Lupus erythematosus tumidus: A case and discussion of a rare entity in black patients

Stephanie M. Gallitano, MD,[a] and Alessandra Haskin, BA[b]
Brooklyn, New York and Washington, DC

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INTRODUCTION

Lupus erythematosus tumidus (LET) is a nonscarring form of cutaneous lupus erythematosus that typically presents in middle-age men as urticarial-like lesions on sun-exposed areas.[1] There is a paucity of information on black patients with LET. To our knowledge, this case marks the fourth published report of LET in a black patient.[2-4] This report also reviews the previously published cases of LET in black patients to promote awareness on how this entity may present differently in darker pigmented patients.

Abbreviations used:

LET: lupus erythematosus tumidus
SLE: systemic lupus erythematosus

CASE REPORT

A 54-year-old black man presented for evaluation of hundreds of asymptomatic soft nodules on his trunk that were present for 3 to 4 years. They were slowly increasing in number and the patient denied seasonal variation of the lesions. Review of systems was negative, and medical history was unremarkable except for active tobacco smoking. Physical examination revealed hundreds of small, flesh-colored to hypopigmented soft nodules on his back, chest, and lateral arms (Fig 1). The scalp and face were unaffected. A biopsy specimen obtained from his chest showed a superficial and deep perivascular and periadnexal lymphocytic infiltrate, with increased dermal mucin. There was no alteration of the dermoepidermal junction (Fig 2, *A* and *B*). The patient had a positive antinuclear antibody with a titer of 1:320. Findings from all other laboratory workups, including complete blood count, comprehensive metabolic panel, and urinalysis, were unremarkable, and a diagnosis of LET was made. The patient was started on hydroxychloroquine 200 mg twice a day and intralesional kenalog injections at 5 mg per mL.

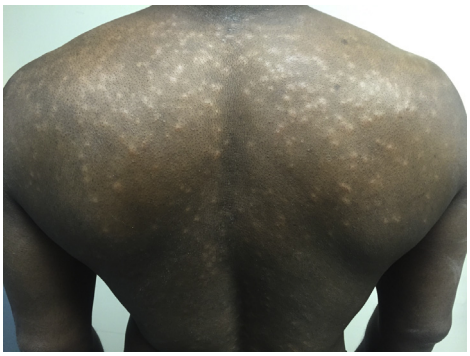

Fig 1. Lupus erythematosus tumidus: hundreds of hypopigmented soft nodules on the back of this 54-year-old black man.

DISCUSSION

Gougerot and Burnier first described LET in 1930 in 2 patients with erythematous, infiltrated, nonscarring plaques.[5] The next 70 years consisted of infrequent case reports in the European literature, until 2000 when Kuhn et al[1] reported the largest case series of LET in 40 patients. Since then, several case series have addressed the epidemiology, treatment, and pathogenesis of this disease; however, there are few reports of its manifestations and outcomes in black patients.

Discussion of these cases is important, as the appearance and distribution of LET may present

From the Department of Dermatology,[a] SUNY Downstate Medical Center, Brooklyn and Howard University College of Medicine, Washington, DC.[b]
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Correspondence to: Stephanie M. Gallitano, MD, SUNY Downstate Medical Center, Department of Dermatology, 450 Clarkson Avenue, Box 46, Brooklyn, NY 11203. E-mail: s.m.gallitano@gmail.com.

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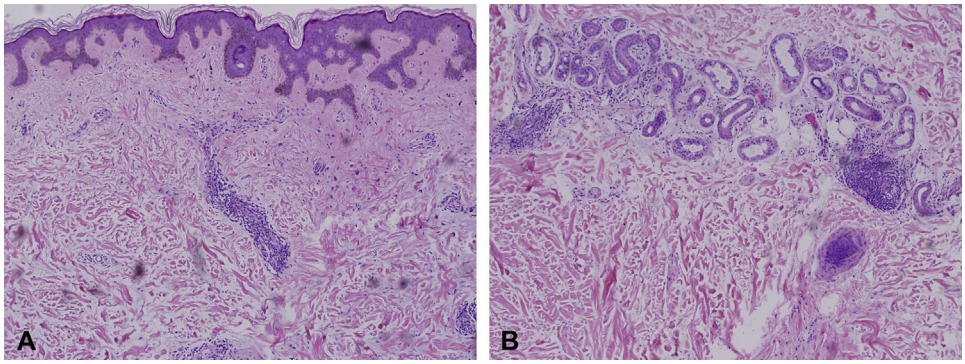

Fig 2. A, Perivascular and periadnexal lymphocytic infiltrate with increased dermal mucin. There is no interface change present. **B**, Lymphocytic infiltrate in the eccrine coil. (**A** and **B**, Hematoxylin-eosin stain; original magnifications: **A**, ×40; **B**, ×100.)

