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BULLOUS PEMPHIGOID: A CASE REPORT

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Abstract:

Bullous Pemphigoid (BP) is an acquired autoimmune disorder characterized by chronic blistering of the sub-epidermal skin. It occurs most frequently in elderly patients and has a rising incidence. The typical clinical features of BP are large, tense bullae preceded by urticarial plaques and severe pruritus. The exact etiological cause of Bullous Pemphigoid is not known. Diverse factors have been reported to play a role in triggering Bullous Pemphigoid and include mechanical trauma, drugs like non-steroidal anti-inflammatory agents, DPP-4 inhibitors, captopril, penicillamine, antibiotics, furosemide and penicillin's, and physical traumas like burns from radiation, sun or heat. The pathogenesis of BP is characterized by tissue bound and circulating IgG autoantibodies against two components of the hemidesmosome of stratified epithelia, BP 230 KD (BPAg1) and BP 180 KD (BPAg2, COL17). A case report of a 54 years old, female patient diagnosed with Bullous Pemphigoid on the basis of clinical findings, histopathology and direct immunofluorescence is presented. Therapy was initiated with Antibiotics, Corticosteroids, topical creams, and calcium supplements.

Key words: *Bullous Pemphigoid (BP), autoimmune, sub-epidermal bulla, hemidesmosome, elderly, corticosteroids.*

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INTRODUCTION:

Bullous Pemphigoid (BP) is an idiopathic auto-immune disease characterized by multiple tense blistering of the skin. It majorly affects the geriatric population in the fifth to seventh decade of life, with the average age of onset being 65 years[1]. It is characterized by the formation of large tense blisters and immunologic finding of C3 and IgG at the basement membrane zone.

About 1 in 10,000 people are affected by this disease in the UK each year. In India, the incidence of sub-epidermal autoimmune disease is comparatively low[2]. Including India, BP in childhood has been reported from various countries. There is no known ethnic, racial, or sexual predilection.

Multiple factors have been reported to play a vital role in triggering BP and include mechanical trauma, drugs like furosemide, penicillin drugs, and physical traumas like burns from radiation, sun or heat. The diagnosis must be established by a skin biopsy. There is an equal occurrence of BP in both genders, and there are no known racial biases.

The diagnosis must be established by a skin biopsy. Immune-fluorescence biopsy may also be desirable. Therapy is focused solely on reprieve of symptoms and avoidance of infection[3]. Therapy involves systemic prednisone alone or in combination of a steroid-sparing agent such as azathioprine, mycophenolate mofetil, rituximab, and tetracycline. Patients with mild symptoms may require topical corticosteroids. The patients with severe disease who can't tolerate the treatment with prednisone may use methotrexate[3,4].

CASE REPORT:

A 54-year-old female presented with blister formation which had appeared 15 days before admission. Initially, the blisters developed on the arms and then spread into other parts of the body, also associated with itching and burning sensation. Blisters were itchy, tense, of varying size ranging from 2-6cm in diameter, which later broke down to become erosions and ulcers.

She had no similar complaints in the past. She has no history of DM, hypertension and pulmonary tuberculosis. She had no previous hospital admission, surgery or history of blood transfusion. There were no aggravating factors. However, there was a history of ingestion of ibuprofen and paracetamol.

On physical examination she was in painful distress, anxious, ill looking, afebrile, dehydrated and showed pale pigmentation of skin. There was no evidence of peripheral lymphadenopathy and pedal enema was absent.

Her integumentary system revealed widespread blistering breaking down to erosions and ulcers all over her body measuring from 2cm to 6cm. Bullae were tense. Her neurological, chest and abdominal findings were normal. Her pulse rate was 78 beats per minute, respiratory rate was 21 cycles per minute and blood pressure was 120/80 mm Hg, Both pulse rate and blood pressure were within normal limits.

Investigations:

Haemoglobin - 12.6g%, Total Count -13730/cu.mm [N-83 %, L- 10 %, E-15 %], PCV- 36.8%, Blood urea - 20 mEq/L, serum creatinine-0.8mg/dl, Serum calcium - 7.8mg/dl, Total Protein-6.2g/dl, serum albumin - 3.3g/dl, serum globulin - 2.9g/dl, sodium concentration-137 mEq/L, serum potassium - 4.4 mEq/L, Random blood sugar- 95mg/dl, HIV 1& 2-Negative.

Histopathology showed sub-epidermal and a light upper dermal lymphocytic inflammation associated with scattered eosinophils. Direct Immunofluorescence (DIF) showed linear IgG and complement deposits in the base of the blister, which provided the evidence of Bullous Pemphigoid.

In this case the patient was treated with following medications:

- I.V. normal saline - To correct dehydration.
- Inj. Avil (2cc) - Pheniramine maleate used to treat itching condition.
- T. Doxycycline (500 mg, BD).
- T. Prednisolone (50mg, OD) - steroid medication used to treat itching.
- Inj. Pantoprazole (40mg) - used as an anti ulcerative.
- Tab. Cetrizine (10mg) - anti-histamine used to relieve symptoms of itching.
- Tab. Calcium (500mg) - Calcium supplementation.
- White soft paraffin - emollient that moisturizes skin.
- Soframycin silver nitrate Ointment - antibiotic used to prevent or treat skin infection.

