Reverse Koebner Phenomenon in Bullous Pemphigoid – A Case Report

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Abstract

Reverse Koebner response is the nonappearance or disappearance of the lesions of particular dermatoses at the site of injury. Herein we report a case of the reverse Koebner phenomenon in bullous pemphigoid in a 35-year-old male patient with sparing of the waist area which could probably be because of the phenomenon of locus majoris resistentiae. The histopathology of the lesion showed subepidermal split with a mixed inflammatory infiltrate, which was composed of predominantly eosinophils, neutrophils, and lymphocytes, but the histopathology of the spared skin showed no abnormalities. However, the direct immunofluorescence from the perilesional area showed linear deposits of immunoglobulin G and C3 in the dermoepidermal junction, but there were no deposits in direct immunofluorescence from the spared skin. This case is being reported for its originality and one of its kind.

Keywords: *Bullous pemphigoid, isotraumatopic non-response, reverse Koebner phenomenon*

Introduction

Heinrich Koebner described the Koebner phenomenon for the first time in psoriasis patients.[1] Since then, it has been reported in various dermatoses. Contrary to the Koebner phenomenon, the reverse Koebner response is the nonappearance or disappearance of the lesions of particular dermatoses at the site of injury. We present to the best of our knowledge the first case of the reverse Koebner phenomenon occurring in a patient with bullous pemphigoid because of pressure.

Case Report

A 35-year-old male presented with a 6-month history of the appearance of multiple discrete fluid-filled lesions over erythematous skin, of size around 0.1–0.2 cm, which gradually increased to a size of around 1–3 cm. The lesions first appeared over the trunk and gradually over other parts of the body. The lesions used to rupture after 5–6 days leaving behind raw erosions, which were nonspreading in nature. The erosions used to get crusted in the next 4–5 days which fall off leaving behind light-colored skin. There was no history of any drug intake before the appearance of the lesion. On cutaneous examination, few tense bullae of size around 0.5 cm were found to be present in the perineal region [Figure 1]. There were multiple healed hypopigmented lesions with perifollicular hyperpigmentation, with some lesions showing crusting over them. A peculiar finding of the distribution of lesions was that the lesions covered the whole body but spared a band-like area in the hip region both in anterior and posterior aspects [Figure 2]. Oral mucosa was not involved. Remaining systemic examinations were within normal limit. A provisional diagnosis of bullous pemphigoid with the reverse Koebner phenomenon was made with the probable cause of sparing being the tightness of clothing in that particular area. Suspecting the same biopsy was sent for histopathology and direct immunofluorescence from both the lesion and the spared area. The histopathology of the lesion showed subepidermal split with a mixed inflammatory infiltrate, which was

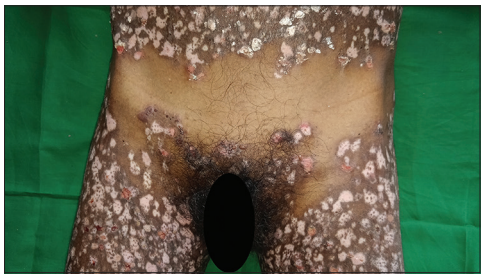

Figure 1: "Bullous pemphigoid" – Tense bullae over the perineal area

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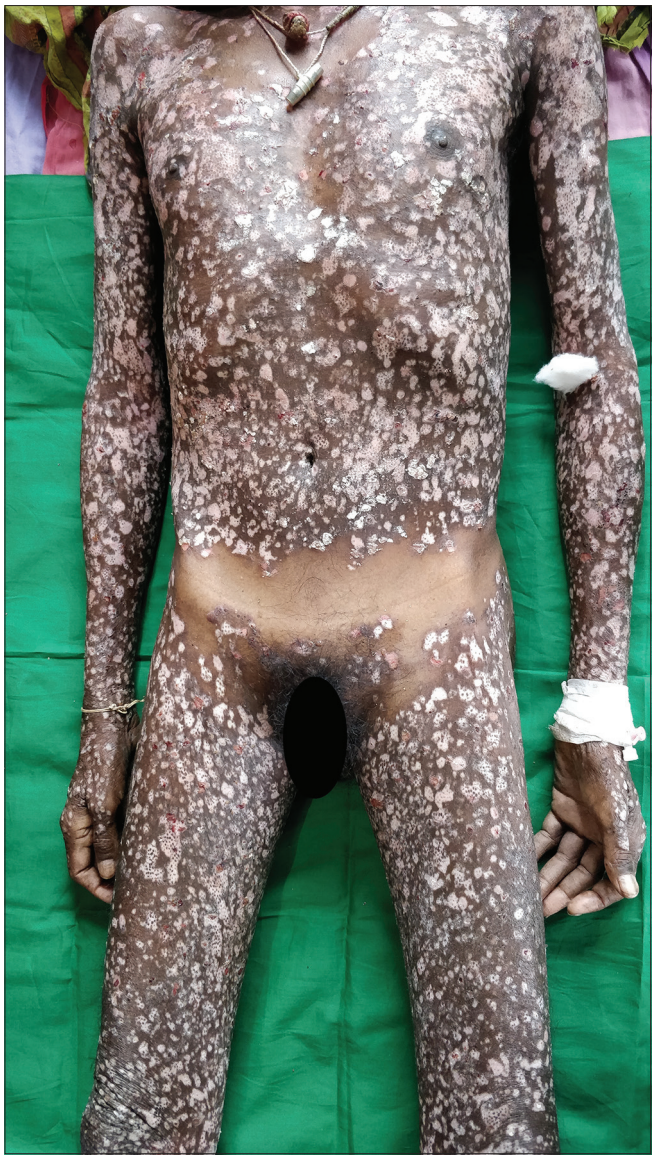

