CASE REPORT

# Infantile Bullous Pemphigoid Following Vaccination

**KAVITA BISHERWAL, DEEPIKA PANDHI, ARCHANA SINGAL AND *SONAL SHARMA**

*From Department of Dermatology & STD and *Pathology, University College of Medical Sciences and Guru Teg Bahadur Hospital, University of Delhi, Dilshad Garden, Delhi, India.*

*Correspondence to:*
*Dr Deepika Pandhi, Professor,*
*Department of Dermatology and STD,*
*University College of Medical Sciences and associated GTB Hospital,*
*Delhi 110 095, India.*
*deepikapandhi@rediffmail.com*
*Received: July 25, 2015;*
*Initial review: November 09, 2015;*
*Accepted: February 02, 2016.*

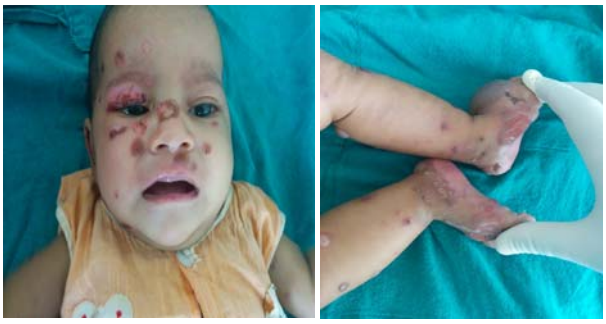
**Background**: Post-vaccination infantile bullous pemphigod is a rare presentation. **Case characteristics**: A 2-month-old girl presented with widespread bullae, erosions, necrotic and targetoid lesions over body and mucosae after vaccination. Histology and direct immunofluorescence (DIF) were consistent with bullous pemphigoid. **Intervention**: Clinical remission with oral steroids and no recurrence with subsequent vaccination. **Message**: Continuation of vaccination may not be contraindicated in infants where bullous pemphigoid onset occurs after vaccination.

**Keywords:** *Adverse events following immunization, Infant, Vaccine.*

Bullous pemphigoid is an acquired autoimmune subepidermal blistering disorder, commonly affecting the elderly population [1]. Pediatric autoimmune blistering disorders including bullous pemphigoid are uncommon and have varied manifestations, thereby leading to difficulty in clinical diagnoses [1]. Pediatric bullous pemphigoid differs from adult variety by more frequent involvement of mucous membranes, face, palms, soles, no association with any neoplasm and rapid resolution with steroids [2]. We report a 2-month-old female infant who presented with bullous pemphigoid two days after receiving DPT and hepatitis B vaccination.

## CASE REPORT

A 2-month-old girl presented to the dermatology outpatient department with complaint of fluid filled lesions over limbs, face, trunk, and groin area of 10 days duration. The lesions initially appeared over the chest region and extended to involve limbs, palms, soles, face, abdomen, oral and genital mucosae, and scalp over the next two days. There was no history of accompanying fever, diarrhea or prior drug intake, and the child was taking feeds normally. She received first dose of DPT and hepatitis B vaccines two days prior to onset of lesions. She was a product of full term normal vaginal delivery born of a non consanguineous marriage, and there was no history of similar complaints in the family members. Examination revealed multiple tense vesicles and bullae predominantly over extremities, palms, soles and face. There were multiple targetoid lesions over trunk and multiple erosions all over body including scalp, oral and genital mucosae (***Fig.* 1**). Tzanck smear showed few eosinophils without acantholytic cells and no organisms were seen on Grams staining. She had been started on intravenous vancomycin with a provisional diagnosis of bullous impetigo, with no response. All hematological investigations were unremarkable and blood culture was negative. A skin biopsy and direct immunofluorescence (DIF) was performed with differential diagnoses of linear IgA dermatosis and infantile bullous pemphigoid. Histological examination revealed subepidermal bulla with dense eosinophilic infiltrate (***Web Fig.* 1*a*)** and DIF showed linear deposits of IgG and C3 and absence of IgA and IgM at dermo-epidermal junction (***Web Fig* 1*b***). With a final diagnosis of infantile bullous pemphigoid, oral prednisolone therapy was initiated at a dose of 1 mg/kg per day. Rapid improvement with resolution of bullae was observed after 2 weeks, and the steroids were slowly tapered. The child's vaccination was continued as per



*(a)* *(b)*

**FIG. 1** *(a) Bullae and crusted erosions over face; and (b) multiple bullae over lower limbs.*

schedule and no recurrence was observed with second dose of DPT and hepatitis B vaccine. The child is in remission at 6 months follow-up.

