Open Journal of
Clinical & Medical
Case Reports

Volume 2 (2016)
Issue 7

ISSN 2379-1039

Lupus Erythematosus Tumidus Mimicking Alopecia Areata

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Abstract

A 19-year-old Chinese female patient presented with recurrent violaceous plaque on the face, and hair loss mimicking alopecia areata on the mid-scalp for two months, without itching or pain. Histologic findings of lesions include a dense, perivascular, periadnexal lymphohistiocytic infiltrate in the reticular dermis and an interstitial mucin deposits in connective tissue. There is no visible damage in the dermo-epidermal junction. With the presentation of a photosensitive rash and the histologic findings, a diagnosis of lupus erythematosus tumidus was considered. Oral hydroxychloroquine sulphate 400 mg and prednisone 20 mg daily was administered for three weeks plus topical corticosteroids (mometasone furoate 0.1% cream) twice-daily for two weeks until the skin lesions cleared completely and hair started re-growing in the affected areas of the scalp. Hydroxychloroquine sulphate and prednisone doses were then reduced to 200 mg and 10 mg per day, respectively, for one month. New hair growth was observed during the 3-month follow-up visit, and no recurrence of cutaneous lesions was seen at the 12-month follow-up visit.

Keywords

Lupus erythematosus tumidus; Lupus Erythematosus; Cutaneous Lupus Erythematosus

Introduction

Lupus erythematosus tumidus (LET) is a subtype of cutaneous lupus erythematosus (CLE) that presents with annular urticaria-like papules and plaques on sun-exposed areas of the body but lacks the typical manifestations of CLE [1]. Among them, LET with lesions mimicking alopecia areata have only been reported in a few occasions [2]. Here, we report a LET case with alopecia areata-like lesions in a Chinese female who was treated successfully with hydroxychloroquine and glucocorticoids.

Case Report

A 19-year-old Chinese female developed a gradually enlarging, recurrent violaceous plaque on left cheek that companied with hair loss on the middle part of her head. Patient has no complaints of either itching or pain for two months. She denied ever taking any medications or having any other lupus-associated symptoms in past years except a history of photosensitive rashes when exposed to sunlight. Examination showed a violaceous edematous plaque of 4-6 cm in diameter on the left cheek and focal hair loss of 3-4 cm in diameter on the scalp without follicular plugs, atrophy, or scales (Figure 1A and B). Routine laboratory tests including erythrocyte sedimentation rate and serum complement levels reveal

no abnormalities. Anti-nuclear antibodies, anti-double-stranded DNA antibodies, anti-Ro/SS-A and anti-La/SS-B antibodies were all negative. Biopsy samples were taken from the plaque on the cheek and the scalp lesion. Pathological findings are similar to CLE, presenting with a dense, perivascular and periadnexal infiltrate of inflammatory cells, predominantly lymphocytes and scattered plasma cells except no changes at the dermo-epidermal junction (Figure 2A and B). The excessive mucin deposition in the dermis was easily visualized by Alcian blue staining (Figure 2C). With mouse anti-human CD4 antibody, the majority of T lymphocytes in infiltrate were CD4 positive (Figure 2D). Direct immunofluorescence staining of specimens was negative. The patient was thus diagnosed with LET. The treatment started with oral hydroxychloroquine sulphate 400 mg daily and prednisone 20 mg daily in combination with topical corticosteroids (0.1% mometasone furoate cream) twice daily for two weeks. The skin lesions had completely resolved after three weeks and hair on the scalp lesion began to re-grow. Hydroxychloroquine sulphate and prednisone were then reduced to 200 mg per day and 10 mg per day, respectively, for one month in order to reduce the withdrawal reaction. Meanwhile, the patient was also advised to avoid exposure to sunlight. At three months, new hair growth was noted in the affected area (Figure 1C and D). No signs of recurrence of cutaneous lesions or hair loss were observed at 12 months.

Discussion

LET is a rare disease that was first described in 1909 and categorized into a subgroup of chronic CLE [3]. Some have suggested separating LET from other variants of CLE according to its typical lesions, extreme photosensitivity, and good response to antimalarials [4].

Caucasian population in Europe seems to have a relatively higher prevalence of LET than other races. The median age of onset for LET is 39-49 years,although children may also be affected [5]. There is no known gender difference of its occurrence. A few cases of LET have been reported among Chinese population with smooth, erythematosus plaques on sun-exposed areas. The case reported here was unique by showing localized hair loss mimicking alopecia areata with a typical plaque on sun-exposed skin.

Clinically, LET is characterized by single or multiple, bright red or violaceous, smooth, indurated, succulent urticaria-like plaques on sun-exposed areas, without typical epidermal changes. Individual lesions may disappear spontaneously, even if the disease recurs chronically. The symptoms of non-scarring hair loss associated with LE include diffuse hair shedding during acute exacerbations, telogen effluvim, alopecia areata, and alopecia secondary to the use of medications such as antimalarials, systemic glucocorticoids, and azathioprine [7]. Scarring alopecia is invariably associated with discoid LE (DLE). Up to date, there is only one case report of a patient with LET who reported a two-year history of discrete areas of hair loss mimicking alopecia areata [2], as seen in our case here. Histopathologically, our patient had a typical histopathological features of LET, which include perivascular and periadnexal cellular infiltrates in the papillary and reticular dermis that are primarily composed of lymphocytes along with upper dermal edema and interstitial mucin accumulation [8]. The diagnosis of LET can be made based on the absence of epidermal atrophy or alteration at the dermo-epidermal junction or its presence only as scattered and focally restricted areas of vacuolar degeneration at the dermo-epidermal junction [9].

