

# Exuberant skin involvement in Systemic Lupus Erythematosus: a clinical case report

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## INTRODUCTION

Lupus erythematosus (LE) is a chronic inflammatory disease with a broad spectrum of cutaneous and systemic manifestations. Skin involvement includes a variety of LE specific lesions subdivided into 3 major categories – chronic cutaneous LE (CCLE), subacute cutaneous LE (SCLE) and acute cutaneous LE (ACLE) – based on clinical and histopathologic examination<sup>1</sup>.

Patients presenting with SCLE constitute 7%–27% of LE cases. These type of lesions are mostly seen in females (ratio 1:4) of caucasian race (>85%) and are clinically characterized by nonscarring erythematous papulosquamous and/or annular skin lesions with a symmetrical distribution typically located in sun-exposed areas<sup>2</sup>. Approximately 50% of patients with SCLE fulfil the American College of Rheumatology classification

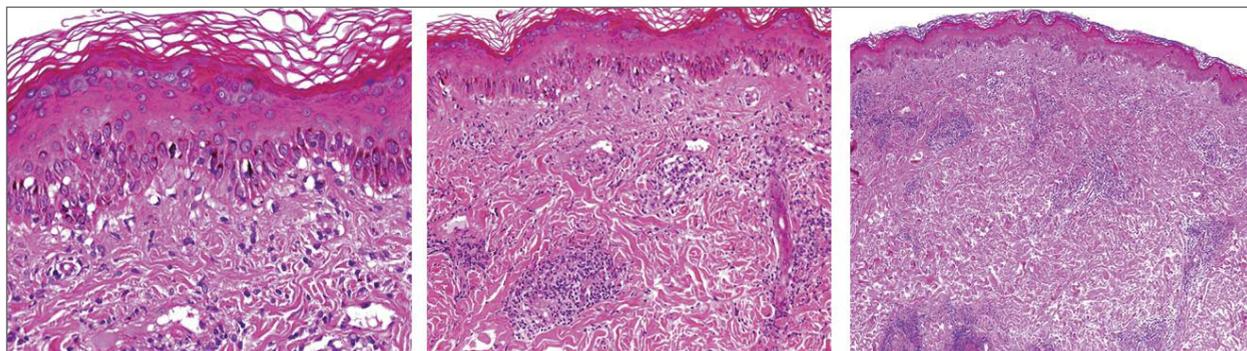
criteria for Systemic Lupus Erythematosus (SLE), but only 10–15% will develop severe clinical manifestations of SLE<sup>3</sup>.

Neonatal LE, with or without congenital heart-block, is also a rare condition seen in 1% of Lupus women who have anti-Ro and/or La antibodies, regardless of their symptoms<sup>4</sup>.

## CASE REPORT

This case refers to a 27 year-old black woman, smoker of 5 cigarettes per day, with a background history of SCLE, diagnosed after a skin biopsy at the age of 20 (Figure 1), and three uncomplicated pregnancies. Seven weeks after her third pregnancy she presented to our clinic with a 4-month history of progressive and widespread erythematosus lesions in a photodistribution, mouth ulcers, alopecia, persistent migraines and depression. Her newborn son had a hyperpigmented skin macule, in remission, on his right cheek (Figure 2). Her two other children also had the same type of lesions when they were born. Fetal echocardiograms of all pregnancies were normal. Examination of the patient revealed scaly erythematosus macules and annu-

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**FIGURE 1.** Apoptotic keratinocytes in the upper layers of the epidermis. In the superficial and deep dermis, there is a perivascular and periadnexal mild to slightly moderate mononuclear inflammatory cell infiltrate



**FIGURE 2.** Patient's baby with cutaneous neonatal lupus syndrome: a large hyperpigmented macule seen in the right cheek

lar dermal plaques with rims of erythema, most prominent on the extremities, but also involving her face, scalp and neck (Figure 3). Her ankles and wrists were tender and swollen. Cardiovascular and respiratory examinations were unremarkable. Blood tests showed ANA of 1/1000 (fine speckled), anti-Ro and anti-La positive, dsDNA of 8 IU/mL (0-40), normal complements and negative antiphospholipid antibodies. Her full blood count revealed normocytic/normochromic anemia (Hb 9.2g/dL), persistent lymphopenia (minimum value of  $0.63 \times 10^9/L$ ) and no thrombocytopenia. ESR was 63 mm and CRP 0.3 mg/dL.

Given the above clinical, laboratory, and previous histopathologic findings, a diagnosis of SLE with SCLE type lesions was made. The patient was started on prednisolone 0.5mg/kg/day, hydroxychloroquine 400mg/day, clobetasol propionate 0.05% cream and tacrolimus ointment, with moderate improvement of skin lesions at the 1-month follow-up.

## DISCUSSION/CONCLUSION

Severe SCLE associated with an active SLE in black people is an infrequent event. It is known that Lupus disease activity may flare during pregnancy or puer-



**FIGURE 3.** Exuberant skin lesions in the patient's lower and upper limbs and face: Hyperpigmented and scaly erythematous annular plaques

perium and more often in African-Americans and current smokers<sup>5</sup>. These facts can substantiate the spectrum of manifestations seen in this patient.

The authors draw attention for the severity and exuberance of the skin involvement in a SLE patient, which is an uncommon presentation in the rheumatology outpatient setting. In addition, they highlight the occurrence of neonatal LE, a very rare condition present all this patient's newborns.

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