

Psoriasis and Bullous Pemphigoid: Co-occurrence

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Abstract

Psoriasis is a chronic recurrent inflammatory skin disorder. Bullous pemphigoid (BP) is an autoimmune disease characterized by formation of blisters in subepidermal zone. The concurrent occurrence of these two disorders is rare. The pathogenesis behind this occurrence however remains unclear. Here, we are reporting a case of 48-year-old male with psoriasis for 20 years presenting with recent onset BP. Histopathological examination and direct immunofluorescence confirmed the diagnosis of psoriasis and BP.

Key words: Bullous pemphigoid, Histopathology, Immunofluorescence, Methotrexate, Psoriasis

INTRODUCTION

Psoriasis is a T-cell mediated inflammatory disease affecting the skin and joints. Psoriasis has been associated with many other autoimmune diseases such as bullous pemphigoid (BP), linear IgA bullous disorder, epidermolysis bullosa acquisita, lichen planus, rheumatoid arthritis, and atopic dermatitis.^[1] Immunobullous disorders occurring with psoriasis is a rare phenomenon, among those, BP is the most common association. BP is a subepidermal autoimmune bullous disorder. Pathogenic mechanisms proposed for occurrence of BP in psoriasis include epitope spreading and Th1 to Th2 response switch.^[2]

A 48-year-old male presented with complaints of fluid filled lesions over the inner aspect of bilateral forearm, thighs, and back for the past 1 month. History of itching presents before the onset of lesions. He is a known case of psoriasis for past 20 years on irregular treatment. No history of drug intake before onset of illness. No history of similar complaints in family members.

Dermatological examination revealed multiple well defined scaly erythematous to hyperpigmented plaques of varying sizes present over the trunk and extremities with positive auspitz sign [Figure 1a]. Multiple tense clear vesicles and bullae of varying sizes present over the inner aspect of forearm, back, and thighs [Figure 1b]. Blisters were present over the normal skin. Few crusted erosions present over the back. Examination of mucosae was normal. Nikolsky sign was negative and bulla spread sign was positive. Biopsy taken from the plaque revealed hyperkeratosis, parakeratosis, acanthosis, and regular elongation of rete ridges [Figure 2a]. Biopsy taken from the intact vesicle revealed sub epidermal split [Figure 2b]. Direct immunofluorescence of perilesional skin showed linear deposits of IgG and C3 at basement membrane zone [Figures 3 and 4].

Based on the clinical findings, histopathological examination and direct immunofluorescence, we arrived at the diagnosis of BP coexisting with chronic plaque psoriasis.

The patient was started on methotrexate 10 mg/week, Doxycycline 100mg o.d., and topical clobetasol propionate for the blisters for 2 weeks. However, the patient continued to develop new blisters, thus methotrexate dose was escalated to 20 mg/week and tablet nicotinamide 250 mg t.d.s. was added. Psoriatic plaques resolved over a period of 6 weeks and there was a significant reduction in bullae after 10 weeks of treatment [Figure 3a and b].

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Figure 1: (a) Multiple scaly hyperpigmented plaques over the back, (b) multiple tense vesicles over the flexor aspect of forearm, (c) few vesicles with scaly papules over the dorsum of hands

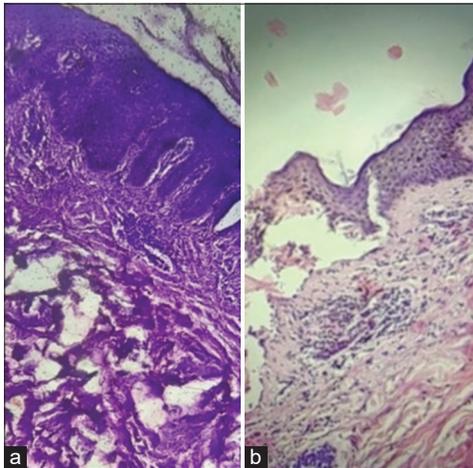


Figure 2: (a) Hyperkeratosis, parakeratosis with regular elongation of rete ridges, (b) biopsy showing sub epidermal split

DISCUSSION

Coexistence of bullous disease and psoriasis was first described by bloom in 1929.^[1] Autoimmune bullous diseases occurring with psoriasis include pemphigus vulgaris, pemphigus foliaceus, pemphigus herpetiformis, cicatricial pemphigoid, BP, linear IgA bullous dermatoses, anti-p 200 pemphigoid, and epidermolysis bullosa acquisita. BP is the most common association with psoriasis vulgaris.

BP patients develop autoantibodies predominantly against BPAG1 (BP 230 kDa) and BPAG2 (BP 180 kDa). Several

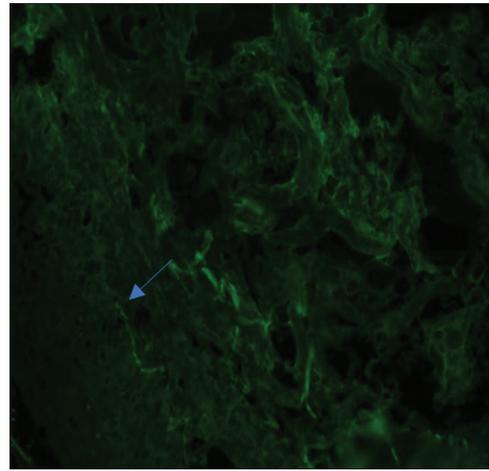


Figure 3: C3 deposits at BMZ

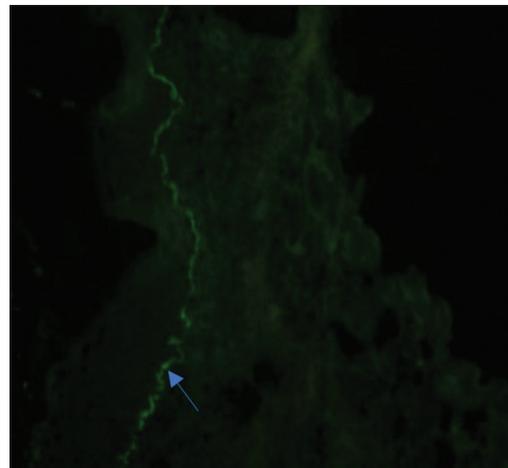


Figure 4: Linear IgG Deposits at BMZ



Figure 5: (a) Hyperpigmented patches over the lower back, (b) resolved vesicles with post inflammatory hypopigmentation

factors such as trauma, drugs, UV light, radiation therapy, and malignancy can trigger BP.

Mechanism behind the concomitant occurrence of psoriasis with BP remains unclear. Theories proposed include, concept of epitope spreading, where the inflammatory process in the epidermis triggers the release of previously unexposed antigen resulting in formation of autoantibodies. Dysregulation of T-cell activity in psoriasis results in induction of specific antibodies to basement membrane antigens. Induction of Th 17 causes switching of Th1 to Th2 response leading to production of autoantibodies. Keratinocytes in both psoriasis and BP produce neutrophil chemo attractants, leading to neutrophil infiltration. Neutrophils produce matrix metalloproteinases, which degrade matrix proteins leading to exposure of surface antigens.^[2]

Methotrexate is an effective drug for BP associated with psoriasis. Other immunosuppressive drugs such as azathioprine, cyclosporine, dapsone, mycophenolate mofetil, and acitretin can be used. Systemic steroids are

usually not advised due to risk of triggering pustular psoriasis.^[3]

CONCLUSION

So far, only few cases of BP occurring concomitantly with psoriasis vulgaris have been reported in literature; hence, we are reporting this case for its rarity.

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