Bullous Lichen Planus vs Lichen Planus Pemphigoides: A Diagnostic Dilemma

Bullous lichen planus (BLP), a variant of lichen planus (LP), and lichen planus pemphigoides (LPP), a variant of bullous pemphigoid (BP) manifest with diverse clinical and histopathological features and hence, pose difficulty in diagnosis. The case is being reported to highlight important clues in history, examination, and investigations, which aid in differentiating these two close mimickers.

A 49-year-old man presented with multiple, itchy raised lesions over upper and lower limbs of 6 weeks duration. Two months later, he developed multiple blisters over the pre-existing lesions, predominately over ankles and wrists. There was no history of any oral lesions/weight loss/fever/any drug intake/aggravation of lesions on sun exposure. On examination, there were multiple discrete, erythematous papules with a violaceous hue, multiple vesicles, and tense bullae on top of these lesions.

Koebner’s phenomenon was also present [Figures 1–3]. Nails, hair, and mucosa did not reveal any abnormality. The patient was evaluated further considering BLP and LPP as two possibilities. Two skin biopsies from a plaque and a bulla were done for HPE and DIF. Histopathology of both was consistent with LP, whereas additionally, the bulla showed a prominent subepidermal blister containing fibrin and mixed inflammatory cells [Figures 4 and 5]. Direct immunofluorescence was negative for IgG, IgM, IgA, or C3 deposits along the dermo-epidermal junction confirming the diagnosis of BLP. The patient was managed by tapering doses of oral steroids and topicals to which he responded well with significant resolution in 3 weeks.

BLP is a rare variant of LP. Both familial and sporadic forms are known to occur, with the former occurring early with prominent nail involvement. BLP is considered as a
hyper-reactive form of LP, where Th1 and Th2 immune reactions mediate the pathogenesis.\(^1\)

Clinically, BLP is characterized by vesicles and tense bullae over pre-existing LP lesions and in the near vicinity with less severe pruritus compared to classical LP. Mucosal lesions usually manifest as shallow erosions as the bullae rupture very fast. Nail involvement in BLP is also frequent and shows characteristic LP features. The course of the disease is variable, and recurrence tends to occur, which can present with classical LP and rarely BLP lesions.\(^2\)

BLP must be clinically and histopathologically differentiated from LPP, which is a sub-epidermal blistering disorder resulting from IgG and C3 autoantibodies directed against basement membrane antigens. First described by Kaposi, LPP was initially considered as a separate entity; however, later it was included as a variant of BP. Prodromal urticarial wheals and pruritus are prominent in LPP, whereas the bullae appear on normal skin not harboring LP lesions. Nail involvement and mucosal LP lesions additionally aid in distinguishing BLP from LPP.\(^3\)

On histopathology, a subepidermal separation is evident, which may be accompanied by interface dermatitis. Colloid and civatte bodies are prominent in BLP, whereas they are rarely seen in LPP. Eosinophils are prominent in LPP, which is a mixed infiltrate in BLP. Direct immunofluorescence is pivotal in distinguishing LPP from BLP and shows C3 and IgG deposits along the basement membrane zone. Immunelectron microscopic studies show deposition of IgG and C3 in the base of bulla and not in the roof as found in BP. On serology, circulating antibodies against BPAG 1 and 2 can also be detected.\(^4\) The course of the disease is less severe than that of BP with less recurrence. Prognosis is good and so is the response to conventional therapy.\(^5\)

It is difficult to distinguish between BLP and LPP solely based on clinical presentation. However, reaching a correct diagnosis is crucial to decide upon the correct management and predicting the clinical course. The aim of this case report was to highlight overlapping features of two rare variants of two very common dermatoses leading to diagnostic dilemmas. The report also emphasizes the importance of DIF studies to differentiate the two entities.

**Ethics approval and consent to participate**

Not applicable.

**Consent for publication**

Obtained.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**Durga M. Tripathy, Deepak Vashisht, Gyanesh Rathore, Prashant Sengupta\(^1\)**

Department of Dermatology, Armed Forces Medical College, Pune, Maharashtra, \(^1\)Department of Pathology, Command Hospital Eastern Command, Kolkata, West Bengal, India

**Address for correspondence:**

Dr. Durga M. Tripathy, Department of Dermatology, Armed Forces Medical College, Pune, Maharashtra - 411 040, India.

E-mail: dmt5861@gmail.com

**References**

1. McCartan BE, Healy C. The reported prevalence of oral lichen planus: A review and critique. J Oral Pathol Med 2008;37:447-53.

2. Wagner G, Rose C, Sachse MM. Clinical variants of lichen planus. J Dtsch Dermatol Ges 2013;11:309-19.
3. Gawkrodger DJ, Stavropoulos PG, McLaren KM, Buxton PK. Bullous lichen planus and lichen planus pemphigoides-clinico-pathological comparisons. Clin Exp Dermatol 1989;14:150-3.
4. Liakopoulou A, Rallis E. Bullous lichen planus–A review. J Dermatol Case Rep 2017;11:1-4.
5. Verma R, Vasudevan B, Kinra P, Vijendran P, Badad A, Singh V. Bullous lichen planus. Indian J Dermatol Venereol Leprol 2014;80:279.