CASE REPORT

Bullous Pemphigoid in an Infant: A Case Report

Oscar Thabouillot;[1,2] Julien Le Coz;[1] Nicolas-Charles Roche[2]

1. Pediatric Emergency Department, Robert Debré Teaching Hospital, Paris, France
2. Bégin Military Teaching Hospital, Saint-Mandé, France

Correspondence:
Oscar Thabouillot
Pediatric Emergency Department
Robert Debré Teaching Hospital
Paris, France
E-mail: thabouillot@hotmail.com

Conflicts of interest: none

Keywords: autoimmune diseases; bullous pemphigoid; emergency department; infant

Abbreviations:
BP: bullous pemphigoid
ED: emergency department
IgG: immunoglobulin G
MMF: mycophenolate-mofetil

Received: December 5, 2017
Revised: February 4, 2018
Accepted: February 5, 2018

Online publication: June 22, 2018

doi:10.1017/S1049023X18000456

Abstract

A seven-month-old girl was referred to the emergency department (ED) after a general practitioner suspected Steven-Johnson syndrome. Actually, the diagnosis of bullous pemphigoid (BP) was made based on biopsies; BP is a rare, autoimmune skin disease involving the presence of blisters known as bullae. The child was efficiently treated with topical steroids. This case shows the importance of the ED physician's prior knowledge of BP so that a differential diagnosis with other autoimmune diseases (dermatosis, pemphigus) can be made.

Thabouillot O, Le Coz J, Roche NC. Bullous pemphigoid in an infant: a case report. *Prehosp Disaster Med*. 2018;33(4):448–450.

Introduction

Bullous pemphigoid (BP) is a disease of autoimmune origin which is clinically characterized by the appearance of large blisters (known as bullae) on erythematous and pruritic zones. The bullae are preferentially present on the limbs, but any area can be affected. Oral impairment is also possible. From a histological point of view, bullae are explained by the abnormal presence of autoantibodies against two proteins (AgPB230 and AgPB180) between the dermis and the epidermis. For this reason, the histological diagnosis of BP is asserted when subepidermal blisters and linear deposits of immunoglobulin G (IgG) and complement are observed in the basement membrane.

Case Report

This report is a case of BP in a seven-month-old female infant, without any significant medical history, who was referred to the emergency department (ED) for fever, macular erythroderma, and some millimeter-sized vesicles on her hands and feet. The parents reported a severe pruritus. The infant had been seen 48 hours beforehand by her general practitioner, who had diagnosed impetigo and had prescribed amoxicillin-clavulanic acid. The mother had not administered the medication to her child. On the day of the consultation in the ED, the mother asked another practitioner for a second opinion. This doctor, who suspected Steven-Johnson syndrome, referred her to Robert Debré Teaching Hospital's (Paris, France) ED.

When examined, the infant was afebrile, without hemodynamic failure. The clinical examination showed erythroderma on the whole body, with inconspicuous millimeter-sized vesicles on the palms of the hands and feet soles (Figure 1; color image available as supplementary material [online only]), with small, yellowish oozing crusts. Considering these symptoms, a bullous impetigo was diagnosed and Steven-Johnson syndrome was dismissed in the absence of epidermal peeling. Amoxicillin-clavulanic acid was prescribed for 10 days, a new medical consultation being scheduled after a 48-hour period for a re-evaluation.

The infant was re-examined 48 hours later. She was febrile with a body temperature of 38.3°C (100.9°F), with no hemodynamic failure. In addition to erythroderma, the clinical examination demonstrated very large bullae on her hands and feet. The bubbles on the feet entirely covered the soles (Figure 2; color image available as supplementary material [online only]). They were popped and the child was hospitalized in the dermatology department.

The biopsies showed linear deposits of IgG in the basal membrane, and thus physicians could assert the diagnosis of BP. The symptoms went away after a few days following a treatment with topical corticosteroids.

