CASE REPORT

Rare Coexistence of Lupus Erythematosus Tumidus With Systemic Lupus Erythematosus: A Case Report

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ABSTRACT

Lupus erythematosus tumidus (LET) is a rare form of cutaneous lupus erythematosus and the coexistence of LET with systemic lupus erythematosus (SLE) also being rare. We present a case of a 43-year-old lady who was recently diagnosed with SLE and concurrent presence of LET. She presented with a skin lesion over the right lateral nasal bridge, which then spread to the right cheek. She also complained of joint pain and hair thinning. Laboratory studies showed positive both antinuclear antibody (ANA) and anti-double stranded DNA (anti-dsDNA) with hypocomplementemia. The diagnosis of LET was confirmed via skin biopsy. She fulfilled the criteria of SLE classification and was started on daily doses of hydroxychloroquine and prednisolone combined with photoprotection.
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Keywords: Lupus erythematosus tumidus, Systemic lupus erythematosus, Cutaneous lupus erythematosus, Antinuclear antibody, Skin biopsy

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INTRODUCTION

Lupus erythematosus tumidus is a rare form of cutaneous lupus erythematosus (1). It is also considered a highly photosensitive inflammatory skin disorder. SLE is an autoimmune disorder that involves multiple body systems and can exhibit various clinical symptoms, including skin involvement(2). However, the coexistence of LET and SLE is an exceedingly rare presentation(3). LET is clinically recognized for its indolent and recurring nature, as well as its ability to heal without leaving scars (1). Histopathologically, LET is characterized by dense infiltration of lymphocytes in the periadnexal and perivascular areas, with an intact basement membrane (3). Low titers of ANA (1:160) and negative titers of other autoantibodies are characteristic of most documented instances of LET. LET shows a marked response to treatment with antimalarial drugs and sunscreen (3). Other therapies, including topical or systemic corticosteroids and immunosuppressive drugs, have also been shown to be effective. Thus, LET is considered a separate entity, owing to its distinctive histopathologic features and notable response to antimalarial drugs and photoprotection.

CASE REPORT

A 43-year-old lady recently diagnosed with SLE presented with a 3-month history of hyperpigmented skin lesion over her right lateral nasal bridge. Subsequently, 10 weeks later, she developed redness and swelling over the same area extending to the right cheek. The skin lesion was itchy and tender on palpation. She also complained of a 7-month history of swelling and pain over the right proximal interphalangeal joint of the second fingers together with the left ankle joint which required analgesics when necessary.

Previously, she was diagnosed with seropositive rheumatoid arthritis (RA) with extraarticular manifestation and was under the care of a follow-up rheumatology team. However, she defaulted to follow-up for the past 2 years because her joint pain was well controlled. She also complained of hair thinning however denied photosensitive rash, oral ulcer, or fever. Clinically. she was alert, pink in appearance, and not in respiratory distress. The skin over the right cheek was erythematous with the presence of edematous plaques up to the right nasal bridge and a hyperpigmented plaque over the right lateral nose (Fig. 1). In contrast, she had no alopecia, oral ulcer, Raynaud's phenomenon, or vasculitis. Synovitis was observed over the right proximal interphalangeal joint of the second finger and the medial side of the left ankle joint.

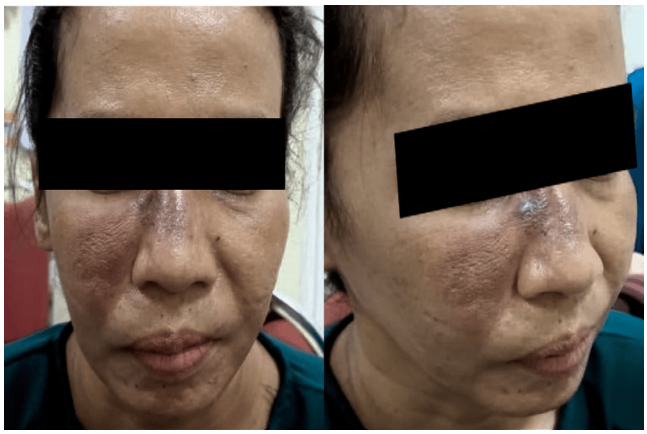

Fig. 1: Erythematous skin lesion over the right cheek with edematous plaques over right nasal bridge and hyperpigmented plaque over the right lateral nose

