

A Case Report on Bullous Pemphigoid- Its Management with Dexamethasone and Its High Efficacy

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Abstract

Bullous pemphigoid is the most frequently occurring skin condition. Increase incidence rate in the past decades has been attributed to the population of drug-induced cases and with aging, that causes large, fluid-filled blisters, so commonly called as a blistering disorder. They tend to develop on areas of skin that often flex such as the hands, legs especially on thighs and feet. The blisters occur because of a malfunction in immune response since it is an autoimmune disorder often triggers by UV Rays exposure. According to the American Academy of Dermatology, UV radiation also impairs the skin's immune system in alarming ways by the reduction of watchdog cells that help recognize and respond to antigens and alters their function so they are as effective as dozing prison guards [8]. Diagnosed based on physical examination, clinical, histopathological, immunological criteria. The histopathological report reveals the presence of a linear band of IgG deposit found on the blister roof (epidermal side of split skin). Multiple treatment modalities are present for this condition, involving medications are anti-inflammatory, drugs to reduce immunoglobulin formation, treatments to increase the elimination of antibodies. This case report aims to present a classic case of this condition, to highlight awareness on dexamethasone efficacy in such rare skin conditions like bullous pemphigoid, and to advocate referral to a dermatologist given its potential severity.

Keywords: Bullous pemphigoid; Dermatology; Blisters; Autoimmune; UV ray's exposure; Dexamethasone

Abbreviations: BP- Bullous pemphigoid, UV- ultraviolet, DIF- Direct immunofluorescence, IgG- Immunoglobulin G, SPF- Sun protection factor, UVB- short wave ultraviolet B-rays, UVA- long wave ultraviolet A-rays, BD- Twice a day, OD- Once a day, SOS- whenever necessary, TID- Three times a day

Introduction

Bullous pemphigoid is a blistering disorder, autoimmune origin triggers often by UV Rays exposure, According to the American Academy of Dermatology, UV radiation also impairs the skin's immune system in alarming ways by the reduction of watchdog cells that help recognize and respond to antigens, and alters their function so they are as effective as dozing prison guards [8], is diagnosed based on clinical, histopathological, immunological criteria [1]. The patient has a medical history mouth canker sore with the complaints of photosensitivity, which shows the triggering factor as UV Rays, played a major role in this bullous pemphigoid case. The aim of this case report is to present a classic case of this condition and its management with dexamethasone.

Case Presentation

A 48-Year-old male presented to the hospital with a worsening fluid-filled blister & raw areas crusting over both feet and hand. The mild eruptions with a minute fluid-filled blisters started one month prior and were started spreading or distributing and the final condition is enlarged bullous reported while the patient presented to the hospital. The patient went to primary care local hospital to a physician on his current place of residence Saudi Arabia, where they burst fluid-filled lesions & applied Ineffectual Fucidin (sodium fusidate) ointment, physician-prescribed oral tablet Montek LC (which is a combination medicine contains montelukast sodium & levocetirizine hydrochloride Anti-allergic) & Neosporin powder (contains neomycin, bacitracin, & polymyxin Antibiotics) and told the patient that he had hypersensitivity reaction. The patient followed the medications for 3weeks without fail, but the bullous continued to worsen and he experienced the pain and redness, less physical activity due to the bullous formation in the feet and hands and some got ruptured and were both crusting and oozing with edema. On admission to our hospital the patient presented with cutaneous, tense bullae & vesicles over hands & feet, lichenoid with papules to plaque & mouth canker sore. On day 1: parenteral drugs such as cefotaxime 1g/BD, pheniramine maleate 2ml/BD, and other supportive therapies like high potency Fucidin ointment (topical), chlorhexidine mouth wash (TID), calcium/