The patient improved during her hospitalisation and was discharged after 11 days after tapering prednisolone regime. Topical soframycin silver nitrate ointment and doxycycline 500mg BD were given as discharge medications. She had two follow-up visits which showed most of the lesions healed. Patient's caregivers (daughter-in-law) complained of the distance that she had to make to come to the hospital for follow-up and requested

for referral to a centre nearer to their place. She was then referred to a hospital nearer to her home.

DISCUSSION:

Bullous Pemphigoid (BP) is an idiopathic disorder. It is life threatening immunopathologic dermatologic disease occurring in people at the age of 50-80 years[5]. There are a large number of medications that have been reported to induce BP or BP-like eruptions[6-9].

Table 1: Medications reported to induce Bullous Pemphigoid

Actin-mycin D, Ampicillin, Amoxicillin, Arsenic, Azapropazone, Anti-influenza vaccine, Captopril, Chloroquine, Clonidine, Dactinomycin, Enalapril, Furosemide, Flupenthixol, Gold thyosulfate, Ibuprofen, Interleukin-2, Mefenamic acid, Methyldopa, Nadolol, Omeprazole, Penicillamine, Penicillin, Phenacetin, Placental extracts, Potassium iodide, Practolol, Psoralens with UVA, Risperidone, Salicylazosulfapyridine, Sulfonamide, Tetanus toxoid, Tiopronin, Tiobutarit, Tolbutamide

In BP patients, there is an auto-reactive response of T cells and B cells to BP antigen-1 (BPAG1/230 kDa) and BPAG2 antigens (180 kDa)[10]. Incidence of Bullous Pemphigoid among the dermatology attendees was found to be 1.8%. A considerable proportion of patients with BP have been younger than 40 years of age. Both sexes are uniformly affected by this condition; studies reported a considerable male to female preponderance as 3:2. It is a rare autoimmune disease (Th2 predominance) observed and confirmed by Satyam et. al[11].

It is a rare chronic intra-epidermal Bullous disease classified as type II hypersensitivity reaction which attacks Desmosomes (components of the skin functioning to keep certain layers of the skin together bound) with the formation of antibodies.

The bullae are formed because of an immune reaction, initiated by the formation of IgG autoantibodies targeting the protein called Dystonin, also called Bullous Pemphigoid Antigen 1 and/or type XVII collagen, also called Bullous Pemphigoid Antigen 2 which is a main component of hemidesmosomes.[10] The symptoms may appear urticarial with visible rash. Extremities are commonly involved. Any part of the skin surface can be affected.

Classically, patients will have pruritus, urticaria or eczema followed by symmetric tense bullae on urticated, erythematous or normal skin on the trunk, inner thigh and flexures. Atypical presentations include an absence of blisters in up to 20% of cases, chronic itch with urticaria or eczematous eruptions, prurigo nodules and excoriations.

The heterogeneity of clinical presentation of BP can result in a delay in diagnosis. Diagnosis consists of at least 2 positive results out of 3 criteria (2-out-of-3 rule): (1) pruritus and/or predominant cutaneous blisters, (2) linear IgG and/or C3c deposits (in an n- serrated pattern) by direct immunofluorescence microscopy (DIF) on a skin biopsy specimen, (3) positive epidermal side staining by Indirect Immunofluorescence Microscopy on Human Salt-Split Skin (IIF SSS) on a serum sample[11-12].

Treatment is focused on reprieve of symptoms and avoidance of infection with minimal adverse events, so that the quality of life of the patient can be improved by the healing of blisters and erosions on the skin[12]. Tetracycline and Minocycline antibiotics are very helpful for mild to moderate disease and can be used with potent topical steroid creams for quick relief. However, in widespread cases or difficult-to-manage cases, systemic prednisone and powerful steroid free immunosuppressant medications, such as azathioprine, methotrexate or mycophenolate mofetil, may be appropriate[12-14]. The recurrence can be seen after six years in mostly 50% of the patients who are cured, as we know that there is no permanent cure.

CONCLUSION:

Bullous Pemphigoid is a rare disease in India as it has low prevalence in our country. It has a pattern of remissions and flare-ups. Confirmation of BP is done by histologic and immunopathologic investigations. Histopathology from skin lesions demonstrates a sub-epidermal blister. Treatment is multidisciplinary. Treatments help to control the itch and blisters and also help the skin to heal. Among all the treatment modalities in India, the most commonly used medications for the BP treatment is steroids, either oral or in the form of pulse therapy. Powerful steroid free immunosuppressive drugs are added as adjuvants to increase the efficacy and to have a steroid sparing effect.

Conflicts of interest

The author declares that there is no conflict of interest to disclose.

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