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A Case of Linear IgA Dermatosis in a Young Child

V. Thadchanamoorthy[1*], V. Vijayakanth[2] and N. Thamilvannan[2]

[1]Faculty of Health Care Science, Eastern University, Sri Lanka.
[2]Teaching Hospital, Batticaloa, Sri Lanka.

Authors' contributions

This work was carried out in collaboration among all authors. Author VT led clinical management of the patient, wrote manuscript and performed literature survey. Author VV edited the manuscript. Author NT confirmed the diagnosis. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Diagnosis of Linear immunoglobulin A (IgA) bullous disease (LABD) is difficult as they present to relevant clinician late. This is the case of a 4-year-old Sri Lankan boy with a 2 months history of non-itching, blistering lesions on his extremities. Medical history was unremarkable. Medical treatments included a visit to a general practitioner, where he was treated with antibiotics unsuccessfully. We performed a skin biopsy and Direct immunofluorescent studies revealed a linear deposition of IgA antibody at the basement membrane. The clinical diagnosis of linear IgA dermatosis was established, with no eliciting cause. Resolution of blisters was achieved with oral prednisolone, dapsone and antibiotic treatment. This is the first case to our knowledge occurring in this age group with involvement of only extremities, sparing genital area.

Keywords: Linear immunoglobulin a bullous disease; linear IgA dermatosis; dapsone; immunofluorescence.

1. INTRODUCTION

Linear immunoglobulin A (IgA) bullous disease (LABD) is a rare mucocutaneous autoimmune subepidermal blistering disease in skin and mucus membrane [1]. In childhood, linear IgA bullous disease typically presents before puberty. It starts abruptly with blistering from genital area

**Corresponding author: E-mail: vijayakumaryt@esn.ac.lk;*

and progress to face and limbs including hands and feet. There might be eye involvements which leads to irritation, dryness, light sensitivity, blurred vision, corneal scarring and even blindness but extremely rare [2]. We report a young child who initially thought to have bullous impetigo, was ultimately diagnosed as Linear IgA Dermatosis (Lings & Bygum, 2015 Venning, 2011).

2. CASE HISTORY

A-4 –year- old boy was admitted to paediatric ward with extensive recurrent bullous lesions for two months. According to the history, blisters have started from lower extremities, then spread to upper limbs with new blisters emerging in normal skin and around the old blisters. They were non-itchy with no history of fever at any time of illness. Initially, the blisters were filled with clear fluid, but later infected with secondary infection following rupture. The child had been given antibiotics by general practitioner without any improvement.

Examination revealed blisters of different ages and mostly distributed in extremities (Fig. 1). It wasn't present in trunk and perineum. Mucosal membranes were intact. Blisters were seen on the erythematous bed. Some of the blisters were grouped around a healed scar like rosette (Fig. 2). Some were grouped giving cluster of jewel appearance. Some vesicles were tensed and contained clear fluid (Fig. 3), Rest of the systemic examination were normal.

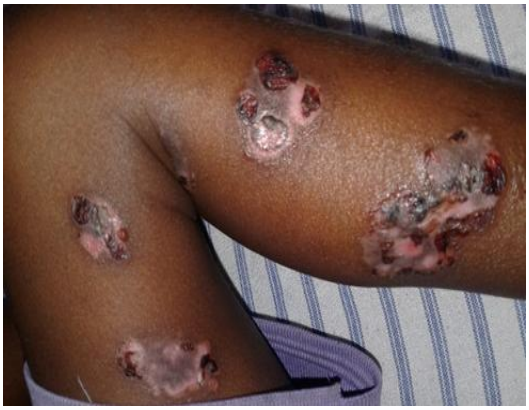

Fig. 1. Various stages of blisters

His full blood count and C-reactive protein were normal. Renal and liver function were also normal. Skin biopsy revealed subepidermal blister with neutrophilic predominance. Linear immunoglobin A deposition in epidermal layer was confirmed by immunofluorescence. Diagnosis of linear IgA dermatosis was made.

Child was treated with oral prednisolone and dapsone. Secondary infection was treated with oral antibiotic. He improved with medication and was discharged with follow up arrangement.

