

CPD

Bullous pemphigoid in a 3-month-old infant after vaccination

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A 3-month-old boy presented with a 6-day history of blisters and vesicles predominantly localized to his palms and soles, and urticarial plaques on his trunk and limbs (Fig. 1a–c). The mucous membranes were not involved. Nikolsky sign was absent and the remainder of the physical examination was normal. The child's medical history was unremarkable and he had not been administered any medication previously. The child's parents were not related, and there was no family history of cutaneous or autoimmune disorders was negative. The patient had received his first vaccination with pneumococcal and hexavalent vaccines [diphtheria, tetanus, pertussis (DTaP), polio, *Haemophilus influenzae* type b (Hib) and hepatitis B virus (HBV)] 2 weeks previously.

Skin biopsy for histology and immunofluorescence was consistent for bullous pemphigoid (Fig. 2a–d).

Laboratory findings showed leucocytosis (18.770/ μ L; normal range 6000–17 500/ μ L) with eosinophilia [19% (3570/ μ L); normal ranges 2.6–5.5% and 40–0650/ μ L]. ELISA was positive for bullous pemphigoid (BP) 180 (titre > 200) and negative for BP230.

Treatment was initiated with oral prednisolone 1 mg/kg/day and topical corticosteroids. However, after 1 month with only slight improvement and development of new blisters, dapsone was added at 0.5 mg/kg/day. Complete remission was achieved after 1 week of dapsone treatment, and prednisolone dose was gradually tapered until discontinuation (Fig. 1d–f). Dapsone was maintained for a period of 6 months. By the 12-month follow-up, the patient had received the second and third doses of the hexavalent vaccine and

also the pneumococcal and meningococcal conjugate vaccines without new relapses of the disease.

BP is a rare bullous disorder in children, with about 150 cases reported in the literature, of which around 80 were infants. The disease is caused by the presence of autoantibodies against the basement membrane proteins BP180 and BP230. In several cases, vaccinations including DTaP, polio, Hib, HBV, meningitis C, rotavirus and influenza, have been reported as preceding the onset of infantile BP, raising the possibility of association between vaccines and infantile BP.¹ Lesions appear between 1 day and 4 weeks after its administration, but recurrences have been observed in subsequent vaccinations only in some patients.² In our case, the lesions started 2 weeks after vaccination, so the vaccine could have acted as a triggering factor. BP has also been described in relation to infections, autoimmune diseases and drugs.³ Based on these observations, it can be hypothesized that bacterial, viral or pharmacological agents act as epitopes, which crossreact with some epidermal antigens that trigger an immune response. Clinically, as in adults, paediatric BP is characterized by multiple tense blisters arising on normal and erythematous skin, often preceded by pruritic and urticarial plaques. However, BP in children presents some peculiarities compared with BP in adults and, depending on the age of onset, there are two clinical presentations. In infants aged < 1 year, involvement of the palms and soles with or without generalized blisters is a typical and practically consistent finding, and mucosal involvement is rare. Distribution of skin lesions in children older than > 1 year old is less uniform, but mucous involvement is more common, with the genital mucosa involved more frequently than the oral mucosa.

Histologically, infantile BP it is identical to the adult form. The presence of anti-BP180 antibodies is more frequent and their levels are associated with the severity of the disease, so they are important both in prognosis and in therapeutic monitoring.⁴ Early diagnosis

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Figure 1 (a–c) Initial clinical presentation with tense blisters on the palms and soles and urticarial plaques on the trunk; (d–f) remission 4 weeks after the start of dapsone.

and treatment are essential to improve prognosis. Corticosteroids are recommended as first-line treatment.⁵ In refractory cases, combination with dapsone, immunosuppressants, immunoglobulin or rituximab may be considered, depending on the severity of the disease.⁶ Unlike in adults, relapses of BP in children are rare and remission is usually achieved within a few months, and often, in less than 1 year.

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CPD questions

Learning objective

To identify the characteristics of bullous pemphigoid in infants.

Question 1. Which of the following clinical features on an infant would alert to probable infantile bullous pemphigoid?

- (a) Blisters located mainly in areas of friction.
- (b) Blisters located mainly in the perioral area.
- (c) Blisters located mainly on elbows and knees.
- (d) Blisters located mainly on the lateral parts of the fingers.
- (e) Blisters located mainly on palms and soles.