Figure 2: "Bullous pemphigoid" – Sparing of a band-like area in the hip region

composed of predominantly eosinophils, neutrophils, and lymphocytes [Figure 3a], but the histopathology of the spared skin showed no abnormalities [Figure 3b]. However, the direct immunofluorescence from the perilesional area showed linear deposits of immunoglobulin G and C3 in the dermoepidermal junction [Figure 4], but the direct immunofluorescence from the spared area showed no deposits. Indirect immunofluorescence and salt splitting could not be done because of resource constraints. Rest of the laboratory investigations were within normal limits. As the diagnosis of bullous pemphigoid was confirmed, the patient was started with oral prednisolone and oral dapsone.

Discussion

Bullous pemphigoid represents the most common type of autoimmune blistering disorder characterized by autoantibodies located at the dermoepidermal junction

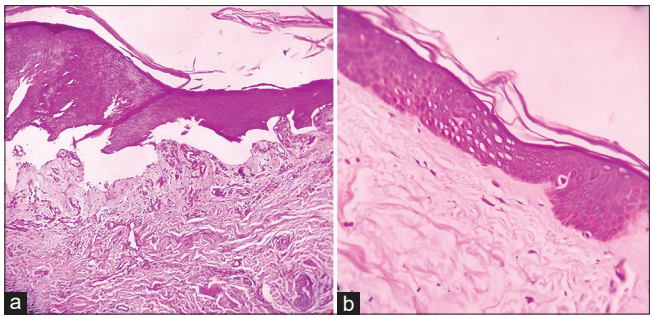

Figure 3: (a) "Bullous pemphigoid" – Histopathology showing subepidermal bulla with a mixed inflammatory infiltrate, which was composed of predominantly eosinophils, neutrophils, and lymphocytes (H and E, 40×). (b) "Bullous pemphigoid" histopathology showing no abnormalities in the spared area (H and E, 40×)

against specific antigens in the hemidesmosomes, thus causing complete separation of the epidermis from the dermis. In bullous pemphigoid, the antigens targeted by circulating autoantibodies have been described as BPAG1 – MW 230 kD and bullous pemphigoid antigen 2 – or type XVII collagen – MW 180 kD.[2] These are key components of the hemidesmosomes and play a major role in cell–matrix adhesion. While both antigens are recognized and targeted by T and B cells in bullous pemphigoid, it has been demonstrated that the NC16A region of BPAG2 is the most commonly targeted by pathogenic antibodies.[2-4] These autoantibodies that bind to the complement lead to the disruption of the dermoepidermal junction.

The exact pathogeneses of Koebner's phenomenon and reverse Koebner's phenomenon are poorly understood.[5-7] Koebner's phenomenon is highly associated with active diseases; however, reverse Koebner's phenomenon is reported in individuals with more stable conditions.[7] A newly defined concept known as locus majoris resistentiae describes a site of the body that offers greater resistance to immunity-related eruptions or skin disorders than the rest of the body owing to a localized immune dysregulation induced by intradermal vaccinations, trauma, infection, mosaicism, radiotherapy, or phototherapy.[6,7] Typically, the sparing phenomenon of locus majoris resistentiae occurs in a previously injured or diseased cutaneous site. Sparing because of local pressure has only been described in one case of leukocytoclastic vasculitis where there was nonappearance of lesions, which was covered by a pressure bandage.[8] We found the sparing phenomenon in bullous pemphigoid in only a single case with sparing of the lower limb and acquired lymphedema secondary to lymph node surgery,[9] but not a single case of sparing because of pressure effect in this available literature. The failure of new lesions to appear at the pressure site could probably be because of the mechanical pressure which led to diminished blood flow in the small vessels of the dermis and the immune complexes failed to deposit in adequate concentration. It was proved by the absence of immune complex deposits in direct immunofluorescence at the spared site.

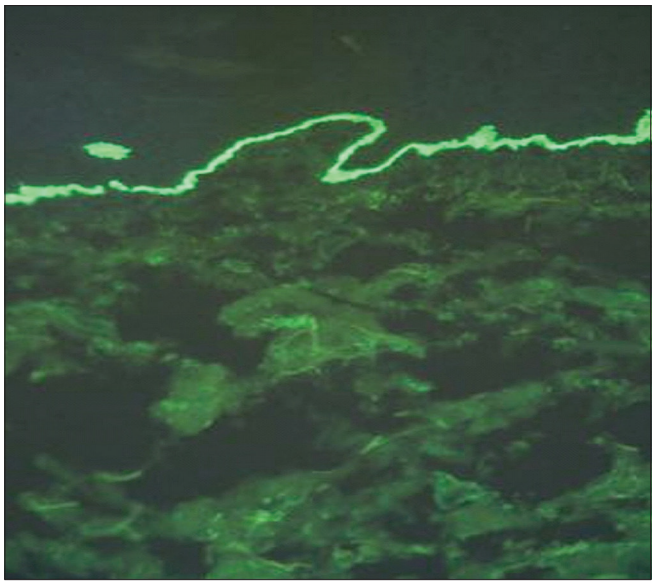

Figure 4: "Bullous pemphigoid" – Direct immunofluorescence study showing linear immunoglobulin G and C3 deposits along the dermoepidermal junction (immunofluorescence, 100×)

Reverse Koebner phenomenon in bullous pemphigoid is a very rare presentation and further studies are required to understand the pathogenesis of this phenomenon.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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