Pruritic skin blisters: what could be the diagnosis?

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INTRODUCTION

Lichen planus is a chronic inflammatory disorder that is seen in skin, oral mucosa and nails, with its bullous form being a rare variant.

Materials and Methods

A 71-year-old female patient, presented with pruritic bullous lesions evolving over 3 years. The skin examination showed violaceous erythematous plaques on the forearms, inframammary fold, abdomen and legs. Legs plaques were surmounted by tense serous filled blisters (figure 1). Nikolsky sign was negative, post-bullous erosions were also noted, with an estimated body surface area involvement of 6%. Pigmented macules were visible on the buccal mucosa. Dermoscopy showed an erythematous-violaceous background, Wickam's striae, grayish blue globules, a pigmented pattern, erosions, whitish scales and a peripheral rainbow-like pattern (figure 2-3).

Histopathological examination revealed acanthotic epidermal lining with notably increased hypergranulosis, covered by lamellar orthokeratotic hyperkeratosis. Subepidermal detachment containing serous and mildly inflammatory content with a few eosinophilic polymorphonuclear cells. Dermis exhibited a lympho-histiocytic inflammatory infiltrate arranged in a discontinuous band-like pattern along the epidermis, sharply abutting the basal layer without subepidermal detachment or keratinocyte necrosis. The infiltrate was associated with focal pigment incontinence. Direct immunofluorescence was negative.

The diagnosis of bullous lichen planus was established and a biological assessment was conducted, revealing normal results. The patient was treated with very high-potency topical corticosteroids, applied once daily for one month on the lesions followed by a gradual reduction.

During the follow-up appointment, the patient reported relief from itching. No new bullous lesions were observed.

