Lichen planus pemphigoides in coexistence with psoriasis: A case report

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Abstract
Psoriasis vulgaris and lichen planus pemphigoid (LPP) represent two clinically well-characterized, chronic, inflammatory skin conditions. The concomitant occurrence of these two entities in a patient is rare. We hereby report a known case of psoriasis vulgaris later on presenting with clinical picture of lichen planus pemphigoides which was confirmed by histopathological examination and direct immunfluorescence. The etiopathogenesis of the coexistence of these two entities is not well known. Both diseases are immunologically mediated involving both cellular and humoral immune system.
We believe that this is the rare case report of psoriasis vulgaris coexistent with lichen planus pemphigoides.

Keywords: lichen planus, Psoriasis vulgaris, Rare bullous disorder, Co-existence, Concomitant, Lichen planus pemphigoides.

Introduction
Lichen planus pemphigoides is described as a rare bullous disorder in concurrence with typical lichen planus features. It is usually acute in onset and generalized in distribution followed by the sudden appearance of large bullae on both lesional as well as normal skin.[1-4]

Case Report
A 66yr old male patient presented with complaints of multiple dark coloured raised lesions over scalp since past 4 years which was insidious in onset and progressive in nature initially on scalp and gradually progressed to all over the body associated with itching. This is followed by appearance of clear fluid filled lesions over the pre existing raise lesions and over the normal skin (Fig. 1,2.) from past 20 days. The lesions were progressive and associated with moderete to severe pruritis. He was known case of psoriasis vulgaris from past 11 years and was on irregular treatment for same (topical corticosteroids for variable period, T. Methotrexate 7.5mg weekly 5-10 weeks multiple times and narrow band phototherapy – details unknown). He also had complaints of joint pain with waxing and waning history. Joint pain is not associated with localised swelling/morning or evening stiffness. History of photosensitivity is present over scalp, bilateral upper limbs and upper back. There was no history of oral ulcers or dental pathology. History of Drug intake prior to onset of initial lesions was not present. No history of pus filled lesions and fever. No associated co-morbidities were present.

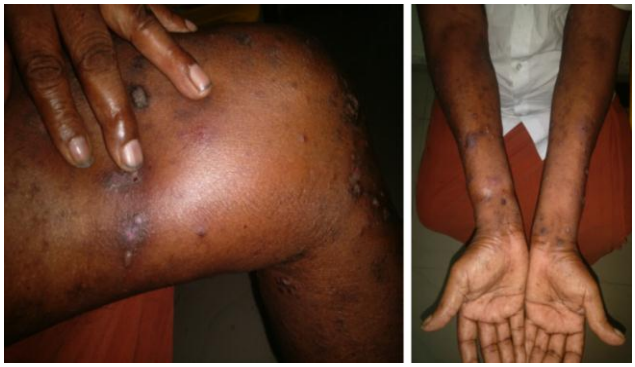

Fig. 1,2: Multiple dark color lesions over thigh (Fig. 1) and both upper limb (Fig. 2).

Clinical Examination
General physical and systemic examinations were normal. Cutaneous examination revealed presence of multiple polygonal, flat topped, scaly violaceous papules and plaques measuring 2cm by 1cm present over anterior aspect of legs, forearms, abdomen, back and the trunk; sparing the face, palms and soles. (Fig. 3). There were multiple vesicles and tense bullae over the pre existing plaques and the normal skin involving flexor aspect of the forearms, abdomen and thighs (Fig. 3-5). Multiple scaly plaques were present over the scalp. Nails and mucosa were unaffected. Oral cavity was normal with poor oral hygiene. Ocular examination was normal with no associated conjunctival congestion or corneal opacity. No abnormality was detected on genital examination. Both the Bulla Spread sign and the Nikolsky sign were negative. The diagnosis of Lichen Planus was made on history and clinical examination. Close differential diagnosis could be psoriasis vulgaris. But morphology of lesions with sites of predilection favours more toward lichen planus.

Patient was explained about the diagnosis alongwith treatment approach and informed consent was taken for further investigations and treatment protocol.

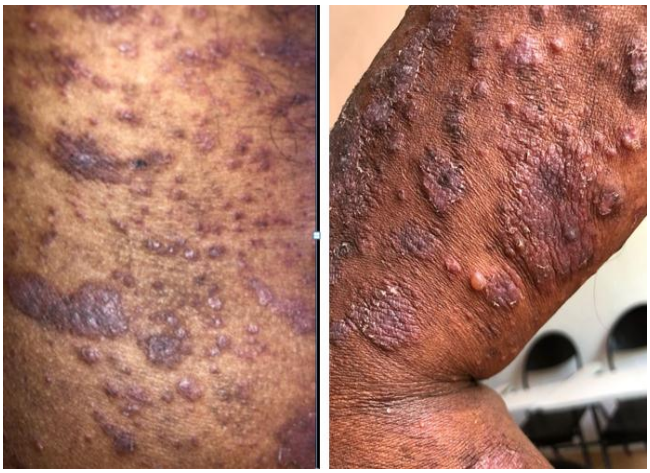

Fig. 3,4: Multiple vesicles and bullae present adjacent to violaceous plaques.

