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Childhood Vesicular Pemphigoid Mimicking Severe Atopic Dermatitis: A Case Report

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Bullous pemphigoid (BP) is an autoimmune blistering disorder that typically affects elderly patients. Rarely, it can occur in childhood. Vesicular pemphigoid is an atypical variant of BP. We report a case of childhood vesicular pemphigoid in an infant aged 6 months that was initially misdiagnosed as severe atopic dermatitis (AD). To the best of our knowledge, only one other case of childhood vesicular pemphigoid has been reported in the literature.

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Bullous pemphigoid (BP) is an autoimmune blistering disorder that typically affects elderly patients.[1] Rarely, it can occur in childhood.[1,2] We report a case of childhood vesicular pemphigoid, an atypical variant of BP, in an infant aged 6 months that was initially misdiagnosed as severe atopic dermatitis (AD).

Case Report

An infant aged 6 months presented with pruritic, erythematous, vesicular lesions that had evolved over the past 2 months. The patient's mother had a history of AD. The lesions first developed on the abdomen (Figure 1) and rapidly spread to the rest of the body, with marked lesions on the trunk, face, palms, and soles. Physical examination revealed minor signs of AD, including Dennie-Morgan fold, facial pallor, infraorbital darkening, hyperlinear palms, and white dermographism. Laboratory test results revealed an elevated eosinophil count (1500/µL; reference range, 0–450/µL) and an elevated IgE level (72.2 mg/L; reference, <1.5 mg/L). Atopic dermatitis was diagnosed based on the Hanifin and Rajka[3] criteria. The patient was administered topical corticosteroids (desonide cream 0.05% then betamethasone dipropionate cream 0.05%) for 1 month without improvement (severity scoring of AD [SCORAD][4] estimated at 60 [score range, 0–103, with a higher score indicating more severe disease]). Because of the unusual evolution of the lesions, a skin biopsy was performed with direct immunofluorescence (DIF). The biopsy showed subepidermal blisters containing eosinophils, spongiosis with exocytosis of eosinophils, and a dense dermal infiltrate of eosinophils. Direct immunofluorescence demonstrated linear deposits of IgG and C3 along the basement membrane zone (BMZ)(Figure 2). A sodium chloride split-thickness test with DIF demonstrated deposits of IgG on the epidermal side of the split. A serum Western blot analysis using total human skin extracts as substrates revealed one major polypeptide with a molecular weight of 180 kDa (BP antigen 180). Thus, the diagnosis of vesicular pemphigoid was finally made. Treatment with oral erythromycin (50 mg/kg daily) with betamethasone dipropionate cream 0.05% twice daily was initiated. Two weeks after initiation of treament, the patient's symptoms improved and

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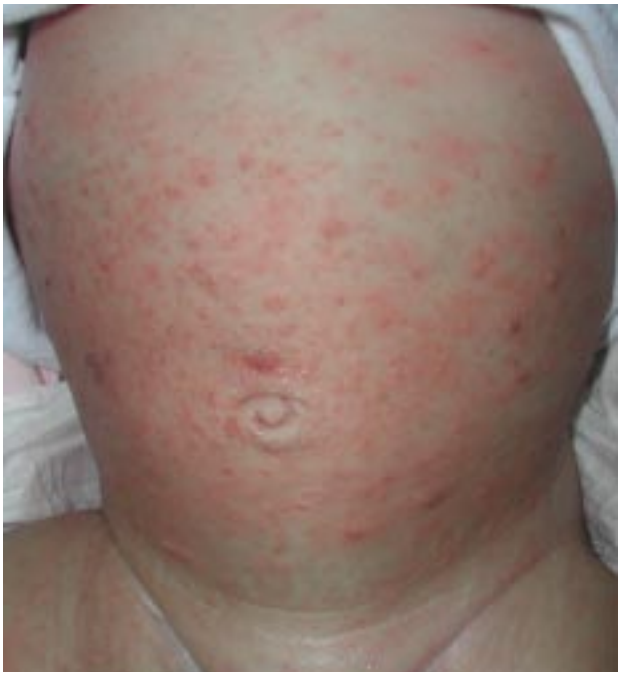

Figure 1. An infant aged 6 months with erythematous vesicular lesions on the abdomen.

