

LUPUS TUMIDUS: UNDERREPORTED VARIANT OF LUPUS ERYTHEMATOSUS (A CASE REPORT AND REVIEW OF THE LITERATURE)Viktoryia Kazlouskaya^{1,2}, Karan Lal³, Alena Khaikova²¹Ackerman Academy of dermatopathology, New York, USA²Gomel State Medical University, Gomel, Belarus²New York College of Osteopathic Medicine, New York, USASource of Support:
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None**Corresponding author:** Viktoryia Kazlouskaya, MD, PhD viktoriakozylovskaya@yahoo.com

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Abstract

Introduction: Lupus erythematosus tumidus (LET) is an underreported variant of lupus erythematosus (LE) that is characterized by soft urticarial-like elements usually located on the sun exposed skin. LET is featured with high photosensitivity. Lesions of LET resolve without scarring and do not cause disfigurement as seen in the discoid LE variants. Lesions of LET may co-exist with other variants of LE: discoid or systemic.

Main observations: The case presents a female patient with LET localized on the lateral infraorbital areas of her face and cheeks. Histopathological evaluation showed a lymphocytic infiltrate in the middle and deeper parts of the dermis. This article also presents a contemporary review of the clinical variants of LET, histopathological features and approaches to the treatment of LET.

Conclusions: LET should be considered in urticarial-like lesions on the skin of the face and other skin exposed areas. Histopathological examination is needed to make a definitive diagnosis of the condition and helps in cases when the clinical presentation is subtle or non-specific.

Key words: lupus erythematosus; lupus tumidus; tacrolimus**Cite this article:**

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Introduction

Lupus erythematosus tumidus (LET) is an underreported variant of lupus erythematosus (LE) that is characterized soft urticarial-like elements usually located on the sun-exposed skin. LET is featured by high photosensitivity. Lesions of LET resolve without scarring and do not cause disfigurement as seen in discoid LE variants. Lesions of LET may co-exist with other variants of LE: discoid or systemic.

Case Report

A 41 year old otherwise healthy female presented with asymptomatic lesions on both sides of her face. The lesions were present for several weeks and had first appeared at the beginning of the summer season. Scaling of the skin and atrophy were absent in the lesions. The patient noticed exacerbation of lesions after increased sun exposure. She denied a family history of similar conditions. On physical examination, poorly differentiated soft urticarial lesions were located on both the right and left sides of her face inferior to the eyes and cheeks (Fig. 1).

Biopsy of the lesions was performed. Histopathological

examination revealed an unchanged epidermis without atrophy or desquamation. Periadnexal and perivascular lymphocytic infiltrates were present with concomitant mucin deposition throughout the dermis (Fig. 2).

Clinical presentation along with histopathologic analysis confirmed the diagnosis of lupus erythematosus tumidus (LET). Laboratory tests including erythrocyte sedimentation rate (ESR), anti-nuclear antibodies (ANA), anti-ds DNA antibodies, and anti-Ro antibodies were insignificant. Tacrolimus ointment (0.01%) and sunscreens were administered to the patient with the remission of the elements after 14 days.

Discussion

LET is the uncommon subtype of chronic cutaneous LE that is distinguished from the other chronic forms by absence of skin atrophy. The condition is commonly under-reported, and its incidence according to Kuhn et al, is around 16% [1]. Incidence of LET among women and men is approximately same [2]. The disease usually presents in the 30-40's age range, but several cases have been reported in kids [3].



Figure 1. Subtle soft elevated violet patches on the both cheeks.

