A Unique Presentation of Lupus Erythematosus Tumidus in an Adolescent Boy

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PRACTICE **POINTS**

- Lupus erythematosus tumidus rarely occurs in the pediatric population.
- Lupus erythematosus tumidus is a unique subset of lupus associated with lack of interface change on histology and large amounts of mucin.
- Lesions typically present on the face and trunk but can very rarely present on the extremities and hands.

To the Editor:

Lupus erythematosus tumidus (LET) is a rarely diagnosed condition that was first described in 1909 by Hoffmann.[1] Limited cases have been reported in the literature, with few documenting the disease in children.[2] We report a unique clinical case of LET in a 14-year-old adolescent boy that was distributed solely on the hands. With slight heterogeneity in regards to clinical presentation and histopathology, there is a need for further exploration with regard to LET.

A 14-year-old adolescent boy presented to the dermatology clinic with progressive bilateral edema of 1 year's duration with plaques and some scaling on the dorsal aspects of the digits and the nail bases predominantly on the right hand (Figure 1) and to a lesser extent on the left hand. The edema, erythema, and tenderness started in the right fifth digit; soon after the edema appeared, plaques began to form at the base of each nail bed, and the edema and erythema progressively spread to the other digits. He denied worsening of symptoms when exposed to cold temperatures. A complete review of systems was negative. The differential diagnoses included chilblain lupus erythematosus, perniosis, dermatomyositis, and polymorphous light eruption. A punch biopsy from the right fourth digit was performed.

The biopsy showed superficial and deep perivascular and periadnexal mononuclear inflammation with large amounts of interstitial mucin deposition (Figure 2). The epidermis exhibited a loose orthokeratotic scale with no signs of interface damage. A diagnosis of perniosis was entertained but was ruled out due to the lack of papillary dermal edema and large amounts of mucin. With the lack of interface change and large amounts of mucin, a diagnosis of LET was favored over chilblain lupus erythematosus, as the latter diagnosis typically demonstrates interface change. The patient was started on hydroxychloroquine 200 mg twice daily and a short course of

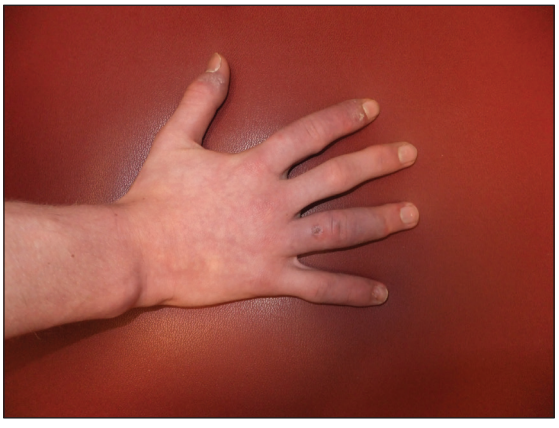

FIGURE 1. Lupus erythematosus tumidus. Digital edema and erythema on the right hand. Plaques/scaling were noted on the dorsal aspects of the first, second, and fourth digits.

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The authors report no conflict of interest.

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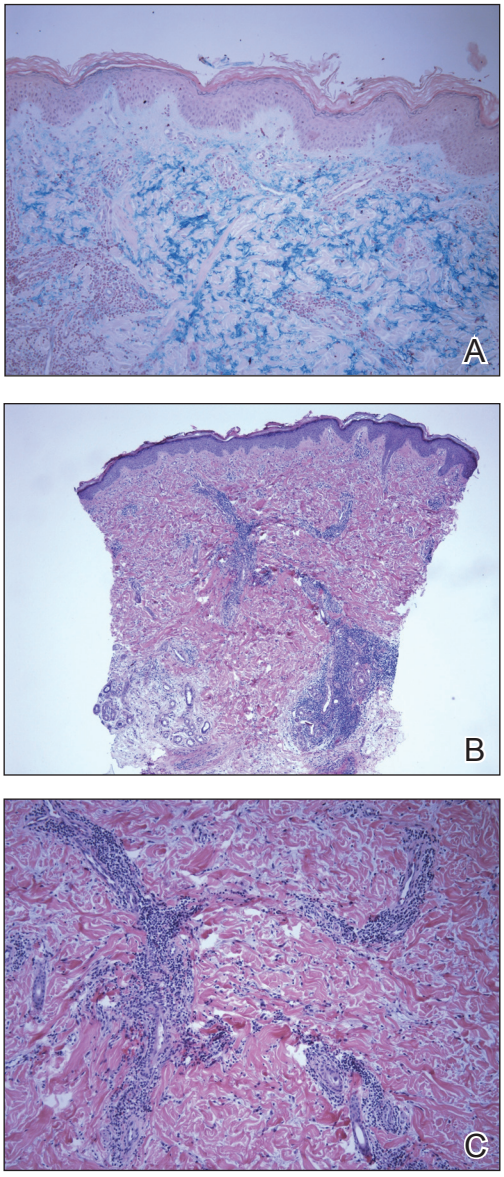

FIGURE 2. Lupus erythematosus tumidus. A, Colloidal iron staining of a biopsy from the right fourth digit showed interstitial mucin deposition (original magnification ×10). B, Superficial and deep perivascular and periadnexal mononuclear inflammation with interstitial mucin deposition and no interface change (H&E, original magnification ×4). C, Perivascular and periadnexal lymphocytic infiltrate (H&E, original magnification ×20).

prednisone, and improvement of the lesions/plaques was noted at follow-up 6 weeks later. Continued improvement was noted 2 years after the initial presentation. His condition recurred when the hydroxychloroquine dosage was reduced to 200 mg once daily after 1 year. The patient did not report any adverse sequelae to treatment.

Histopathologic findings of superficial and deep perivascular and periadnexal lymphocytic infiltrates and interstitial dermal deposition of mucin in LET have remained consistent in the literature. Direct immunofluorescence has not revealed any complement or immunoglobulin deposition on the basement membrane.[3,4] The epidermal characteristics are not as uniform, with the majority of cases in one review showing no epidermal changes and a minority showing minimal epidermal changes (eg, epidermal atrophy, hyperkeratosis, parakeratosis, acanthosis, spongiosis).[5] When working up patients for LET, blood work usually is unremarkable, as LET rarely is associated with antinuclear antibodies or anti-Ro, anti-La, and anti-DNA antibodies.[3,4] Lupus erythematosus tumidus generally is an independent process, but it has been reported to coexist with discoid lupus erythematosus and systemic lupus erythematosus in rare cases.[6]

The lesions of LET have been consistently described in the literature as photosensitive, erythematous, nonscarring, annular plaques and papules commonly occurring on the head/neck and other sun-exposed areas that do not cause hypopigmentation.[3] Treatment of LET consists of systemic treatment with antimalarial drugs, sunscreens, and topical steroids for flares.

Lupus erythematosus tumidus is rare in children, with few case reports noted in the literature. Sonntag et al[2] documented the disease in 3 children ranging from 3 to 8 years of age. Furthermore, Ruiz and Sanchez[7] reported a case of LET in a 16-year-old adolescent girl. Our case is unique in that the lesions only occurred on the hands, whereas most case reports document distribution of the lesions on the head, neck, face, arms, back, and chest. Our patient's age and the location of the lesions make it a unique clinical presentation of LET.

Reports in the literature show evidence of heterogeneity in the presentation, classification, and some of the histopathologic features of LET; however, there are minimal data on childhood LET. Further research and investigations are needed to more precisely define this condition.

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