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Pityriasis lichenoides acuta (PLEVA) pemphigoides: A rare bullous variant of PLEVA

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Abstract

Although the clinical presentations of patients with pityriasis lichenoides et varioliformis acuta (PLEVA) may vary, bullae are not usually part of the clinical spectrum. To date, only two other cases of a bullous variant of PLEVA with evidence of autoantibodies against hemidesmosomal antigens have been reported. The term PLEVA pemphigoides was suggested for this unique clinical, pathological and serological combination of both PLEVA and bullous pemphigoid.

KEYWORDS

blisters, BP180, dapsone, Pityriasis lichenoides

1 | INTRODUCTION

Pityriasis lichenoides et varioliformis acuta (PLEVA) is a rare skin disease of unknown etiology. Although the clinical presentations may vary, bullae are rarely seen in PLEVA.[1]

2 | CASE REPORT

A 6-year-old, otherwise healthy boy presented with a two-month history of pruritic skin eruptions on the neck and upper trunk. He had no history of fever, infection, immunizations, or medication use during the two months prior to the onset of the skin eruption. On examination, there were generalized erythematous macules, papules, and papulovesicles as well as erosions, crusts, and scaling, predominantly on the trunk and neck (Figure 1). The white blood cell count, C-reactive protein as well as creatinine and liver enzyme levels were all in normal range. Skin biopsy including lesional and perilesional skin on the neck showed an interface dermatitis with necrosis of the epidermal basal layer with no intra- or subepidermal blistering (Figure 2). Direct immunofluorescence (DIF) showed IgM antibodies as well as C3 staining of the dermoepidermal junction. Enzyme-linked immunosorbent assay (ELISA) was negative. Given the clinical and histopathological findings, a diagnosis of PLEVA was made. Topical steroids were initiated, but despite this, the skin lesions progressed. Therefore, therapy was escalated to systemic prednisolone (0.5 mg/kg daily) and erythromycin (30 mg/kg/d). After initial improvement, two weeks later, the patient developed tense bullae mainly on the trunk and legs (Figure 3). Repeat ELISA detected autoantibodies against the NCA16 domain of BP180 (45 U/mL, norm < 20 U/mL). Repeat skin biopsy for DIF was declined by the patient's parents. Erythromycin was discontinued and low-dose oral dapsone (0.5 mg/kg daily) was initiated, while oral prednisolone was slowly tapered. Rapid improvement ensued with dapsone, with no new lesions, and gradual resolution of existing lesions, leaving only hypopigmented macules.

3 | DISCUSSION

Although the clinical presentation may vary, bullae are not typically part of the clinical spectrum of PLEVA.[1] A comprehensive PubMed search, using the search terms “pityriasis lichenoides pemphigoides,” “pityriasis lichenoides” and “blisters” or “bullous” revealed

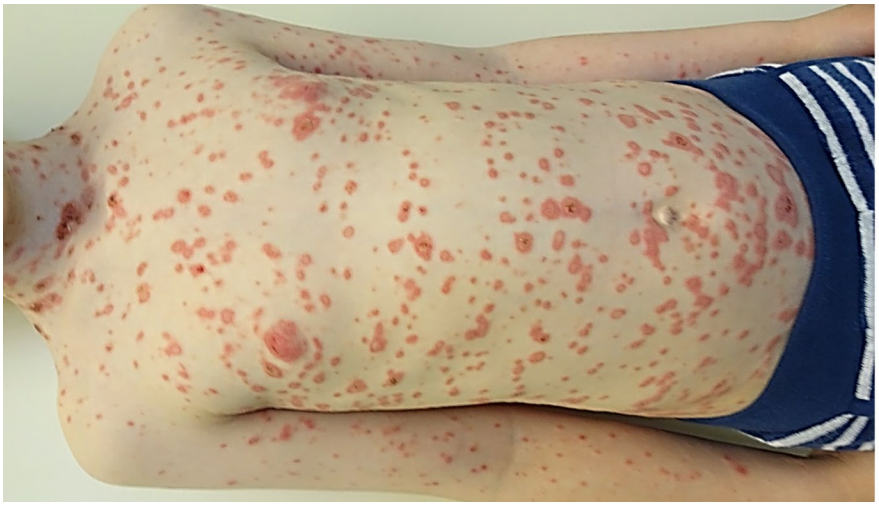

FIGURE 1 Initial presentation with erythematous macules, papules, and papulovesicles as well as erosions, crusts, and scaling

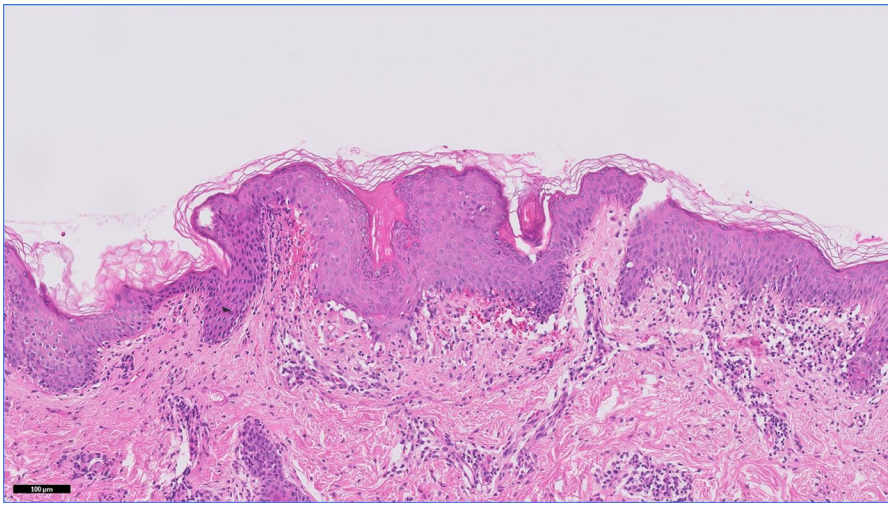

FIGURE 2 Histopathology (hematoxylin and eosin stain) of lesional skin showing interface dermatitis with necrosis of the epidermal basal layer with no intra- or subepidermal blistering (magnification ×10)

