

Case Report

Lupus erythematosus tumidus: a man presented with erythematous papules and plaques

腫脹型紅斑狼瘡：呈紅色丘疹及斑塊的男患者

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A 27-year-old man presented with asymptomatic erythematous papules and plaques over the back, upper chest and face. Skin biopsy showed abundant mucin in dermis with dense lymphocytic and plasmacytic perivascular infiltrate in deep dermis. Based on clinical features, histological findings and laboratory investigations, the diagnosis of lupus erythematosus tumidus was made.

患者男性，27歲，於背部上胸和面部出現無症狀性丘疹及斑塊。皮膚活檢發現真皮內有大量黏蛋白，在真皮深層的血管周圍有密集淋巴細胞和漿細胞浸潤。根據其臨床表現，組織學發現及實驗室檢查診斷為腫脹型紅斑狼瘡。

Keywords: Lupus erythematosus tumidus

關鍵詞： 腫脹型紅斑狼瘡

Introduction

Lupus erythematosus tumidus (LET) is a rare subset of cutaneous lupus erythematosus (CLE) with distinct clinical features and histological findings that should be considered as a separate entity and differentiated from other variants of CLE. It

was first described by Gougerot and Burnier in 1930.¹ Recently, Kuhn et al² proposed the diagnostic criteria of this disorder as namely: (1) presentation with erythematous, succulent, urticarial-like, non-scarring plaques in sun-exposed areas; (2) perivascular and periadnexal lymphocytic infiltrate, interstitial mucin deposition without alteration of the dermo-epidermal junction and epidermis; (3) reproduction of skin lesions after UVA and/or UVB; and (4) a dramatic response to antimalarials. We reported below a man with a diagnosis of LET based on clinical, histological and laboratory findings.

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Case report

A 27-year-old man who enjoyed good past health

presented with asymptomatic erythematous papules and plaques over the back and upper chest for four months. He then developed similar lesions over the pre-auricular areas. He had no systemic symptoms and denied photosensitivity. There was no significant drug history.

Physical examination showed blanchable erythematous papules and plaques with induration over the back, upper chest and both pre-auricular areas (Figures 1 & 2). There was no epidermal involvement. Some papules arranged in a reticular pattern. The differential diagnoses included lupus erythematosus, reticular erythematous mucinosis (REM), Jessner's lymphocytic infiltration, polymorphous light eruption (PLE).

Investigations including complete blood picture and biochemistry showed a mild decrease of lymphocyte count of $0.9 \times 10^9/L$ ($1.2-3.5 \times 10^9/L$) and mildly raised alanine aminotransferase of 63 U/L (<54 U/L). ANA titre was 1:80 (≤ 40) and anti-DNA was markedly raised to >300 iu/ml (<30 IU/ml). Anti-ENA screen was positive (anti-other) as well as ANCA (atypical). C3/C4 complements were normal. IgG was mildly raised to 1830 mg/dL (819-1725 mg/dl). Urinalysis was normal.



Figure 1. Multiple erythematous reticular papules and plaques over the back.

Skin biopsy on the back showed basal squamatization in the epidermis with borderline thickening of basement membrane. There was abundant mucin in the dermis with dense lymphocytic and plasmacytic infiltration in the deep dermis. There was no evidence of vasculitis. Immunofluorescent studies were all negative (Figures 3 & 4). The clinical and histological features were compatible with lupus erythematosus tumidus.

Discussion

Lupus erythematosus tumidus (LET) is a rare subset of cutaneous lupus erythematosus (CLE) which is characterised by distinct clinical and histological features differentiating it from other variants of cutaneous lupus erythematosus (Table 1). The rarity of reporting might be because other authors did not consider LET as a distinct entity. In a recent study,² the prognosis in patients with LET is generally more favourable than in those with other forms of CLE because none of the LET patients showed 4 or more of the American Rheumatism Association criteria for the diagnosis of systemic



Figure 2. Erythematous papules over the pre-auricular area.

