Lichen planus pemphigoides: A case report

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Lichen planus pemphigoides (LPP) is a rare autoimmune blistering disease that is characterized by the development of vesico-bullous skin lesions in patients with lichen planus. The histopathology of LPP reveals a sub_epidermal blister with linear deposition of IgG and /or C3 along the dermal_epidermal junction (DEJ) upon direct immunofluorescence (DIF). We hereunder describe a case of lichen planus pemphigoides in a 69-year-old otherwise healthy male who initially presented with typical lesions of lichen planus (LP), predominantly on the lower extremities; then, bullous lesions developed on the normal skin.

keywords: bullous pemphigoid, lichen planus pemphigoides, lichen planus, bullous disease, direct immunofluorescence

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INTRODUCTION

Lichen planus (LP), the prototype of lichenoid dermatoses, is an idiopathic inflammatory disease of the skin and mucous membranes. Bullous eruptions in LP was first described in 1892 by Kaposi and since then, two distinct forms of LP with bullae have been described [1]. Bullous or vesiculobullous lesions can develop just within pre-existing LP lesions or more randomly, including on the previously uninvolved skin, The former is called bullous LP, while the latter is referred to as LP pemphigoides [2].

CASE REPORT

A 69-year-old otherwise healthy Iranian male initially presented with typical lesions of lichen planus (LP), predominantly on the lower extremities. After 4 months, tense vesicles and bullae appeared on previously uninvolved areas on the shins and thighs (Figure 1,2). The patient denied systemic symptoms and had not taken any new medications. Laboratory results including liver function, urine analysis and stool smear were normal. Three biopsy specimens (lichenoid lesion, bullous and peribullous lesion) were taken for histopathologic examination and immunofluorescence study. Histopathologic examination of a bullous lesion demonstrated typical subepidermal bullae formation without any evidence of acantholysis (Figure 3). Histopathologic examination of lichenoid lesions demonstrated band-like infiltrations, a mild perivascular lymphocytic infiltration and hydropic degeneration of the basal layer as the histologic findings of conventional lichen planus. Direct Immunofluorescence (DIF) of the peribullous skin showed linear deposition of IgG and C3 at the base of a bulla along the dermoepithelial junction (DEJ) (Figure 4). He was treated with oral prednisolone

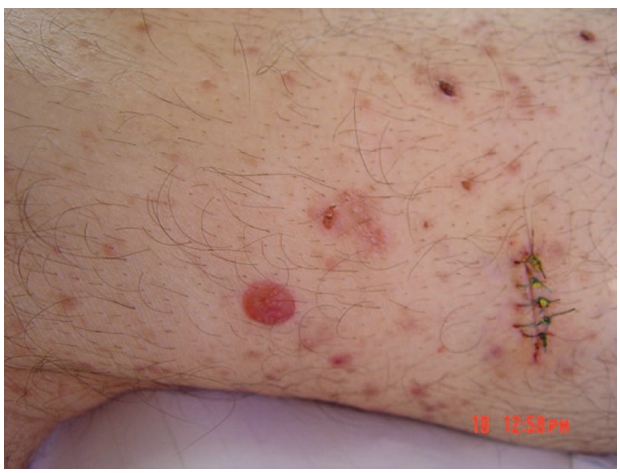

Figure 1. A tense bullae and lichenoid papuleson the lower leg

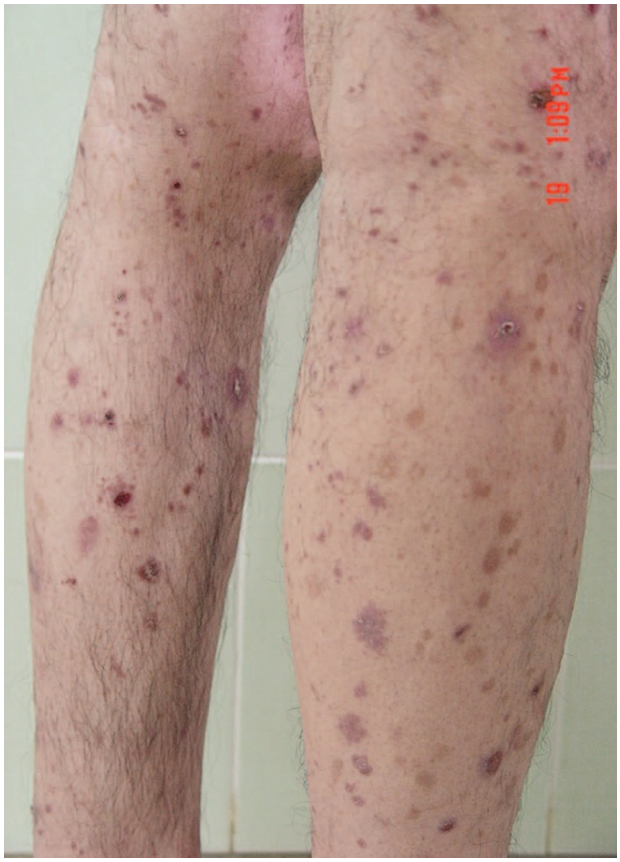

Figure 2. Lichenoid papules, plaques and crusted lesions on the lower extremities.

