Case report

J Shaheed Suhrawardy Med Coll, 2012;4(1):35-37 ISSN 2226-5368

Lichen Planus Pemphigoides: A Case Report

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Abstract

Lichen planus pemphigoides describe a rare subset of patients who usually have typical lichen planus 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 resemble either lichen planus or bullous pemphigoid. Lichen planus pemphigoides has been triggered by medication & PUVA. Pruritus may be severe and lesions may evolve to resemble pemphigoid nodularis. Histopathologically lichen planus lesions show lichen planus and bullous lesion shows the features of bullous pemphigoid. DIF is positive in a linear pattern with IgG and C3 along the basement membrane zone, at the roof of saline split skin. The antigen targeted by the autoantibody in Lichen planus pemphigoides is located in the same region as the bullous pemphigoid antigen (at the basal hemidesmosomes). Lichen planus pemphigoides tends to follow a benign and chronic course, even when compared to bullous pemphigoid. We diagnosed a case of Lichen planus pemphigoides on the basis of history, clinical examination, histopathology & DIF. The patient was treated with systemic & topical steroid, Dapsone. After 2 month of treatment steroid was withdrawn, but Dapsone continue with no relapse.To our knowledge this is the first diagnosed and treated case in this hospital.

Key Words: Lichen planus pemphigoides, bullous lichen planus, Dapsone

Introduction

Lichen planus pemphigoides (LPP) is a rare condition characterized by the coexistence of bullae and lichen planus, in which the bullae may occur on both clinically normal skin and that involved by lichen planus, sub-epidermal bullae in histopathology and linear deposists of IgG and C3 along Basement membrane zone on DIF[1]. Although Lichen planus pemphigoides (LPP) and bullous pemphigoid manifest in different ways clinically, the same antigen may be involved in the immunopathogenesis of these two diseases.

Case Report

In 10th April 2012, a 40 years old lady presented to the Department of Dermatology and Venereology, Shaheed Suhrawardi Medical College & Hospital , Dhaka, Bangladesh with 3 months history of pruritic papulo-vesico bullous skin lesion on both lower extremities with history of diabetis mellitus. She had been treated with various drugs, like antibiotics, antihistamine, analgesic and topical steroid cream with no improvement. On examination we found bilateral nearly symmetrical multiple ,discrete,violaceous ,prutitic ,papules,plaques and

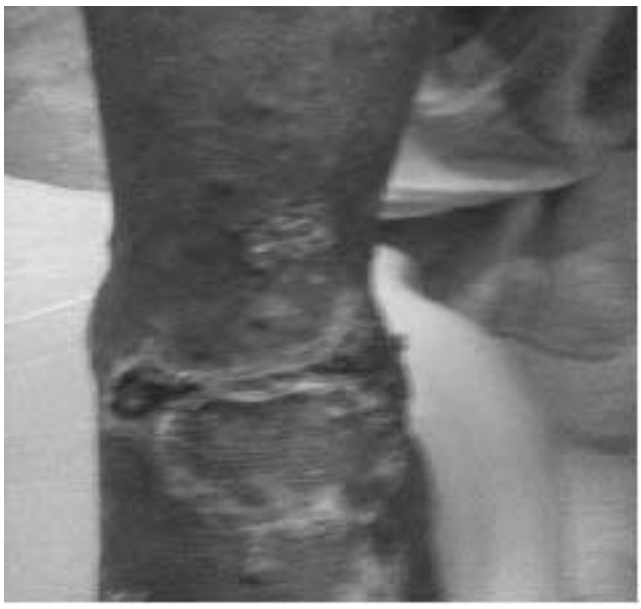

Figure 1a: Violaceous plaques & eroded areas before treatment.

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tense bullae of various size and shape located over the both lower extremites.Bullae appear on the papules,plaques and uninvolved skin.Some of the bullae ruptured and produce oozing, crusting.