## DISCUSSION

In pediatric bullous pemphigoid, large tense bullae are seen predominantly over inner thighs, groin, abdomen, forearms, axillae, palms and soles and mucous membranes [1]. Infantile bullous pemphigoid has more widespread clinical lesions with less mucosal involvement and predominant acral involvement [3,4]; whereas, in childhood bullous pemphigoid, involvement is severe, less uniform, and may be localized to genital area [5]. Palmoplantar lesions are considered as a diagnostic clue of infantile bullous pemphigoid [5]. Schwieger-Briel, *et al*. [5] proposed the following minimal diagnostic criteria for infantile bullous pemphigoid: typical clinical picture (plaques and blisters, acral distribution) and linear IgG and/or C3 deposition at the basement membrane in DIF.

The cause of pediatric bullous pemphigoid is unknown and the possible triggering factors reported include non-specific maternal antibodies and foreign antigen (infectious agents, drugs, vaccines) [1,6]. Vaccinations implicated include diphtheria, tetanus, pertussis, poliomyelitis, influenza, hepatitis B, meningococcal C, pneumococcus, BCG and rota virus with a latent period of 1 day to 4 weeks between vaccination and onset of disease [1,5,6]. Most cases of infantile bullous pemphigoid have been described after first dose of vaccination [5,7]. Vaccination may unmask subclinical bullous pemphigoid by enhancing an autoimmune response in immunologically predisposed individuals [4]. It is also postulated that the inflammation induced in the skin by vaccination – rather than the vaccines themselves – might generate anti basement membrane antibodies consequent to disruption of the basement membrane as no structural similarities have been seen between the implicated vaccines and basement membrane proteins [8]. This might explain rarity amongst children despite frequency of vaccinations. Moreover, no recurrence has been seen with the reintroduction of the vaccination schedule and continuation of vaccination as per schedule, is recommended [4].

The clinical and histological differential diagnoses of bullous pemphigoid include chronic bullous disease of childhood, dermatitis herpetiformis, bullous lupus erythematosus, epidermolysis bullosa acquisita (EBA), porphyria and bullous impetigo [5]. EBA is rarely seen in children, and the histological differentiation is by presence of neutrophilic infiltrate and IgG deposits only on the dermal side (base of the blister) in EBA. The prognosis of pediatric bullous pemphgoid is good with a possibility of spontaneous remission and infrequent relapses triggered by infections or tapering of corticosteroids [5]. Steroids (1 to 2 mg/kg) are the first-line treatment; tapered slowly to prevent rebound [1,5]. Steroid sparing agents include dapsone, IVIGs, mycophenolate mofetil, erythromycin, methotrexate-, cyclophosphamide, azathioprine, rituximab or omalizumab but their use warrants further investigation [5].

*Contributors*: All authors contributed to the acquisition, analysis or interpretation of clinical and histological (SS) data; and; contributed to the drafting the work or revising it critically for important intellectual content. All authors approved the final version.
*Funding*: None; *Competing interests*: None stated.

## REFERENCES

1. Wu KG, Chou CS, Hsu CL, Lee ML, Chen CJ, Lee DD. Childhood bullous pemphigoid: A case report and literature review. J Clin Exp Dermatol Res. 2013;S6:010.
2. Barreau M, Stefan A, Brouard J, Leconte C, Morice C, Comoz F, *et al*. Infantile bullous pemphigoid. Ann Dermatol Venereol. 2012;139:555-8.
3. Waisbourd-Zinman O, Ben-Amitai D, Cohen AD, Feinmesser M, Mimouni D, Adir- Shani A, *et al*. Bullous pemphigoid in infancy: Clinical and epidemiologic characteristics. J Am Acad Dermatol. 2008;58:41-8.
4. Reis-Filho EG, Silva Tde A, Aguirre LH, Reis CM. Bullous pemphigoid in a 3-month-old infant: case report and literature review of this dermatosis in childhood. An Bras Dermatol. 2013;88:961-5.
5. Schwieger-Briel A, Moellmann C, Mattulat B, Schauer F, Kiritsi D, Schmidt E, *et al*. Bullous pemphigoid in infants: characteristics, diagnosis and treatment. Orphanet J Rare Dis. 2014;9:185.
6. Baykal C, Okan G, Sarica R. Childhood bullous pemphigoid developed after the first vaccination. J Am Acad Dermatol. 2001;44:348-50.
7. Erbagci Z. Childhood Bullous Pemphigoid Following Hepatitis B Immunization. J Dermatol. 2002;29:781-5.
8. Cohen AD, Shoenfeld Y. Vaccine-induced autoimmunity. J Autoimmun. 1996;9:699-703.