Clinical Case Report

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A case report of lupus erythematosus tumidus converted from discoid lupus erythematosus

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Abstract

Rationale: Lupus erythematosus tumidus (LET) is an uncommon type of cutaneous lupus erythematosus (CLE) that is rarely associated with other forms of lupus erythematosus.

Patient concerns: We report a 62-year-old Chinese man presented with recurrent erythematous facial plaques for 1 year and a low-grade fever for 1 week. He had been diagnosed as discoid lupus erythematosus (DLE) 1 year before. Physical examination showed diverse lesions, including prominent swelling of the eyelids, a few erythematous, edematous plaques on the left forehead, face, and neck, and 2 hairless macules. The histopathologic findings reveal liquefaction degeneration of the basal cells, perivascular, and periadnexal infiltration of lymphocytes, and interstitial mucin deposition in the superficial, and deep dermis.

Diagnoses: A diagnosis of LET was made on clinical and histological features.

Interventions: The patient started treatment with prednisolone (1 mg/kg. d), combined with hydroxychloroquine (200 mg twice daily), and topical tacrolimus.

Outcomes: The cutaneous lesions completely cleared after a period of 3 months. No adverse effects or clinical evidence of recurrence had been found during the 6-month follow-up period.

Lessons: We report a case of LET converted from DLE with diverse lesions, unusual pathologic findings and slow response to the treatment of corticosteroids combined with hydroxychloroquine. We speculate that a continuous spectrum may include DLE, LET, and systemic lupus erythematosus (SLE), these 3 entities could potentially, convert between each other.

Abbreviations: CCLE = chronic cutaneous lupus erythematosus, CLE = cutaneous lupus erythematosus, DLE = discoid lupus erythematosus, LET = Lupus erythematosus tumidus, SLE = systemic lupus erythematosus.

Keywords: discoid lupus erythematosus, hydroxychloroquine, lupus erythematosus tumidus

1. Introduction

Lupus erythematosus tumidus (LET) is a rare type of cutaneous lupus erythematosus (CLE), which has commonly, been underestimated, and neglected in clinical practice. LET is generally, an independent disease, although it has been reported to co-exist with discoid lupus erythematosus (DLE), and systemic lupus erythematosus(SLE).[1,2] Only 2 cases have been described in which LET lesions converted to DLE type lesions over time.[3,4] We are unable to find any previous reports of DLE converted to LET. Herein, we report a case of LET converted from DLE with diverse clinical manifestations and unusual pathologic findings.

2. Case report

A 62-year-old Chinese man presented with a 1-year history of recurrent erythematous facial plaques. He had developed dark red linear plaques on his left forehead 1 year previously (Fig. 1 A), and was diagnosed with DLE based on the typical pathologic features (Fig. 1B). The lesions essentially, disappeared after treatment with hydroxychloroquine at a dose of 200 mg twice daily for 1 month. Six months prior to admission, persistent erythema reappeared, and subsequently, spread to much of the left side of the patient's face, and neck. Exposure to sunlight worsened the condition. Swelling of the eyelids had been seen 2 months before, which was diagnosed as erysipelas at the local hospital, but remission was not achieved with the use of antibiotics. He was referred to our hospital for a low-grade fever and myalgia that had persisted for 1 week. The physical examination revealed 2 hairless macules, 3.0 cm × 3.0 cm and 1.0 cm × 3.0 cm in size, on the left occipital scalp, and arrayed in a linear form (Fig. 2A), swelling of the eyelids (Fig. 2B); erythematous, edematous plaques on the left forehead, face, and neck [Fig. 2C]; 2 well-demarcated, reddish-brown plaques on the left flank; telangiectasia, and necrosis on the fingertips. Routine laboratory examinations, erythrocyte sedimentation rate, and

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Ethical approval: It is merely an analysis of clinical and histological data of a patient in this manuscript. No new examinations or treatments had been conducted in the patient. For this reason, we think the ethical approval is not necessary.

