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Dermatology

A RARE CASE REPORT OF CHRONIC BULLOUS DISORDER OF CHILDHOOD

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ABSTRACT Chronic Bullous disease of childhood is a rare autoimmune condition resulting in clusters of blisters resembling a 'STRING OF PEARLS' often on the face extremities or genitals.A Case report of a 5yr old male child who presented with chronic bullous disorder of childhood confirmed by biopsy and immunofluorescence.

KEYWORDS : chronic bullous disorder of childhood, IgA dermatosis, blistering disease of children, immunobullous disorder of childhood, string of pearls

INTRODUCTION:

Chronic bullous disease of childhood (CBDC) is a rare self-limiting disease of young children which resolves within months to years. Clinically, it is characterized by tense vesicles or bullae, on histopathological shows subepidermal blister with a predominantly neutrophilic infiltrate and immunofluorescence showing linear IgA deposits. The disease is usually associated with significant morbidity and usually requires standard systemic therapy. Treatment is aimed at controlling blistering while avoiding adverse reactions.

CASE REPORT:

A 5-year-old boy presented to The Department of DVL., Government Medical College, Kadapa with blistering over the lower limbs, abdomen and groin associated with intense pruritis.

History revealed that the lesions usually burst open on its own within two to three days, forming erosions with serous discharge and crusting which was followed by new blisters appearing around the healing erosion. There was no history suggestive of mucosal involvement, photosensitivity, prior drug intake, trauma, consanguinity or any similar lesions in the family.

General physical and systemic examination were unremarkable.

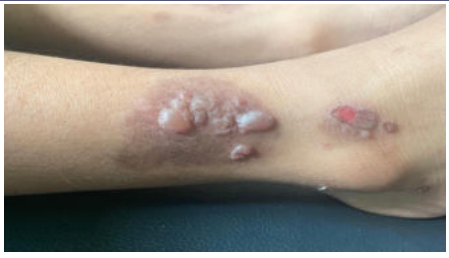
Cutaneous examination revealed multiple grouped vesicles, bullae, and erosions involving the entire body, predominantly the legs perineal, and lower trunk with sparing of palms, soles, and mucosa. The characteristic annular arrangement of the vesicles / bullae over a hyperpigmented background, described as 'string of pearls sign' or 'cluster of jewels', was seen in many sites Routine blood investigations, Gram stain, and Tzanck smear were normal. Histopathology showed subepidermal blistering dermatitis with numerous neutrophils within the blister cavity. The blister roof showed mild spongiosis. Periphery of the blister showed mild spongiosis with neutrophils and collection of few neutrophils within the papillary dermis. The floor of the blister shows moderately dense perivascular interstitial infiltrate of neutrophils and eosinophils.

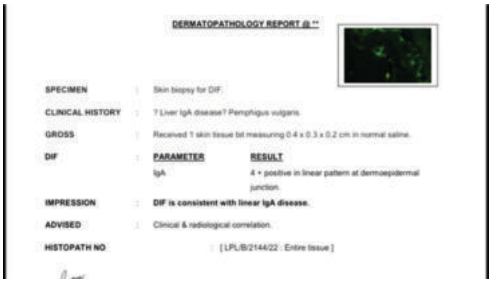
Direct immunofluorescence of the perilesional skin revealed linear IgA [++++], and C3, along the basement membrane zone.

Taking into account of clinical, histopathological, and immunopathological features, a diagnosis of chronic bullous disease of childhood was made.

After G6PD assay being normal, patient was started on Oral Dapsone 25 mg / day (body weight was 15 kg) and oral Prednisolone 15mg/day along with symptomatic management.

The lesions started clearing by 72hrs and clearance of 90% lesions was observed by 10 days, and the patient is on continued medication till date (1 month since the start of therapy}. Baseline investigations were repeated on week 2 and week 4, which showed no derangement.





DISCUSSION

Although rare, CBDC is the most common immunobullous disorder seen in childhood which was initially described in 1970. CBDC is the childhood form of linear IgA bullous disease (LABD) of adults. It is characterized by linear deposition of IgA directed against a fragment of collagen XVII (linear IgA disease antigen LAD-1), at the epidermal basement membrane zone.

The age of CBDC onset is usually below 5 years. Our patient presented at the mean age of onset for CBDC, around age 5 years. A classical CBDC skin lesion is a central bulla surrounded by smaller vesicles-a pattern known by several descriptive names: “rosette”, “string of pearls” and “cluster of jewels”. The most commonly involved sites in children are the lower trunk, perianal, perioral and upper thighs as seen in our case. Recurrence rates are higher in children.

Onset of CBDC lesions is usually insidious.

Our patient's initial lesions suggestive of CBCD, confirmed by histopathological examination . The differential diagnosis included bullous impetigo, bullous erythema multiforme, bullous pemphigoid and pemphigus vulgaris. A smooth, linear pattern of immunoglobulin A deposition in the basement membrane zone on direct immunofluorescence is considered the gold standard for establishing a diagnosis, so as in our case.

First-line treatment includes oral sulfone, dapsone or sulfapyridine, with an oral corticosteroid. The risk of sulfone-associated severe haemolytic anaemia is high in people with inherited G6PD deficiency, so a normal screening G6PD level is required before initiating treatment which was followed in our patient.

Oral corticosteroids provide short term relief, but chronic use is not advisable due to long term side effects.
Conflict of interest: Nil.

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