Drug-induced IgA bullous dermatosis: a rare presentation of SLE

Sirs,

Linear immunoglobulin A dermatosis (LAD) is an autoimmune subepidermal vesiculobullous disease, histopathologically characterised by linear deposition of IgA at the basement membrane zone (1). Here, we describe a child with drug-induced severe IgA dermatosis associated with systemic lupus erytematosus (SLE), which is reported very rare (2).

A 7-year-old girl was admitted with swelling and limited motion of knee, ankle and metacarpophalangeal joints at a local hospital and was given ibuprofen and oral corticosteroids, 1 mg/kg/day with a diagnosis of oligoarticular juvenile idiopathic arthritis. However, she was out of follow-up and continued prednisone treatment with a dose of 5 mg/day. One year later, she was applied with diffuse vesiculobullous lesions 20 days after cephaclor treatment due to acute bronchitis. She appeared generally ill, wasted and exhausted. Physical examination revealed tenderness, warmness and swelling of knee, elbow and wrist joints. There were numerous bullae and vesicles on a background of urticated plaques affecting the back, abdomen, neck, arm and groin region (Fig. 1). Haemoglobin was 10.3 g/dl, white blood cell 2700/mm^3, platelets 940.000/mm^3, erythrocyte sedimentation rate 120 mm/h, C-reactive protein 17.7 mg/dl. Serology for hepatitis B and C virus were negative. Complement C3 and C4 levels were normal (87.8 and 16.2 mg/dl). She also had autoimmune thyroiditis with elevated thyroid stimulating hormone (291 μIU/ml), low free thyroxine (0.75 ng/dl) and elevated human antithyroglobulin (880 IU/ml) levels. IgA dermatosis was diagnosed with a skin biopsy showing linear IgA deposition with subepidermal blisters. Pulse methylprednisolone (30 mg/kg/d) and subsequently oral prednisone 2 mg/kg/d were administered. Dapsone (1 mg/kg/d) and colchicine (1 mg/day) were also added. As it was resistant, methotrexate (10 mg/m^2/week) was given one month after admission, after when the lesions begin to regress. Nine months after admission, she was referred with widespread blistering from head to foot with oral ulcers. Laboratory investigations revealed low C3 (66.7 mg/dl) and normal C4 (15 mg/dl) levels, positive anti dsDNA antibody, positive antihistone antibody, haemolytic anemia (Hb: 9 g/dl) and thrombocytosis (1.352.000/mm^3). Anticardiolipin antibody and lupus anticoagulant were negative. A therapy with prednisone, 2 mg/kg/d, and cyclophosphamide, 2 mg/kg/d, p.o. was administered. All lesions were resolved in four weeks. After dischargement from hospital, she was lost to follow-up.

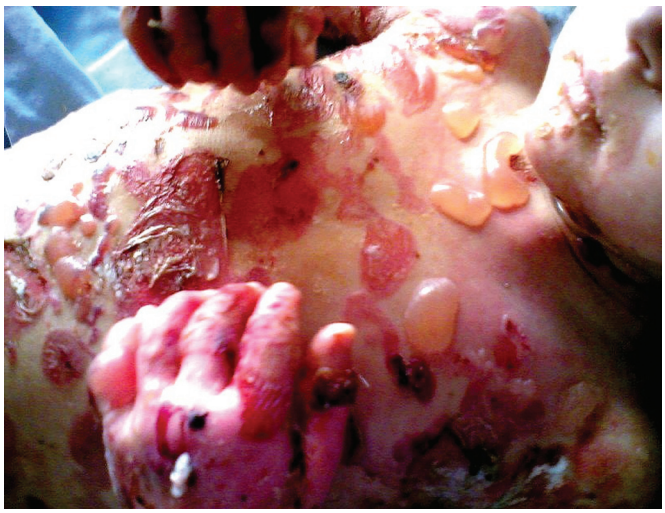

Fig. 1. Large and tense bullae in a 7-year-old girl who developed linear IgA bullous dermatosis and systematic lupus erythematosus.

As far as we know, the association of LAD and SLE was reported in only one child in the literature (2). Tobon *et al*. presented a 9-year-old girl with LAD who developed features of SLE similar to our patient (2). However, the use of antibiotic treatment before the onset of symptoms and positive anti-histone antibodies, both of which are compatible with drug-induced SLE, and also the severe progression with widespread blisters and unresponsiveness to conventional treatment in our patient are features distinct from that described by Tobon *et al*. LAD may be idiopathic or associated with infections (6), malignancy (7), SLE (2), rheumatoid arthritis (8) and certain drugs such as vancomycin, phenytoin, acetaminophen and captopril (9). In children, the onset is usually acute; papules and urticarial plaques with blisters are localised mainly on the face, perineum, trunk, limbs and sometimes on mucous membranes. In idiopathic cases, the treatment should be started with dapsone (10). In resistant cases whose treatment has failed, systemic corticosteroids should be used.

The present case had features of SLE such as anemia, leukopenia, arthritis and oral ulcers in addition to positive anti-dsDNA antibodies and complement consumption. Bullous SLE may rarely be the initial clinical manifestation of SLE (3). Corticosteroids are commonly used for the treatment of childhood-onset SLE; however, there appears to be important between-centre differences in the use of intravenous and oral corticosteroids for SLE therapy (4). Deaths of patients with SLE result from organ involvement, thromboses, infections and therapy-related events (5).

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