Discussion

As illustrated by this case, physicians working in a pediatric ED must know BP in its broad outline. The differential diagnosis of BP is mainly with other autoimmune bullous diseases, namely linear IgA dermatosis and pemphigus. According to Kong in a retrospective study of

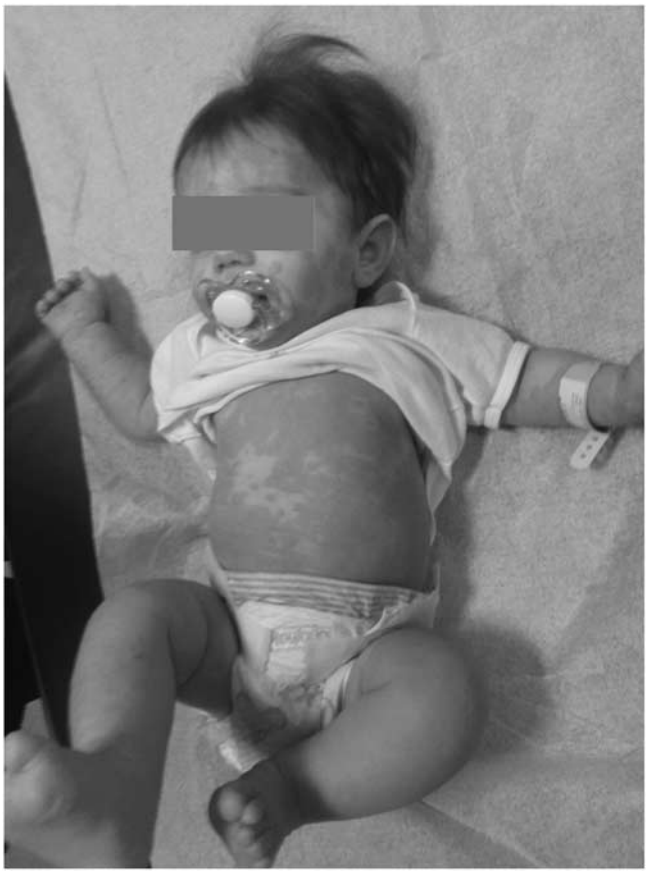

Figure 1. Erythroderma without Bullae.

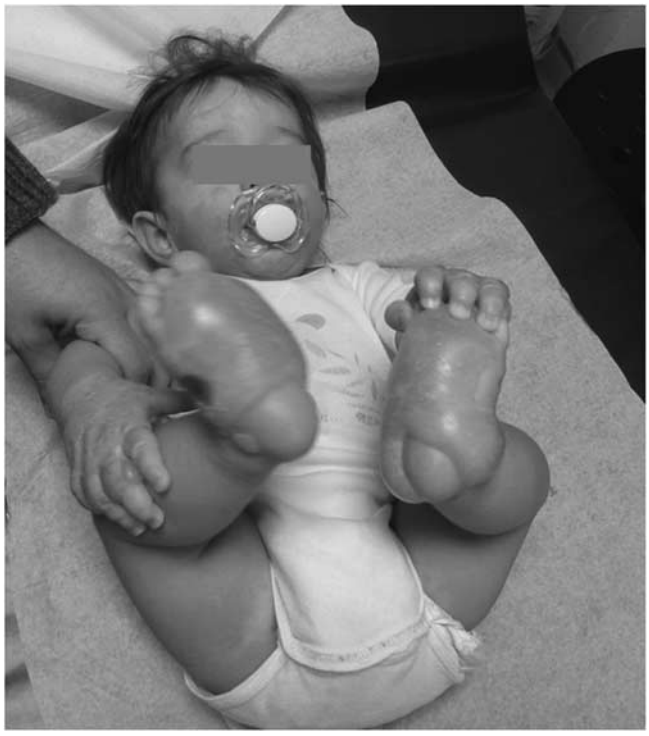

Figure 2. 48 Hours After the First Examination in ED.
Note: Large, swollen bullae were observed on the feet, following erythroderma.
Abbreviation: ED, emergency department.