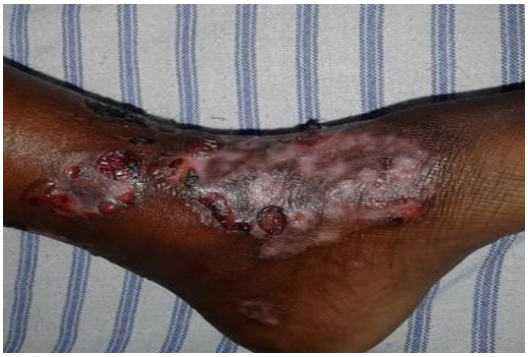

Fig. 2. Rosette like lesion

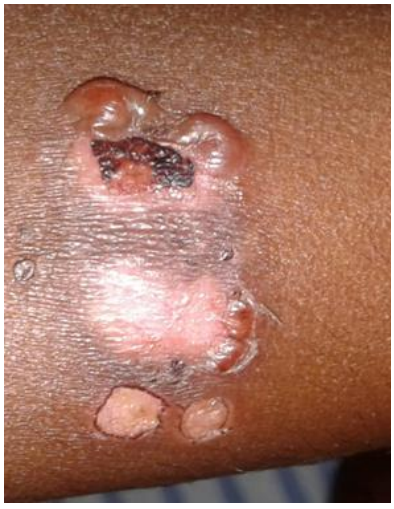

Fig. 3. Tense bullae

3. DISCUSSION

Linear immunoglobulin A (IgA) vesiculobullous dermatosis (LABD) was first discovered by Bowen in 1901 as an autoimmune disease arising in all age group [3]. It is more prevalent in developing countries [2]. The prevalence of LABD in childhood is still not known. Although the age of onset in children varies from 1 to 10 years, the majority of childhood cases present by the age of 6 to 8 years. Our child presented at age of 4 years.

The exact pathogenesis of LABD is still unclear. It is considered to be a heterogeneous disorder or associated with various stimuli such as infections, drugs, haematological diseases, rheumatological conditions, inflammatory bowel disease, solid tumours and some malignancy such as lymphoma [2,4]. It is caused by IgA autoantibodies directed against different antigens of the basement membrane zone (BMZ) of the skin and mucosae. It creates blistering at the site of splitting between the epidermis and the dermis [2,5].

LABD clinically presents with widespread vesiculobullous eruption on both upper and lower limbs involving the dorsa of the hands and feet, groin area, in the face including perioral area and mucus membranes which include eye and oral mucosa and the gravity of itchiness may vary [2]. However, our patient had lesions on the extremities only. It was non-itchy and there is no mucous membrane involvement. Our patient also had bullae which typically organized in rings (annular lesions) or form a target shape. The predisposition for new blisters to arise in a ring around an old one is called the string of beads sign, and groups of small blisters is described as a cluster of jewels. Secondary lesions consist of crusts, scratch-marks, sores and ulcers [5]. There are conditions which mimic LABD include bullous impetigo, dermatitis herpetifomis, epidermolysis bullosa, bullous popular urticarial, childhood pemphigoid, infected scabies and chicken pox [6].

Diagnosis is by biopsy and presence of a continuous linear IgA deposit along the basement membrane zone is characteristically seen on direct immunofluorescence [7]. Our patient also has similar report.

The treatment of choice in LABD is dapsone alone or in combination with steroids [4] than topical corticosteroid treatment [4,7]. Our patient was treated with dapsone and steroid. Although it has a chronic cause in children [2]. Our child is free from recurrence on one year follow-up.

4. CONCLUSION

LABD is usually mimic like other bullous disease. So careful inspection of lesions will guide us to diagnose it. Yet biopsy with direct immunofloracence is the confirmatory test.

AVAILABILITY OF DATA

The data that support the findings of this case report are available from Medical Records Department, Batticaloa Teaching Hospital, but restrictions apply to the availability of these data, which were used under license for the current report and so are not publicly available. Data are, however, available from the authors upon reasonable request and with permission of Medical Records Department, Batticaloa Teaching Hospital, Sri Lanka.

CONSENT

Written informed consent was obtained from patient's legal guardian for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

ETHICAL APPROVAL

It is not applicable.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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