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CASE REPORT AND LITERATURE REVIEW

Bullous Pemphigoid Masquerading as Erythrodermic Psoriasis

Pemphigoïde Bulleuse Déguisée en Psoriasis Érythrodermique

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ABSTRACT

Bullous Pemphigoid occurs more commonly in the elderly with a rare occurrence in infancy, childhood and adolescence. The uniqueness of this presentation in the adolescents warrants this report. Both Erythrodermic Psoriasis and Bullous Pemphigoid are autoimmune skin disorders that differ in presentation though some of the symptoms may overlap. While Erythrodermic Psoriasis presents with massive scaling, Bullous Pemphigoid presents with vesiculo-bullous lesions and blisters which heal and keep spreading leaving burn-like areas. Bullous Pemphigoid is the most frequent subepidermal autoimmune bullous skin disease and could have a polymorphic presentation. At presentation there was massive scaling with intense itching however in the course of treatment, vesicles, blisters and bullae became apparent and the histology result was consistent with the diagnosis of Bullous Pemphigoid. Bullous Pemphigoid was therefore masquerading as Erythrodermic Psoriasis. **WAJM 2022; 39(12): 1319–1323.**

Keywords: Erythrodermic Psoriasis, Pemphigoid, Bullae, Auspitz sign.

RÉSUMÉ

La pemphigoïde bulleuse se produit plus souvent chez les personnes âgées, mais rarement chez les nourrissons, les enfants et les adolescents. Le caractère unique de cette présentation chez l'adolescent justifie ce rapport. Le psoriasis érythrodermique et la pemphigoïde bulleuse sont des troubles cutanés auto-immuns dont la présentation diffère, bien que certains symptômes puissent se chevaucher. Alors que le psoriasis érythrodermique se manifeste par une desquamation massive, la pemphigoïde bulleuse présente des lésions vésiculo-bulleuses et des cloques qui guérissent et s'étendent en laissant des zones semblables à des brûlures. La pemphigoïde bulleuse est la maladie cutanée bulleuse auto-immune sous-épidermique la plus fréquente et peut avoir une présentation polymorphe. Au moment de la présentation, il y avait une desquamation massive avec des démangeaisons intenses, mais au cours du traitement, des vésicules, des cloques et des bulles sont apparues et le résultat de l'histologie était cohérent avec le diagnostic de la pemphigoïde bulleuse. La pemphigoïde bulleuse se faisait donc passer pour un psoriasis érythrodermique. **WAJM 2022; 39(12): 1319–1323.**

Mots clés : Psoriasis érythrodermique, pemphigoïde, bulles, signe d'Auspitz.

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INTRODUCTION

Over 2000 different skin diseases which cuts across all age groups are encountered in the field of Dermatology but majority are non-neoplastic lesions. Although some of them are easy to diagnose clinically, majority of them require good history and histopathologic diagnosis for proper management.[1] Generally with the paucity of Dermatopathologists in Nigeria, West Africa and sub-Saharan Africa there is usually an interaction between the attending physician and the general histopathologist for challenging cases.[2] This patient had a challenging diagnosis at presentation because the presentation was similar to that of Erythrodermic Psoriasis and thus a histopathologic examination proved handy.

Erythrodermic Psoriasis is a rare variant of Psoriasis affecting 1–2.25% of the Psoriatic population. It presents with skin lesions like erythema, edema, pruritus, ill-defined psoriatic plaques, scaling, hair loss, occasional exudative lesions and palmoplantar or diffuse desquamation.[3] This was part of the presentations of this patient warranting the initial difficulty in the clinical diagnosis of Bullous Pemphigoid.

Psoriasis and Bullous Pemphigoid are two entities that rarely occur concomitantly in a patient and they represent two well-characterized, chronic inflammatory diseases. Thus immunomodulatory therapy may positively influence shared as well as distinct inflammatory processes in both Psoriasis and Bullous Pemphigoid.[4] Therefore treatment at a point may be similar however results of skin biopsy will enable definitive treatment.

