Juvenile Bullous Pemphigoid in a Patient with Atopic Dermatitis and Trisomy of Chromosome 9

Christine R. Mehren, Patricia L. Danielsen and Robert Gniadecki
Department of Dermatology and Venereology, Bispebjerg University Hospital, Bispebjerg Bakke 23, DK-2400 Copenhagen NV, Denmark. E-mail: c_mehren@hotmail.com

Juvenile bullous pemphigoid in combination with atopic dermatitis in a 13-year-old girl is described below. It was easily cured and the disease is in general much easier to manage than adult bullous pemphigoid.

Bullous pemphigoid (BP) is an acquired autoimmune subepidermal blistering disorder, caused by autoantibodies of the IgG4 subclass against hemidesmosome components, namely protein BP 180 (collagen type XVII) and BP 230. The peak incidence occurs between 60 and 80 years of age, but a juvenile form (JBP) has also been described (1–3).

In JBP the lesions are often localized rather than widespread, mucous membrane involvement is more common and prognosis is more favourable than in classic BP (1–3).

To the best of our knowledge, the number of published cases of JBP up to December 2010, is 97. We report here a case of JBP in a 13-year-old girl with atopic dermatitis (AD), trisomy of chromosome 9, and allergy presenting with a vesicobullous rash. The skin lesions resolved on treatment with topical steroid and systemic dapsone.

Case report

A 13-year-old Danish girl with trisomy 9 was referred to our institution because of a 1-month history of an itchy eruption predominantly localized to the lower extremities. She had atopic eczema since infancy, together with asthma, and a type I allergy to *Dermatophagoides pteronyssinus* (dust mite) and animal hair.

Palm-sized areas consisting of crusted and haemorrhagic vesicles, and a few bullae containing clear fluid arranged in clusters arising from inflamed skin, were observed on the inner thighs and the anterior side of the left lower leg. Annular eczema-like plaques were seen on the thorax and right heel. There was no mucosal involvement.

Histology showed dermal inflammatory infiltrate with eosinophil dominance. Gliadin antibodies were negative, and routine laboratory tests were normal, except for peripheral blood eosinophilia of 0.61×10^9/l (reference value $\leq 0.5 \times 10^9$/l). Direct immunofluorescence of perilesional skin revealed linear IgG and C3 depositions along the base membrane zone, which, in the salt-split skin, were located on the epidermal side of the blister diagnostic of BP.

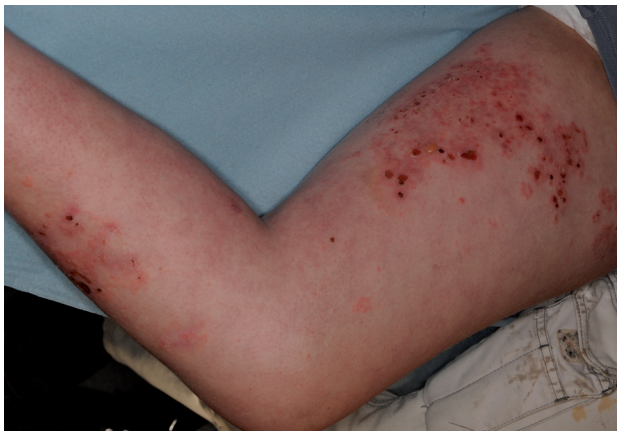

Fig. 1. Haemorrhagic vesicles and bullae on the left lower limb.

Treatment with topical steroids group III (mometasone furoate) was initiated upon admission, supplemented with dapsone, 50 mg daily. The lesions resolved within 2 weeks.

Discussion

To the best of our knowledge, 97 cases of immunofluorescence-confirmed JBP have been reported. One case describes a patient with an atopic condition and JBP (4). The unusual feature of our case was a constellation of JBP and AD in a patient with a trisomy 9. To our knowledge, there is no association between chromosomal defects or AD and JBP. However, atopic conditions and BP may be associated, since peripheral blood and tissue eosinophilia and increased IgE accompany both diseases.

One of the known triggers of JBP is vaccination (5). However, our patient had no history of vaccination. Systemic corticosteroids remain the mainstay of treatment of JBP in moderate to severe cases. Since JBP is usually a self-limiting disease, corticosteroid-sparing agents are preferred, such as dapsone

or intravenous immunoglobulin. Our patient had a rapid response to dapsone, underscoring the fact that JBP is easier to manage than adult-onset BP.

Conflict of interest

The authors have no conflicts of interest to declare.

References

1. Gajic-Veljic M, Nikolic M, Medenica L. Juvenile bullous pemphigoid: the presentation and follow-up of six cases. J Eur Acad Derm Venerol 2010; 24: 69–72.
2. 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: 41–48.
3. Toyama T, Nakamura K, Kuramochi A, Ohyama B, Hashimoto T, Tsuchida T. Two cases of childhood bullous pemphigoid. Eur J Dermatol 2009; 19: 368–371.
4. XiaoT, Li B, Wang Y-K, He CD, Chen HD. Childhood bullous pemphigoid treated by i.v. immunoglobulin. J Dermatol 2007; 34: 650–653.
5. Majmudar V, Herath D, O'Toole EA, Harrison A. Bullous pemphigoid of childhood: a rare disease with diagnostic and management challenges. Clin Exp Dermatol 2010; 35: 213–214.