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Case Report

Case report on localized bullous pemphigoid in two young females

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ABSTRACT

Bullous pemphigoid (BP) is a chronic autoimmune bullous eruption that is characterized by pruritic, tense sub-epidermal bullae. It commonly affects those aged 60 years and above. It has no gender predilection. It can be generalized or localized. This is a case report of 2 female patients with localized BP presenting as recurrent skin rash of more than 1 year duration which were later referred to the dermatology clinic. The patients were both females less than 40 years of age who have been having recurrent skin rash that have been associated with intense itching localized to a unilateral lower limb. The first patient had obvious bullae and ulcers while the second didn't have any obvious bullae. Initial clinical diagnoses did not consider BP. Punch skin biopsy was done in both cases that revealed histologic features confirming BP. Clinicians should have high index of suspicion of BP even in cases of localized chronic wound or recurrent ulcers irrespective of the age of patient.

Keywords: Bullous pemphigoid, Chronic, Localized, Ulcer

INTRODUCTION

Bullous pemphigoid (BP) is a chronic autoimmune bullous eruption that is characterized by pruritic, tense sub-epidermal bullae. BP is the commonest autoimmune skin disease. It is characterized by the production of antibodies against BP180 and BP230 proteins, which are normal components of hemidesmosomes found in the dermo-epidermal junction.[1] It commonly affects those aged 60 years and above. It has no gender predilection. It can be generalized or localized.[1] Localized (LBP) is known to be a rare variant of BP.[2] Few case reports of LBP have emerged from different parts of the world.[3] It can precede the generalized form.[4]

The aim is to create awareness that LBP can be a cause of recurrent wound ulcers.

CASE REPORT

Case 1

A 34 year old female administrator complained of recurrent spontaneous ulcer at the right ankle of 1year duration. Current episode of right ankle ulcer refused to heal with the use of antibiotics and ulcer dressing; she subsequently developed two bullae on the left sole and left ankle which broke down to become ulcers. Ulcers were painful, pruritic and discharging offensive serosanguinous fluid. More bullous eruptions were noted on both legs. Physical examination showed oedematous left leg extending to mid shin, ulcers measuring 12×8 cm, 10×8 cm, 2×2 cm on the right ankle, left ankle and left sole respectively. Surrounding edges were hyperpigmented, tender with raised edges and floor was

covered with slough. BP was not considered by the accident and emergency doctor and patient was referred to the surgeons on a diagnosis of cellulitis. There was a high index of suspicion on the part of the surgeons and dermatologist's review was requested. Results of investigations showed negative results of wound swab microscopy, culture and sensitivity (MCS), GeneXpert MTB/RIF and retroviral screening. Fasting blood glucose (FBG) was within normal limits. The erythrocyte sedimentation rate (ESR) was elevated. Punch biopsy of the skin revealed acanthosis and inflammatory cells infiltration with fibrocollagenous reticular dermis which is in keeping with BP. She was counselled and managed with oral prednisolone and daily wound dressing with good response with lesions completely disappearing from lower limbs after 6 months on follow up.

Case 2

A 24 year old undergraduate student referred from the general outpatient department to the dermatology clinic with complains of recurrent skin rash of 10 years duration. The current episode of rash started 6 months prior to presentation. Skin rash was located at the left anterior ankle and was associated with severe intense itching. It heals spontaneously however the current episode had persisted. Physical examination revealed hyperpigmented patch at the dorsal surface of the left foot with areas of lichenfication. Psoriasis and necrolytic acral erythema were initially considered. Histopthology showed hyperkeratosis, papillomatosis and acanthosis. The papillary dermis displayed inflammatory aggregates while the reticular dermis contained perivascular inflammatory infiltrates and it was densely fibrocollagenized suggestive of BP. Patient was counselled and placed on prednisolone and antihistamine (loratidine). Since then she is yet to return for follow up.

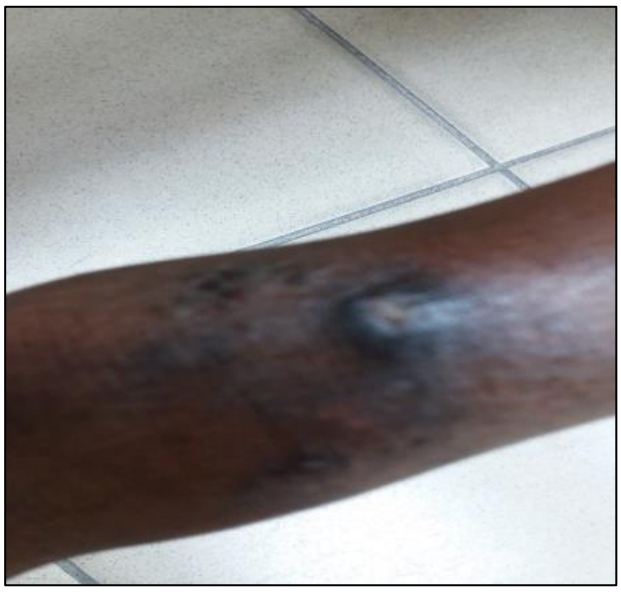

Figure 1: Lichenified recurrent ulcer on left lower limb.