CASE REPORT

This was a 17-year-old girl referred to the clinic on account of inability to walk, massive body scaling with intense itching, generalized body weakness and bed sores of three months duration. Her problem started three months prior to presentation with intense body itching and blisters on the legs. She went to a primary health care clinic where she was given some medications with no significant improvement.

Her grandmother and relatives said it was not a disease that could be treated in the hospital and that no injection should be given to her because that might 'kill her'. This resulted in her being taken for traditional treatment. She was treated with oral herbs and warm compress was applied to the blisters. The problem worsened as the blisters would heal but new ones kept erupting. She developed pressure sores on the medial aspect of both elbows and on the right side of the buttock. There was generalized massive scaling, scalp hair loss, amenorrhea, severe weight loss, pedal swelling and scaling of the soles of both feet.

On examination there were severe generalized silvery, loose scales covered with dusting powder (Figure 1), pressure sores and bruises painted with gentian violet solution, severe wasting with dehydration and inability to ambulate but with no mucosal involvement. The Auspitz sign was positive.

The management and nature of the disease was explained to her and the mother. She was told she will be admitted and that she had an autoimmune skin disorder which will be managed with intravenous drugs, oral medication and topical creams. She was told that the disease could be managed in the hospital and that injections were not forbidden in the treatment. She was requested to do a retroviral screening after counseling, serum urea, electrolytes and creatinine (U/E/Cr), full blood count (FBC) and liver function tests (LFT). A punch biopsy of the skin for histology was done. The retroviral screening was negative, U/E/Cr, LFT and FBC results were:

LFT: Total protein 70g/l, Albumin 39g/l, Total bilirubin 8µmol/l, Direct Bilirubin 2.0µmol/l, SGPT 53µ/l, SGOT 59µ/l, Alkaline Phosphatase 129µ/l

U/E/Cr: Sodium 138mmol/l, Potassium 2.9mmol/l, Chloride 98mmol/l, Bicarbonate 26mmol/l, Urea 2.2mmol/l, Creatinine 40µ/l, Calcium 2.40mmol/l

FBC: PCV 32.1%, WBC 9.3 x 10^9 mm/l, Neutrophil 42.1%, Lymphocytes 51.5%. Monocytes 4%, Basophils 2.4%.

She was given intravenous normal saline one liter every 12 hours for 24 hours, intravenous Ceftriaxone 1g every 12 hours for 48 hours, topical Clobetasol cream after 20–30 minutes of emollients (blue seal Vaseline) daily and low dose oral Prednisolone 10 mg daily. She remarkably improved 48 hours after admission as the scaling, and itching reduced and her appetite had improved (Figure 2). She was then started on oral Cephalexin 500 mg every 12 hours for five days and wound dressing was done using honey.

Ten days on admission, she was ambulant but developed tensed bullae on the abdomen and multiple blisters on the back (Figure 3). There was scalded skin with erosions but no slough. A diagnosis of Bullous Pemphigoid was made. This was explained to her and her mother. Sixteen days on admission she was much better, ambulant, feeding well, pressure sores were healed and there was no more scaling. Histology result was received and she was asked to repeat U/E/Cr, FBC and LFT to prepare her for immuno-suppressants.

