Suspected case of chronic bullous disease of childhood in a rural area of Southern Mozambique

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DESCRIPTION

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] although CBDC tends to occur in children less frequently, usually as a response to neoplasms or drug hypersensitivity.

The clinical hallmark of this condition is the abrupt appearance on normal or erythematous skin of many large, tense bullae, filled with clear or sometimes haemorrhagic fluid and acquiring the typical rosette-like pattern of '*string of pearls*' (figure 1), with or without associated pruritus. Bullae predominantly appear in the genital and buttocks area, although they can also appear in the trunk, extremities and face (figure 2). Mucous membranes may also be affected. Secondary impetiginisation may be frequent, but scarring sequelae are not common. The disease is often self-limiting, but may persist for months or even years, with occasional recurrences, before complete resolution, usually by puberty.

Diagnostic confirmation requires direct immunofluorescence studies, where IgA linear deposition at the dermal–epidermal junction of peri-lesional skin areas can be observed. In resource-constrained settings, however, where such diagnostic methods are seldom available, diagnosis remains clinical, but requires the exclusion of other diseases such as bullous pemphigoid, dermatitis herpetiformis or erythema multiforme.[2 3]

The majority of cases respond well to long-term oral dapsone, or sulfapyridine. Alternatively, oral or systemic corticosteroids or tacrolimus can be

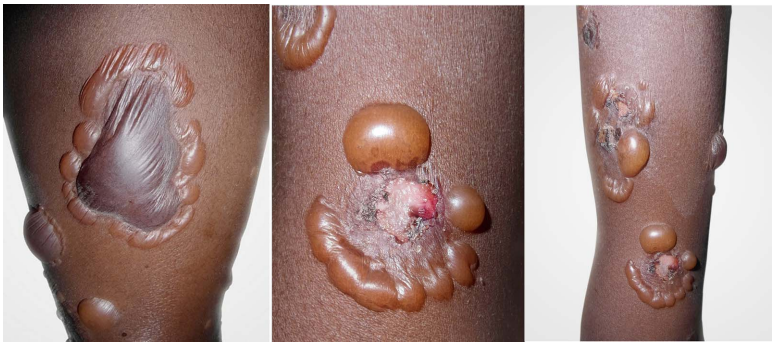

Figure 1 Classic '*string of pearls*' or '*cluster of jewels*' pattern of lesions with tense vesicles arising at the periphery of old lesions, located in the extremities and genital area of an HIV-negative male Mozambican child aged 8 years, with no other significant diseases.

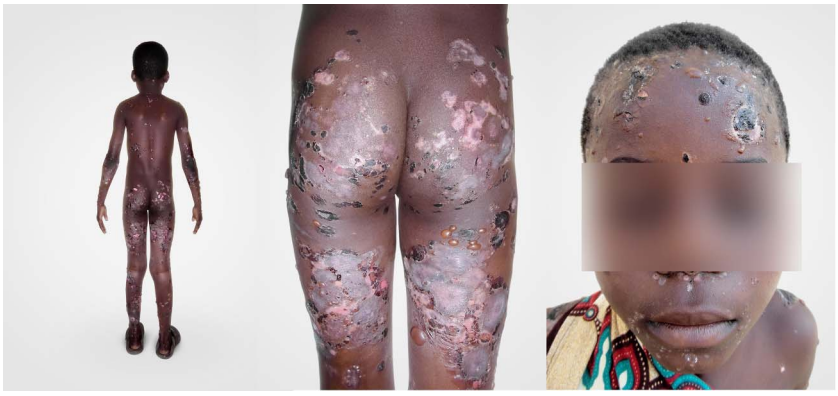

Figure 2 Generalised distribution in the same child of a heterogeneous set of cutaneous lesions, including vesicles, tense bullae and impetiginised erosions. Lesions resolved completely about 4 months after the initiation of oral corticosteroids, as dapsone was unavailable in Mozambique.

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effective, and sometimes may need to be complemented with topical or oral antibiotics.

Learning points

- Albeit rare, chronic bullous disease of childhood is the most frequent non-hereditary autoimmune blistering disease in childhood.
- In African settings, where diagnostic methods remain scarce, it is important to recognise clinically this disease, and initiate a specific long-term treatment with dapsone or corticosteroids.
- In spite of its impressive appearance, the disease is often self-limited and disappears by puberty.

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Competing interests None declared.

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