

## CASE REPORT

# Scaly Ear Rash as the Herald of a Young Girl with Juvenile Systemic Lupus Erythematosus

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Juvenile systemic lupus erythematosus (JSLE) is an autoimmune-mediated multiorgan disease. The cutaneous manifestation is one of the most common initial presentations in JSLE. A typical lesion is a facial malar rash, but a patient may sometimes present with nonclassical lesions. Herein, we report two cases of JSLE with similar persistent scaly ear rashes as the heralding cutaneous symptom preceding systemic symptoms. Identifying this atypical and underestimated cutaneous rash in juvenile patients might help the clinician make the correct diagnosis and provide earlier intervention, which may help prevent disease progression. (*Ann Dermatol* 23(S3) S333~S337, 2011)

## -Keywords-

Juvenile systemic lupus erythematosus, Ear, Rash

## INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune condition involving inflammation of multiple organs. In contrast to adult SLE, the prevalence rate of juvenile SLE is very low, less than 1/100,000<sup>1</sup>, although black children have a higher incidence<sup>2</sup>. Juvenile SLE (JSLE) is clinically similar to adult SLE, but there are some differences bet-

ween JSLE and adult SLE. We, herein, report two rare cases of JSLE involving a similar scaly ear rash followed by a facial rash and constitutional symptoms.

## CASE REPORT

The first patient was a 12-year-old girl who presented with an asymptomatic skin rash on her left ear that had persisted for more than 4 weeks. About 2 weeks before presenting to our clinic, she experienced constitutional symptoms such as dyspnea, sore throat, and a low-grade fever of 38°C. A skin rash was noted on her face, chest, upper neck, and dorsal aspect of the forearms. The patient was initially diagnosed with an acute viral infection at a local clinic. However, the symptoms worsened and more skin lesions appeared on her body. Because of the persistent symptoms, the patient was transferred to our center for evaluation. A review of the patient's medical history showed no significant findings, except that her mother had a history of idiopathic hemolytic anemia.

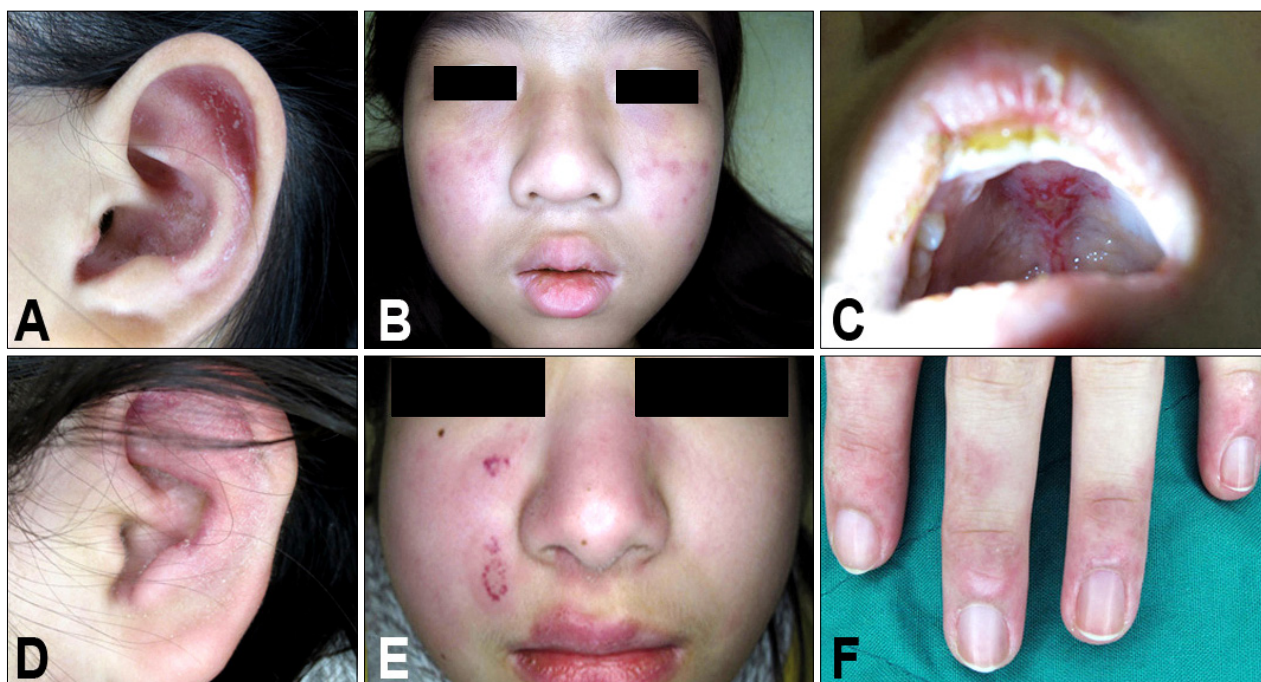
On dermatological examination, a well-demarcated scaly erythematous plaque was found on the patient's left ear (Fig. 1A). Numerous erythematous papules with scales were noted on the anterior neck, chest, and both forearms. The characteristic confluent symmetric erythema and edema were centered over the malar eminence and bridge of the nose, but the nasolabial folds were spared (Fig. 1B). Other extracutaneous lesions included an oral ulcer on the hard palate (Fig. 1C) and erythema with telangiectasia around the proximal nail fold.