HISTOLOGY RESULTS

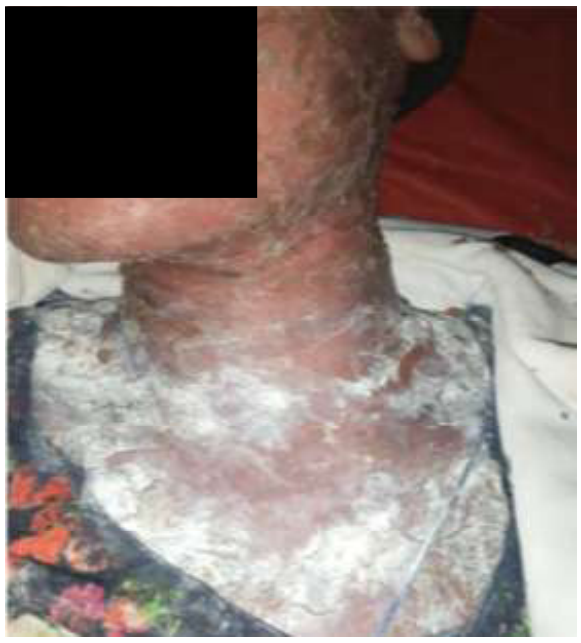
Histologic sections of skin biopsy tissue show keratinizing stratified squamous epithelium exhibiting multiple foci of acantholysis with some vacuolated epithelial cells and eosinophils. Underlying these are seen foci of subepithelial blister cavities containing mixed inflammatory infiltrates with few eosinophils and necrotic debri. This is consistent with Bullous Pemphigoid.

LFT: Total protein 61g/l, Albumin 36g/l, Total Bilirubin 8µmol/l, Direct Bilirubin 2.0µmol/l, SGPT 14µ/l, SGOT 17µ/l, Alkaline Phosphatase 60µ/l.

U/E/Cr: Sodium 140mmol/l, Potassium 4.7mmol/l, Chloride 99mmol/l, Bicarbonate 30mmol/l, Urea 2.3mmol/l, Creatinine 40µ/l, Calcium 2.30mmol/l.

FBC: PCV 36.09%, WBC 10.47 x 10^9 mm/l, Neutrophils 61.4%, Lymphocytes 29.6%. Monocytes 5%, Basophils 5%.

She was placed on 2.5mg Methotrexate tablets on Mondays, tablets Folic acid 10mg on Fridays and topical Clobetasol with emollients. She was discharged home and told of the need to repeat the investigations electrolytes, urea and creatinine, liver function test and