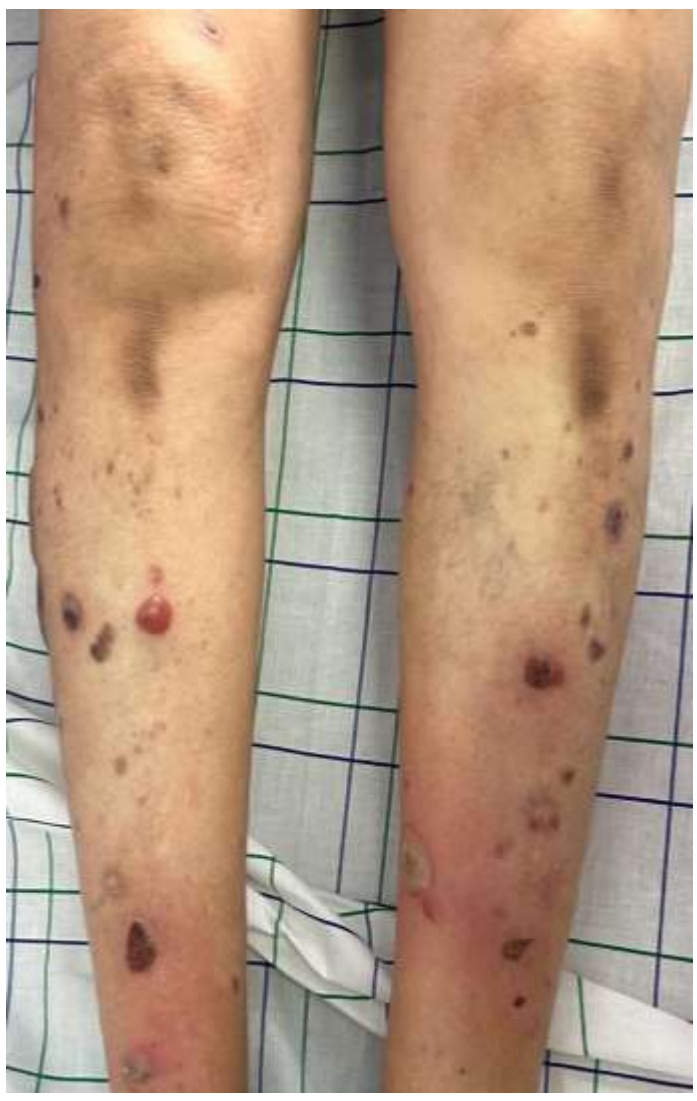

Figure 1

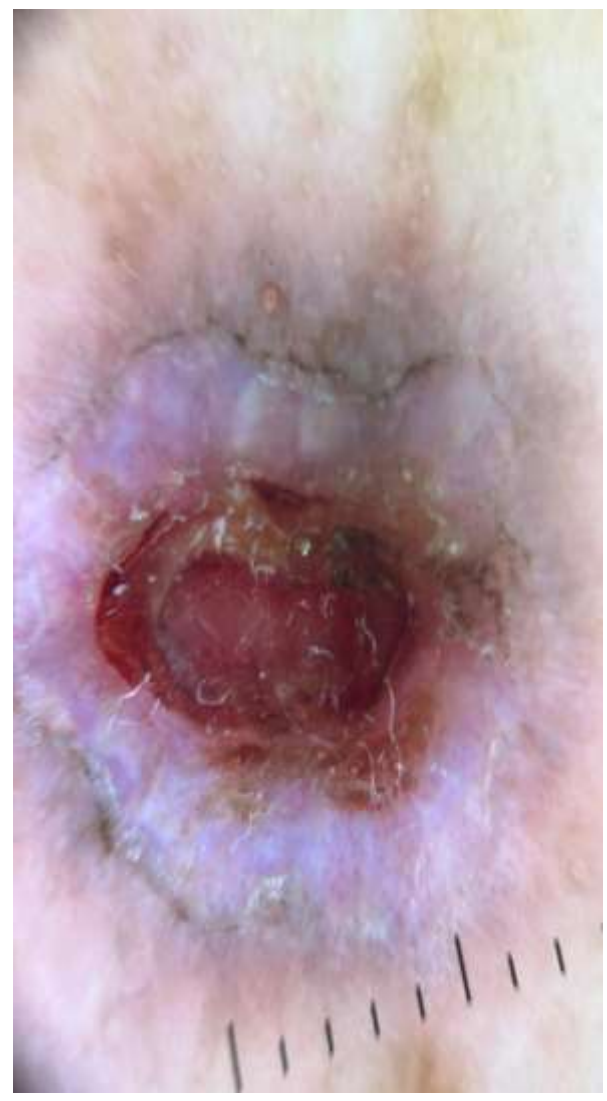

Figure 2

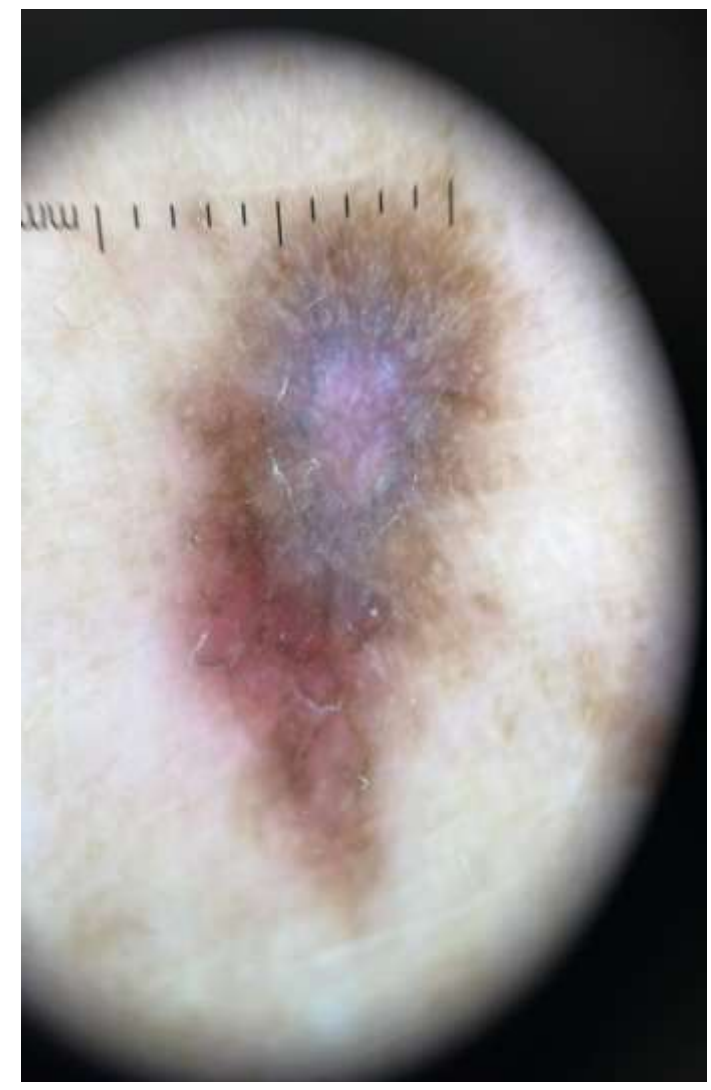

Figure 3

DISCUSSION

Bullous lichen planus is a rare variant of Lichen Planus. Both familial and sporadic forms are known to occur[1].

Clinically, BLP is identified by the presence of tense blisters or vesicles over pre-existing LP lesions accompanied with less sever pruritus compared to classical LP[2].

The characteristic histopathological features of LP consist of hyperkeratosis, increased granular cell layer, acanthosis, liquefaction of basal cell layer and presence of band-like inflammatory cell infiltrate at the dermo-epidermal junction. Apoptotic keratinocytes can be found at the lower dermis and superficial epidermis. Vacuolar degeneration of the basal layer is typical. Often there is presence of pigment incontinence with numerous dermal macrophages[3]. The direct immunofluorescence is negative.

The main differential diagnosis is lichen planus pemphigoides.

There is no established treatment of choice for BLP. Topical potent corticosteroids, systemic corticosteroids, dapsone and acitretin have been described as efficient[4].

CONCLUSION

Bullous lichen planus is a rare entity, and its clinical resemblance to pemphigoid lichen lanus creates a diagnostic dilemma in dermatology.

Only a meticulous clinical, dermoscopic and histological approach allows for an informed differentiation between these two conditions.

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