IJBPAS, September, 2023, 12(9): 4211-4214

ISSN: 2277–4998

International Journal of Biology, Pharmacy and Allied Sciences (IJBPAS)

'A Bridge Between Laboratory and Reader'

www.ijbpas.com

CHRONIC BULLOUS DISEASE OF CHILDHOOD - A CASE REPORT

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Received 26th July 2022; Revised 19th Sept. 2022; Accepted 11th Jan. 2023; Available online 1st Sept. 2023

https://doi.org/10.31032/IJBPAS/2023/12.9.7415

ABSTRACT

Chronic Bullous Disease of Childhood (CBDC) is an acquired autoimmune blistering disorder of childhood, and it occurs without preferance to race and gender. CBDC may even temporarily improve with the brief course of antibiotics. Disease onset usually occurs at the age of 6 months to 10 years with a mean age of 4.5 years. However, disease onset has been reported as early as within 24 to 48 hours after birth and up to the age of 12 years. The 'clusters of jewels' pattern is typical of CBDC. In this pattern of distribution tense vesicles arise at the periphery of old lesions. Tense pruritic blisters of variable sizes, small clear-filled vesicles normal appearing skin or at periphery of an annular erythema, along with crusts, excoriations and erosions may be present in the patient. It is characterized by the deposition of linear band of IgA along the dermal-epidermal junction. CBDC is thought to be the result of a humoural response to a normal constituent of the epidermal basement membrane zone (BMZ). There may be genetic susceptibility to CBDC with an increased incidence of HLA-B8, HLA-DR3 and HLA-DQW2. However, the molecular basis of CBDC has not yet been clearly determined. This condition is seen in all ethnic groups seems to be more common in developing countries.

Despite its good prognosis, most children are offered treatment to decrease the disease severity and shorten its duration. The most frequent medication is dapsone, starting at 0.5 to 1 mg/kg of body weight per day increasing up to2 mg/kg depending on the response.

Keywords: Chronic Bullous Disease of Childhood, Linear IgA dermatosis

INTRODUCTION:

Chronic bullous disease of childhood (CBDC) is the most common, non-hereditary, autoimmune blistering disorder of childhood. This rare condition, characterised by linear IgA staining on direct immunofluorescence of the basement membrane of the squamous epithelium, has been considered the paediatric variant of adult linear IgA disease **[1]**. Autoimmune blistering disorders comprise a series of conditions in which autoantibodies target components of the skin and mucous membranes, leading to blister and bullae formation. Most conditions in the spectrum of autoimmune blistering disorders are uncommonly seen in the pediatric population, even the most common ones, such as chronic bullous disease of childhood and dermatitis herpetiformis; however, they often come into the differential diagnosis of other more common pediatric entities. In addition, prompt recognition and treatment avoids unnecessary morbidity and improves ultimate outcome **[2]**.

CASE REPORT:

A 5 years old male patient reported to the hospital with the chief complaints of skin lesion over whole body. The patient is on Rx for the same complaints since 1.5 years. Clinical examination revealed that the patient is having same condition with the development of clusters of blisters all over the body.

Patient developed clear fluid filled lesion over left leg which gradually increase in size and number and gradually developed over B/L back, abdomen, B/L hands, face, mouth. Lesion ruptured by its own within 3-4 days leaving behind raw area. Lesion associated with itching after rupture, was mild in intensity, intermittent and relieved by medication without diurnal variation.

Skin lesion was associated with mild fever without chills and rigors without diurnal variation. Under macroscopic examination, received skin biopsy of 0.3 cm. Multiple sections show epidermal ulcerations with massive number of neutrophils and acantholytic squamous cells mixed with fibrinous debris. Relatively under microscopic examination, adjacent epidermis show irregular rete ridges with marked atypia and prominent intraepidermal neutrophils. Dermal perivascular inflammation is also seen. Epidermal blister roof is not seen. Various examination revealed the presence of chronic bullous disease of childhood.

Patient was examined under proper light and with consent. Based on different examination examined under general and cutaneous condition and other hematological test, patient was given treatment with TESS Buccal paste (LABD)

and xylocaine viscous gel (LABD) before meal.

DISCUSSION

Linear IgA dermatosis or chronic bullous disease of childhood (CBDC) is generally a rare, nonhereditary, autoimmune disease. It is, though, the most common chronic bullous disease during the first decade of life. Bowen described in 1901 the first six cases of linear IgA bullous disease of childhood, by that time considered as dermatitis herpetiformis **[3]**. A changing terminology for this entity existed for almost 80 years. In 1979, chronic bullous disease of childhood or linear IgA dermatosis of childhood was classified as a subepidermal bullous disease characterized by the presence of continuous linear deposits of IgA along the basement membrane zone and as a separate entity from bullous pemphigoid or dermatitis herpetiformis[4]. Controversies on this disease continue regarding the age - related forms. Recently, Haneef *et al* **[4]** proposed that all pediatric cases showing the typical clinical picture of 'cluster of jewels' or 'string of pearls' sign to be included under the broad term 'chronic bullous disease of childhood,' irrespective of the nature of the immune deposits, as there are cases which share this clinical feature but show IgG predominance. The authors claim that linear IgA dermatosis of childhood is a separate entity with variable immune deposits but unique clinical features, while the adult disease has variable clinical features and the linear deposition of predominantly IgA is an essential diagnostic finding[4]. Linear IgA dermatosis with adult onset and linear IgA dermatosis of childhood share the same basement membrane zone antigens and thus, they are currently considered as variants of the same disease **[5-6]**.

CONCLUSION:

Chronic bullous disease is a unique variant of linear antigen bullous disease which is characterized by linear deposition of basement membrane zone –specific IgA. Clinically, it can be separated into an adult and childhood form. Immunopathologically, the IgA antibodies have been reported to react with numerous basement membrane antigens. The therapy of choice is dapsone.

CONFLICT OF INTEREST:

The author declares no conflict of interest.

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