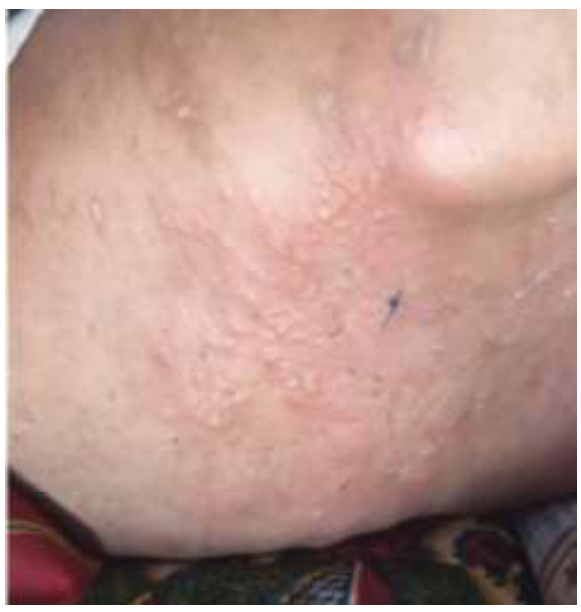


A. At Presentation

B. At Presentation

Fig. 1

Fig. 2: 48 hours after Admission



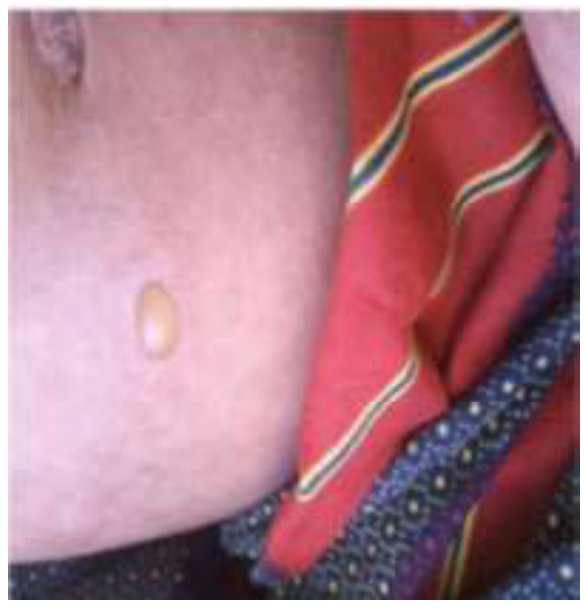


Fig. 3: 10th Day on Admission and Treatment.

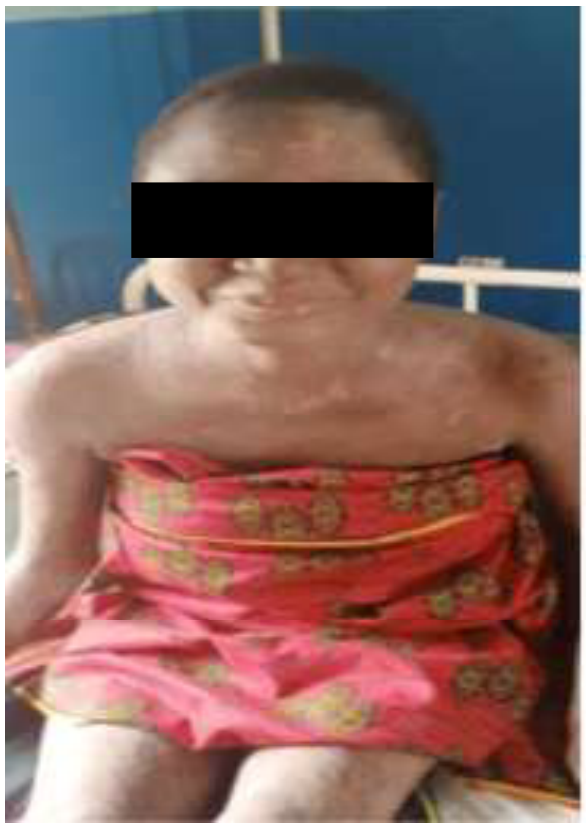


Fig. 4: The Patient after 4 weeks of Treatment.

full blood count every fortnight and that there could be remission of the problem. She was also told that if the problem persisted, she could be placed on biologics. She returned for follow up in four weeks with minimal lesions on her body (Figure 4).

DISCUSSION

Bullous Pemphigoid (BP) is one of the most common autoimmune subepidermal blistering skin disorders in which the body produces antibodies against hemidesmosomal proteins of the skin and mucous membrane.[5] Autoantibodies are thought to be produced against two autoantigens, BP180 (also called type xvii collagen) and BP 230 (a plakin) which are components of hemidesmosomes that promote epithelial-stromal adhesion in various tissues including the skin. This binding of autoantigens to antibodies leads to a cascade of events like recruiting inflammatory cells (neutrophils and eosiniphils) and release of proteases. This causes proteolytic degradation of extracellular matrix and destruction of the desmosomes resulting to sub-epidermal blisters.[5] The term pemphigoid was introduced by Lever in 1953 to describe a disease characterized by formation of blisters resulting from detachment of subepidermal layer which distinguishes it from pemphigus an intraepidermal blistering induced by acantholysis.[5,6]

It affects mainly the elderly in the 7th or 8th decade of life without ethnic, race or social predilection, however there are rare case reports in adolescents and children.[5,10] The index patient was a 17 year old female. There is no gender predilection though it seems to be commoner in men.[5,6] Studies in Europe show an incidence of 2.5–42.8 cases/ million/year while in Asia annual incidence ranges from 2.6–7.5.[7] A histologic study in Ibadan, Nigeria on 209 non-neoplastic skin diseases showed that Bullous Pemphigoid comprised 45.5% of bullous disorders.[8] Though BP is the commonest bullous disorder, even in our environment it poses a diagnostic challenge to the non-dermatologist because of the low index of suspicion leading to misdiagnosis.[9] Before referral to our facility this patient was managed for suspected Steven Johnson's syndrome.