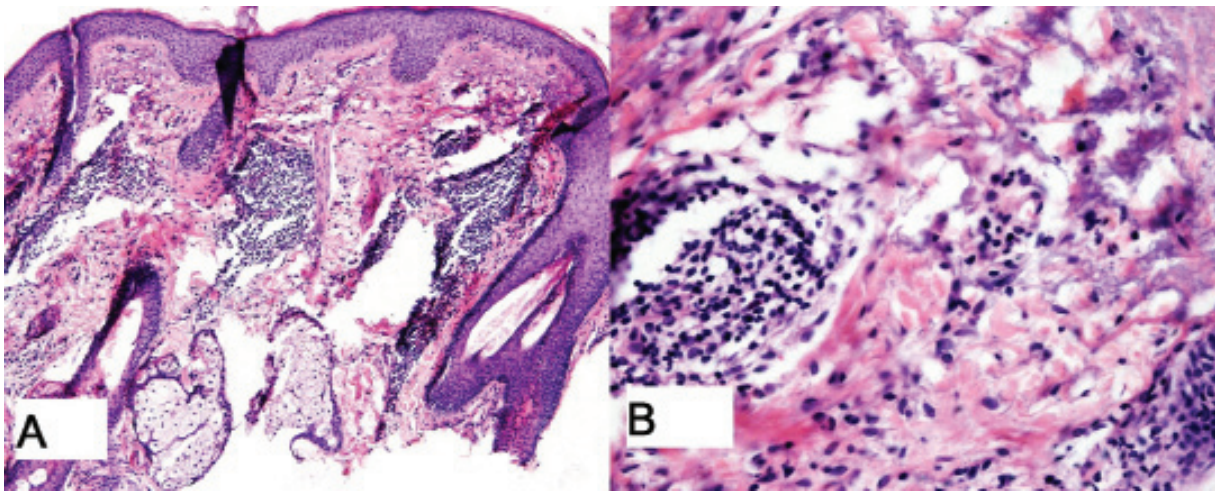


Figure 2. Histopathology of lupus tumidus. A. Deep perivascular and periadnexal lymphocytic infiltrate, Hematoxylin&Eosin, magnification $\times 40$. B. Perivascular lymphocytic infiltrate and mucin deposition in the dermis, hematoxylin eosin, magnification $\times 400$.

Pathogenesis of LET is similar to the other forms of LE, but is featured by high photosensitivity [4]. Photosensitivity is less frequent in LET patients with dark skin [5]. Patients with LET and positive ANA more frequently show signs of photosensitivity [4]. LET may be provoked by the use of certain medications such as antiviral agents [6], bortezomib [7], adalimumab [8], and ACE-inhibitors [9]. A case of LET has been described in a patient who had undergone a sex reassignment operation and was on hormone therapy [10].

Some authors argue the fact that LET should be considered a form of LE on the basis of its indolent course, absence of atrophy and changes of the basal membrane [11]. On the other hand, LET often co-exists with other forms of LE or systemic LE (SLE) and that confirms the common etiology. This has led to the further classification such that LET was classified among other forms of chronic cutaneous LE by a group of dermatologists from Dusseldorf, Germany [12].

LET has a unique clinical presentation. Disease usually manifests during months of maximum sun exposure. Uncovered skin areas (face, neck, upper trunk) are typical between locations. Elements of LET resemble urticaria, in that they are elevated and have no scaling. Their color may vary from pink to violet. Lesions may form annular processes with symmetrical

distribution, sometimes resembling targets and scaling is not seen in such lesions. Usually the lesions are asymptomatic, but light pruritis may be present. Rare manifestations of LET include unilateral eyelid edema [13] and linear LET distribution following Blaschko lines [14].

Histopathological examination is essential in the establishing diagnosis of LET. Characteristic features include a deep lymphocytic perivascular and a periadnexal infiltrate and the presence of mucin [15]. The epidermis is always intact. LET is identified by the presence of ICAM-1 in the epidermis only, in comparison to SLE where ICAM-1 expression is predominant throughout all layers of the skin [16]. Data on direct immunofluorescence (DIT) is controversial. Kuhn et al. reports that DIT was negative in all 80 patients with LET from his study with the exception of five that had IgG depositions along the basal membrane [15].

The study by Cozzani et al. in contrary show that DIT was positive in 16 from 19 patients [17]. According to their data patients presented a mixed pattern of positive DIT: deposition of IgA was seen in 2 patients, IgM – in 8 patients, IgG – in one patient and C3 in 9 patients [17]. Only 10% of all patients with LET show positive ANA [4].

Differential diagnosis of LET is extensive and includes dermatoses with the predilection on the open areas of the skin (polymorphous light eruption, rosacea, sarcoidosis, granuloma faciale, porphyria cutanea tarda), annular skin eruptions and erythemas (tinea corporis, subacute LE, superficial form of erythema annulare by Darier, erythema migrans, and erythema marginatum). Urticarial and urticarial vasculitides also should be considered. Several conditions, previously described in medical literature as separate ones, namely Jessner's lymphocytic infiltrate and deep variant of erythema annulare centrifugum of Darier are considered to be variants of LET by some authors based on identical histopathological pictures and very similar clinical presentations [18,19]. The same situation probably concerns reticular erythematous mucinosis as well [20].

Treatment of LET depends on the severity of clinical presentation and presence of other forms of LE. Usually in limited skin eruptions, local therapy and prophylaxis are sufficient for the management. Corticosteroids are traditionally used, but should be carefully monitored for the development of skin atrophy, especially on the face. Several studies as well as our case report suggest the efficacy of local tacrolimus ointment in treatment of LET [21]. In severe cases antimalarials remain the standard of treatment. It should be remembered that lower efficacy may be registered in smoking patients [22]. Prophylaxis of LET is the same as in other forms of LE and includes use of sun screens with SPF 50 or more. The course of LET is usually milder than that other forms of LE, but remission of elements may occur.

Conclusion

LET is a specific variant of LE. It should be suspected in photosensitive urticarial-like lesions on the sun exposed areas. Tacrolimus ointment and prophylaxis serve as safe and sufficient measures for treatment of localized forms of LET.