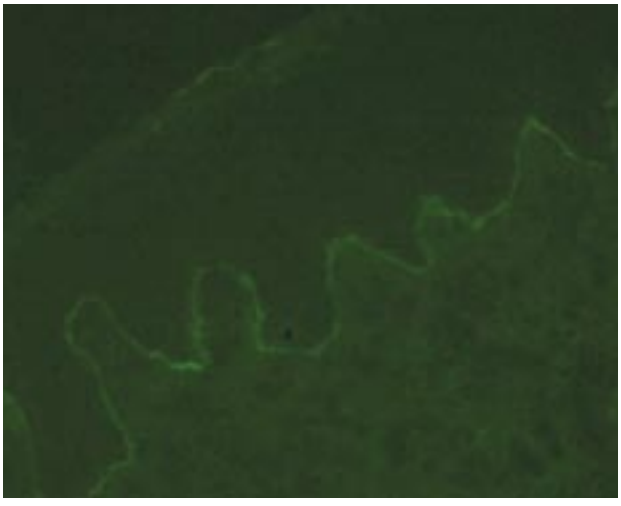

Figure 2. Direct immunofluorescence demonstrated linear deposits of IgG along the basement membrane zone (original magnification ×100).

complete resolution occurred within 1 month. Oral erythromycin and betamethasone dipropionate cream were discontinued and there was no relapse during a follow-up period of more than 1 year.

Comment

Childhood BP usually is diagnosed on the basis of the following criteria: (1) the patient is younger than 18 years with bullous skin lesions, with or without mucous membrane involvement; (2) there are characteristic histopathologic features of BP (subepidermal bullae with variable amount of eosinophils); and (3) DIF shows linear deposits of IgG and/or C3 at the BMZ, or indirect immunofluorescence demonstrates IgG antibodies reacting with the BMZ.[2] It is a rare condition. Fisler et al[2] reviewed the literature in 2003 and identified only 53 cases of childhood BP. Since 2003, approximately 20 more cases have been reported in the literature.[5-15] Age of onset varies from 2 months to 16 years, and there is no sex predilection.[2,5] Childhood BP is considered a true variant of adult BP.[16] However, Nemeth et al[1] reported that mucous membrane involvement was more frequent in childhood BP than in the adult form, and all infants 1 year and younger had marked palmoplantar and facial involvement. In our patient, mucous membranes were not affected. Moreover, 15 of the 20 cases of childhood BP with specified target antigens reviewed by Trüeb et al,[17] Edwards et al,[18] and Arechalde et al[19] showed no mucosal involvement, nor did the 4 most recent cases of infants with BP.[14]

Our patient presented with small pruritic vesicles rather than large bullae. The atypical clinical presentation of the vesicles, family history of AD, and elevated IgE level led to the diagnosis of AD. However, elevated IgE levels have been reported in patients with BP and seem to correlate with disease activity.[20] Atopic dermatitis with vesicles also would suggest secondary impetigo or eczema herpeticum, which were both ruled out by the skin biopsy results with DIF in our patient. Vesicular pemphigoid is an atypical variant of BP characterized by multiple small tense vesicles with a symmetrical distribution.[5,21-24] To the best of our knowledge, only one case of childhood vesicular pemphigoid in a 16-year-old boy has been reported.[5] Thus, our patient is the youngest case of childhood vesicular pemphigoid reported in the literature.

Oral corticosteroids (1–2 mg/kg daily) are the treatment of choice in childhood BP.[2] Dapsone,[25] sulfapyridine,[19] erythromycin,[26,27] cyclosporine,[17] and immunoglobulins[17] have been tried with encouraging results. Oral erythromycin seems to have been effective in our patient. However, most reported cases of childhood BP involved resolution for 1 year or less,[1] thus a spontaneous resolution could not be excluded in our patient.

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