Psoriasis pemphigoides – a rare co-occurrence

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ABSTRACT
Psoriasis and bullous pemphigoid are immunologically mediated diseases. Their concurrent occurrence has been described rarely in literature. The pathogenesis behind concomitant occurrence however remains unclear. The treatment modality of such patients differs from usual. We report a case of a 72 years old man with psoriasis vulgaris presenting with tense bullae, erosions and crusts. Histopathological examination was done to confirm the diagnosis of psoriasis pemphigoides, but the exact etiology could not be identified.

Introduction:
Psoriasis is an inflammatory disease that is immune mediated. Bullous pemphigoid is an autoimmune skin disease which can be acute or chronic, causing formation of blisters between the epidermis and dermis of the skin. Multiple diseases have reported to concurrently occur with psoriasis, such as, lichen planus, atopic dermatitis[1], vitiligo, rheumatoid arthritis and lupus erythematosus[2]. The concurrent occurrence of bullous pemphigoid has been reported in very few cases. Treatment modalities significantly differ in these cases. In the past, such patients have been treated successfully with immunosuppressive agents like methotrexate, dapsone, azathioprine.

Case report:
A 72 years old man presented to our OPD with complaints of scaly lesions all over the body for the past 3 years. There was no history of treatment for the skin complaints, except application of liquid paraffin. He gave history of development of fluid filled skin lesions over the trunk, left thigh and palms for the past 2 weeks, each blister lasting for more than 24 hours before spontaneously eroding. There was no history of itching. No history of any new drug intake prior to onset of skin lesions. No history of loss of weight or loss of appetite. There was no history of similar complaints in the family members. He is a known case of type 2 diabetes mellitus, hypertension, hypothyroidism and coronary artery disease, under treatment for more than 5 years.

Dermatological examination revealed multiple erythematous, scaly plaques over the trunk, all four limbs, and scalp, with positive Auspitz sign. Multiple tense vesicles and bullae were noted over the trunk, left thigh, and bilateral palms, some containing clear fluid and some, hemorrhagic[Figure 1]. Minimal crusting and erosions were noted. Examination of oral mucosa, soles, nails and external genitalia were normal. Nikolsky sign was negative. Bulla spreading sign revealed rounded border. Systemic examination was normal. A clinical diagnosis of psoriasis vulgaris with concomitant occurrence of a bullous disorder, was made. Hematological and biochemical parameters were normal. Two skin biopsies were taken from tense hemorrhagic vesicle over the left thigh and clear fluid filled bulla over the trunk, which revealed hyperkeratosis, parakeratosis, with neutrophilic infiltrates at certain areas, and sub-epidermal bulla containing eosinophilic infiltrates [Figure 2], confirming a diagnosis of psoriasis pemphigoides (pemphigoid in psoriatic plaques).

Discussion:
Concurrent occurrence of bullous disease and psoriasis was first recorded by Bloom in 1929[3]. Since then, several autoimmune bullous disorders associated with psoriasis have been reported in literature, such as pemphigus vulgaris, pemphigus foliaceus[4], myasthenia gravis, Crohn’s disease, ulcerative colitis, linear bullous dermatoses[5], epidermolysis bullosa acquisita[6], rheumatoid arthritis and lupus erythematosus. The first case to be reported in India, was by Raghavendra et al, wherein, the term psoriasis pemphigoides was proposed [7]. The pathogenesis behind co-occurrence of psoriasis and bullous pemphigoid remain unclear. Whether the treatment of psoriasis[8], immune mediated activity in psoriasis itself, or a coincidence of multiple factors precipitate the autoimmune bullous disease remains unclear. The coexistence has been seen in patients who received wide range of treatment for psoriasis as well as in untreated patients, hence it has been difficult to conclude at a single agent as a potential causative factor in the etiology of bullous pemphigoid. In psoriasis, the reduced barrier function of the epidermis, combined with the irritant effects of therapies for psoriasis, such as anthralin, coal tar, and ultraviolet light, together with an immune mediated basement membrane zone insult, may provoke blister formation. Ultraviolet-B rays used in treatment of psoriasis may trigger bullous pemphigoid. PUVA therapy is also known to induce blisters that resemble bullous pemphigoid[9]. Histopathological and immunohistochemical studies aid in making a diagnosis of psoriasis pemphigoides in such cases.
Selection of a treatment modality that will be effective against both bullous pemphigoid and psoriasis vulgaris, is crucial and challenging, especially when patient has co-morbidities like obesity and metabolic syndrome. It is well established that obesity correlates with a higher incidence of psoriasis vulgaris and more severe disease. Obesity also influences response to therapy. Although systemic corticosteroids are effective in bullous pemphigoid, they are contraindicated in view of rebound phenomenon of psoriatic lesions and risk of precipitation of pustular psoriasis. At high-doses, corticosteroid therapy may be life-threatening, particularly in patients with co-morbidities such as obesity, hypertension and diabetes mellitus. Taking into consideration the above-mentioned conditions, it is important to make a choice of suitable treatment modality.

Methotrexate has been used with good effect to treat BP-associated psoriasis. Steroids are not preferred, with the fear of triggering pustular psoriasis. Other drugs that have shown to aid in recovery include dapson, azathioprine, cyclosporine, mycophenolate mofetil[10] and acitretin.

Legends and Figures:

Figure 1
Clinical photographs showing erythematous scaly plaques with tense vesicles and hemorrhagic bullae over trunk(A), lower limbs(B & C), and palms (D).

Figure 2
Photomicrograph showing sub-epidermal split in low power magnification(A), with blister fluid containing collection of eosinophilic infiltrates in high power magnification(B).

References:

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