AUTOIMMUNE BULLOUS DISEASES

CHRONIC BULLOUS DISEASE IN CHILDHOOD SHOWING REMARKABLE IMPROVEMENT WITH DAPSONE: A CASE REPORT

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Introduction: Chronic bullous disease of childhood (CBDC) is a rare autoimmune blistering disease, non-hereditary, and is characterized by immunoglobulin A (IgA) deposits at the basement membrane zone. It has been considered the pediatric variant of adult linear IgA disease.

Case report: A 8-year old girl presented with multiple vesicles and tense bullae on the lower legs, some gradually evolved to eryterythematous plaques with erosions on the oral mucosa, neck, both axillary area, buttock and extremities. Skin punch biopsy showed subepidermal blister containing fibrin, many neutrophils and eosinophils. Direct immunofluorescence showed strong linear deposits of IgA (+2), weak linear deposits of IgG (+1) and C3 (+1) in basement membrane zone. Patient was initially given oral glucocorticoids, and treated with doxycycline as steroid sparing agent with little improvement. Resolution of lesions were noted under maintenance dapsone 100mg daily for several months.

Conclusion: CBDC is a recurrent blistering disease with a potential to become generalized or to severely affect the mucosa. This disease is often mistaken for other bullous diseases such as impetigo (children) or bullous pemphigoid (adults). It is important to make the correct diagnosis, based on the clinical and histopathological, immunohistochemistry in order to provide the optimum treatment and care for the patient. The principle of therapeutic decision is to achieve long lasting remission with the minimum prescription of a systemic drug.

Commercial funding N/A
Conflict of interest : N/A
Keywords: Chronic Bullous Disease of Childhood, Immunoglobulin A, blistering disease