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CASE REPORT

Bullous pemphigoid in infants and linear IgA bullous dermatosis, the main condition considered in its differential diagnosis

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Abstract

Bullous pemphigoid is an autoimmune subepidermal dermatosis which is difficult to be differentiated from other bullous dermatosis, like the linear IgA bullous dermatosis, rarely seen in children (there are less than 15 cases reported in the literature). A 3 months and 24 days infant was admitted at our urgent care service due to bullous lesions all over the body. Initially managed as chickenpox, the hypothesis was discarded after injury progression. Skin biopsy of two lesions were performed, and the diagnosis of bullous pemphigoid was confirmed. Treated with oral corticotherapy, complete remission of injuries was noticed after 90 days.

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INTRODUCTION

Bullous pemphigoid (BP) is a subepidermal autoimmune bullous dermatosis rarely seen in children. BP usually affects individuals aged 70+ years, with no predilection for ethnicity or sex. It cannot be distinguished clinically from other pediatric bullous dermatoses such as linear IgA bullous dermatosis. The first case of the disease in children was described in 1970. Since then, fewer than 15 cases involving children have been reported in the literature[1-3].

This report describes the case of an infant aged 3 months and 24 days brought to our emergency unit with generalized sudden-onset bullous lesions initially considered as a case of linear IgA bullous dermatosis. Pathology tests and the development of signs and symptoms eventually confirmed a diagnosis of bullous pemphigoid, which subsided after the initiation of therapy.

CASE REPORT

A male infant aged 3 months and 24 days was admitted to our pediatric emergency unit in September 2020 presenting eruptions with erythematous papules on his trunk and limbs, associated with fever beginning the day before he was admitted. Initially diagnosed with chickenpox, the infant was discharged with a prescription to manage symptoms and orientation. The infant was brought in again the next day with worsening vesiculobullous lesions, mainly in the acral regions (Figure 1-A). He was admitted and started on antibiotic therapy (oxacillin 200 mg/kg/day) due to the risk of secondary bacterial infection. Diagnostic investigations considered a possible case of linear IgA bullous dermatosis or bullous pemphigoid.

In the following days, the lesions spread over his body, mainly to the hands and feet, with erythematous plaques and large tense blisters, some with bloody content and sparing mucosae (Figure 1-B).

The infant was in good general condition. There was no evidence of active bacterial infection and he did not have a fever during hospitalization. Laboratory tests showed no metabolic anomalies or signs of involvement of other organs. A polymerase chain reaction test for COVID-19 was negative.

His mother (G2 P2 A0) had an uneventful obstetric history. The infant was exclusively breastfed until three months and had since been on the formula.

A biopsy of active lesions was performed due to the suspicions around autoimmune diseases such as linear IgA bullous dermatosis and bullous pemphigoid. C-reactive protein was negative and glucose-6-phosphate dehydrogenase and immunoglobulins (IgA, IgE, IgM) were within normal range. The patient was started on prednisolone 1mg/kg/day.

Histopathology examination including a specimen collected after eight days of treatment revealed subepidermal cleavage with a predominance of eosinophils, confirming the diagnosis of bullous pemphigoid. Immunohistochemistry analysis of the same material revealed positive staining for anti-IgG, anti-IgM, and anti-C3, and negative staining for anti-IgA and anti-fibrinogen in the dermal-epidermal basement membrane, which also confirmed the diagnosis of bullous pemphigoid and ruled out IgA bullous dermatosis.

The infant showed signs of improvement on the ninth day of treatment, with new lesions ceasing to appear and old lesions healing. He was discharged and placed on outpatient follow-up care with a prescription of prednisolone (Figure 2-A). In a return visit 20 days after discharge, the lesions were almost completely healed, and the patient gradually discontinued prednisolone therapy. After three months of therapy, the lesions remitted and healed completely (Figure 2-B).



Figure 1. Vesiculobullous skin lesions; A. Initial bullous lesions; B. Tense bullous lesions with bloody content.

DISCUSSION

Bullous pemphigoid may be clinically indistinguishable from other bullous dermatoses in children, including linear IgA dermatosis. This is why supplementary tests are needed to establish diagnosis and treatment, with biopsy as the gold standard diagnostic method.

In linear IgA bullous dermatosis, also a non-hereditary autoimmune disease, subepidermal blisters form due to the linear deposition of immunoglobulin A (IgA) in the basement membrane area. Cutaneous and mucosal involvement is observed, and short-duration nonspecific prodromal disease is common, accompanied at times by systemic symptoms such as fever and anorexia. A typical sign of this condition is the sudden appearance of tense blisters on apparently normal skin, or occasionally localized or disseminated urticaria-like lesions, mainly on the face, extremities, trunk, and genitals. Our patent had no mucosal or genital involvement, although widespread skin lesions appeared suddenly[4].

Bullous pemphigoid also develops with the appearance of subdermal vesiculobullous lesions, which sit on top of erythematous or normal skin. Hands and feet are often involved at the onset of the disease, as seen in our patient, with lesions disseminating to other parts of the body and affecting mainly the extremities. Facial lesions often appear. In the limbs, the distribution of lesions is predominantly flexural. Although oral, nasal and conjunctival mucosae are affected in about 30% of cases, our patient had no such involvement during follow-up. Initially, the disease may manifest with itching, erythematous plaques that develop into large tense blisters, sometimes with bloody content[5].

Usual findings include leukocytosis, eosinophilia, and increased total serum IgE. Some authors have described signs of bullous pemphigoid that may contribute to diagnostic elucidation, such as resolution of blisters without scarring. As the lesions improve, transient pigmentary changes appear, usually evolving with complete resolution without permanent scars. Diagnosis is based on skin biopsy and direct immunofluorescence[6].



Figure 2. Healing lesions; A. Healing bullous lesions; B. Remission of the lesions and completion of the healing process.

The histology of linear IgA bullous dermatosis is characterized by subepidermal bullous lesions with a predominantly neutrophilic infiltrate in the papillary dermis, although mononuclear cells and occasional eosinophils might also occur. Immunofluorescence tests performed on perilesional skin is the gold standard diagnostic method to demonstrate linear IgA deposition along the basement membrane. Treatment is designed to control the bullous lesions while avoiding adverse effects. Dapsone is the drug of choice with an initial low dose of 0.5 mg/kg followed by gradual dose escalation until the symptoms and bullous lesions have been controlled (usually 2 mg/kg/day)[3].

In bullous pemphigoid, biopsy shows subepidermal bullous lesions, while direct immunofluorescence tests with perilesional skin finds IgG deposition along the basement membrane in almost every patient. C3 deposits are also seen in almost all cases. The clinical development of bullous pemphigoid in children is benign and the treatment of choice is systemic therapy with steroids (prednisone or prednisolone 1 to 2 mg/kg/day). Sulfapyridine, dapsone, and azathioprine may be used as steroid sparers. The condition regresses within one year (average of five months) and prognosis is excellent. Recurrences, as a rule less intense than the original episode, are frequent[6].

CONCLUSION

Bullous pemphigoid is a rare disease that is difficult to differentiate from others such as linear IgA bullous dermatosis based on clinical signs alone. Diagnosis can only be confirmed via further examination and biopsy of the lesions. Steroids are the mainstay of treatment, but keeping the lesions clean and treating secondary bacterial infections are of utmost importance for a good outcome. The present case shows the importance of considering this condition in the differential diagnosis of children, performing biopsy for diagnostic confirmation, and starting treatment early on. If treated properly, the disease carries a favorable prognosis.

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