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Childhood bullous pemphigoid

Amarouch Hajar, Benzekri Leila, Ismaili Nadia, Senouci Karima, Badredine Hassam

CASE REPORT

A two-year-old girl was presented with vesiculobullous eruptions initially on the palmoplantar region then extend to the whole body (Figure 1). On examination multiple tense vesicles, bullae and crusted erosions touching the whole tegument were seen. Nikolsky's sign was negative. Mucous membrane examination revealed erosions over buccal mucosa and hard palate. There was no regional lymphadenopathy. Histopathological examination revealed subepidermal blister and a mild inflammatory infiltrate on the dermis. Direct immunofluorescence (DIF) showed linear deposition of IgG and C3 at the dermoepidermal junction. The specific blood test searching for cutaneous basement membrane antibodies was positive. Based on clinical, histological and immunopathological criteria, the diagnosis of childhood bullous pemphigoid was made. The patient was treated with prednisone 1 mg/kg/day associated with methotrexate 5 mg/week which led to a rapid resolution of the skin lesions within two months. Topical steroids and dapsone were ineffective in this case.

DISCUSSION

Bullous pemphigoid (BP) is an acquired autoimmune blistering disorder affecting mostly elderly people and rarely children [1]. There have been fewer than 100 cases of childhood bullous pemphigoid reported. Clinically, it is similar to adult bullous pemphigoid, with tense vesicles and bullae of variable size and localization. Urticarial papules with pruritus are usual in bullous pemphigoid [1]. However, mucous membrane involvement, thus hand and feet lesions are more frequent in children [2]. Histology and direct immunofluorescence are fundamental for the diagnosis. It presents as a subepidermal bullae with variable amount of eosinophils; associated to a linear deposition of IgG and/or C3 at the basement membrane zone. The presence of IgG antibodies reacting with the

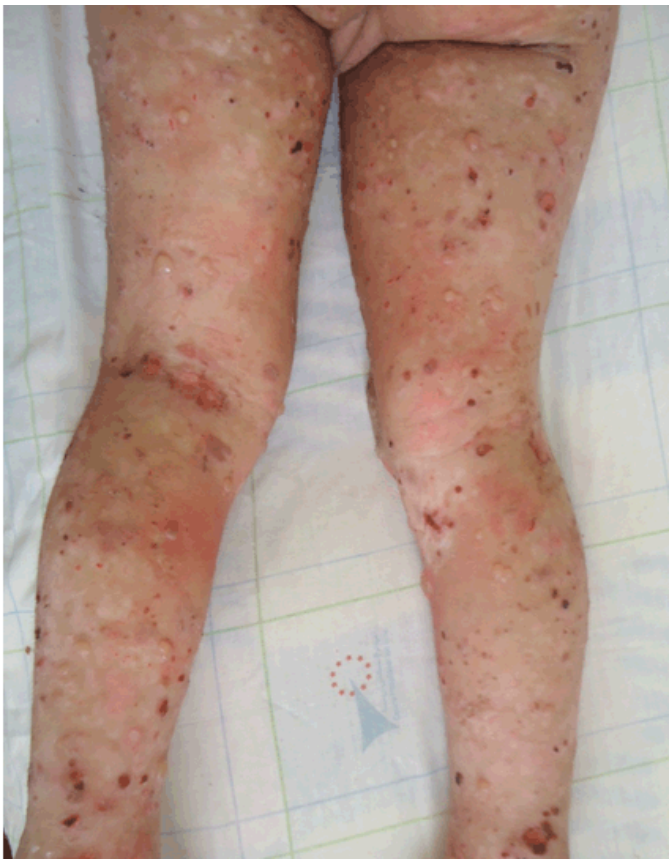

Figure 1: Presence of multiple tight, widespread vesicles bullae and crusted erosions.

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basement membrane zone is considered to be diagnosis criterion. Clinical differential diagnoses include linear IgA dermatosis, bullous impetigo, dermatitis herpetiformis, mastocytosis, epidermolysis bullosa, and porphyria. In our case clinical and immunopathological criteria made the diagnosis. Topical or systemic corticosteroids represent the first line treatment at a dose of 0.5–1 mg/kg/day. Other immunosuppressive drugs frequently used are methotrexate, azathioprine, cyclosporine, cyclophosphamide, and mycophenolate mofetil [3].

CONCLUSION

In conclusion, a diagnosis of childhood bullous pemphigoid should be considered in any infant or child presenting with vesiculobullous eruptions, especially on the palms and soles. Early onset of appropriate treatment must be done to avoid infections, fluid and electrolyte loss.

Keywords: Childhood bullous pemphigoid, Corticosteroids, Dapsone, Methotrexate

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Author Contributions

Amarouch Hajar – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Benzekri Leila – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Ismaili Nadia – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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