Linear immunoglobulin A (IgA) bullous dermatosis

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Introduction

Linear immunoglobulin A (IgA) bullous dermatosis (LABD), first described by Bowen in 1901, is an autoimmune disease occurring in both children and adults[1,2]. Around two thirds of cases are induced by antibiotics, nonsteroidal anti-inflammatory agents and diuretics[2]. Childhood LABD is synonymous with chronic bullous disease of childhood (CBDC)[3].

Case report

A 3 year old boy presented with blisters all over the body, more over the lower limbs, upper limbs and ear lobes, since 10 days associated with severe itching and mild pain (Figure 1).

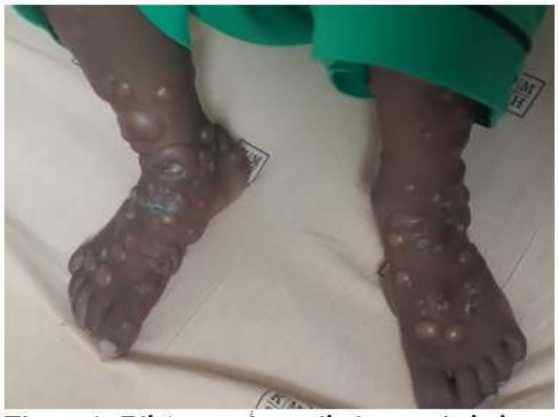

Figure 1: ***Blisters on lower limbs on admission***

Ruptured blisters were also present. There was no history of fever. The blisters started as small papules on the lower limbs and spread all over the body.

Total white blood cell count was 11,200 /cu mm (24% polymorphs, 50% lymphocytes, 24% eosinophils, 2% monocytes). Erythrocyte sedimentation rate was 8 mm in the first hour, the haemoglobin (Hb) level 12.7 g/dl and the platelet count 458,000 /cu mm. Serum immunoglobulin E (IgE) level was 112.4 IU/ml (normal up to 60 IU/ml), serum immunoglobulin A (IgA) level 72 mg/dl (normal 19-220 mg/dl), serum immunoglobulin M (IgM) level 167 mg/dl (normal 40-180 mg/dl) and the serum immunoglobulin G (IgG) level 800 mg/dl (normal 500-1300 mg/dl). The gram stain of blister fluid showed a few pus cells but no organisms. Skin biopsy taken from the leg was consistent with linear IgA bullous dermatosis. Direct immunofluorescence showed a linear basement membrane zone band with IgA, confirming linear IgA disease. Glucose-6-phosphate-dehydrogenase (G6PD) level was 11 units/g Hb (normal range 4.6-13.5 units/g Hb). He was successfully treated with intravenous methylprednisolone (1.5mg/kg/day) for 5 days followed by oral steroid (1mg/kg) and oral dapsone (1mg/kg). He is on regular follow up and his skin lesions have improved (Figure 2).

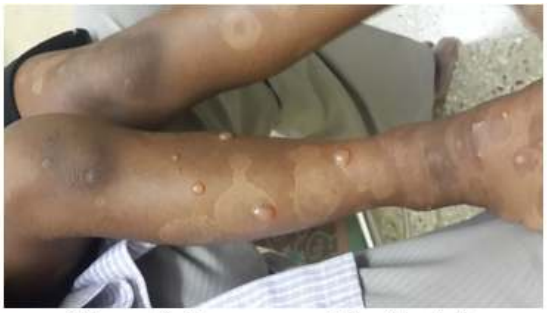

Figure 2: ***Improvement in skin lesions***

Discussion

Prevalence of LABD in children is unknown. Age of onset in children ranges from 1 to 10 years[4]. Our patient was 3 years old. Childhood LABD has rarely been associated with ulcerative colitis and autoimmune lymphoproliferative disease[5,6]. LAPD characteristically has linear IgA deposits in the basement membrane zone of the skin, circulating basement membrane zone antibodies being detected in over 80% of patients[3]. Our patient showed a linear basement membrane zone band with IgA on immunofluorescence. How drugs stimulate the immune system to produce IgA antibodies against the basement membrane in LABD is still unknown[5]. Both idiopathic and drug-induced LABD can be triggered by physical trauma, burns, or infections[3]. There were no triggering factors noted in our case. Typically, there is an interval of 2 to 28 days between the drug involved and the bullous eruption[2].

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Lesions of drug-induced LABD resolve spontaneously within 2 to 7 weeks following withdrawal of the drug. No drug was incriminated in our patient. In a study by Wojnarowska F, et al. of 25 cases of childhood LABD, 64% remitted within 2 years and only in 12% was the disease active after puberty[8]. The treatment of choice in LABD is dapsone alone or in combination with systemic steroids[9,10]. Our patient was treated with dapsone and steroids. Dapsone may cause haemolysis in G6PD deficient individuals. In our patient the G6PD level was normal. Flucloxacillin may be considered as an alternative therapy[11]. In dapsone resistant cases, tacrolimus ointment may be beneficial[12].

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