A history of photosensitivity is often present in patients with LET. Also, provocative photo-testing

reveals that patients with LET were more photosensitive than patients with other forms of CLE. Kuhn *et al*. found that characteristic LE skin lesions can be induced by UV radiation in 72 % of LE patients [4]. When UVA and UVB irradiations were used together, 32 % of LE patients developed LET [6]. The positive photo-testing reaction in LET patients often appeared at an average of 7.5 days (SD, 4.9) after the last irradiation, which implies that sun protection is one of important therapeutic regimens for LET. Antimalarial treatment is highly effective for LET treatment. Other therapeutic options include topical and systemic corticosteroids, topical tacrolimus, and methotrexate [10]. Our patient had complete remission of the plaque on the cheek and hair on the affected alopecic plaque showed signs of re-growth even after three weeks of treatment with hydroxychloroquine and prednisone.

Figures

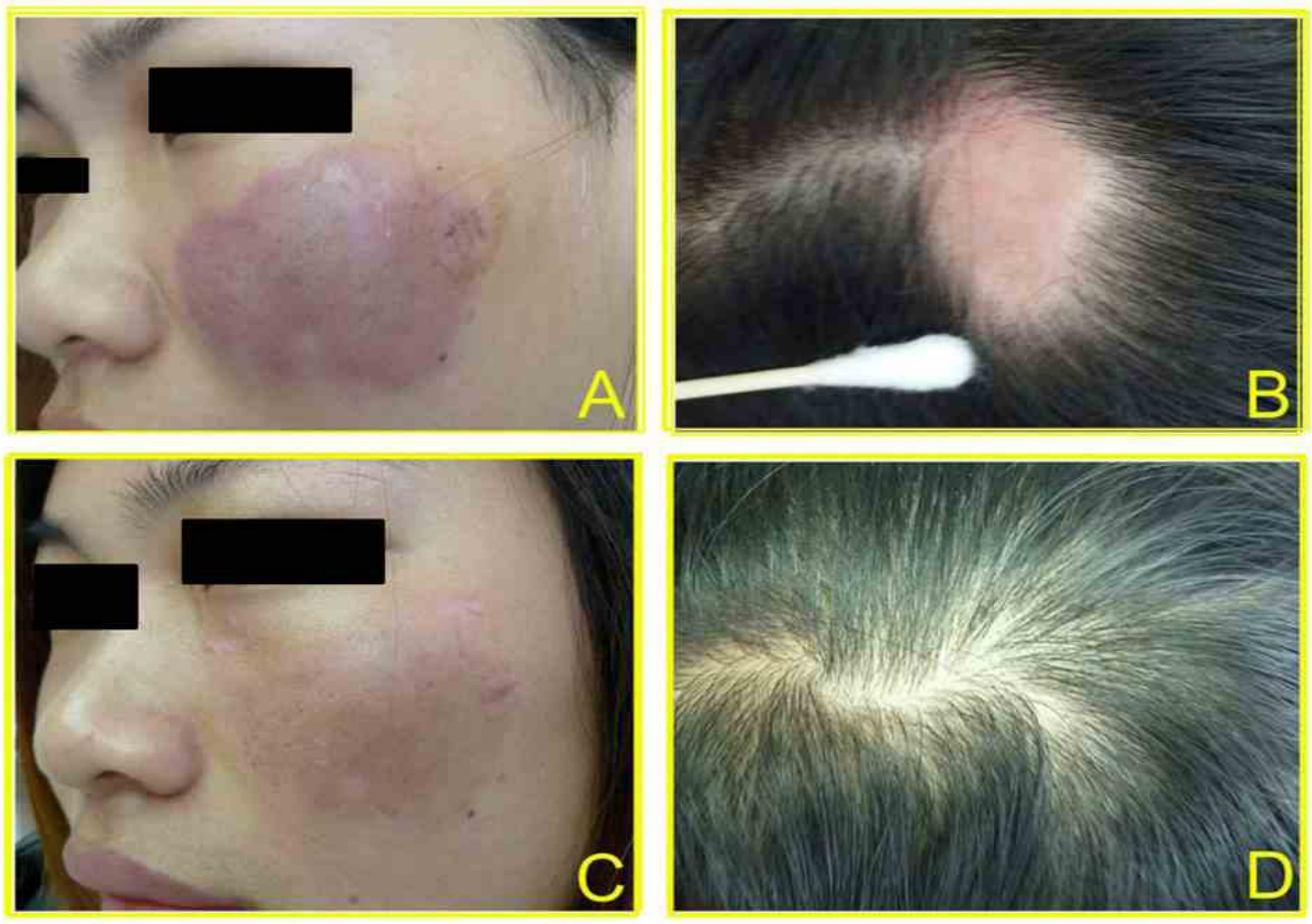

Figure 1: (A) The patient initially presented with a violaceous plaque on the cheek. (B) Hair loss on the middle scalp. (C) The skin lesion was completely resolved after three weeks of treatment. (D) New hair growth appeared on the affected scalp after three months of treatment.

