

Picture Story

Linear IgA dermatosis in a 7 year old child

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Case report

A 7-year old girl was assessed for generalized, recurrent blisters of 2 months duration. The blisters were filled with clear fluid and many had healed spontaneously. However, new blisters appeared in normal skin and around healed lesions. Despite receiving multiple courses of antibiotics for presumed bullous impetigo, her condition deteriorated. Examination revealed blisters of different ages, mostly in lower and upper limbs, perineal region and on lower trunk. Vesicles and bullae were seen on healthy skin with an erythematous base. Some lesions clustered around previously healed annular scars forming a rosette like pattern (Figures 1 & 2).



Figure 1: Sausage shape bullae arranged in annular or rosette like pattern around a central crust.



Figure 2: Lesions at different ages

These vesicles were tense and filled with serous fluid. She did not complain of itchiness and did not have mucosal lesions. Clinical differential diagnoses included linear IgA dermatosis, bullous pemphigoid and epidermolysis bullosa. Her complete blood count, renal and liver function tests were normal. Subsequently a biopsy was performed. Histopathological examination revealed a sub-epidermal blister with neutrophilic and lymphocytic infiltrate in perivascular locations. Linear IgA deposition in the epidermal basement membrane was confirmed with direct immunofluorescence (Figure 3).

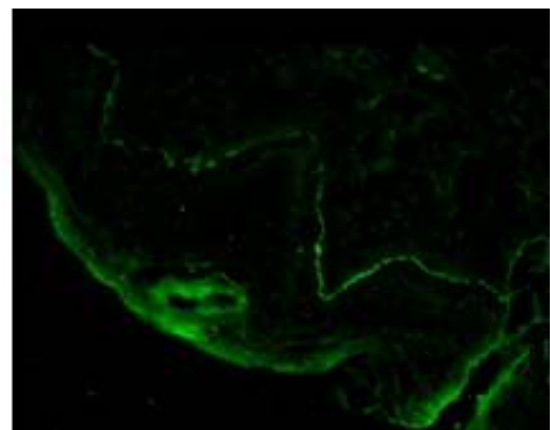


Figure 3: Direct immunofluorescence showing linear IgA deposition in epidermal basement membrane

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Oral prednisolone (1mg/kg/day) was initiated and a partial response was seen. With the addition of dapsone there was complete remission of symptoms (Figure 4).



Figure 4: Clinical improvement following dapsone and corticosteroid therapy

Discussion

Children with linear IgA dermatosis (LAD) typically present with tense bullae or vesicles distributed over the lower trunk, thighs, buttocks and face¹. Affected superficial dermis is infiltrated predominantly with neutrophils as a result of a burst of inflammatory mediators released following a reaction between IgA auto-antibodies and basement membrane antigens of the skin². Linear deposits of IgA in the basement membrane by direct immunofluorescence confirms the diagnosis². First line treatment is dapsone³.

In LAD there are typically two variants⁴. In children, the onset is commonly seen in the first decade of life with the peak incidence during the preschool ages⁴. Lesions are bullae/vesicles filled with serous fluid or blood, and often clustered together and formed on erythematous or normal base⁴. Some lesions classically form a rosette like pattern around the healed lesions⁴. It rarely persists after puberty³. In adults, peak incidence is after 60 years of age⁴. Mucosal lesions are more common in adults⁵. Painful oral ulcers, conjunctival ulceration, blindness and breathing difficulties due to laryngeal involvement are some of the complications of mucosal involvement⁶. Itchiness may be seen in both adults and children⁷.

Circulating IgA targets antigens in the lamina lucida of the epithelial basement membrane⁸. IgA1 subclass is involved in LAD¹. Antigens are components of the dermo-epidermal junction adhesion complex². With deposition of IgA, complement activation leads to attraction of neutrophils which release enzymes and weaken the basement membrane, resulting in sub-epidermal blister formation⁷. LAD is drug induced or idiopathic⁹. Vancomycin, cephalosporins, penicillins and nonsteroidal anti-inflammatory

drugs are implicated^{10,11}. There are reports showing an association with autoimmune disorders like rheumatoid arthritis, ulcerative colitis, and Crohn's disease^{12,13}.

The first line medication is dapsone³. Main adverse effects of dapsone are methaemoglobinaemia and G6PD deficiency¹⁴. Other drugs that may be used are oral steroids, azathioprine, mycophenolate mofetil and methotrexate³. It is reported that antibiotics such as erythromycin and doxycycline help in treatment¹⁵. Usual disease course is 2-6 years⁴. However, some get recurrences or even persistence of the disorder¹⁶. LAD should be considered in the differential diagnosis of blistering disease in children.

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