Bullous Pemphigoid has a polymorphic presentation which could be

classic, non-bullous, exfoliative erythroderma, infantile and childhood types. The Classic type mainly affects the intertriginous areas like axillary folds, lower abdomen, inguinal areas and inner parts of the thigh. It is characterized by intense itching, tense blistering on normal skin or background erythematous or edematous skin on the trunk or extremities sparing the mucosal membranes in most cases.[5,10] This was the presentation of the patient, however, 30-40% of oral, esophageal and genital involvement of the lesions have been reported.[2,10]

Exfoliative erythrodermic BP is a rare form and presents with generalized erythema and desquamation without blisters and thus diagnosis can be challenging as other lesions such as pemphigus foliaceus, eczema, psoriasis and drug reaction can present this way.[11] This made her initial diagnosis challenging as the presentation was akin to erythrodermic psoriasis. Occasionally infants and children are affected with two peaks at 4 months and 8 years of age. Lesions commonly occur on the face (62%), palms and soles (75%) with increased generalized lesions requiring prompt use of systemic steroids.[6] The Non-bullous BP has raised debates as to whether it should be considered as a variant of BP or as a prodromal phase because according to a systematic review published in 2017 only about 9.8% of these patients develop blisters in the course of the disease and would rarely develop mucosal lesions. Common clinical findings include pruritus (100%), urticarial rashes (52.3%), excoriations (22.7%), papules and nodules (20.5%).[2,12] This patient presented with scaling, severe itching, excoriations and desquamations. In the course of treatment the patient developed tensed bullae and blisters giving the clinical diagnosis confirmed by histology.

Treatment requires the use of oral steroids, topical potent steroids, immunosuppressants, antibiotics and intravenous fluids. Glucocorticoids have been the mainstay of treatment to reduce inflammatory response, however, to reduce its systemic effects, immuno-suppresants like methotrexate, azathioprine, cyclophosphamide and cyclosporine are used as steroid-sparing agents.[10, 13] Another study showed that treating extensive lesions with potent topical steroids yielded better survival rates, and less severe complications compared to oral prednisolone.[5,13] Few control studies exist for treatment of BP, hence most treatment regimens are based on clinical experience.[13,14] A systematic review of literature showed that treating with lower doses of systemic steroids and potent topical corticosteroids is effective, also treatment with tetracyclines and niacinamide are associated with less serious adverse effects including death.[15] Immunosuppressants like methotrexate are well tolerated when used alone or in combination with systemic steroids. It is also cheap and easy to administer and usually taken once a week.[5]

Left untreated patients can recover within few months to years however these patients have prolonged morbidity due to intense itching and bullous-eroded skin.[16] In this patient morbidity was prolonged. The BP patients manifest psychological problems due to body dysmorphism associated with skin lesions. Counseling opens a window of opportunity for physicians or primary health care givers to provide information about the disease, encourage patients and have follow-up visits.[17]

This patient was treated with intravenous fluids, intravenous antibiotics, and bedside barrier wound dressing for bed ulcers. The parents were counseled and educated on the nature of the disease. Their perception about the disease was changed and the need for them to support her throughout the treatment was emphasized.

CONCLUSION

The diagnosis of Bullous Pemphigoid can be challenging, more so, it is commoner in the elderly. Its presentation in young adults, adolescents and children is very rare, therefore, there should be a high index of suspicion in these age groups. A combination of clinical history, examination and histopathologic skin examination is crucial for diagnosis and proper management. Counseling of the parents and care givers will help to give support to patients and avoid erroneous beliefs about the disease and disease progression.

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