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Case Study

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BULLOUS PEMPHIGOID: A CLASSICAL CASE REPORT MANAGED WITH PULSE THERAPY

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ABSTRACT

Bullous pemphigoid is an autoimmune blistering disorder that typically presents in elderly patients as pruritic tense subepidermal blisters. It presents clinically with diffuse eczematous, pruritic, urticaria-like lesions, with the later appearance of tense bullae or blistering lesions typically filled with clear fluid. It primarily affects elderly individuals with an average age of onset being 65 years.Bullous Pemphigoid (BP) can be diagnosed clinically with a high index of suspicion with the aid of skin biopsy and histology.[1] A 64 year old female patient with known comorbidities of Systemic hypertension and type2 diabetes mellitus, presented with complaints of multiple vesicles all over the body, pain and burning sensation all over the body since 20days. She was initially treated in nearby clinic for the same. She had past surgical history of lump excision on scalp few years back. On physical examination, some of the lesions had ruptured and were both pruritic and painful. Baseline blood investigations done. Skin biopsy was done revealed Bullous Pemphigoid. Dermatologist opinion was sought in view of painful blisters on she advised for 3 days cycle Pulse therapy. The pathogenesis of BP is complex and its treatment and management still represents a challenge due to higher frequency of several comorbidities in this group of patients which may leads to tolerance to Bullous Pemphigoid treatments. Hence an early diagnosis and a prompt treatment are mandatory to reach better clinical outcome.The aim of this case report is to present a classic case of this condition, to highlight an awareness of differing treatment options, and to advocate referral to a dermatologist given its potential severity.

KEYWORDS: Bullous Pemphigoid, Pulse therapy, Autoimmune-disease.

INTRODUCTION

Bullous pemphigoid is a rare autoimmune blistering disorder that typically presents in elderly patients as pruritic tense subepidermal blisters. It is brought on by tissue-bound and circulating autoantibodies that are directed against either bullous pemphigoid antigen 1 or 2 or both. It presents clinically with diffuse eczematous, pruritic, urticaria-like lesions, with the later appearance of tense bullae or blistering lesions typically filled with clear fluid.[3]

The pathogenesis of BP is characterized by tissue-bound and circulating IgG autoantibodies against two components of the hemidesmosome of stratified epithelia, BP230kD and BP180kD. A cytoplasmic protein called BPAg1 aids in the cytoskeleton's anchoring of intermediate filaments and a transmembrane adhesion molecule called BPAg2 has a number of collagenous extracellular domains. Antibodies to BPAg2 appear to be important in blister formation.A transmembrane adhesion molecule called BPAg2 has a number of collagenous extracellular domains. Activation of complement system attracts inflammatory cells to the basement membrane which leads to release of proteases which inturn cause degradation of hemidesmosomal proteins leading to blister formation. The bullae are usually filled with clear fluid but may be hemorrhagic. Oral and ocular mucosal involvement rarely occurs. During the early phase lesions are generally pruritic erythematous, eczematous or urticarial.[7]

It has been recognized that a combination of genetic predisposing factors such as class 2 HLA and environmental influences such as UV radiation, trauma and drugs may contribute to the loss of immune tolerance towards the antigens.[4] Neurologic and neurodegenerative disease such as multiple sclerosis and Alzheimer's disease[5] have an increased risk of developing BP. The risk of thrombosis[6] and vitamin D deficiency were found to have augmented in BP patients.

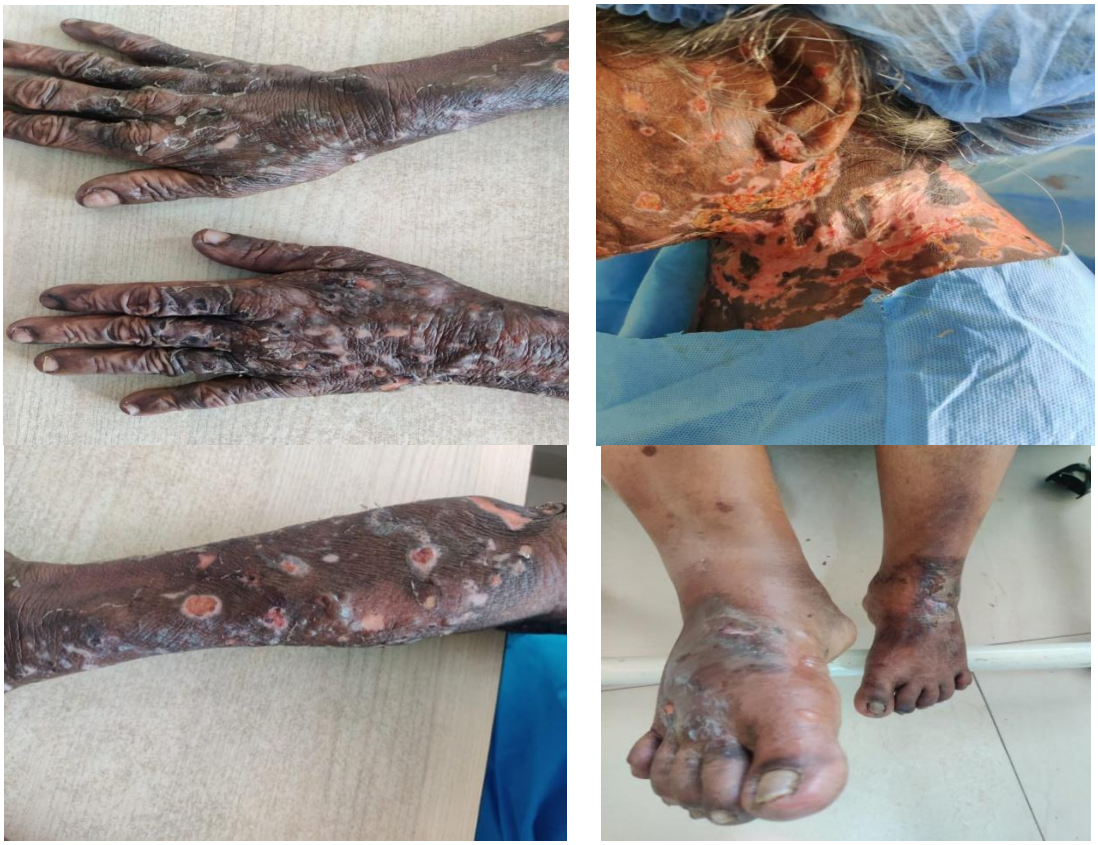
The mainstay of therapy for bullous pemphigoid is systemic corticosteroids, with or without topical corticosteroids, and/or systemic dapsone or immunosuppressants. In fact, the dyshidrosiform bullous pemphigoid lesion shape resembles a number of different disorders, such as allergy and irritating contact dermatitis, that are characterised by blisters on the hands