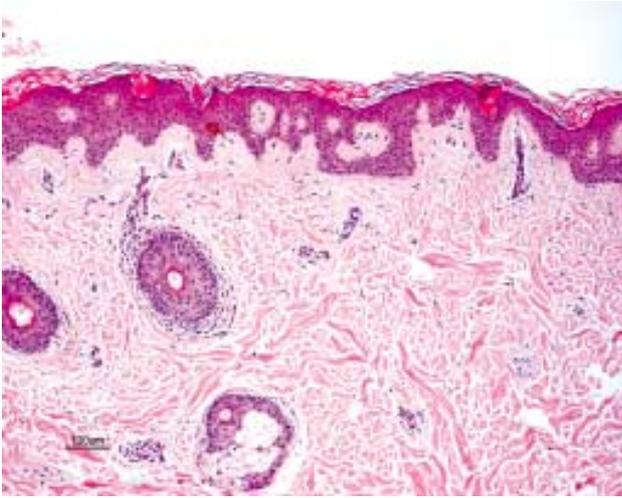


Figure 3. There is normal basket weave pattern in the stratum corneum and little interface change except around the hair follicle. (H&E)

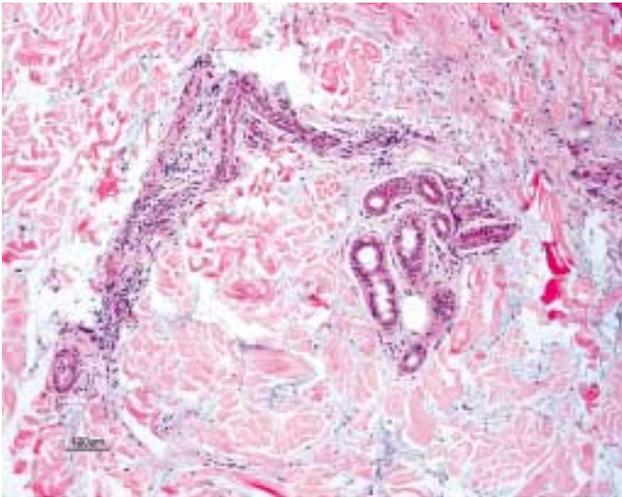


Figure 4. In the deep dermis, there is prominent increase in stromal mucin and perivascular lymphocytic infiltration. (H&E)

LE.³ However, the coexistence of LET and other features of LE, especially discoid lupus erythematosus (DLE) and SLE, has been recorded.^{4,5}

According to Kuhn's review of forty patients with LET in 2000,² 55% were male revealing a significant sex difference compared with SLE

patients. The mean age at onset of the disease was 36.4 years while the mean duration of disease was 7.8 years. The mean interval between onset of the disease and confirmation of the diagnosis was 4.9 years. There was no significant difference in disease duration between male and female patients with LET but the onset of the disease seems to be earlier in male patients.

The clinical features of LET is characterised by erythematous, succulent, oedematous, urticarial-like and non-scarring single or multiple plaques with no surface changes such as follicular plugging, scarring or atrophy that are typical features of DLE. The lesions may be mildly pruritic or asymptomatic. They involve sun-exposed areas (e.g., face, upper back, V area of the neck, extensor aspects of the arms, and shoulders) sparing the knuckles, inner aspect of arms and axilla and have never been detected below the waist. Some patients have lesions imitating the annular type of subacute cutaneous lupus erythematosus (SCLE). The lesions can disappear spontaneously and recur chronically.