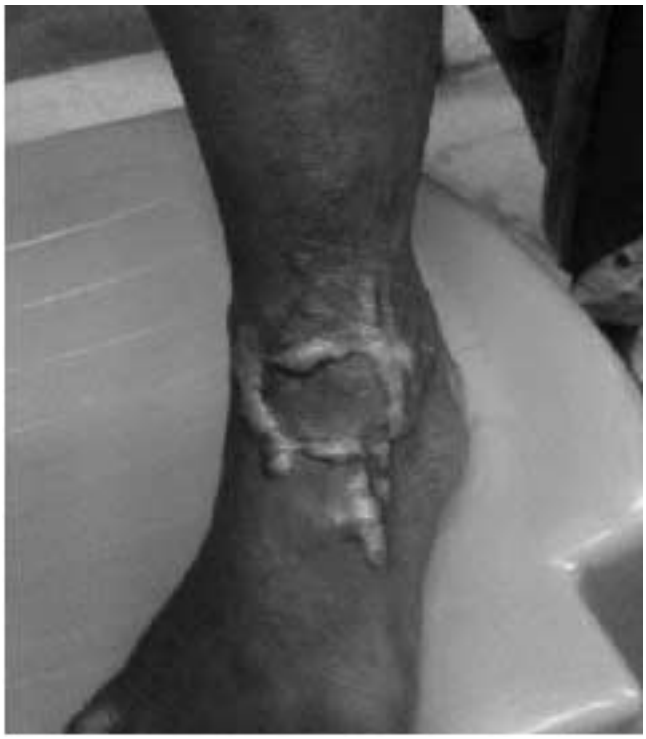

Figure 1b : Development of multiple keloids after treatment

A Skin biopsy was taken from a bullae on the plaques showed hyperkeratosis, focal hypergranulosis, irregular acanthosis and sub epidermal bullae containing fibrin and inflammatory cells including neutrrphils and eosinophils . Direct immunofluorescence of perilesional skin found linear deposition of IgG autoantibodies and C3 along the basement membrane zone (BMZ). The histopathology and immunofluorescence was consistent with the diagnosis of lichen plnus pemphigoides(LPP). Circulating anti-BMZ antibodies titre detected by indirect immunofluorescence technique in LPP could not be done due to unavailability in our country. Other haematology and biochemical parameter were normal except fasting blood sugar. Based on clinical and laboratory, a diagnosis of Lichen planus pemphigoides (LPP) with diabetis mellitus was made. Systemic treatment with prednisolone 50mg/day, dapsone 100mg/day and topical treatment with clobetasol with mupirocin and injection insulin and other supportive treatment resulted in regression of the disease. The dose of steroid was gradually tapered and stopped after 2 months. No clinical relapse was found. The dose of dapsone still continues as 100mg/day.

Figure 1c: Multiple tense bullae appearing on normal skin

Discussion

Lichen planus pemphigoides (LPP) is characterized by development of tense blister atop lesions of lichen planus or development of vesicles denovo on uninvolved skin[1,23]. LPP has been reported to be induced by medication such as cinnarizine, Captopril, ramipril, psoralen & Ultraviolet A therapy[3,4,5]. The aeitology of LPP remains unclear but the current controversy centres on whether LPP is a distinct entity or whether it represents the coexistence of lichen planus and bullous pemphigoid[2,4]. The primary event may be non-specific cell mediated basal layer damage which then exposes immunogenic auto antigens or alternatively specific auto antibody binding may lead to dermoepidermal separation[2,4]. In comparison with classical bullous pemphigoid, the course of LPP is less severe[5,6,7].

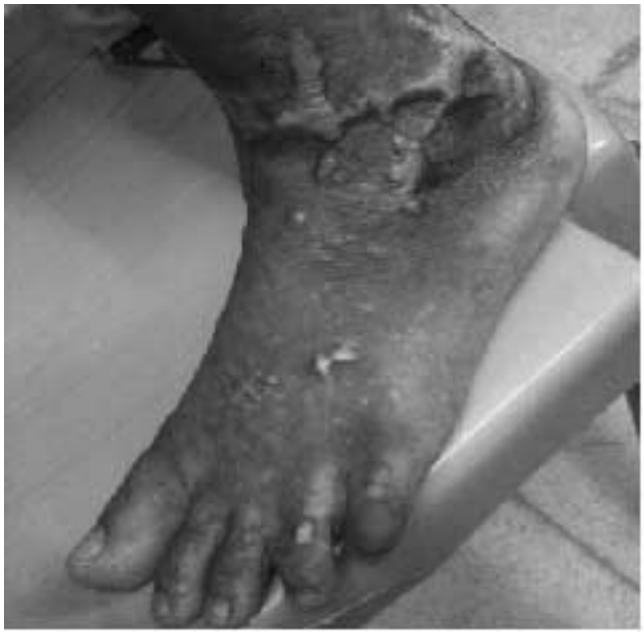

Figure 1d: Development of multiple keloids and post inflammatory hyperpigmentation after treatment

LPP may predominantly affect a younger age group and is responsive to standard treatments used in acquired auto-immune bullous disease[8]. LPP may be treated with topical steroids, systemic steroids , tetracycline & nicotinamide, isotretinoin, dapsone with immunosuppressive drugs with variable success[5,7,9,10,11].

This patient started with systemic prednisolone 50 mg/day, Dapsone 100mg/day, Inj. Insulin , topical clobetasol with mupirocin and other supportive treatment resulted regression of the disease. The steroid was tapered gradually and stopped after 2 month but Dapsone continues as starting. No clinical relapse was seen during the follow up period and her diabetic status also controlled. There was brownish pigmented area as well as little keloid formation at some of the lesion areas.

Conclusion

There is a suggestion that any patient with lichen planus may turn into or will develop Lichen planus pemphigoides (LPP). Bullous lichen planus pemphigoid may develop post inflammatory hyperpigmentation and/or keloid.

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