Initial laboratory testing showed the following values: hemoglobin, 8.1 g/dl (reference range, 12~16 g/dl); white blood cell (WBC) count, 3,400/mm<sup>3</sup> (4,500~1,0000/mm<sup>3</sup>); aspartate transaminase, 81 U/L (<32 U/L), alanine transaminase, 52 U/L (<31 U/L); total bilirubin, 1.5 mg/dl (<1.2 mg/dl); indirect bilirubin, 0.5 mg/dl (<0.2 mg/dl); and erythrocyte sedimentation rate, 99 mm/h (<20

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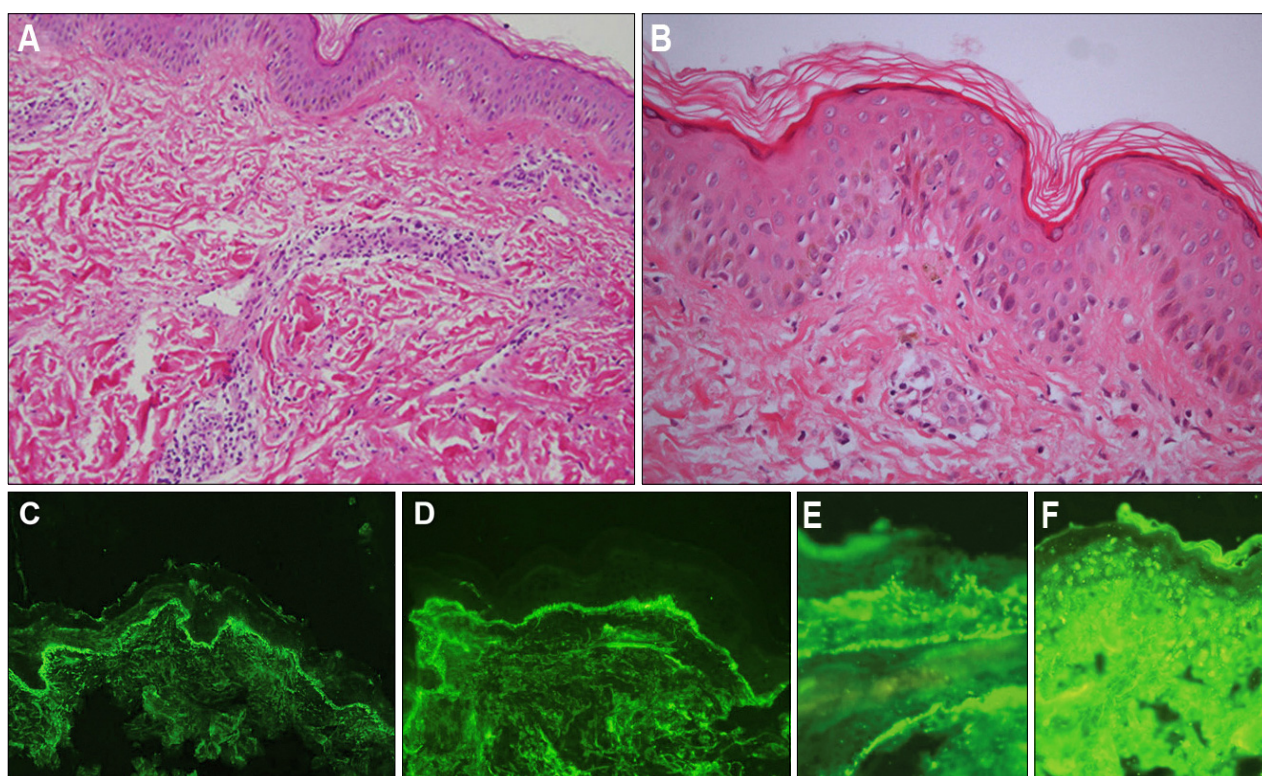
**Fig. 1.** Patient 1. (A) Scaly erythematous plaque on the left ear. (B) Typical malar rash. (C) Painful ulcer on the hard palate. Patient 2. (D) Edematous, erythematous plaque with scales on the left ear. (E) Facial malar rash, discoid rash with petechiae. (F) Periungual erythema over the fingers.

mm/h). Forty-eight hours after admission, a follow-up blood test showed severe leukopenia (WBC count,  $780/\text{mm}^3$ ) and anemia (hemoglobin, 7.8 g/dl) with reticulocytosis.