12 patients, the most common disease in children would be linear IgA dermatosis, affecting 41.7% of patients, BP and pemphigus being on equal terms with 16.7% each.[1]

Of note, BP is a disease whose prevalence increases with age. In France, while the prevalence estimate is 1.00 out of 40,000 for all ages combined, it is 1.00 out of 3,000 after 70 years of age. Although much rarer, this disease is also present in children. Waisbourd-Zinmman estimates the incidence of childhood BP in Israel at 2.36 out of 100,000 inhabitants per year.[2] In addition, the disease seems to include certain peculiarities in children when compared to the adults' form. Thus, for example, more than one-half (58%) of the cases of BP begin before the first year of life, and the predisposition for reaching the extremities is higher in children under one year of age (79% versus 17%). The practitioner must keep in mind certain atypical and serious manifestations affecting children. Akin described the case of a 16-day-old infant clinically presenting BP associated with respiratory distress.[3] A fiberoptic examination showed bullae in the upper airways and the epiglottis, thus explaining the presented symptomatology.

The French National Authority for Health (HAS; Saint-Denis, France) recommends topical corticosteroids as the first line treatment for BP.[4] Systemic corticosteroids are indicated only in the event of topical corticosteroids' failure. On a case-by-case basis, other immunosuppressants such as methotrexate, mycophenolate-mofetil (MMF), or azathioprine can be added to systemic corticosteroids. Fine noticed that the three main causes of relapse in the first year were significant dermal involvement prior to treatment, dementia, and high levels of anti-BP180 antibodies (greater than 23 U/mL) after 150 days of treatment.[5] In children, several authors reported cases of BP resistant to treatment with corticosteroids. For example, Ister described the case of a 3-month-old infant who developed BP one week after administration of a hexavalent vaccine.[6] The disease resisted local and oral corticosteroids and required the use of immunoglobulins. These allowed a complete control of the disease and the absence of recurrence, including after the next injection of the hexavalent vaccine. In addition, Seminario-Vidal described the case of a child for whom a treatment with MMF was required to control BP resistant to local and systemic corticosteroids.[7] The use of this treatment once again allowed a complete control of the disease.

Conclusion

This case shows the importance of the ED physician's prior knowledge of BP so that a differential diagnosis with other autoimmune diseases can be made.

Supplementary Material

To view supplementary material for this article, please visit https://doi.org/10.1017/S1049023X18000456

References

1. Kong YL, Lim YL, Chandran NS. Retrospective study on autoimmune blistering disease in paediatric patients. *Pediatr Dermatol.* 2015;32(6):845-852.
2. Waisbourd-Zinman O, Ben-Amitai D, Cohen AD, et al. Bullous pemphigoid in infancy: clinical and epidemiologic characteristics. *J Am Acad Dermatol.* 2008;58(1):41-48.

3. Akin MA, Gunes T, Akýn L, et al. A newborn with bullous pemphigoid associated with linear IgA bullous dermatosis. *Acta Dermatovenerol Alp Pannonica Adriat*. 2009;18(2):66-70.
4. Joly P, et al. Haute Autorité de Santé. April 2016. Protocole national de diagnostic et de soins de la pemphigoïde bulleuse. http://www.has-sante.fr/portail/upload/docs/application/pdf/2016-06/pnds_-_pemphigoide_bulleuse_pb.pdf [in French]. Accessed December 1, 2017.
5. Fine JD. Clinical and immunological factors associated with bullous pemphigoid relapse. *JAMA Dermatol*. 2014;150(1):34.
6. Ister M, Pouessel G, Ythier H, et al. Post-vaccinal, corticosteroid-resistant bullous pemphigoid in infancy: treatment with intravenous immunoglobulin. *Pediatr Dermatol*. 2014;31(4):e94-e95.
7. Seminario-Vidal L, Sami N, Miller J, et al. Mycophenolate mofetil therapy for pediatric bullous pemphigoid. *Dermatol Online J*. 2015;21(8).