Bullous Pemphigoid in a 32 year old lady at Delta State University Teaching Hospital, Oghara

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ABSTRACT

Bullous pemphigoid is a chronic autoimmune, sub-epidermal blistering skin disease commonly observed in the elderly (over 70 years old).

It is characterized by the presence of immunoglobulin G autoantibodies specific for the hemi-desmosomal bullous pemphigoud antigens BP 230 (Bp Ag1) and BP 180m (BP Ag2).

In this study, we report the case of a 32 year old Urhobo lady with bullous pemphigoid and its frequently presents as a diagnostic and therapeutic challenge.

Key words: pemphigoid, bullous, skin disease, immunoglobulin, young adult.

INTRODUCTION

BULLOUS pemphigoid is the most common subepidermal autoimmune blistering disease.

It generally affects people older than 70 years with no ethnic, racial or social predilection[1,2,3]. Lesions occur mostly along large skin folds and on the abdomen and may initially appear as urticarial and itchy lesions with blisters eventually erupting.

Bullous pemphigoid is the most common subepidermal autoimmune blistering disease. It generally affects people older than 70 years with no ethnic, racial or social predilection.

Lesions are located on erythematous, erythematous-edematous or apparently healthy skin. They are tense and their content is sero-heamorrhagic. Mucous membranes are rarely affected; they are affected in 10-35% of cases[2,3].

There are clinical variants which includes classic, localized, nodular and drug induced[1,2,3].

There is a paucity of reports of bullous eruption especially bullous pemphigoid and reported cases of bullous pemphigoid in patients under 60 years is rare [1,4].

We report a case of bullous pemphigoid in a 32 year old lady.

CASE REPORT

A 32 year old female patient from Sapele, Delta State, Nigeria; presented with intense itching associated with blisters urticarial plaques, bullae, papular lesions and lichenified plaque which had been present for 4 months.

The bullae with serous and haemorhagic content were located on apparently healthy or erythematous skin on the trunk and upper and lower limbs (Tables 1-3).

She had no mucosal lesions. Her general health was good.

She denied use of medications.

Histolopathological examination of the bullous lesion showed subepidermal cleavage with a mixed inflammatory infilterate (polymorphonuclear cells and lymphocytes). Direct immunofluorescence of perilesional skin showed linear, continuous and intense deposition of IgG and C_3 in the basal membrane zone.

A complete blood count revealed eosinophilia.

She was treated with oral prednisolone 40mg daily. Betnovate (Betamethasone propionate) scalp lotion and topical Clobetasol dipropionate 0.05% cream.

Figure 1. Bullae on the back

Figure 2. Bullae on the legs

Figure 3 Bullae on the arms and hands

DISCUSSION

Bullous pemphigoid is a chronic autoimmune subepidermal blistering skin disease that rarely involves mucous membranes.

It is characterized by the presence of immunoglobulins (IgG) autoantibodies specific for the hemidermosonal bullous pemphigoid antigen BP 230 (BPAg 1) and BP 180 (BPAg 2).

It is an autoimmune skin condition, the incidence of bullous pemphigoid was found to be 4.3 cases per 100,000 persons per year in UK[7,12].

It is a chronic inflammatory disease, if untreated; it can persist for months or years with period of spontaneous remission and exacerbations.

Patients with aggressive or widespread disease, as the case of the reported patient, those requiring high dose steroids and immunosuppressive agents have increased morbidity and risk of death[6]

The risk of death for bullous pemphigoid patients was found to be twice as great as the controls[12]. There is no racial predilection. The risk of bullous pemhpigoid appears to be equal in men and women and occurs usually in age group above 60 years.

Bullous pemphigoid can be triggered by drugs such as Captopril, enalapril, furosemide, spironolactone, amiodarone, losartan, β-blockers, ibuprofen, fluoxetine, sulphasalazine, chloroquine, ampicillin, D-penicillamine, ciprofloxacin, gabapectin.

IgG autoantibodies bind to the skin basement membrane in patients with bullous pemphigoid. The binding of autoantibodies at the basement membrane activates complement system which is thought to play a critical role in attracting inflammatory cells to the basement membrane. These inflammatory cells are postulated to release proteases which degrade dermosomal proteins and lead to blister formations. Eosinophils are characteristically present in human patients.

Interleukin 16 is a major chemotactic factor responsible for recruiting CD_4^+ helper T cells to the skin. Significantly higher levels of interleukin 16 were detected in sera and blisters of bullous pemphigoid patients compared with healthy subjects[7]

In a study of 27 bullous pemphigoid patients (reported in 2006), serum level of monokines induced by interferon gamma (Th 1- type chemokine) and serum levels of CCl 17 and CCl 22 (Th2-type chemokine) were significantly increased in bullous pemphigoid patients compared to healthy patients[9].

Matrix metalloproteinases (MMP2, MMP9, MMP13) were significantly increased in lesional bullous penphigoid skin compared with healthy subjects[10]

In 2008, a role of IgE class of autoantibodies, particularly those that target BP180 has been found to be related to Bullous Pemphigoid. The higher levels of IgE anti Bp 180 correlated with a more severe clinical phenotype[11].

Generalized bullous forms is the most common presentation. Bullae arise on any part of the skin surface, with a predilection on the flexural areas of skin. The bullae heal without scarring. Other forms include the vesicular, vegetative, generalized, erythrodermic, urticarial, acral and nodular forms.

Histological examination of the bullous lesions show subepidermal cleavage with mixed inflammatory infiltrates. Direct immunofluorescence of the perilesional skin shows linear and intense deposition of IgG and C_3 in the basal membrane zone.

We highlight in this case report the importance of considering the possibility of a diagnosis of bullous pemphigid in patients younger than the age group usually affected by bullous pemhigoid.

The clinicopathological features in our patient are compatible with the diagnosis of Bullous Pemhigoid.

It is fundamental to perform direct immunofluorescence and possible indirect SSS (salt split skin test) for differential diagnosis between bullous pemphigoid and other subepidermal vesiscobullous eruptions such as dermatitis herpetiformis, and inflammatory epidermolysis bullosa acquisita.

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