Because more skin lesions appeared on the patient's upper limbs, a skin biopsy was performed to exclude viral exanthema or other forms of inflammatory dermatosis. Five days following admission, a rheumatology test was positive for anti-nuclear antibody (ANA) with a speckled pattern (1:320), anti-dsDNA antibody (67.8 IU/ml; reference range,  $<10$  IU/ml), and anticardiolipin immunoglobulin M (IgM) ( $>100$  U/ml; reference range,  $<15$  U/ml). The complement 3 (C3) concentration was 22.5 mg/dl (reference range, 90~180 mg/dl), and the C4 concentration was 6.2 mg/dl (10~40 mg/dl). Histopathology of the skin showed typical lupus dermatitis with characteristic thickening of the basement membrane and subtle interface changes, perivascular and periadnexal lymphocytic infiltration, and a positive lupus band (Fig. 2A~D). Based on the clinical, histopathological, and immunopathological findings, the final diagnosis was JSLE. During hospitalization, the patient's hemoglobin level declined progressively, but transfusion of packed red blood cells was prohibited because of positive results on direct and indirect Coombs tests. Based on the final diagnosis and deteriorating clinical course, induction im-

munosuppressive therapy with intravenous pulsed-steroid therapy (500 mg methylprednisolone/day) and oral medication (50 mg azathioprine/day) were prescribed. The patient became afebrile 3 days after treatment. Leukocyte and hemoglobin levels increased, and liver enzyme and bilirubin levels returned to normal. The cutaneous lesions resolved after 1 week. The medications were shifted to maintenance regimens with oral corticosteroid (30 mg/day), azathioprine (50 mg/day), and mycophenolate mofetil (250 mg/day). The disease was controlled, the patient's condition improved, and she was discharged from the hospital. At follow-up 10 months after discharge, the disease was well controlled without cutaneous or systemic sequelae.

The second case was a 14-year-old girl who presented with a 1-month history of asymptomatic skin lesions on both ears. Three weeks later, she developed facial erythema with edema and constitutional symptoms including fever, myalgia, and arthralgia. A physical examination at admission showed scaly erythematous plaques with edema on both ears (Fig. 1D). A facial malar rash and round well-demarcated plaques with petechiae on both cheeks were seen (Fig. 1E). She also had several erythematous papules on the back and cyanotic changes with periungual telangiectasia on all fingers and toes (Fig. 1F). Mucosal ulceration was observed on the hard palate.



**Fig. 2.** Histological sections of skin biopsies, patient 1 (A) thickened basement membrane zone with dermal perivascular lymphocytic infiltrate (B) subtle vacuolar degeneration and melanin incontinence (H&E, original magnification, A:  $\times 20$ , B:  $\times 40$ ), immunofluorescence staining revealing positive for lupus band (C) IgG and (D) IgM (original magnification, C:  $\times 20$ , D:  $\times 40$ ). Patient 2 (E) positive lupus band of IgM and (F) nuclear staining of IgG (original magnification, E:  $\times 20$ , F:  $\times 20$ ).

The laboratory examination showed pancytopenia: WBC count,  $560/\text{mm}^3$ ; hemoglobin, 8.1 g/dl; and platelet count,  $99 \times 10^9/\text{L}$  (reference range,  $150 \sim 400 \times 10^9/\text{L}$ ). ANA with a speckled pattern ( $>1:1,280$ ) and a low C3 concentration (44 mg/dl) were noted. Biochemical values were within the normal limits. A histopathological examination of the biopsy specimen taken from a papule on the back showed similar findings as those of patient 1. An immunofluorescence study showed granular deposition of IgG, IgM, IgA, and C3 at the epidermal-dermal junction as well as nuclear staining of IgG (Fig. 2E, F). The patient was diagnosed with JSLE, and she was treated with intravenous pulsed-steroid therapy (500 mg methylprednisolone/day). The fever subsided 2 days after treatment, and the skin lesions disappeared.

