BULLOUS LICHEN PLANUS: A CASE REPORT

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ABSTRACT

Lichen planus (LP) is a chronic inflammatory dermatosis involving mucocutaneous surfaces and nails. Bullous lichen planus is a rare variant which usually presents with blisters occurring over typical lesions of lichen planus. Rarely, a few lesions occur in the adjoining skin. It is to be differentiated from lichen planus pemphigoides in which the blisters are more generalized and extensive and the course is prolonged. Here we describe a case of bullous lichen planus in a 40-year-old otherwise healthy female who initially presented with typical lesions of lichen planus predominantly on the lower extremities; later followed by the development of bullous lesions.

KEYWORDS

Lichen Planus, Bullous Lesions, Lichen Planus Pemphigoides, 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. 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.

CASE REPORT

A 40-year-old female patient presented with a 4-months history of insidiously developing multiple, itchy, hyperpigmented, raised lesions over both the legs that gradually spread to the trunk and all extremities over a few weeks. Three months later, a few bullae developed over the pre-existing lesions as well as on normal skin. The bullae ruptured in 2-3 days leaving behind raw areas. There were painful oral lesions since 4 months. No drugs were taken prior to development of the eruption. There was no history of weight loss, fever or exacerbation of the eruption with sun exposure.

There were multiple, discrete to confluent, erythematous to violaceous papules coalescing to form plaques on the extremities and back of the trunk. [Figure 1]. There were few tense bullae filled with clear and haemorrhagic fluid present over the extremities. These bullae were present on the violaceous lesions as well as on normal skin [Figure 2]. Bulla spread sign and Nikolsky's sign were both negative. Oral cavity showed reticulate lesions over buccal mucosa. Nails and hair were normal. General and systemic examination did not reveal any abnormalities. Based on these findings, the differential diagnoses of Bullous Lichen Planus and Lichen Planus Pemphigoides were made and the patient had been investigated.

Routine blood tests and urinalysis were normal. Tzanck smear from the bullous lesion negative for acantholytic cells. Skin biopsy from a papule showed features of lichen planus. Biopsy of a tense blister overlying a violaceous papule revealed parakeratotic epidermis with subepidermal bullae & clefts showing few RBCs and eosinophils, upper dermis showed moderate perivascular infiltrate of lymphocytes along with occasional eosinophils. There were lichenoid changes with basal cell vacuolar degeneration, colloid bodies and dermo-epidermal lymphocytic infiltrate. Direct immunofluorescence study showed no immunoreactants at the basement membrane zone. A final diagnosis of bullous lichen planus was made and the patient was treated with oral prednisolone 40 mg/day, which was tapered off in 6 weeks.