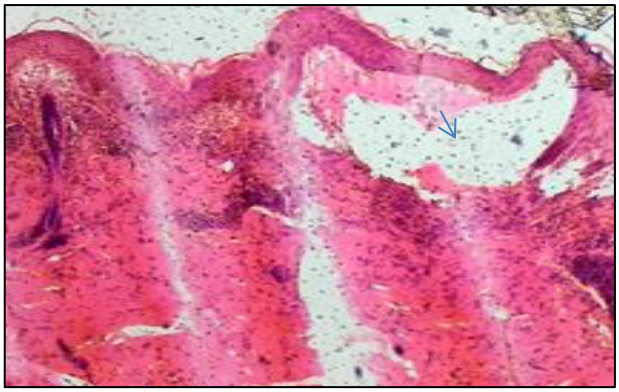

Figure 2: Hematoxylin and eosin stain (x40) arrow showing sub-epidermal split.

DISCUSSION

The patients were both females less than 40 years of age who have been having recurrent skin ulcers that have been associated with intense itching localized to the lower limbs. It is known that BP tends to occur in those that are elderly as seen in the literature, however this case report showed two cases of females that are younger when compared to case report by Otike-Odibi et al from similar region that reported generalized BP in an older adult and that of Noorily and Ren that reported LBP in an 80 year old man.[5,6] The limbs have been commonly implicated in LBP as reported by Wang et al and as well as other case reports.[2,3,6,7] Hasegawa et al had a case of recurrent of lesions on the palms and soles in an elderly patient that had it initially on the right lower leg.[2] The cases seen in the study by Stander et al. noted that LBP constituted 2.5% of all BP cases seen within the ten year period and all were >60 years old with an average age of 68.1±8.2 years with a female predominance; with the upper limbs and abdomen being the most common sites while that of Corbella-Bagot et al showed a male predominance with similar number and age range.[7,8] Intense itching was a common presentation as seen in other case reports.[2-4] This complaint is the most disturbing and causes poor quality of life as seen in the study carried out by Kalinska-Bienias et al. The pattern of pruritus of BP has been characterized by long itch period lasting more than 10 minutes, worse in the evenings and nighttime, aggravated by sweating and stress and relieved by cold.[9] The mechanism of pruritus has been attributed to autoimmune response initiated by loss of self-tolerance against cutaneous autoantigens resulting in the induction of IgG autoantibodies although the exact pathogenesis of pruritus in BP remains unknown.[9] In this case report there was no established aggravating or relieving factors in the patients. Risk factors are yet to be fully established for LBP however certain triggers have been identified such as burns, chronic venous stasis, radiotherapy, surgical scars, and trauma, have been implicated.[4,7,8] BP has been known to have these risk factors such as non-bullous chronic inflammatory diseases such as psoriasis and lichen planus, drugs such as non- steroidal anti- inflammatory

drugs (NSAIDs) like ibuprofen, captopril and oral anti-diabetic agents such as vildagliptin, ultraviolet radiation and vaccination.[1,5] LBP has also been associated with stoma sites as seen in the reports by Stander et al and Salomon et al.[8,10] The first patient had obvious bullae and ulcers while the second didn't have any obvious bullae. Initial clinical diagnoses did not consider BP in both cases. The inability to detect LBP from initial clinical findings is not peculiar to this study. LBP initially has been thought to be acute contact dermatitis as reported by Wang et al and Salomon et al.[4,10] Misdiagnosis is not rare as noted by the literature review of case reports carried out by Corbella-Bagot et al.[7] Punch skin biopsy was done in both cases that revealed histologic features confirming BP. This is recommended.[1-4] Punch biopsy and histology is available in our setting and can be used to make a diagnosis.[5] LBP diagnostic criteria has been proposed by Corbella-Bagot et al which includes having any three of these four : bullous eruption at any localized site with a possible trigger which has not progressed to generalized form within three months of occurrence(clinical criteria), positive IgG antibodies against the epidermal side of basement membrane zone (BMZ) by indirect immunofluorescence (IIF) and/or positive IgG antibodies reacting with BP180 and/or BP230 by ELISA, IIF, immunoblot, or immunoprecipitation (serologic criteria); subepidermal blister with the presence of eosinophils (histologic criteria) and positive direct immune-fluorescence (DIF) with linear deposits of IgG and/or C3 along the BMZ (DIF criteria).[7] DIF and IIF are recognized as gold standards in making diagnosis and excluding other causes of autoimmune blistering disorders as reported by Kitajima et al.[11] DIF and IIF availability is currently limited in the setting of this case report hence skin punch biopsy is recommended in addition to clinical history of recurrent ulcers with or without blistering with intense pruritus to make diagnosis.[5] Oral prednisolone was used for the two patients and their was improvement as seen in other case reports.[2-4] Potent topical steroids such as 0.05% clobetasol propionate ointment has also been used with good response and is recommended as reported in other case reports.[6-11] LBP progressing to the generalized BP has been reported in few cases just as it was in this case report.[2-4,6-12] Koebnerization or isomorphic phenomenon is the occurrence of new lesions at different site elicited by trauma. It has not been established if this might be a factor in LBP progressing to the generalized form.[12]

CONCLUSION

Clinicians should have high index of suspicion of BP even in cases of localized chronic wound or recurrent ulcers irrespective of the age of the patient.

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