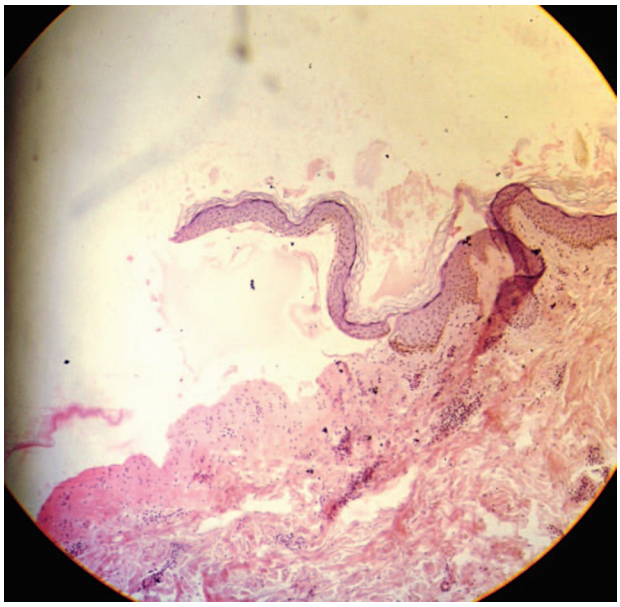

Figure 3. Histopathology view showed subepidermal blister with inflammatory cells (H&E*10).

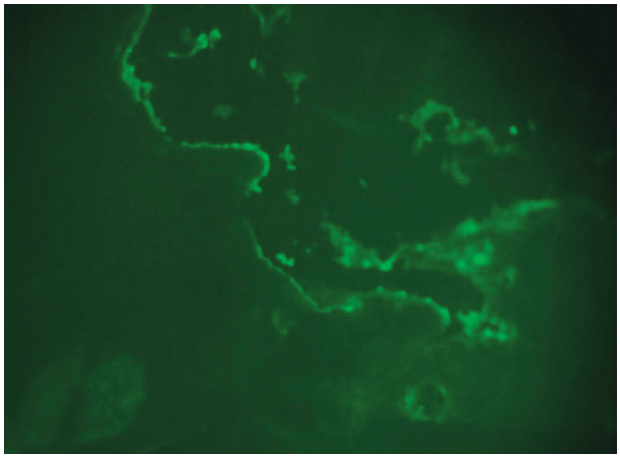

Figure 4. Direct Immunofluorescence (DIF) of the peribullous skin showed linear deposition of IgG and C3 at dermoepithelial junction.

0.5 mg/kg/day, hydroxyzine (25mg/twice daily) and topical clobetasol. Prednisolone was gradually tapered and then discontinued upon healing of the lesion after 8 weeks.

DISCUSSION

Lichen Planus Pemphigoides (LPP) is a rare bullous disease. It may occur at any age, and the incidence of LPP disease is greater in male, (male to female ratio in LPP is 3:2 in adults) [3]. Diagnosis of LPP is based on clinical, histologic, and immunopathologic evaluations. The clinical differential diagnosis of LPP includes Bullous LP and classical bullous pemphigoid [4]. The mean age of onset in LPP is lower than bullous pemphigoid, and the course of the LPP is also less severe than BP [5].

LPP is usually idiopathic, but has been reported after treatment with multiple drugs including Cinnarizine, Captopril, Ramipril, Simvastatin, Furosmide, antituberculous medications, and phototherapy (PUVA) [2,6,7]. It is also associated with internal malignancy [3].

In LPP, histology shows a subepidermal bulla that is not distinguishable from BP [8]. LPP reveals a linear deposition of IgG and/or C3 against 180 kDa (type XVII collagen) BP antigens. However, cases detecting a 200kDa BP antigens have also been reported [9,10-12]. Bouloc et al, reported that the target antigen in LPP was not uniqe [13]. Circulating autoantibodies against basement membrane zone

(BMZ) are often found in more than 50% of cases upon using indirect immunofluorescence (IIF) [13,14]. The cutaneous lesions mostly clear with a low dose of prednisolone [9]. Other treatment options include topical corticosteroids [3], dapsone [3] and azathioprine [5].

Review of recent literature suggests that LPP has a mean age of about 48 years [15] while our case was 69 years old. The distribution of the blisters shows a marked predilection for the distal extremities, similar to our case. Although we did not perform immunoblotting studies, our patient's clinical, histological, and immunofluorescence features were compatible with LPP.

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