Informed consent: Written informed consent was obtained from the patient for publication of this case report and its accompanying images.

The authors report no conflicts of interest.

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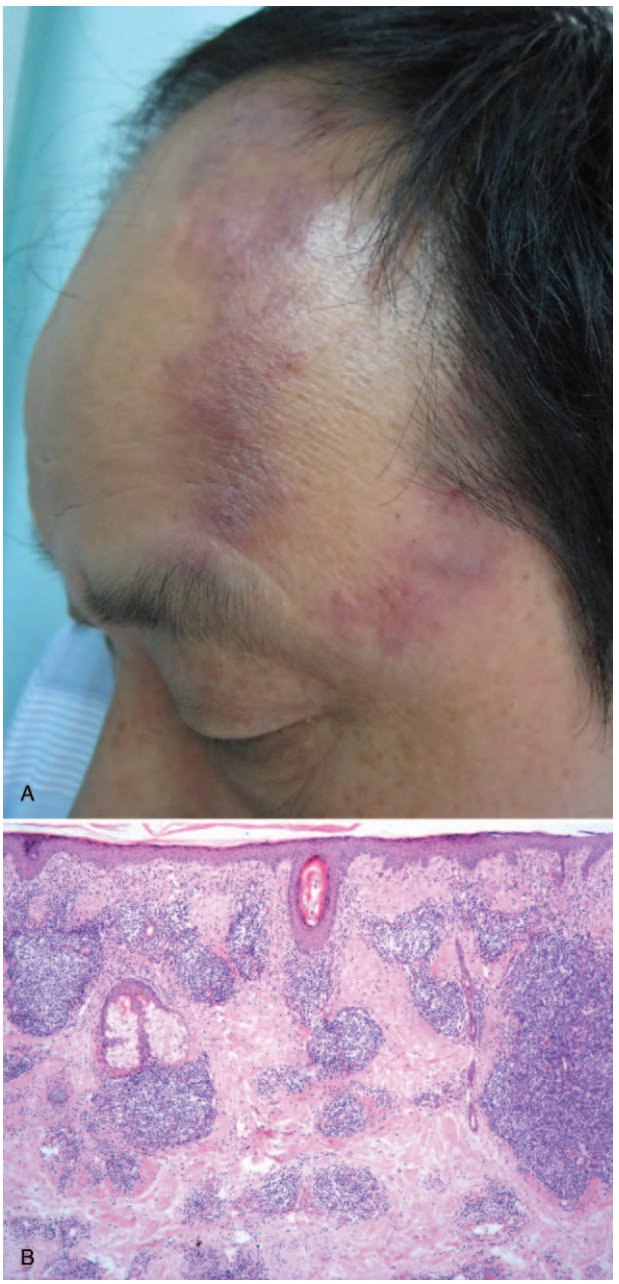

Figure 1. (A) a dark red linear plaques on the left forehead 1 year before, (B) histological features 1 year before: follicular plugging, vacuolar degeneration of the dermoepidermal junction. (hematoxylin-eosin, original magnification: ×100).