vitamin D3(OD), paracetamol 650mg (SOS), Tab. A to Z (vitamin/mineral supplement) is given. But still, the condition of bullous pemphigoid was ameliorated accompanied by complaints of new lesions with severe itching. On day 2: skin biopsy was suggested then along with the day 1 drug parenteral Dexamethasone 2ml/OD was add on, then the reduction in new lesion formation was notified on the physical examination which shows a drastic improvement with the dexamethasone (Anti-inflammatory drug) and other supportive treatment such as doxepin (Tricyclic anti-depressants) for the sleep disturbance. On day 3: the same drugs were followed. On day 4: skin biopsy report arrived Impression: direct immunofluorescence study performed on a perilesional skin biopsy specimen from a patient with bullous pemphigoid detects a linear band of immunoglobulin G deposit with dermal side split skin along the dermo-epidermal junction confirms the bullous pemphigoid. On day 5: the patient became stable, decreased number of tense bullae condition were improving side by side, so discharge was advised by the dermatologist with the following drugs such as calcium-vitamin D3, vitamin/mineral supplements, Fucidin ointment, chlorhexidine mouth wash as supportive care therapy and oral tablets such as doxycycline and niacinamide (Vit-B3) were prescribed. As a clinical pharmacist suggestion patient was referred to a dermatologist to discuss adding a high potency oral & Topical corticosteroid such as dexamethasone.

Discussion

Pemphigoid is derived from the Greek words pemphix (bulla, blister) and eidos (form). BP is the most common of blistering disorders, with an incidence of 0.2 to three cases per 100,000 persons each year [1]. BP is an autoimmune disorder characterized by the presence of any of these IgG, IgA, IgM & C3 antibodies directed against the structural components of the keratinocytic hemidesmosomal proteins BP180 & BP230 [2]. Both the antibody and compliment components deposit along the basement membrane at the level of the dermo-epidermal junction, triggering a destructive inflammatory response with the formation of the characteristic blistering lesions [2]. Antibody levels will directly correlate with the disease activity [3]. Focal separation of the epidermis and dermis results in tense subepidermal blister formation. The clinical characteristic of BP may resemble a variety of other skin conditions. It is important to use clinical, histopathological, and direct immunofluorescence (DIF) findings to differentiate BP from other blistering disorders like dermatitis herpetiform, epidermolysis bullosa acquisita, pemphigus Vulgaris, etc. [3,8].

Diagnostic Criteria

DIF studies demonstrate in vivo deposits of antibodies and other immunoreactions, such as complement. DIF tests usually demonstrate IgG (70-90% of patients), IgA, IgM, and complement C3 deposition (90-100% of patients) in a linear band at the dermal-epidermal junction [6]. Bullous pemphigoid can be differentiated from these conditions by incubating the patient's skin biopsy sample in 1 mol/L salt prior to performing the DIF technique. This process induces cleavage through the lamina Lucida. DIF on salt-split skin reveals IgG on the blister roof (epidermal side of split skin) in patients with bullous pemphigoid, while, in CP and EBA, the IgG localizes to the blister floor (dermal side of split skin). The optimal location for DIF testing is normal-appearing perilesional skin. False-positive results can be observed when it is performed on lesional skin. Rarely, skin biopsy samples placed in transport media may yield false-negative results. This observation makes the use of fresh tissue the preferred substrate for DIF studies [6].

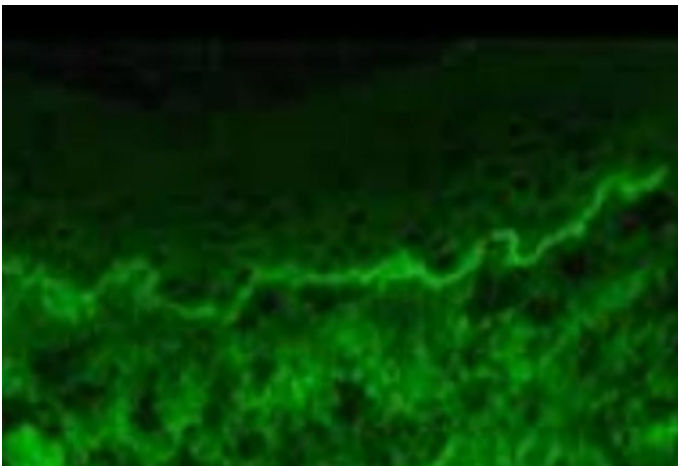


Figure-1: A DIF study performed on tense bullae perilesional skin biopsy specimen of from a patient with bullous pemphigoid detects a band of immunoglobulin G&C3 Granular deposit along the dermo-epidermal junction (DEJ).



Figure-2: A part of the thigh region with bullous pemphigoid lesions on the day of admission.