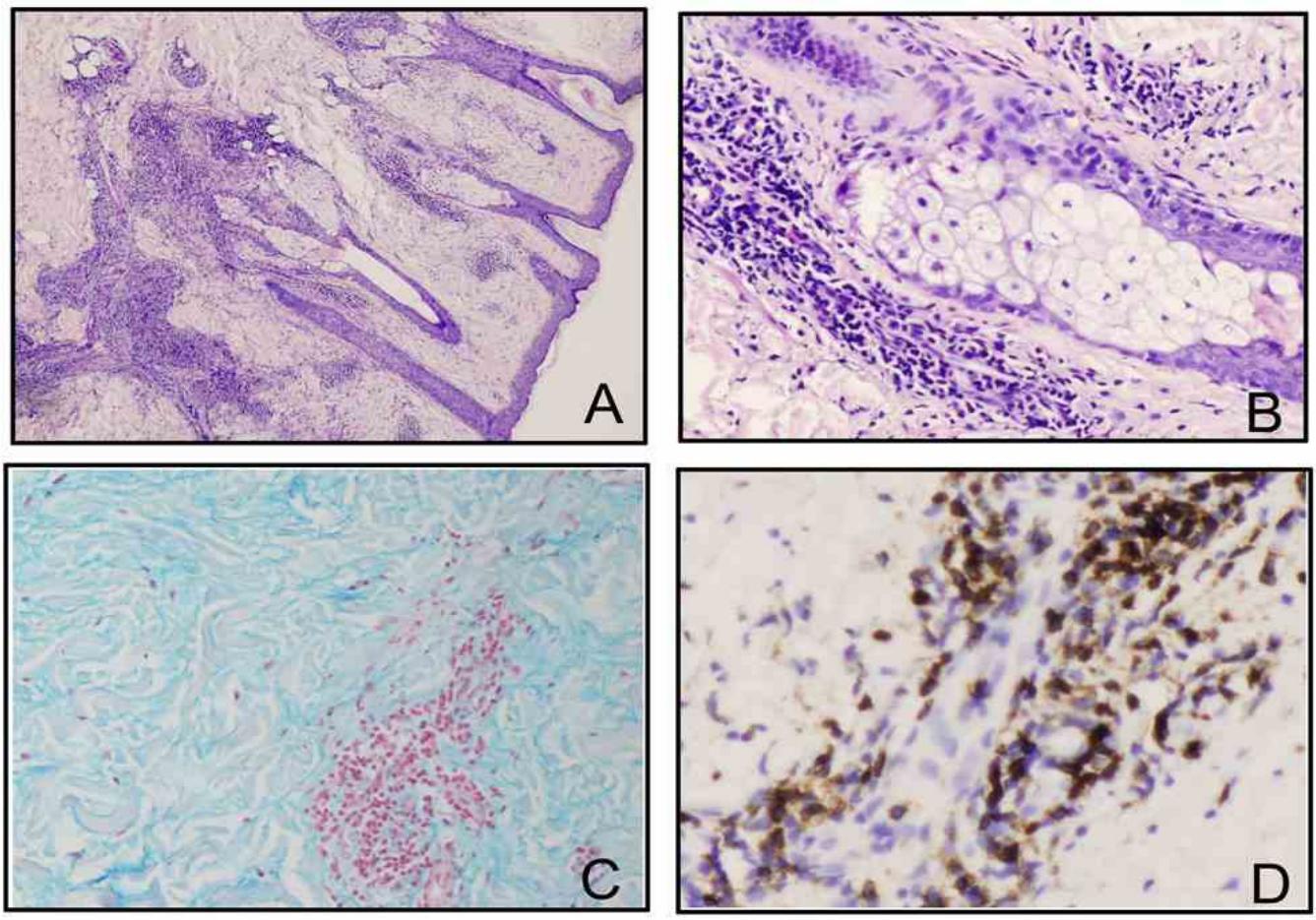

Figure 2: Histological findings of skin lesions from the scalp: (A and B) Perivascular and periadnexal infiltrate is predominantly comprised of lymphocytes and scattered plasma cells. (C) Alcian blue staining reveals interstitial mucin deposition. (D) The infiltration of $CD4^+$ cells are demonstrated by mouse anti-human CD4 antibody.

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Manuscript Information: Received: January 14, 2016; Accepted: April 08, 2016; Published: April 15, 2016

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Citation: Shi M, Yan H, Wang G, Xu X, Jia H. Lupus erythematosus tumidus in a Chinese patient mimicking alopecia areata. Open J Clin Med Case Rep. 2016; 1101

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