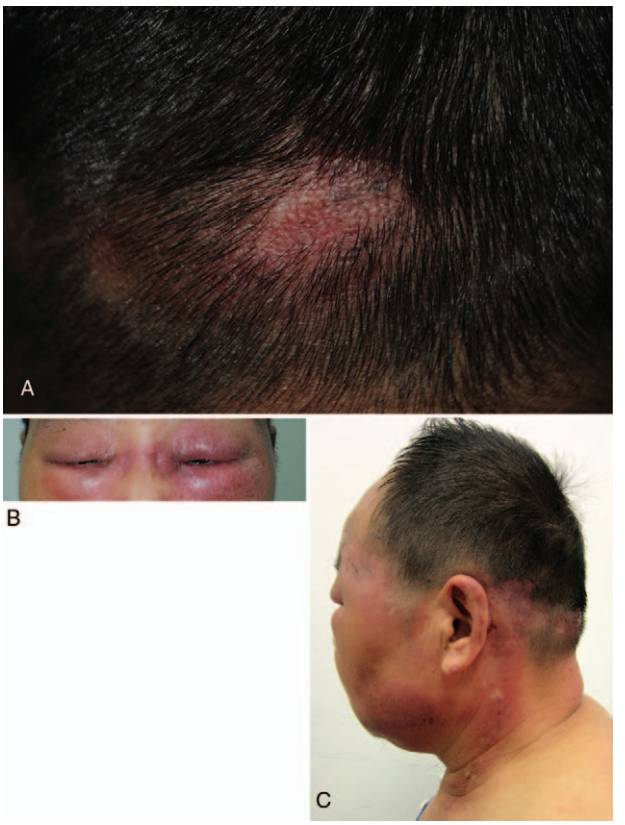

Figure 2. (A) indurate, hairless macule on the left occipital scalp, (B) swelling of the eyelid, (C) edematous plaques on the left forehead, face, and neck.

serum complement levels were normal. Antinuclear antibodies were suspicious positive, while anti-double-stranded DNA antibodies, anti-Ro/SS-A, and anti-La/SS-B antibodies were undetectable. No abnormalities were observed in chest X ray examination, abdomen color Doppler ultrasound, electrocardiogram, and electromyography. Biopsy specimens were taken from the forehead, face, flank, hairless macule, and knuckle. The histopathologic findings were similar, and demonstrated liquefaction degeneration of the basal cells, perivascular, and periadnexal infiltration of lymphocytes [Fig. 3A], and interstitial mucin deposition in the superficial, and deep dermis [Fig. 3B]. The direct immunofluorescence examination was negative.

On base of the clinicopathologic findings, a diagnosis of LET was made. Treatment was initiated with prednisolone (1 mg/kg. d), combined with hydroxychloroquine (200 mg twice daily), and topical tacrolimus. Fever subsidized within 2 days, and the cutaneous lesions completely, cleared after a period of 3 months. The prednisone dose was reduced gradually, and discontinued after 4 months. The patient had a complete clinical remission, and was instructed to avoid sun exposure. After a follow-up period of 6 months, no relapse of LET was observed.

3. Discussion

LET is a rare disease first described by Erich Hoffman in 1909.[5] It is widely, accepted as a rare variant of chronic cutaneous lupus erythematosus (CCLE). It has recently, been believed to be a new separate entity of CLE because the lesions are intermittent, and resolve spontaneously. This condition has been named intermittent CLE.[6]

According to Kuhn,[7] LET is the most photosensitive variant of cutaneous lupus. The major clinical features include non-scarring erythematous, edematous plaques presenting predominantly, on sun-exposed areas, and resolving without scarring, or hyper/hypopigmentation. In addition to the clinical morphology of the lesions, other criteria for LET include characteristic histopathologic findings, and rapid, effective systemic treatment with antimalarials. The characteristic histopathologic features of LET are firstly, a perivascular, and periadnexal lymphohistiocytic infiltrate in the superficial, and deep dermis; secondly, mucin deposition in the dermis; and thirdly, a lack of alteration of the

