

Lupus Erythematosus Tumidus Gets Nailed Down

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DÜSSELDORF, GERMANY — After years of relative obscurity, lupus erythematosus tumidus is getting dragged out into the light and painstakingly classified as a distinct subtype of cutaneous lupus erythematosus.

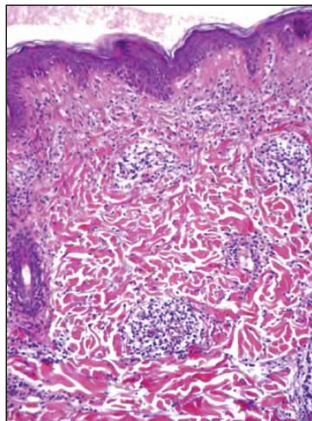
Ever since it was first described in 1930, investigators publishing in non-English language journals have sought to more precisely define the rare and elusive subtype. Meanwhile, English-language journals made no mention of lupus erythematosus tumidus (LET) before 2000.

Today, the collective clinical, photobiologic, histologic, and immunohistochemical evidence appears to firmly classify the disease as a subtype of cutaneous LE, Annette Kuhn, M.D., said at an international conference on cutaneous lupus erythematosus.

Differential skin conditions, such as discoid LE, subacute cutaneous LE, polymorphic light eruption, Jessner's lymphocytic infiltration, and reticular erythematosus mucinosis, share a variety of LET features, so "the correct diagnosis demands attention to rather subtle detail,



This patient with LET has erythematous, succulent, nonscarring plaques.



Perivascular and periadnexal lymphocytic infiltration is seen on histology.

characteristic clinical features, as well as the course of the disease," said Dr. Kuhn, who for the last decade has been seeking to characterize the disease with her colleagues at the University of Düsseldorf, Germany.

Clinically, LET is characterized by erythematosus, urticaria-like, nonscarring plaques with a bright and smooth surface. The swollen appearance of the lesions and the absence of clinically evident epidermal involvement are the most

important features of this LE subtype.

In general, skin lesions of lupus tumidus involve sun-exposed areas, such as the face, upper back, arms, and neck, but these lesions have never been detected below the waist, Dr. Kuhn said.

The lesions can disappear intermittently even when the disease is chronic. In some cases, the skin lesions slough in the periphery and flatten in the center, producing an annular configuration. The skin lesions can also appear as multiple, con-

fluent papules, sometimes indicating an eruption.

LET following the lines of Blaschko also has been described (Lupus 2002;11:388-91).

Mostly occurring in adults, only four children have been reported to have LET during the last 10 years.

One 8-year-old boy had erythematosus lesions on his ears that first appeared at 9 months of age. In the following years, he developed lesions on sun-exposed areas of

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the skin (Dermatology 2003;207:188-92).

Photoprovocation testing can be helpful in diagnosing LET, since the cutaneous subtype may be the most photosensitive type of LE, she said.

Characteristic skin lesions arose in 72% of 60 patients with LET after following a standardized protocol of UVA and UVB irradiation in a study conducted by Dr. Kuhn and her associates (Photochem. Photobiol. 2001;73:532-6). Some of these patients reacted to only UVA (50%) or UVB (48%) or both (63%). About 70% of the patients with a positive phototest reaction denied that sun exposure had an impact on their disease, "which might be due

to the latency in developing these lesions after UV irradiation and the difficulty of the patient to relate their skin disease to sun exposure," Dr. Kuhn said. The investigators also positively correlated the production of antinuclear antibodies with positive phototest reactions.

Both primary and experimentally UV-induced skin lesions display perivascular and periadnexal superficial and deep lymphocytic infiltra-

tion containing mostly leukocytes and scattered neutrophils.

These findings lend weight to the theory that LET represents a distinct subset of cutaneous LE with a similar pathogenic mechanism.

In a study of skin biopsy samples from 80 patients, Dr. Kuhn and colleagues found that in contrast with other forms of cutaneous LE, the epidermis is not involved in LET. No changes to the dermal-epidermal junction were found. Colloidal iron staining revealed abundant subepidermal deposits of mucin between collagen bundles (J.

Am. Acad. Dermatol. 2003;48:901-8).

The expression of epidermal surface molecules such as intercellular adhesion molecule-1, histocompatibility class II molecules (HLA-DR), and a distinct marker of cell activation and differentiation (27E10) are upregulated in skin biopsy segments of patients with LET to a degree similar to that seen in patients with subacute chronic LE and discoid LE (Br. J. Dermatol. 2002;146:801-9).

Such findings lend weight to the theory that LET represents a distinct subset of cutaneous LE with a similar pathogenic mechanism, rather than a distinct disease, she said. ■



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