A healthy 5-year-old boy presented with a three-month history of recurrent episodes of pruritic vesiculobullous lesions predominantly in the genital and inguinal areas. Clinical examination revealed a linear distribution of vesicles, bullae, papules and erosions in the retro-auricular region and upper trunk. In the genital and inguinal areas the lesions had an annular and herpetiform conformation (Fig. 1). Skin biopsy revealed numerous subepidermal blisters with a mixed inflammatory infiltrate (Fig. 2). Direct immunofluorescence showed linear deposits of immunoglobulin (Ig) A throughout the basal membrane zone (Fig. 3). In view of these findings a diagnosis of linear IgA bullous dermatosis (LABD) was established. The patient was treated with sulfasalazine (35 mg/kg/day), but recurrent outbreaks were seen, and so colchicine (1 mg/day) was added, which was well tolerated. After one year of treatment, the patient remains asymptomatic.

LABD, also known as chronic bullous disease of childhood, is a rare immune-mediated vesiculobullous disease. In children, the peak incidence is at 4-5 years of age and it rarely persists after puberty. Although spontaneous remission can occur, most cases require treatment. The first-line treatment is dapsone or sulfapyridine. In selected patients, oral prednisone or another corticosteroid-sparing agent such as colchicine may be used to achieve complete control of the disease.

**Figure 1.** Genital and inguinal areas with erosions and vesicles on erythematous skin with an annular conformation (crown of jewels conformation).

**Figure 2.** Subepidermal blister with a mixed inflammatory infiltrate, predominantly neutrophils and eosinophils (haematoxylin-eosin staining, x100).

**Figure 3.** Linear deposits of IgA throughout the basal membrane zone (original magnification x20).

**Palavras-chave:** Criança; Dermatose bolhosa IgA linear/tratamento e diagnóstico
Keywords: Child; Linear IgA Bullous Dermatosis/diagnosis and therapy

WHAT THIS CASE TEACHES
- The presence of tense bullae and vesicles on normal or urticarial skin in the anogenital region, perineum and lower abdomen should raise suspicion of linear IgA bullous dermatosis.
- Diagnosis relies on histology and direct immunofluorescence.
- Spontaneous remission can occur, but most cases require treatment with dapsone or sulfapyridine.

Conflicts of Interest
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Protection of human and animal subjects
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Confidentiality of data
The authors declare that they have followed the protocols of their work center on the publication of patient data.

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