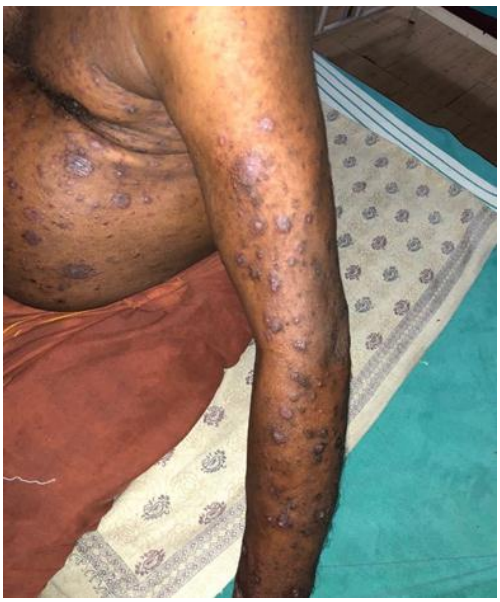

Fig. 5: Multiple erosions and raw areas with few pustules (distal forearm) over the violaceous plaques present on abdomen and right upper limb.

Investigations

Complete hemogram and Other relevant biochemical tests, such as complete urine analysis, serum creatinine, blood urea, uric acid, lipids profile, liver function test, Hepatitis B surface antigen (HbsAg) and anti-HCV (hepatitis C virus) antibodies were within physiological limits. 5mm punch biopsy was taken from the intact vesicle over the plaque on right forearm. Histological examination showed hyperkeratosis and irregular acanthosis in the epidermis and a subepidermal bulla with fibrinous exudates. There were scattered neutrophils, eosinophils and lymphocytes. Upper dermis showed moderate perivascular lymphocytic infiltrate (Fig. 6). Direct immunoflouresence revealed linear deposits of IgG with complement C3 at dermoepidermal junction. Details were not known as immunofluoresence was done outside.

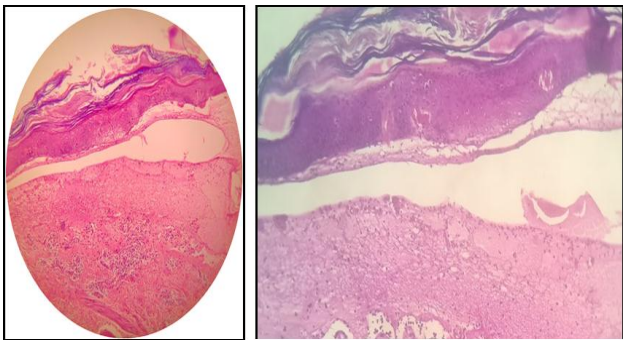

Fig. 6: Photomicrograph showing hyperkeratosis and irregular acanthosis in the epidermis and a supepidermal bulla (H&E stain,×10).

Based on clinical features, morphology of lesions and histo-immunological studies, a final diagnosis of lichen planus pemphigoides was made. Hence the treatment with oral prednisolone (30mg/day) along with topical application of potassium permanganate solution over lesions thrice daily (Condy's crystals mix with warm water in ratio of 1:10000) was initiated. Lesions started resolving as the induration and scaling started reducing without emergence of new lesion. Blisters also started resolving with symptomatic improvement of the patient. Therefore, the dose of prednisolone was tapered gradually (20mg/day). After two days, patient developed multiple erythematous papule and plaques over bilateral legs and trunk with scaling. Skin biopsy from scaly papule showed acanthosis, parakeratosis with elongated club shaped rete ridges and supra papillary thinning (Fig.7). Hence reoccurrence of psoriasis vulgaris was confirmed. Oral prednisolone was stopped after tapering

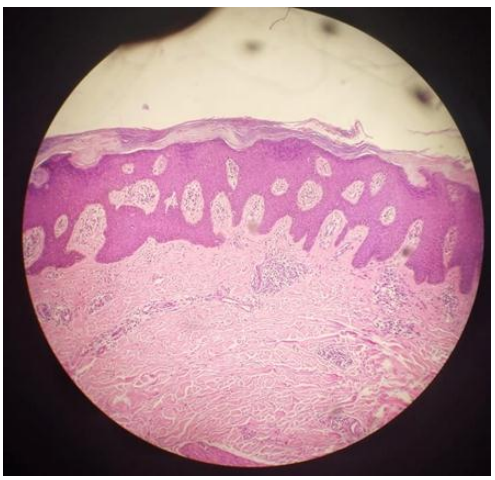

Fig. 7: Photomicrograph showing acanthosis, parakeratosis with elongated club shaped rete ridges and supra papillary thinning (H&E stain, ×10).

And further management was switched to oral methotrexate (7.5mg/week) after its test dose of 5mg oral weekly once and assessing hepatic and renal profile. After 1 month of follow up, the lesions regressed well with residual hyper

pigmentation. Hence, a final diagnosis of Psoriasis vulgaris coexisting with lichen planus pemphigoides was made.