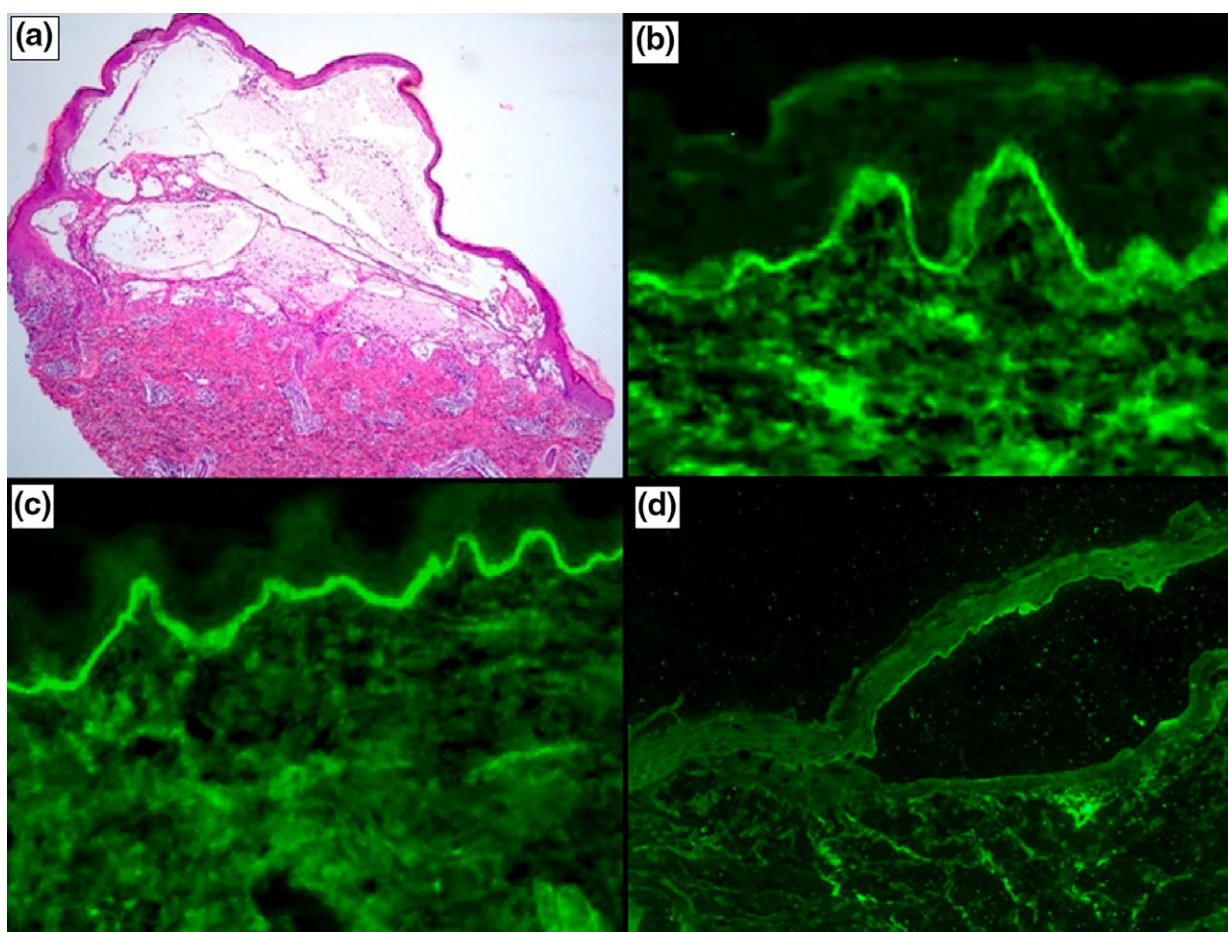


Figure 2 (a) Subepidermal blister with perivascular lymphocytic and eosinophilic infiltrates in the superficial dermis (haematoxylin and eosin, original magnification $\times 40$). (b,c) Direct immunofluorescence staining showing linear deposits of (b) IgG and (c) C3. Original magnification (b,c) $\times 200$. (d) Indirect immunofluorescence showed IgG deposits on the roof of the bullae (original magnification $\times 200$).

Question 2

Which of the following statements is true of infantile bullous pemphigoid?

- (a) Gluten has been implicated as a trigger.
- (b) The malignant neoplasms has been implicated as a trigger.
- (c) The repeated traumas has been implicated as a trigger.
- (d) The stress has been implicated as a trigger.
- (e) The vaccination has been implicated as a trigger.

Instructions for answering questions

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- Read the article in print or online, paying particular attention to the learning points and any author conflict of interest disclosures
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