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CASE REPORT

Bullous pemphigoid - A case report

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Abstract

Bullous pemphigoid (BP) is a chronic, self-limiting, blistering autoimmune disease that mainly affects the elderly. Its occurrence in children is rare. It is characterized by generalized bullous eruptions and tense blisters with either serous or hemorrhagic content. This is a case report of a 2-month-old female infant presenting a history of bullous lesions, initially on the palms and plantae. Approximately 1 week after the appearance of these lesions, an intense erythema appeared, associated with purulent secretion in some locations. The patient was hospitalized and received intravenous antibiotic therapy. The bullous lesions persisted. A biopsy of one of the lesions was performed, and the material was sent for histopathological study and direct immunofluorescence. The results were compatible with BP. Treatment was started with prednisolone, with progressive improvement. This type of bullous dermatosis has a benign clinical course. Systemic corticosteroids are the basis of treatment. The disorder regresses within 1 year and has an excellent prognosis. Because it is a rare condition, knowledge of its characteristics is important for the differential diagnosis of other bullous diseases in childhood.

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INTRODUCTION

Bullous pemphigoid (BP) is a chronic, autoimmune, self-limiting, blistering disease that mainly affects older adults of all ethnicities[1]. It rarely occurs in children, with 60 cases reported, among which 10 occurred in infants. It is characterized by generalized vesiculobullous eruption and tense bullae, with serous and/or hematic content. Herein, the authors describe the case of a 2-month-old infant who was hospitalized to investigate vesiculobullous skin lesions. Direct immunofluorescence (DIF) led to the diagnosis of BP. The treatment comprised administration of oral corticosteroids, with a favorable progress.

CASE REPORT

The patient was a white female infant, 2 months and 28 days old, with bullous lesions initially on the palms of her hands, later spreading to the entire body. Her parents were young, healthy, and non-consanguineous Figures 1 and 2. There was no family history of atopy or autoimmune diseases. Obstetric history featured a cesarean section at 39 weeks, with a history of maternal preeclampsia. Personal history featured no relevant diseases. Approximately 1 week after the emergence of the lesions, they showed intense erythema associated with purulent discharge in some spots. The parents sought medical care, and hospitalization and intravenous antibiotics were indicated due to the extent of the condition. There was no damage to mucous membranes. Laboratory tests upon admission showed the following results: hemoglobin: 11.6 g/dL, hematocrit: 32.8%, leukocytes: 9,300/mm^3, segmented cells: 25%, lymphocytes: 58%, band cells: 1%, eosinophils: 10%, monocytes: 6%, platelets: 292,000/mm^3, C-reactive protein: 0.5 mg/dL, albumin: 3.9 g/dL, globulin: 2.3 g/dL, sodium: 136 mEq/L, potassium: 4.9 mg/dL, calcium: 10.8 mg/dL, magnesium: 2 mg/dL, *Venereal Disease Research Laboratory (VDRL*) not reactive, and rapid HIV test: not reactive. Crystalline penicillin and oxacillin were administered for 10 days. Although the medication improved the inflammatory symptoms, the bullae persisted. Therefore, a biopsy of the lesions was conducted, and the material was examined by histopathology and DIF. DIF evidenced linear IgG and C3 deposits in the epidermal basement membrane. Histopathology showed subepidermal bullous dermatosis. Both results are compatible with BP. After these results, treatment with prednisolone (1 mg/kg/day) was initiated, with progressive improvement of the lesions. Physical examination at 9 days after beginning corticotherapy revealed that the patient did not show any more lesions in the palmoplantar region. Medication (prednisolone) was continuously administered for 3 months, when gradual weaning was initiated.

DISCUSSION

BP is a subepidermal autoimmune dermatosis that usually affects older adults aged over 70 years, with no ethnic or sexual predilection[2]. The first case during childhood was reported in 1970. Since then, few cases involving the pediatric population have been observed[3].

Figure 1. Intact and ruptured bullae.

This autoimmune disorder is involved with autoantibodies of the IgG class targeted against antigens (230 kD and 180 kD, designated as 230 BP Ag1 and 180 BP Ag2, respectively) located on the hemidesmosome plaque, resulting in the emergence of subepidermal vesiculobullous lesions.

Clinically, the bullae are tense, with serous and/or hematic content, appearing on erythematous or normal skin. On the limbs, the lesions have a predominantly flexural distribution. The oral, nasal, and conjunctival mucosae are affected in approximately 30% of cases[4]. Initially, the disease can be manifested with pruritus and erythematous plaques that progress to large, tense bullae, occasionally with hemorrhagic content. Nikolsky's sign is characteristically absent, and the bullae do not extend or increase in size, as they do in patients with pemphigus vulgaris.

BP can be clinically indistinguishable from other bullous dermatoses in infants, thereby necessitating the use

Figure 2. Intact and ruptured bullae.

of ancillary tests. Leukocytosis, eosinophilia, and increased serum total IgE are nearly always present. Some authors list clinical peculiarities of BP in childhood that can contribute to the differential diagnosis, such as frequent involvement of hands and feet, which is usually the initial presentation of the disease. The occurrence of facial lesions is also very common.[5] The bullae resolve without scarring.

The lesion biopsy shows subepidermal bullae. DIF shows C3 and IgG deposition, being positive in 80% of the cases. The clinical course of BP in children is benign. In most cases, it is useful to resort to pharmacological treatment to shorten its duration and minimize the associated symptoms. Systemic corticosteroids are the basis of the treatment (prednisone or prednisolone 1 to 2 mg/kg/day). Sulfapyridine, dapsone, and azathioprine may be used as steroid-sparing agents. The disorder regresses within 1 year (mean time: 5 months) and has an excellent prognosis. Recurrences are frequent but are of lower intensity than that of the initial episode.

In 1990, Anhalt described paraneoplastic pemphigus. This autoimmune dermatosis is caused by autoantibodies that are targeted against the epidermis and are apparently produced by tumor cells[6]. Since then, the association between bullous dermatoses and malignancy in adults has been researched.[7] This association has not been reported in children.

CONCLUSION

BP is a rare disease, with few cases described in the pediatric population. Knowledge of its characteristics is important for the differential diagnosis of bullous diseases in childhood. Corticotherapy is the basis of the treatment, but cleaning the affected area and treating secondary infections are of utmost importance. This case highlights the importance of considering this diagnosis in the pediatric population.

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