Discussion

Lichen Planus pemphigoides has been speculated as a variant of LP, a coincidental existence of Lichen Planus with Bullous Pemphigoid. Kaposi coined the term "Lichen ruber pemphigoides" in 1892 for combination of a case of typical Lichen Planus with bullous eruption.[5] Lichen Planus Pemphigoides is an acquired autoimmune bullous dermatosis with rare occurrence.[6] LPP is commonly affects males with age of fourth or fifth decade of life.[7] Clinical features includes sudden onset of tense, dome-shaped bullae before, during, or after the evolution of Lichen Planus in a patient. The vesicles or bullae may present over normal skin or over pre existing lichenoid lesions.[8] Here the lesions are present over distal extremities or may present as generalized form. Some reports shows oral and conjunctival mucosa involvement also.[9] Majority of the patients of LPP on an immunoblotting technique showed 180 kD antigen.[10] On histopathologic examination, LPP shows typical histological picture of lichenoid tissue response with subepidermal bullae. Direct immunofluorescence (DIF) of peribullous skin demonstrates linear IgG and C3 deposits along the basement membrane zone (BMZ).[11] Differentials of LPP can be Bullous Lichen Planus which can be differentiated clinically by presence of blisters only over the lesion of Lichen Planus. Bullous LP affects older age group than LPP. We can rule out Bullous LP with help of Histopatholgy, Immunofluorescence (DIF).

Psoriasis is a chronic, multifactorial, T-cell mediated inflammatory disease associated with increase in the epidermal cell turnover rate and hyperproliferation of keratinocytes in the epidermis. Psoriasis may involve multiple environment triggering factors, e.g. trauma, infections, medications, psychological stress along with polygenic predisposition, immune dysfunction and skin barrier disruption and are commonly associated.[13] Systemic corticosteroid is contraindicated in the treatment of psoriasis due to the risk of flaring up of existing psoriasis or sometimes evolution of generalized pustular psoriasis.[14] The concomitant occurrence of Psoriasis and bullous pemphigoid, as two well-characterized, chronic inflammatory skin diseases was first described in the literature in 1929 by Bloom et al. Since then, less than 100 cases were described worldwide.[15]

The concomitance occurrence of Psoriasis and Lichen Planus Pemphigoides is very rare. The co-existence of two diseases is rare. It may be due to the genetic predisposition towards psoriasis providing an unfavourable predisposition to the development of LPP, and vice versa, although evidence for this possibility is not yet proved. Shiohara et al described a case report of coexistence of psoriasis and lichen planus in a patient and based on his immunohistochemical findings suggested a putative role for CD4 T cells and several lymphokines could be involved in the pathogenesis of two diseases.[16] Considering the pathophysiology of lichen planus pemphigoides, epitope-spreading phenomenon may play an important role. LPP could be an association between Lichen Planus and Bullous Pemphigoid, a unique entity, or a heterogeneous response to self antigen of Basement Membrane Zone (BMZ). Epitope spreading phenomenon may present with exposure of different antigens and triggers, like BMZ aggressions, leading to subepidermal blistering.[4] Epitope spreading phenomenon explains the progressive formation of antibodies against more than one antigenic determinant during the course of an autoimmune disease. In the course of disease, progressive tissue injury caused by primary antibodies may results in the unmasking of neighbouring proteins which will lead to generation of secondary antibody responses. These new antibodies may manifest with atypical clinical picture. For example, the transformation of pemphigus foliaceous into pemphigus vulgaris or change from the mucosal form of pemphigus vulgaris to mucocutaneous form. Direct Immunofluroscence (DIF) in lichen planus shows fibrinogen at the basement membrane zone (BMZ) in a shaggy pattern with IgM deposition on the colloid bodies (CB) and shaggy fibrinogen along BMZ, pemphigus group shows IgG in intercellular space in all cases, pemphigoid shows linear deposits of IgG±C3 at BMZ, mucous membrane pemphigoid shows IgG and C3 arranged as continuous band over BMZ and lichen planus pemphigoid shows linear deposits of IgG and the third component of complement at the epidermal BMZ. Indirect immunofluroscence (IIF) presents in lichen planus as MHC Class I antigen called lichen planus specific antigen (LPSA), pemphigus group as Anti-ICS (intercellular space) antibodies in 80% of cases with target antigen as Desmoglein.[3] Pemphgoid shows Anti-BMZ antibodies in 70-80% on IIF with target antigen as BP230 and BP180. LPP under IIF shows unique 200 kD antigen.

Conclusion

This case showed that lichen planus pemphgoides may co-exist with psoriasis vulgaris rarely. This significantly points towards common underlying etiopathogenesis or immune mechanism pathway of two distinct dermatological diseases which needs further research.

Conflict of Interest: None.

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How to cite this article: Khushboo, Subramanian S, Venkateswaran S, Nydile SV, Navein RK, Lichen planus pemphigoides in coexistence with psoriasis: A case report. *Indian J Clin Exp Dermatol* 2019;5(2):176-179.