## DISCUSSION

Hematological and cutaneous signs are the most common initial manifestation of JSLE<sup>3</sup>. Compared with adult SLE, the initial presentation of JSLE is sometimes nonclassical and can be more severe<sup>3</sup>. Diagnosis and treatment may be delayed in this age group. In 1981, Gilliam and Sonth-

**Table 1.** Association between cutaneous lupus erythematosus and systemic disease activity<sup>5</sup>

Disease	Systemic disease association
ACLE	++++
LE-nonspecific disease	++++
SCLE	+++
Generalized DLE	++
Localized DLE	+
Hypertrophic LE	+
LE panniculitis	+
Tumid LE	+

ACLE: acute cutaneous lupus erythematosus, LE: lupus erythematosus, SCLE: subacute cutaneous lupus erythematosus, DLE: discoid lupus erythematosus. Correlation between cutaneous lesion and systemic disease activity: + (rarely correlate), ++ (sometimes correlate), +++ (usually correlate), ++++ (always correlate).

eimer proposed that the cutaneous signs of lupus erythematosus (LE) be divided into LE-specific and LE-nonspecific lesions<sup>4</sup>. Acute cutaneous LE, subacute cutaneous LE, and chronic cutaneous LE are three specific lesions seen in SLE. In contrast, nonclassical presentations such as vasculitis, Raynaud's phenomenon and periungual telan-

**Table 2.** Differences between juvenile SLE and adult SLE<sup>1</sup>

Clinical/laboratory manifestations	Juvenile SLE	Adult SLE
Sicca symptoms	+	++
Fever	++	+
Arthralgia	+	++
Anemia (hemolytic)	++	+
Chilblains LE	++	+
SCLE	++	+
DLE	+	++
Kidney involvement	++	+
Anti-dsDNA	++	+
Anti-ribosomal P	++	+
Antihistone	++	+

SLE: systemic lupus erythematosus, LE: lupus erythematosus, SCLE: subacute cutaneous lupus erythematosus, DLE: discoid lupus erythematosus. One plus sign (+) indicates that a characteristic occurs frequently, 2 plus signs (++) indicate that a characteristic occurs more frequently.

gicctasia are defined as LE-nonspecific lesions. Although not unique to LE, the nonspecific lesions are as important as the butterfly rash, whose appearance correlates strongly with disease activity (Table 1)<sup>5</sup>. All of these lesions may occur in patients with SLE or in patients who do not fit the criteria for SLE<sup>6</sup>. These different types of lesions may occur simultaneously in a patient with SLE<sup>6</sup>.

Our patients might have ignored the erythematous and scaly changes on the ear because they may have considered them to be eczema or physiological flushing. However, we suspected that the persistent scaly erythema might reflect disease activity, because it was caused by inflammatory hyperemia<sup>7</sup>. The scaly ear rashes were not sampled for histological examination, and we could not identify whether the rashes were LE specific or LE non-specific. However, the ear rashes may provide an important dermatological clue for diagnosing JSLE early, regardless of which specialized group of cutaneous LE the ear lesion falls into. Hence, if a juvenile patient presents with a scaly ear rash followed by fever, leukopenia and anemia, we suggest that JSLE should be considered promptly.

The clinical and laboratory presentations of JSLE are similar to those of adult SLE<sup>1</sup>, although the clinical manifestations and prevalence of autoantibodies vary between the two forms. Clinically, fever and organ involvement are significantly more frequent in JSLE, whereas arthralgia and sicca symptoms are more common in adult SLE<sup>1</sup>. The most commonly affected organs are the brain and kidneys, particularly in black children<sup>8</sup>. A laboratory examination will show that hemolytic anemia and antiribosomal P, anti-dsDNA, and antihistone antibodies are found more

often in JSLE (Table 2)<sup>1</sup>.

In contrast to adult SLE, children usually have a more severe clinical course, and large studies have revealed higher mortalities<sup>9</sup>. In an international multicenter cohort study, the mortality rate was almost eight times greater for patients with SLE who were younger than 24 years of age compared with older patients<sup>9</sup>. The complications of JSLE caused by infection or organ involvement including lupus nephritis determine the prognosis of JSLE. The therapeutic plan should focus on reducing morbidity and mortality rates. The overall treatment in juvenile patients is similar to that in adult patients and depends on the clinical manifestations and the major organs involved. There are two parts to modern therapeutic strategies of JSLE<sup>10</sup>. The first part is induction therapy to control disease activity; potential organ-threatening or life-threatening disease must be managed aggressively. After remission, the second part is maintenance therapy to avoid relapse and to control the disease, and most patients need to be treated for several years<sup>10</sup>. Immunosuppressants are the drugs of choice in both parts of treatment, although efficacy and side effects must be balanced.

We believe that scaly ear rash seems be overlooked in the literature, because it can be an underestimated and easily ignored skin lesion in children with SLE. This lesion could be an initial sign preceding other systemic symptoms. Hence, when patients present with this type of skin lesion, they should be followed up regularly to provide early diagnosis and management.

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