Table I. Clinical and histologic characteristics of lupus erythematosus tumidus reported in black patients

Clinical characteristics	Current case	Dekle et al[3]	Jolly et al[4]	Perez et al[2]
Age at presentation	54	25	38	42
Duration of lesions	3-4 y	7 mo	Unclear	3 y
Location of lesions	Back, chest, and lateral arms	Arms and lower back	Arms and upper back	Within a scar on the upper back
Description of lesions	Hypopigmented, non-scaly, soft nodules	Hypopigmented macules and plaques with mild scale	Fleshy, subcutaneous nodules with adherent scale in some areas	Erythematous plaques
Interface dermatitis on pathology	No	No	Minimal	No
Concomitant diagnosis of SLE	No	No	Yes; before diagnosis of LET	No

differently in skin of color. Table I outlines some of the characteristics and histopathologic findings in the few reported cases of LET in black patients.[2-4] Most of these patients exhibited lesions that appeared hypopigmented or flesh colored, as opposed to the erythematous plaques seen in patients with lighter skin tones. Additionally, all of these patients lacked lesions in common sun-exposed areas such as the head and neck, which predominate in patients with lighter skin. It is possible that these differences in distribution and secondary characteristics may be unique to black patients with LET.[3]

Clinically, these lesions may resemble papulonodular mucinosis, which is seen in the setting of active systemic lupus erythematosus (SLE). In those patients, serum autoantibody titers parallel the amount of papulonodular mucinosis. Papulonodular mucinosis is identified on biopsy by extensive dermal mucin but lack of an inflammatory infiltrate.[6] Many other inflammatory disorders appear hypopigmented in patients with skin of color, and a broad differential should be considered, including sarcoidosis, mycosis fungoides, and granuloma annulare. Skin biopsies are useful diagnostic tools in these circumstances.

Although there are no specific histologic criteria, most investigators agree that LET shows perivascular and periadnexal lymphocytic infiltrate with interstitial mucin.[1] However, more controversy exists on the role of dermal-epidermal changes. Some investigators consider interface changes to be acceptable within the spectrum of LET, and others consider them to be criteria for exclusion.[7-9] Two recent retrospective studies explored this question and found that mild epidermal changes were not associated with a worse prognosis or increased risk of progression to SLE.[8,9] These findings suggest that looser criteria be used with regard to dermal-epidermal changes in LET.

The exact cause of LET is likely multifactorial with ultraviolet exposure, genetics, and environmental exposures all playing a role in disease pathogenesis. The role of smoking has been explored in several case series. Up to 78% to 80% of patients with LET

are smokers, which is significantly higher than age-matched controls, in which approximately 25% to 36% of the population are smokers.[10,11] Smokers with cutaneous lupus erythematosus do not respond as well to antimalarial therapy as do nonsmokers.[10] Although no studies have looked at the effect of smoking cessation on LET clearance, Kreuter et al[10] noted that 13 patients who quit or reduced smoking within the period of their study had complete clearance of their skin lesions. Our patient was a smoker; however, the smoking status of the other reviewed cases was not reported. Smoking status should be obtained in patients with LET, and those with a positive history should be counseled on smoking cessation.

The course of LET is typically benign. Most case series found that progression to SLE does not occur in patients with LET, although a few reports indicate concurrence of disease in up to 11% of patients.[1,7-10,12] Patients typically respond well to topical corticosteroids, systemic antimalarial drugs, and photoprotection.

The current case highlights the importance of recognizing that LET can present differently, in terms of appearance and location, in skin of color. Patients may present with hypopigmented papules on covered areas. Given the close association between smoking and disease activity, patients should be screened for their smoking history, and cessation should be encouraged. The few published cases of LET in black patients may be because of underreporting.

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