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A Case of Linear IgA Bullous Dermatosis in a Child

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TO THE EDITOR,

A 16-year-old girl was referred to our department, complaining of pruritic vesicles which had been present for four years. The vesicles primarily affected her lower legs soon after having a high fever and abdominal symptoms at the age of 12 years. Since then, vesicles had repeatedly appeared on the trunk and upper limbs. There was no family history of cutaneous disease and no drug intake prior to the onset of this symptom. Physical examination revealed tense vesicles with crusts scattered on the trunk and elbows (**Figure 1**). The patient told us at the physical examination that her fingers had been swollen repeatedly. A histopathological examination of a biopsy specimen from the elbow showed a subepidermal edema, neutrophilic abscess in the papillary layers, and inflammatory cellular infiltrates in the upper dermis (**Figure 2a**). Positive linear deposition of IgA and C3 at the dermo-epidermal junction was evident with direct immunofluorescence study (**Figure 2b**). She was diagnosed as having linear IgA bullous dermatosis (LABD). She was treated with dapsone, initially at a dose of 75mg/day. The lesions gradually resolved within three weeks of starting treatment.

LABD is an autoimmune blistering disease that is characterized by IgA deposits at the basement membrane zone [1]. It mainly affects children at preschool age, although newborns and adolescents have been affected. The childhood form is characterized by tense annular or arcuate bullae and vesicles with serous or hemorrhagic fluid. Classic sites of LABD during childhood are the trunk and iliosacral region; further regions such as the legs, face, or large body folds can also be affected.

To date, more than 70 cases of childhood LABD have been reported. Of these, we could obtain information on age and gender of 51 patients. The mean age was 7.4 years (range: 4 days-15 yrs), and 32 patients (63%) were boys. Extracutaneous symptoms of LABD are mucosal involvement, especially of the oral mucous membranes. In our case, vesicles appeared at the age of 12, and she repeated swelling of the fingers during the course, but examination by magnetic resonance imaging did not detect inflammation. Previously, a similar case was reported [2], in which spondylarthropathy was associated with LABD. However, there are very few reports on the co-existence of LABD and arthritides, and thus the relationship between LABD and joint manifestation is currently unknown.

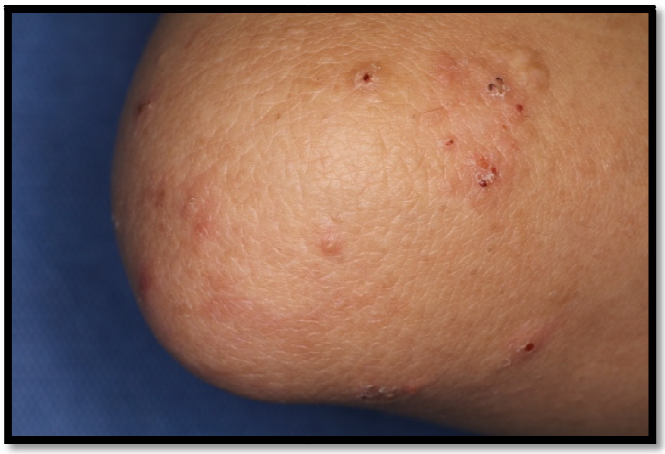

Figure 1. Clinical feature of the primary cutaneous lesion showing tense vesicles and scales scattered on the elbows.

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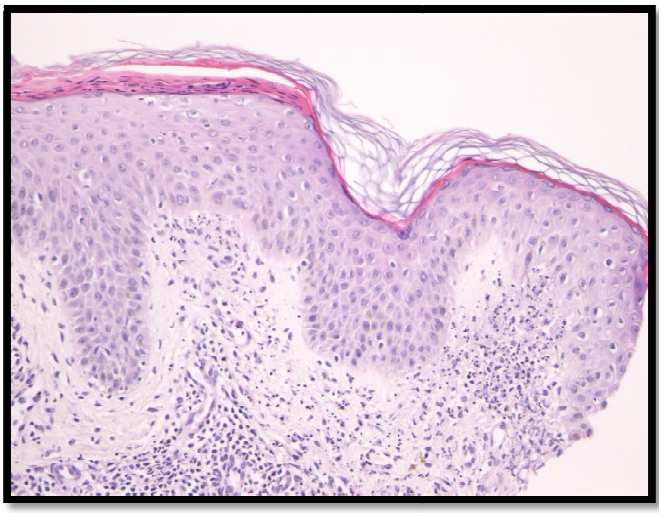

Figure 2. (a) Histology demonstrating neutrophilic abscess in the papillary edematous layers.

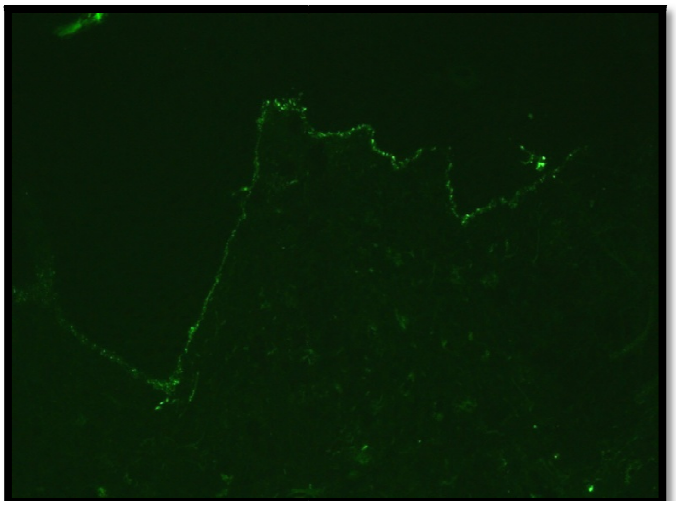

Figure 2. (b) Immunofluorescence findings in LABD demonstrating positive linear deposition of IgA and C3 at the dermo-epidermal junction.

The eruption is typically described as self-limiting, clearing within a few months or several years. However, for cases without spontaneous regression, therapies for LABD include dapsone or a combination therapy. Sulfapyridine, colchicines, erythromycin, oxacillin sodium, and dicloxacillin have also been tried with variable results [3]. Our patient showed a good response to treatment with dapsone. Three weeks after the initiation of the treatment, the lesions gradually resolved and the finger swelling completely disappeared. Dapsone can be tapered but not be stopped, because the vesicles relapse when it is discontinued. She is now well-controlled with low-dose dapsone (25 mg per day).

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