized separation of the epidermis and dermis [4,5].

The formation of tense bullae that are often filled with clear fluid, followed by the development of extensive eczematous, pruritic, urticarial-like lesions, distinguishes BP. The trunk, flexor compartments of the extremities, and the axillary area are the most common sites for lesions. Blisters are normally healed without leaving scars and develop in a waxing and waning pattern [6].

BP can arise in atypical forms. As localized pemphigoid, most of these are found in the lower limbs, genital region, and trauma regions. It can be extended much more generically. The Dyshidrosiform Bullous Pemphigoid is a rare variant of BP which affects the palms and foot, erythrodermic pemphigoid, nodular pemphigoid, and lichen planus pemphigoid is some of the other types of pemphigoids. Mucosal Involvement is rare and usually affects only the mouth if it occurs.

The clinical features of BP may be reminiscent of those of many other skin conditions. By using clinical, histologic, and Immunologic Direct Immunofluorescence (DIF) findings will help critical to distinguish Bullous Pemphigoid from other blistering conditions like Dermatitis Herpetiformis, Epidermolysis Bullosa Acquisita, Pemphigus Vulgaris, and others [7]. Misdiagnosis can lead to inefficient treatment and, as a result, poor results for persons with potentially treatable diseases.

Clinical, histologic, and immunologic criteria are used to identify BP. A group of French writers advocated that the clinical predictors of BP are the absence of atrophic scars, restricted neck or head involvement, absence of mucosal involvement and age >70 years old as clinical predictors of BP. The presence of 3 of these 4 Bullous Pemphigoid criteria resulted in a sensitivity of 90 percent, specificity of 83 percent, and positive predictive value of 95 percent [8].

Two punch biopsies should be performed to confirm the diagnosis through histology. One skin biopsy should be collected from the margin of an unbroken blister preserved in formaldehyde for Haematoxylin and Eosin (H&E) staining and it revealed that A frequent finding is a subepidermal blister with a perivascular inflammatory infiltrate and eosinophils and A common sign of BP is the presence of Eosinophilic Spongiosis. A second skin biopsy from normal Perilesional tissue should be taken and placed in saline or Michel's solution medium for Immunologic Direct Immunofluorescence {DIF}. The most common DIF findings are linear IgG and C3 deposits along the basement membrane. Immunologic Direct Immunofluorescence {DIF} is deemed the gold standard for diagnosis [2,9].

The antigenic specificity of antibodies can be determined using a variety of immunologic techniques. Bullous pemphigoid antibodies can be tested using an enzyme-linked immunosorbent assay (ELISA). Anti-BP180 levels have been discovered to have a direct association with disease activity [3,8].

Recent French research of 502 individuals found that BP is a potentially lethal condition, with a 1-year mortality rate of thirty-eight percent [6]. A study of 115 individuals with Bullous Pemphigoid discovered probabilities of death. Individuals diagnosed with BP had a threefold greater mortality rate than age & sex-matched patients in the general community [7]. According to the finding above, it is critical to correctly diagnose and treat BP in this patient population to improve outcomes.

Three types of medicines can be used to treat BP [2,4]. The first category includes anti-inflammatory drugs including topical steroids and Sulfonamides as well as antibiotics with anti-inflammatory properties like tetracycline. Inhibitors of antibody production include Systemic Steroids, Azathioprine, Methotrexate, Mycophenolate, Cyclosporin and Rituximab. Lastly, therapies like Plasmapheresis and Intravenous Immunoglobulin G (IVIG) can help with antibody clearance [10].

6. CONCLUSION

Bullous Pemphigoid is a potentially threatening condition with a high prevalence that can be treated with the proper level of care. Given the possible complexity of the disease, this case study intends to raise disease awareness, highlight various therapies, and promote consultation with a Dermatologist.

FOLLOW UP

Physician advice follows up for Pneumocystis carinii pneumonia prophylaxis as well as on discharge Tablet Prednisolone 60-40-20

mg/dose for 1 month on a tapering regimen with Tablet Sulfamethoxazole/ Trimethoprim 160 mg daily was given to the patient to prevent bacterial infection. A dermatologist was recommended to the patient to discuss adding a Steroid- Sparing drug like Methotrexate / Azathioprine.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

CONSENT

While preparing case reports for publication parental informed consent has been taken from the patient.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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