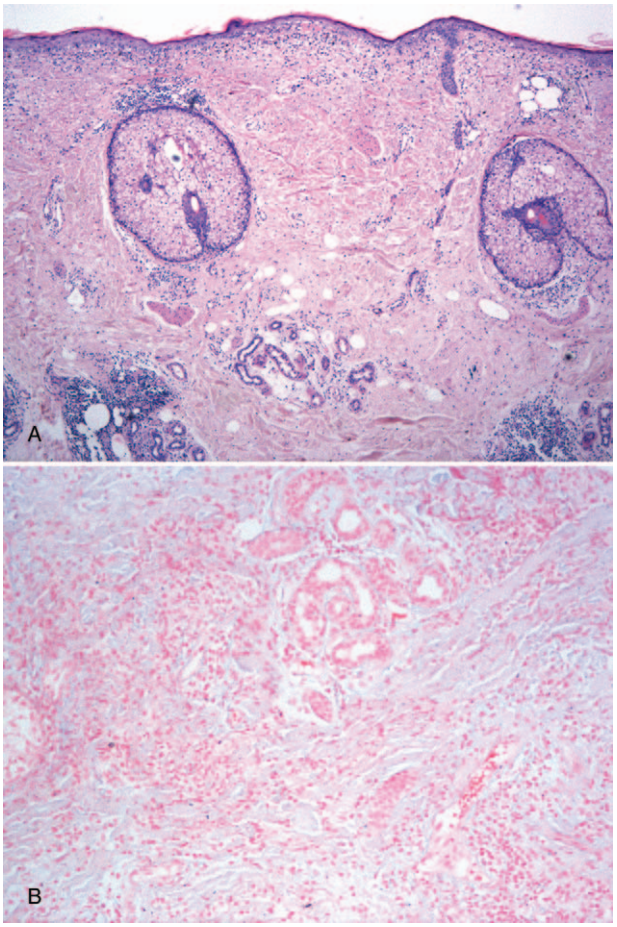

Figure 3. (A) liquefaction degeneration of the basal cells, perivascular, and periadnexal infiltration of lymphocytes. (hematoxylin and eosin, original magnification ×100), (B) interstitial mucin deposition in the superficial, and deep dermis. (alcian blue stain, original magnification ×200).

dermo-epidermal junction, and epidermis, in contrast to DLE, and SLE.[8]

In our case, the diagnosis of LET was established based on typical clinical (erythematous, edematous plaques distributed on sun-exposed areas), and histological features (perivascular, and periadnexal lymphohistiocytic infiltrate, and mucin deposition in the dermis). What called for special attention were the following diverse features that were different from ordinary LET cases.

Firstly, the lesions of our case, compared with common LET, presented with extraordinary diversity. Alopecia is a frequent clinical feature of DLE, and SLE that is rarely, described in LET.[9] Unexpectedly, in this case, non-scarring alopecia areata were pronounced on the occipital scalp. Both biopsy specimens taken from the areas mentioned above were consistent with LET. We propose that LET is a multifaceted disease with a wide spectrum of manifestations. Alopecia areata might be infrequent signs of LET.

Secondly, contrary to DLE, and SLE, the lack of alteration at the dermoepidermal junction is generally considered a prominent feature of LET, and to date only a slight vacuolar degeneration of the dermoepidermal junction has been described in 2 cases in the worldwide literature.[7] Therefore, it was rather striking that app: addword:thereforethe marked liquefaction degeneration of the basal cells was present in our case.

Thirdly, antimalarials are highly, effective in controlling LET, and a complete resolution of the lesions is normally, observed in 4 to 6 weeks.[10] In our case, despite treatment with a combination of corticosteroids, and hydroxychloroquine, the improvement was much slower than we had anticipated.

The distinctive clinicopathologic manifestations mentioned above inevitably, remind us of certain similarity with DLE, and SLE. A few findings even signify a probable progression toward SLE, such as telangiectasis, and necrosis of the fingers, and an association of fever, and myalgia with the progression of skin lesions.

4. Conclusion

Considering the history of LET being transformed from DLE, we speculate that a continuous spectrum may include DLE, LET, and SLE, and these 3 entities could potentially, convert between each other.

Author contributions

Conceptualization: Xiaomei Chen, Li Li.
Data curation: Xiaomei Chen.
Formal analysis: Xiaomei Chen.
Supervision: Li Li.
Writing – original draft: Xiaomei Chen, sheng Wang.
Writing – review & editing: Xiaomei Chen, Li Li.

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