and feet, dermatophyte infection, dyshidrosis or pompholyx, epidermolysis bullosa acquisita, erythema multiforme, herpes gestationis, scabies, and systemic contact dermatitis.[2]

The management of pemphigus has long been a hot topic. Prednisolone was once a frequent treatment for pemphigus, but it has serious steroid side effects. However, Paricha and Gupta introduced dexamethasone-cyclophosphamide pulse (DCP) therapy in 1984.[10] Since a few years ago, pulse therapy has become popular for treating pemphigus. Pulse therapy refers to intravenous infusion of high doses of steroids for one or more days for better efficacy and to decrease the side effects of long-term steroids.Our study represents a descriptive summary of 65-year woman with Bullous Pemphigoid covering history, physical examination and management.

CASE PRESENTATION

A 64 year old female patient with known case of hypertension and type2 diabetes, presented with complaints of multiple vesicles all over the body, pain and burning sensation all over the wound since 20days. She went to nearby clinic and was prescribed:T.Xyzal.M-(10/5)mg-BD, Liquid paraffin ointment-HS, Clonate F ointment-HS, C.Dalacin C-300mg-BD, T.Bact ointment- BD, T.Wysolone-40mg-OD, T.Wysolone-40mg-OD. She had past surgical history of lump excision on scalp. On physical examination, she was conscious, oriented, afebrile and some of the lesions had ruptured and were both pruritic and painful.



Baseline blood investigations done and shows elevated neutrophils (89%), elevated CRP (78mg/L), elevated HbA1C (8.8%), anti-nuclear antibody tested positive. Skin biopsy was done revealed Bullous Pemphigoid. Dermatologist opinion was sought in view of painful blisters on she advised for 3 days cycle Pulse therapy.

PULSE THERAPY- 3 DAYS CYCLE	
DAY 1	Inj.Dexa 100mg+ 10units Insulin in 5%Dextrose 500mL over 2 hours
DAY 2	Inj.Dexa 100mg+100mg cyclophosphamide+10 units soluble insulin in 5%Dextrose 500mL over 2 hours.
DAY 3	Inj.Dexa 100mg+ 10units Insulin in 5%Dextrose 500mL over 2 hours

She was managed with antibiotics (Inj.Amoxicillin+clavulanic acid-1.2g and Inj. Clindamycin-600mg), steroids (Inj. Dexamethasone-100mg), immunomodulators (T.Cyclophosphamide-50mg) and other supportive medications. She improved gradually and symptomatically better and was discharged.

DISCUSSION

Pemphigoid is derived from the Greek word pemphix (blisters) and eidos (forms).Based on the cases reported so far, the condition was slightly more common in women and the onset of the disease, for most of the patients, occurred between the ages of 61 and 94 years.[2] A group of french authors proposed the following clinical predictors of BP: absence of atrophic scars limited neck or head involvement, absence of mucosal involvement and age greater than 70years. The presence of three out of these four criteria, BP had a sensitiviy of 90%, a specificity of 83% and a positive predictive value of 95%.[8] Three categories of drugs may be used to treat BP[9] the first category is anti-inflammatory drugs such as topical steroids, sulfonamides and antibiotics with anti-inflammatory properties like tetracycline. Another drug class consists of those that decrease the production of antibodies such as systemic steroid, azathioprine, methotrexate, mycophenolate, cyclosporin and rituximab. Finally, treatment that increase the elimination of abnormal antibodies like plasmapheresis and intravenous immunoglobulin (IVIG) can be performed.

CONCLUSION

As highlighted in this study, the pathogenesis of BP is complex and its treatment and management still represents a challenge due to higher frequency of several comorbidities in this group of patients which may leads to tolerance to BP treatments. Hence an early diagnosis and a prompt treatment are mandatory to reach better clinical outcome. This case report aims to increase disease awareness and highlight different treatments, as well as to advocate referral to a dermatologist, given its potential severity.

HUMAN ETHICS

Informed Consent was acquired for this case report. No identifying information is included.

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