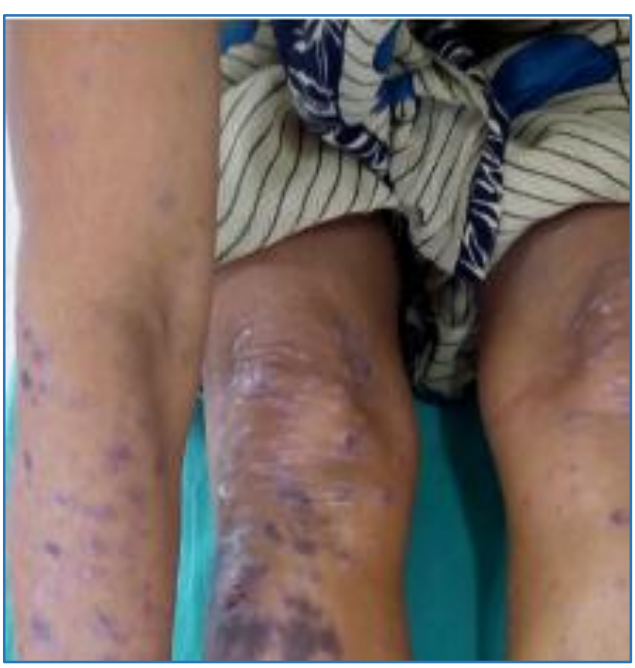

Fig. 1

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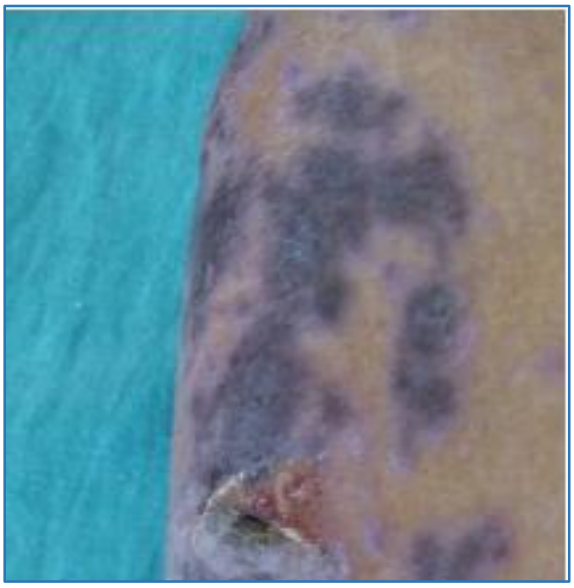

Fig. 2

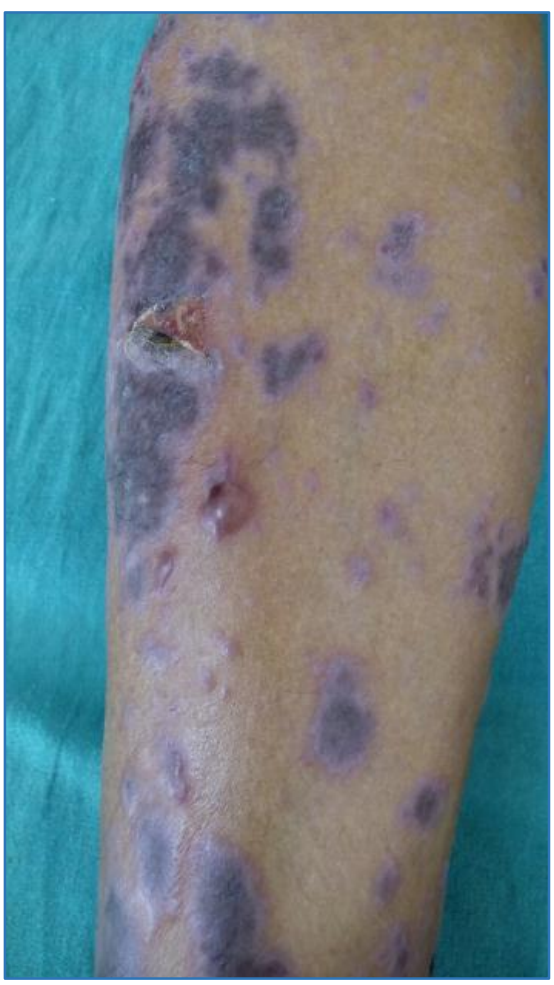

Fig. 3

DISCUSSION

Lichen Planus (LP) is a relatively common chronic inflammatory, papulosquamous and presumably autoimmune disease that affects the skin, mucous membranes, nails, and the scalp.[1] There are many clinical variants of LP: actinic, annular, atrophic, LP hypertrophic (LPH), guttate, linear, LP pigmentosus, erosive (Ulcerative), follicular (lichen planopilaris), bullous LP and LP pemphigoides (LPP).[2]

Bullous lichen planus is commonly seen on the legs. Blisters are tense and may be multilocular. Bulla formation may be due to the extensive liquefaction and vacuolation of the basal layer. Histopathologically, biopsy from the bullous lesion is characterized by a subepidermal bulla accompanied by classical changes of lichen planus. Direct and indirect immunofluorescence testing is negative.[3]

Lichen planus pemphigoides is characterized by bullae developing over lesions of lichen planus and also on normal appearing and erythematous skin, especially during an acute episode of lichen planus. As in bullous lichen planus, the bulla is subepidermal but features of lichen planus are not evident and inflammatory cells are mainly neutrophils and lymphocytes with a few eosinophils. Direct immune-fluorescence testing of perilesional skin shows linear deposits of C3 and IgG along the basement membrane zone.[4] Circulating antibodies have been demonstrated against 130, 200 and 180 kda antigens.[5] Damage to the basal cells could unmask hidden antigenic determinants or create new antigens thus leading to autoantibody formation. Disturbances in local control of immune regulation may also be responsible. Healing may leave behind post-inflammatory hyperpigmentation.

In our patient, the appearance of bullous lesions on normal appearing skin away from the papules of lichen planus created a clinical impression of lichen planus pemphigoides. However, histopathology and immunofluorescence confirmed the diagnosis of bullous lichen planus.

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