REFERENCES

- Kuhn A, Richter-Hintz D, Oslislo C, Ruzicka T, Megahed M. Lupus erythematosus tumidus. A neglected subset of cutaneous lupus erythematosus: report of 40 cases. *Arch Dermatol.* 2000;136:1033-41.
- Alexiades-Armenakas MR, Baldassano M, Bince B, Werth V, Bystryn JC, Kamino H, et al. Tumid lupus erythematosus: criteria for classification with immunohistochemical analysis. *Arthr Rheum.* 2003;49:494-500.
- Sonntag M, Lehmann P, Megahed M, Ruzicka T, Kuhn A. Lupus erythematosus tumidus in childhood. Report of 3 patients. *Dermatology.* 2003;207:188-92.
- Kuhn A, Sonntag M, Richter-Hintz D, Oslislo C, Megahed M, Ruzicka T, et al. Phototesting in lupus erythematosus tumidus--review of 60 patients. *Photochem Photobiol.* 2001;73:532-36.
- Choonhakarn C, Poonsriaram A, Chaivoramukul J. Lupus erythematosus tumidus. *Int J Dermatol.* 2004;43:815-8.
- Chamberlain AJ, Hollowood K, Turner RJ, Byren I. Tumid lupus erythematosus occurring following highly active antiretroviral therapy for HIV infection: a manifestation of immune restoration. *J Am Acad Dermatol.* 2004;51(5 Suppl.):161-5.
- Aguayo-Leiva I, Vano-Galvan S, Carrillo-Gijon R, Jaen-Olasolo P. Lupus tumidus induced by bortezomib not requiring discontinuation of the drug. *J Eur Acad Dermatol Venereol.* 2010;24:1363-4.
- Sohl S, Renner R, Winter U, Bodendorf M, Paasch U, Simon JC, et al. Drug-induced lupus erythematosus tumidus during treatment with adalimumab. *Hautarzt.* 2009;60:826-9.
- Schepis C, Lentini M, Siragusa M, Batolo D. ACE-inhibitor-induced drug eruption resembling lymphocytic infiltration (of Jessner-Kanof) and Lupus erythematosus tumidus. *Dermatology.* 2004;208:354-5.
- Zandman-Goddard G, Solomon M, Barzilai A, Shoenfeld Y. Lupus erythematosus tumidus induced by sex reassignment surgery. *J Reumatol.* 2007;34:1938-40.
- Sontheimer RD. Questions answered and a \$1 million question raised concerning lupus erythematosus tumidus. *Arch Dermatol.* 2012;136:1044-9.
- Kuhn A, Ruzicka T. Classification of cutaneous lupus erythematosus. In: *Cutaneous lupus erythematosus.* Berlin, Germany: Springer; 2004 p. 55.
- Vassallo C, Colombo G, Canevari R, Brazzelli V, Ardigo M, Carrera C, et al. Case report monolateral severe eyelid erythema and edema as unique manifestation of lupus tumidus. *Int J Dermatol.* 2004;44:858-60.
- Sabat M, Ribera M, Bielsa I, Mangas C, Fernández-Chico N, Ferrándiz C. Linear lupus erythematosus following the lines of Blaschko. *Int J Dermatol.* 2006;49:1005-6.
- Kuhn A, Sonntag M, Ruzicka T, Lehmann P, Megahed M. Histopathologic findings in lupus erythematosus tumidus: review of 80 patients. *Am J Dermatopathol.* 2003;48:901-8.
- Kuhn A, Sonntag M, Sunderkötter C, Lehmann P, Vestweber D, Ruzicka T. Upregulation of epidermal surface molecule expression in primary and ultraviolet-induced lesions of lupus erythematosus tumidus. *Br J Dermatol.* 2002;146:801-9.
- Cozzani E, Christana K, Rongioletti F, Rebera A, Parodi A. Lupus erythematosus tumidus: clinical, histopathological and serological aspects and therapy response of 21 patients. *Eur J Dermatol.* 2010;20:797-801.
- Lipsker D, Mitschler A, Grosshans E, Cribier B. Could Jessner's lymphocytic infiltrate of the skin be a dermal variant of lupus erythematosus? An analysis of 210 cases. *Dermatology.* 2006;213:15-22.
- Ziemer M, Eisendle K, Zelger B. New concepts on erythema annulare centrifugum: a clinical reaction pattern that does not represent a specific clinicopathological entity. *Br J Dermatol.* 2009;160:119-26.
- Thareja S, Paghdal K, Lien MH, Fenske NA. Reticular erythematous mucinosis-a review. *Int J Dermatol.* 2012;51:903-9.
- Kuhn A, Gensch K, Haust M, Schneider SW, Bonsmann G, Gaebelein-Wissing N, et al. Efficacy of tacrolimus 0.1% ointment in cutaneous lupus erythematosus: a multicenter, randomized, double-blind, vehicle-controlled trial. *J Am Acad Dermatol.* 2011;65:54-64.
- Kreuter A, Gaifullina R, Tigges C, Kirschke J, Altmeyer P, Gambichler T. Lupus erythematosus tumidus: response to antimalarial treatment in 36 patients with emphasis on smoking. *Arch Dermatol.* 2009;145:244-8.