Histologically, the tumid form of lupus erythematosus shows superficial and deep perivascular lymphohistiocytic infiltrates and abundant mucin deposition in the reticular dermis. Alteration of the dermo-epidermal junction and epidermis is frequently subtle or absent. Direct immunofluorescence staining of lesional skin specimens has mostly been negative for immunoglobulin or complement components. These findings are supported by recent reviews of the histopathologic findings in LET.^{4,6,7} In contrast, early reports suggested that more than 90% of DLE lesions have granular deposition at dermo-epidermal junction and that about 60% of SCLE lesions show immune deposits in a bandlike pattern at the dermo-epidermal junction.^{8,9}

According to Kuhn's study,² there was a positive history for UV sensitivity in 50% of patients and

Table 1. Differential diagnosis of lupus erythematosus tumidus (modified from Kuhn et al)²

Disease	Clinical features	Localisation	Histology (DIF)	Presence of ANA, Ro, La (Phototesting)	Antimalarial treatment	Systemic involvement
LET	Erythematous, succulent, non-scarring plaques	Sun-exposed area	Perivascular and periadnexal lymphocytic infiltrate with interstitial mucin, no epidermal change (Negative)	ANA in 10% Anti-Ro, anti-La in 5% (+ve in 70%)	Effective in 80%	None
DLE	Coin-shaped, erythematous, hyperkeratotic lesion with scaly surface, central atrophic scarring	Face, scalp, ears, V area of neck, extensor aspect of arms	Hyperkeratosis, follicular plugging, vacuolar degeneration of dermoepidermal junction (+ve in 90%)	ANA in 30-40%, Anti-Ro, anti-La occasionally (+ve in 50%)	Effective in 50%	5-10% of patients
SCLE	Non-scarring erythematous annular or papulosquamous lesions	Sun-exposed area	Focally thinned epidermis with vacuolar degeneration at DEJ, superficial perivascular and interstitial lymphocytic infiltrate (+ve in 60%)	ANA in 60-80% Anti-Ro in 40-100% Anti-La in 12-42% (+ve in 65%)	Effective in 80%	50% of patients Severe SLE in 10%
REM	Erythematous indurated plaques or reticulated macular erythema	Central chest, upper back	Copious mucin deposition with perivascular and periadnexal superficial and deep lymphocytic infiltrate (Negative)	Negative (Sun-inducible but provocative phototesting often negative)	Usually effective	None
PLE	Papular or plaque-like lesions	Sun-exposed area	Perivascular and periadnexal lymphocytic infiltrate, no mucin, dermal oedema (Negative)	ANA in 10-14% No anti-Ro/ anti-La (+ve in 75%)	No improvement	None
Jessner's lymphocytic infiltration	Papulonodular erythematous non-scarring lesions	Face, back, chest, arms	Perivascular and periadnexal lymphocytic infiltrate, no mucin, dermal oedema (Negative)	Negative (Negative)	No improvement	None

positive phototest reactions in 75% of those patients. Thus patients with LET were more photosensitive than those with SCLE or DLE as previously described.¹⁰

Our patient had the clinical and histological features that were compatible with LET. He had marked elevation of anti-DNA. There were no systemic involvements detected but the patient would be monitored regularly as there was report that some patients exhibited systemic involvement in the later course of their disease.⁵

The major differential diagnoses of LET include REM, PLE and Jessner's lymphocytic infiltration of the skin (Table 1). REM is also considered to be a variant of DLE or LET by some authors.^{11,12} In contrast to LET, majority REM patients are young to middle aged women with lesions mainly localised on the central chest or upper back. They ranged from erythematous, indurated papules to reticulated, macular erythema. Compared to PLE, LET shows a much more delayed photoreaction and interstitial mucin deposition in the reticular dermis is not detectable in PLE. In Jessner's lymphocytic infiltration of skin, the papulonodular lesions classically involve the face and are usually unrelated to UV exposure. The histological features show no interstitial mucin deposition.

Most patients with LET respond to antimalarials and sunscreens.^{2,4,5} However, even though the disease recurs chronically, the cutaneous lesions can disappear spontaneously within days or weeks.^{2,5}

In summary, our patient had the clinical and histological features that were compatible with the diagnosis of LET. Since patients presenting with LET may rarely develop into SLE during their disease course, close follow-up of the patient is important.

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