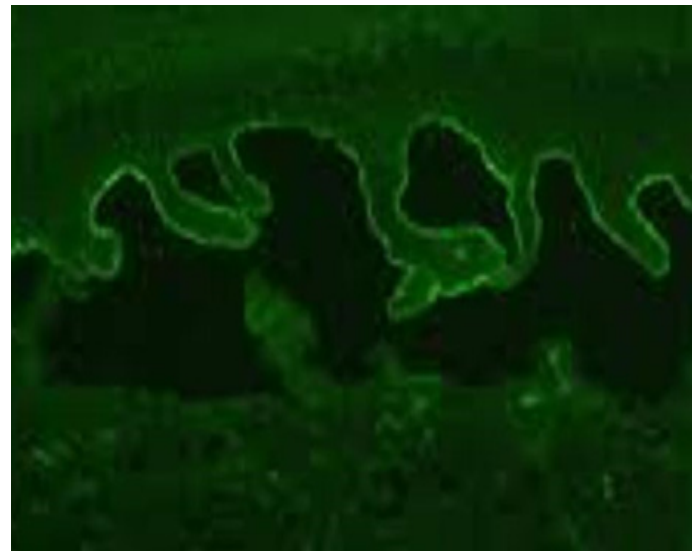


Figure-3: Deposits of circulating anti-basement membrane antibodies in peripheral blood observed by Indirect Immunofluorescence using salt-split human skin substrate (original magnification x200, its mandatory for definite diagnosis of BP.

Bullous pemphigoid histological findings vary according to the duration of the lesion and the clinical appearance of the biopsy site [8,9]. This is important, as misdiagnosis can lead to mistreatment and eventually worse outcomes for patients with a potentially treatable condition. In this case from day, 2nd dexamethasone is been administered and the drastic improvement is been observed and the patient became stable on the 5th day and planned to discharge [5]. The discharge drugs include oral doxycycline and niacinamide (Vit-B3) is shown to have an increased efficacy along with dexamethasone. The treatment of bullous pemphigoid with tetracycline and niacinamide (nicotinamide) is effective and accompanied by less serious adverse effects [4]. Another supportive nutritive therapy is been prescribed and as a clinical pharmacist suggestion sunscreen lotion can also be advised to prevent further relapse due to UV ray's exposure to the disease trigger zone. According to American Academy of dermatology "Dermatologists recommend using a sunscreen with an SPF block of at least 30, Which blocks 97% of the sun's rays & just to make sure it offers a broad spectrum (UVA & UVB) protection, an SPF of 30 or greater, and is water-resistant" [9].

General treatment guidelines

The objective of therapy in bullous pemphigoid include decreasing blister formation, promote healing of blisters and erosions, and to determine the minimal dose of medication necessary to control the disease thereby decreasing inflammatory response and autoantibody production. If the disease is severe and the patient doesn't respond to the treatment, immunosuppressants should be started which also reduces the risk of secondary (systemic) infection [2]. Pharmacological therapy includes

Anti-inflammatory agents such as Corticosteroids, tetracyclines, dapsons, clobetasol, and immunosuppressants like azathioprine, methotrexate, mycophenolate mofetil, and cyclophosphamide. Literature suggests that Omalizumab can be used here as a corticosteroid-sparing agent. As per the studies, treatment with doxycycline was proved to be effective with fewer adverse effects when compared to prednisone. Localized bullous can be treated with high potency topical corticosteroids along with systemic anti-inflammatory agents like tetracycline and nicotinamide can be given [4,10]. In severe cases, it is proposed to begin systemic steroids with immunosuppressants for better outcomes. In extreme cases where the disease becomes very difficult to control an anti-CD20 Antibody - Rituximab can be prescribed. The remission rate of 1.5 to 5 years after the treatment was reported in a few studies [7].

Conclusion

Overall, BP is a potentially fatal disease in this current decade with a high prevalence, which is treatable when under the appropriate level of care. This case report aims to increase disease awareness and highlight differing treatments & significance of the high potency drugs like corticosteroids. Majorly all severe inflammatory conditions treated by the steroids and high potency Fucidin, nicotinamide, as well as to advocate referral to a dermatologist, given its potential severity.

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