Dear Editor,

Bullous pemphigoid (BP) usually affects the elderly, but BP in infancy has also been reported. Here, we report our observation in a case of childhood BP in Taiwan.

A 15-month-old boy presented with recurrent blisters on his face, trunk, and extremities 1 month later after receiving measles, mumps, rubella, and varicella vaccination (Figure 1A–1D). A few vesicles were also seen on his tongue and oral mucosa, and the palms and soles were also involved (Figure 1E). Direct and indirect Nikolsky signs were negative.

Laboratory examinations showed an elevated immunoglobulin G (IgG) level (1290 mg/dL), low Complement component 3 (C3) (76.6 mg/dL), and negative findings for antinuclear antibody. Indirect immunofluorescence testing revealed positive anti-basement membrane zone antibody (1:40) and a negative result for anti-intercellular substance antibody. Histopathologic examination of a blister on the left hand revealed a subepidermal bulla containing neutrophils, eosinophils, and lymphocytes, with papillary edema and congested dermal vessels surrounded by mononuclear cells (Figure 2A–2B). Direct immunofluorescence (DIF) study showed strong linear deposits of IgG and C3 along the dermoepidermal junction and weaker IgA and complement component 1, q subcomponent (C1q) deposits, which supported a diagnosis of BP (Figure 2C–2D). The patient was admitted to our hospital and the lesions on the trunk and extremities responded well to systemic steroid treatment. However, the oral blisters persisted after 3 weeks of intravenous corticosteroid (prednisolone 1–2.5 mg/kg/d) treatment. A dose of intravenous immunoglobulin (IVIG) with 1.8 g/kg was administered for the refractory oral lesions, after which the oral mucosa improved. BP was well-controlled by low-dose systemic steroid use.

BP is an autoimmune bullous disease that is typically seen in the elderly and is very rare in infants and children. The first peak occurs, more frequently from the 1st year of life (53.2%) and the second peak was reported at the age of 8 years (8.8%). Childhood BP is more frequent in girls. As in adults, lesions usually begin as urticarial pruritic papules and plaques followed by tense bullae on an erythematous background. In infancy, BP usually presents with blistering of the palms, soles, and face; mucosal involvement is uncommon. However, mucosal involvement is more frequent among older children, particularly genital involvement in girls.

It is difficult to diagnosis childhood BP on the basis of clinical features alone since there are a wide variety of bullous disorders with similar features. The differential diagnosis includes chronic bullous disease of childhood (or linear IgA bullous dermatosis), epidermolysis bullosa acquisita, bullous arthropod assault reaction, and bullous systemic lupus erythematosus. A BP diagnosis is based on histopathologic examination and DIF study. Examination of biopsy specimens typically reveals subepidermal bullae formation and a variable number of eosinophils. Deposition of linear IgG and/or C3 is typically seen on the basement membrane, and weaker deposition of IgM, IgA, IgD, and/or IgE is occasionally present. IgM deposition is more frequently seen in childhood BP than in infant BP. DIF study helps to differentiate BP from linear IgA bullous dermatosis with linear basement membrane zone IgA deposition and bullous systemic lupus erythematosus with positive lupus band test. Indirect immunofluorescence may reveal IgG antibodies along the epidermal side of salt-split skin in BP. In contrast to BP, IgG antibodies were deposited along the dermal side in epidermolysis bullosa acquisita.

Immunoblotting with serum reacts to BP antigens at 180 kDa and 230 kDa. Autoantibodies of both the IgA and IgG class to the noncollagenous 16A (NC16A) domain of 180-kDa antigen (BP180) were demonstrated in the sera from patients with childhood BP.

Systemic corticosteroids are the best initial treatment for childhood BP. Sulfapyridine, dapsone, and azathioprine are usually used in combination with corticosteroids in refractory cases. IVIG therapy is a valuable treatment option for intractable BP in both infants and adults. Our patient responded well to a single dose of IVIG for the persistent oral mucosal involvement. Childhood BP usually has a good prognosis and remission typically is achieved within weeks to a few months, although the course is less benign in some children.

BP in infancy has been reported within hours to weeks after vaccination, including vaccinations for hepatitis B, pneumococcus, tetanus–diphtheria–pertussis, and poliovirus. BP in adults and infants was reported to be associated with vaccination, but it is difficult to establish an association between vaccination and BP in infancy, because of the high rate of vaccinations during the first year of life.

In conclusion, we report a case of childhood BP with generalized blisters 1 month after vaccination. Intractable oral lesions responded well to IVIG treatment. Although childhood BP is rare, it should be included in the differential diagnosis of bullous disease.
in children. IVIG treatment could be an optional treatment for refractory childhood BP cases.

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