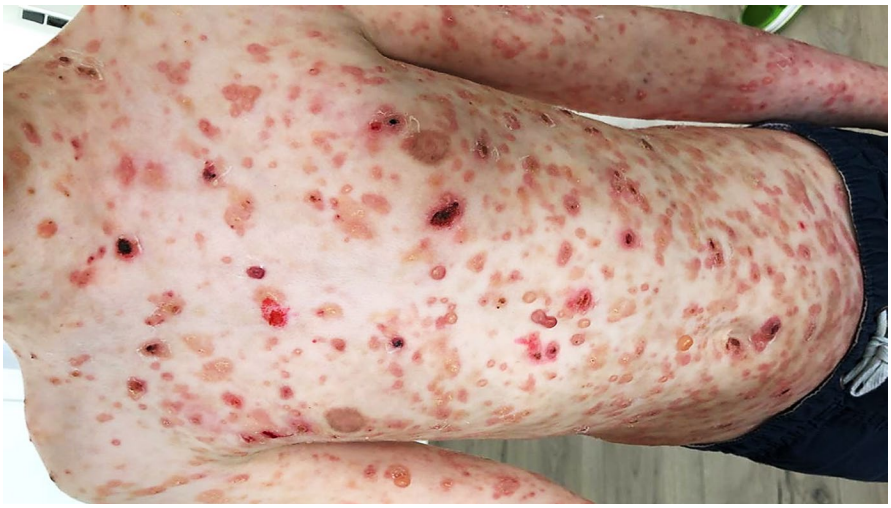

FIGURE 3 Development of tense blisters on the trunk and legs

only two other cases of PLEVA with bullae in children (Table 1).[2,3] Histopathology in these two cases was consistent with pityriasis lichenoides, showing epidermal necrosis and basal layer degeneration. In one case, DIF revealed C3 staining at the dermoepidermal junction without binding autoantibodies. However, ELISA detected circulating IgG autoantibodies to BP180 in both cases. In our case, initial ELISA testing in the early, pre-bullous stage of the disease was negative. When bullae developed, repeat ELISA detected circulating

TABLE 1 Known cases of PLEVA pemphigoides

Authors/Year	Nobbe (2013)[6]	Jiao (2018)[7]	Current patient
Age (y)/Gender	12/male	3/male	6/male
Histopathology	Interface dermatitis and apoptotic keratinocytes	Epidermal necrosis with hydropic degeneration of basal layer	Interface dermatitis and necrosis of the epidermal basal layer
DIF	C3 staining at dermoepidermal junction	Not documented	IgM and C3 staining at dermoepidermal junction
ELISA	BP180 IgG antibodies	BP180 IgG antibodies	BP180 IgG antibodies
Systemic symptoms	Sore throat, fever	Fever	None
Systemic treatment	Prednisolone (0.5 mg/kg/d) and dapsone (1.5 mg/kg/d)	Prednisolone (10 mg/kg/d) followed by methotrexate (5 mg/wk)	Prednisolone (0.5 mg/kg/d) and erythromycin (30 mg/kg/d) followed by dapsone 0.5 mg/kg/d)
Outcome	Complete remission	Complete remission	Complete remission

autoantibodies against the NCA16 domain of BP180, as seen most frequently in childhood bullous pemphigoid (BP).[4] We also found an additional report describing very similar vesiculobullous lesions in a case of PLEVA. This patient had severe systemic symptoms and was diagnosed with the febrile ulceronecrotic variant of Mucha-Habermann disease. The authors in this case suggested that the bullous lesions resulted from massive lymphocytic infiltration of the epidermis leading to intraepidermal edema. Neither DIF or IIF was conducted in that case.[5]

In our patient, despite initial therapy with systemic corticosteroids and erythromycin, disease progression occurred with the development of tense bullae. Due to the clinical and diagnostic similarities to BP, dapsone, a known safe and effective treatment for childhood and adult BP, was initiated with a successful outcome.[4] Of the other two published reports, one patient received oral prednisolone and dapsone, while the other was given oral prednisolone and methotrexate, both with clinical resolution (Table 1).

Nobbe et al suggested the term PLEVA pemphigoides for this rare clinical variant of PLEVA with bullae, analogous to lichen planus pemphigoides, which has overlapping clinical, pathological and serological features of lichen planus and BP.[6] The initial lack of autoantibodies in the IIF in our patient supports the hypothesis of Nobbe et al as well as Jiao L. et al that the phenomenon of epitope spreading, caused by inflammation, cell necrosis and the release of cellular structures, likely led to the synthesis of autoantibodies against hemidesmosomal structures[2,3] This phenomenon has been reported in other diseases such as isolated BP.[7] Another possibility, albeit less probable, would be the concurrent existence of PLEVA and BP, independently of each other. Although this cannot be ruled out entirely, the chronology of the skin lesions favors the hypothesis of a distinct disease entity, PLEVA pemphigoides, due to epitope spreading, which in our case responded readily to a combination of a systemic glucocorticoid and dapsone.

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