

CASE REPORT

A RARE CASE OF LICHEN PLANUS PEMPHIGOIDES

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ABSTRACT: Lichen planus pemphigoides describe a rare subset of patients who usually have typical lichen planus and then develop blistering on their lichen planus lesions and in normal skin. Less commonly the blistering antedates the lichen planus. They clinically appear to be a combination of lichen planus and bullous pemphigoid. Oral disease may occur and may resemble either lichen planus or bullous pemphigoid. Histopathologically, lichen planus lesions show features of lichen planus and bullous lesions show features of bullous pemphigoid. D/F is positive in linear pattern with IgG and C3 along the basement membrane zone. Lichen planus pemphigoides is less severe and responds faster and better to steroids as compared to bullous pemphigoid.

KEYWORDS: Lichen planus pemphigoides, Lichen planus, Bullous pemphigoid, Steroids.

INTRODUCTION: Lichen planus pemphigoides is also known as lichen ruber pemphigoides. It was first described by Kaposi in 1892. Lichen planus pemphigoides is characterized by the development of tense blisters atop lesions of lichen planus or the development of vesicles de novo on uninvolved skin or oral mucosa. It can affect all age group, though most commonly seen in 30 -50yrs age. The male to female ratio is 3:2.

CASE REPORT: A 27years old female, homeopath by profession presented in the OPD with complaints of itching all over the body since 8days and blisters all over the body since one day. Patient was alright 8days back when she developed itching all over the body. On day 4, she developed reddish elevated lesions all over the body and on 7th day developed blisters all over the body, predominantly on extremities. There was difficulty in swallowing and burning in micturition. There was no history of malaise or arthralgia. Obstetric history was uneventful.

There was no history of varicella till date. She was diagnosed with lichen planus about one and a half year back. It improved with application of steroid cream and within 3 weeks she was off medications. She developed similar lesions (lichen planus) one month back. This time she took single does of homeopathic drug (details not available), all the lesions subsided. She was asymptomatic for three weeks and then all of a sudden she started getting itching, followed by red colored lesions in 3-4 days and then blisters all over the body.

General examination was within normal limits. Cutaneous examination showed bullae and vesicles on erythematous base as well as on normal skin all over the body, predominantly on forearms and lower limbs. There were violaceous papules and plaques on forearms and legs. On few plaques there were bullae and vesicles. Hyper pigmented patches with erosions were present on buccal as well as genital mucosa. Post inflammatory hyperpigmented patches were present on lower extremities suggestive of previous healed lichen planus. Palms and soles were involved. Nails and hair were normal. General and systemic examinations were within normal limits.

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After history and clinical examination differential diagnosis of Bullous drug reaction and lichen planuspemphigoides were considered. Routine blood investigation was normal. Tzank smear was negative. Serology for Herpes, Hepatitis B, Hepatitis C were negative.

Biopsy specimens were taken from violaceous papule, vesicle on plaque and from perilesions skin of bulla. Histopathological examination & papule was suggestive of lichen planus; from vesicle showed subepidermal cleft with predominant lymphocytic infiltrate in upper dermis. Perilesional lesions also showed dense subepidermal lymphocytic clefts and subepidermal infiltrates suggestive of bullous pemphigoid.

DIF from perilesional skin was showing linear deposition of IgG and C3 at DEJ. From clinical examination, histopathology and immunofluorescence study final diagnosis of lichen planuspemphigoides was made.

Pt. was treated with inj dexamethasone 12mgs daily and switched over to oral steroids on 7th day and gradually tapered and stopped in five weeks. Pt was also put on Dapsone 100 mgs daily and continued for 3 months. Patient responded to the treatment and almost all the lesions subsided within 14 days leaving PIH. At present patient is in remission and following up with us on regular basis.

DISCUSSION: Lichen planuspemphigoides (LPP) is characterized by development of tense blister atop lesions of lichen planus or development of vesicles denovo on uninvolved skin.¹ LPP has been reported to be induced by medication such as cinnarizine, captopril, ramipril, PUVA therapy. Itching sensation is usually an associated symptom. The distribution of blisters is preferentially on distal extremities. Intraoral involvement is a rare occurrence which was present in our case.²

The pathogenesis of LPP involves epitope spreading. Degeneration of basal epidermal cells leads to exposure of the basement membrane antigens. This stimulates the production of circulating autoantibodies, which are similar to those of bullous pemphigoid (BP). These autoantibodies react with an epitope located on the C-terminal Ncl6A domain of BP180kDa antigen with BP200k Da antigen.³ LPP occurs most commonly in adults although it has also been described in children.⁴

The histopathological features of bullae in LPP are variable. Features of both BP and lichen planus may be seen in a bullous specimen. The dermal infiltrate is diffuse, sparse, lichenoid or perivascular. Eosinophilic spongiosis may be featured. The dermal papillary outline i.e. saw tooth appearance is not uncommon.⁷ Cytoid bodies, a feature of lichen planus may also be seen.

