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Stevens-Johnson Syndrome-Like Skin Lesions in a Patient with Juvenile Systemic Lupus Erythematosus

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Dear Editor:

Skin manifestations in systemic lupus erythematosus (SLE) are very heterogeneous. Recently, "Stevens-Johnson syndrome (SJS)/toxic epidermal necrolysis (TEN)-like lupus erythematosus (LE)" has been reported. Clinically, this condition presents with photodistributed exanthema and minimal mucositis. The onset of the rash is insidious with a prolonged clinical course[1].

A 16-year-old Korean woman presented with a 4-week history of gradually progressive development of painful, annular, erythematous, flattened, and blistering eruptions. Crusting presented on the face, V-neck area, upper back, and dorsum of the hands, and involved less than 10% of body surface area. The lips had erosions and crusts (Fig. 1). Eye and genital examinations were unremarkable. Previous medical history included malar rash and diffuse alopecia. No relevant medications had been newly administered.

Skin biopsy from the back revealed a subepidermal blister with exocytosis and superficial perivascular infiltration of lymphocytes and neutrophils. Marked necrosis of keratinocytes of the suprabasal layer and interface dermatitis were manifested (Fig. 2). Direct immunofluorescence microscopy was remarkable in that linear deposits of immunoglobulin G, C3, and C1q were shown at the basement membrane zone.

Laboratory investigations demonstrated anemia (hemoglobin 10.9 g/dl), leukopenia (3,600/mm^3), and a normal platelet count. The antinuclear antibody titer was positive (1:640 homogeneous pattern), anti-double-stranded DNA was positive, anti-Ro/SS-A was positive and anti-La/SS-B was negative. Anti-Sm was positive, anti-nucleosome, anti-histones, and rheumatoid factor were negative. C3 and C4 levels were markedly low at 47.2 g/dl and 13.6 g/dl, respectively. Plasma creatinine was 0.65 mg/dl and the 24-hour urine proteinuria was 1,589 mg/day. Lupus nephritis class IIA was diagnosed from kidney biopsy. Liver

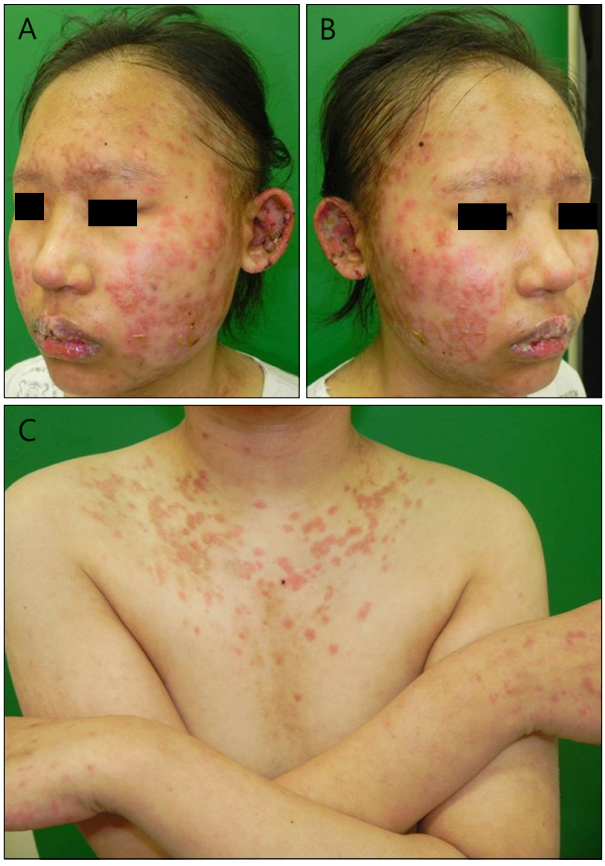

Fig. 1. Photodistributed erythematous flattened vesiculobullous eruptions with crusts on the face, V-neck area and the dorsum of hands and erosion on the lips.

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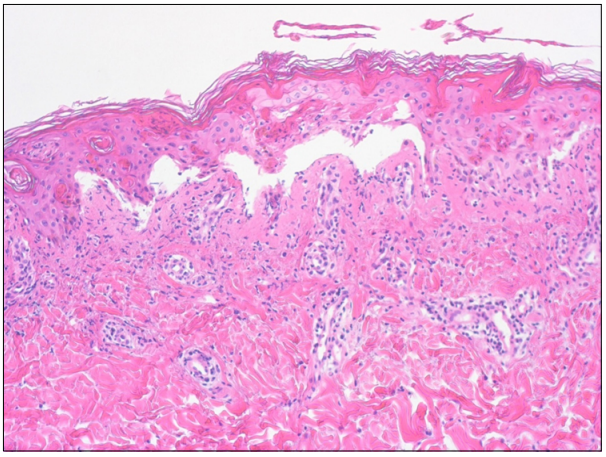

Fig. 2. Subepidermal blister with exocytosis and superficial perivascular infiltration of lymphocytes and neutrophils, marked necrosis of keratinocytes of the suprabasal layer and interface change (H&E, ×100).

function tests were unremarkable. Eventually, SJS-like cutaneous presentation with juvenile SLE was diagnosed based on the above findings.

Treatment regimen included oral hydroxychloroquine 200 mg, deflazacort 48 mg per day with gradual tapering. Lesions improved after dressing on the denuded area and applying topical corticosteroid.

Skin is the second most commonly affected organ system in SLE[2]. A bullous eruption in the setting of SLE can be a diagnostic challenge. Classic SJS/TEN, SJS/TEN-like LE, drug-induced cutaneous LE, bullous LE, Rowell's syndrome, and immunobullous diseases need to be considered in the differential diagnosis[3]. Indirect immunofluorescence is an effective method to diagnose bullous LE, but the present patient did not undergo this test. However, bullous LE appears with various-sized tense blisters, which is clinically different from the patient's presentation. Usually, SJS/TEN-like LE develops as a result of severe inflammatory epidermal basal layer damage. Vacuolar degeneration leads to subepidermal blistering and almost full-thickness epidermal necrosis[1,4].

SJS/TEN-like LE is often triggered by excessive ultraviolet exposure with an underlying predisposition to SLE. These patients often have anti-Ro antibody. Anti-Ro may be associated with increased risk of photosensitive rash[4].

The standard treatment of SJS/TEN-like LE remains controversial. Corticosteroids were prescribed in the present case because of continuing lupus activity. Chloroquine was combined for immunomodulation. In previous reports of TEN-like acute cutaneous LE, intravenous immunoglobulin, and plasmapheresis were successful[5].

To our knowledge, SJS-like skin lesions with juvenile SLE have rarely been reported in the literature. This was the first case reported in Korea.

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