The lichenoid lesions typically show histopathologic features indistinguishable from those of lichen planus: orthokeratosis, hypergranulosis, irregular acanthosis, basal cell hydropic degeneration with cytoid body formation, dense chronic inflammatory infiltration and pigment incontinence.⁵ In our case biopsy of the bullae revealed a subepidermal bulla with inflammatory cell infiltration mainly by lymphocytes and eosinophils.

The histological features of lichenoid lesions were similar to lichen planus. The inflammatory cell infiltrations in upper dermis were mainly lymphocytes and eosinophil. The immunofluorescence findings of LPP are similar to those of BP.⁶ Linear staining of BMZ with IgG and C3 has been the commonest findings, although staining with IgM may also be present. In our patient, linear deposition of IgG and C3 at DEJ was found.

LPP differs from BP; clinically the distribution of blisters in LPP is preferentially on the distal extremities, while BP characteristically involves the abdomen and flexor aspects of upper extremities.

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The predominance of IgG1 subclass contrast with the usual finding of IgG4 in classical BP.⁷ Circulating autoantibodies against BMZ are detected in 50% of patients with LPP, while they can be detected in 70% or more of patients with BP.⁸

LPP also differs from bullous lichen planus (BLP). BLP is the term reserved for vesiculobullous lesions that appear in direct relation to present or previous lesions of lichen planus. On the contrary, lesions of LPP could arise on lesions without lichenoid change. Histology of bullae of LPP could show features of BP with no evidence of lichen planus, while BLP lesions show exaggerated BMZ destruction by a typical lichenoid band like upper dermal lymphohistiocytic infiltrate.⁹ In BLP the direct immunofluorescent findings are those of lichen planus with fibrin and patchy IgM and C3, associated with cytoid bodies' formation. However DIF of LPP shows dermoepidermal linear deposition of IgG and C3 in both lichenoid and bullous lesions.

Systemic steroids appears to be the the most effective treatment for LPP.¹⁰

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**Bullae on normal and on lichen planus
Lesions of arms and forearms.**



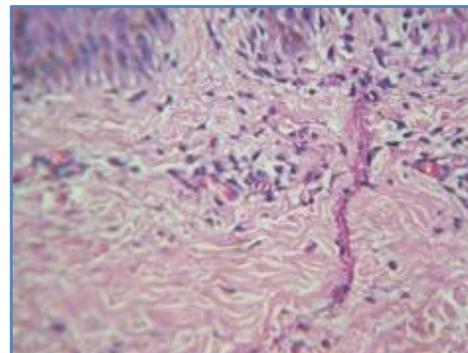
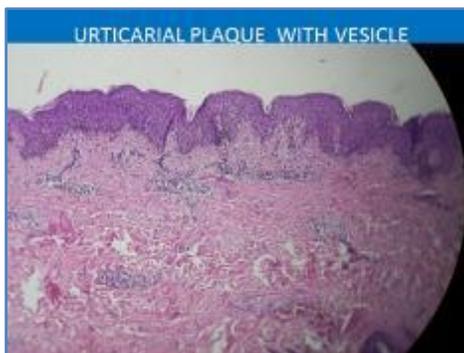
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Bullae on normal and lichen planus lesions of thighs and feet.

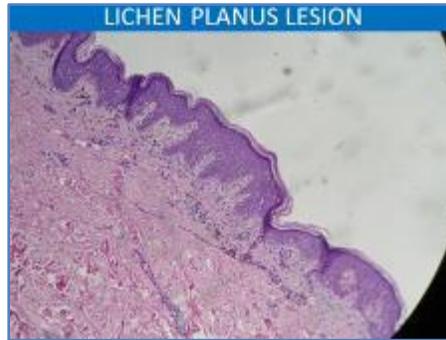


Lichen planus lesions on palms and soles



Histo: 10X

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Histo: 10X

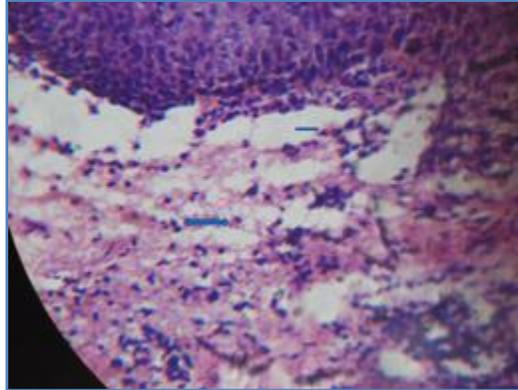


Post treatment photographs



Histo: 10X showing subepidermal vesicle

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Histo : 40X subepidermal vesicle with eosinophils

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