Laboratory investigation showed a normal white cell count; 7.0 10^9/L, normal platelet count; 356 10^9/L, and hypochromic microcytic anemia with hemoglobin of 8.9 g/L. The ANA and anti-dsDNA were positive with a titer of 275 and a high erythrocyte sedimentation rate (ESR); 63mm/hour. The complements C3 and C4 were low, whereas the extractable nuclear antigen (ENA) was negative. The serum creatinine level in the renal function test was normal, and the urinary tests showed no proteinuria. The diagnosis of LET was validated by skin biopsy, with the presence of infiltration of both superficial and deep perivascular and periadnexal lymphocytes with plasma cells (Fig. 2). The epidermis showed slight spongiosis and focal hyperkeratosis. No interface dermatitis or basal vacuolar degeneration was seen. There was also an associated increased in dermal mucin plus positive immunofluorescent study to Ig A, Ig G, Ig M, and C3.

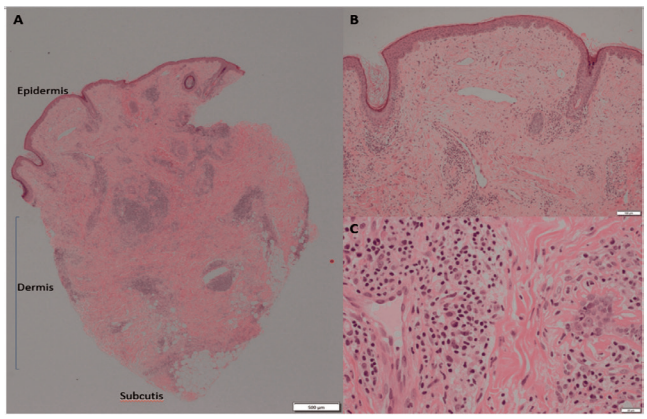

Fig. 2: Image of skin biopsy tissue (Image A, Hematoxylin & Eosin (H&E) x20) with epidermal minimal spongiosis and focal hyperkeratosis (Image B, H&E x100) accompanied by dermal infiltrates of lymphoplasma cells (image C, H&E x400)

The patient fulfilled the criteria for SLE according to the European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) 2019 criteria with positive for ANA and anti-dsDNA, hypocomplementemia with musculoskeletal manifestation, and mucocutaneous lesions. The patient was treated with oral hydroxychloroquine 200mg once daily and prednisolone tapering dose of 25mg to 5mg daily. A potent topical steroid was administered to the skin lesion along with sunscreen and emollient. The cutaneous lesions were improved through the above treatment.

DISCUSSION

The coexistence of LET and SLE is rare. Over the past two decades, LET has been classified as intermittent cutaneous lupus erythematosus (ICLE) and considered a distinct subtype of cutaneous lupus erythematosus with an intermittent and benign clinical course (1). As she was previously being diagnosed with extraarticular RA, the current diagnosis of SLE can also be associated with RA, although the association of both autoimmune diseases in the same patient is a rare phenomenon (4).

LET presented with urticarial-like erythematous, edematous, and succulent plaques that predominantly occur in sun-exposed areas (2). A palpable, non-scarring nodule that is often pink to violaceous in color is the clinical marker that distinguishes LET from other forms of chronic cutaneous lupus (3). In addition, unlike other types of chronic cutaneous lupus, skin lesions heal without atrophy, scarring, or post-inflammatory dyspigmentation. Because it primarily affects the skin, LET is less likely to coexist with SLE compared with another form of chronic cutaneous lupus (3). In this case, the initial presentation was distinct, as the lesion started with hyperpigmentation of the skin for 3 months and then later developed redness and edematous, rather than the common initial presentation of LET which is without hyperpigmentation. The lesion occurred in the face area, which can be missed as melasma or another diagnosis. The tendency to biopsy the lesion may be missed if there is no high suspicion.

Histopathologically, the biopsy sample for this case coincided with the classic finding of LET, which demonstrated the existence of perivascular and periadnexal lymphocytic infiltrate with plasma cells and the associated increased in dermal mucin (1). Serologically, LET typically has no association with the presence of ANA or only with a low titer of ANA (1). A study by Pona et al (5) revealed that the prevalence of ANA positivity in patients with LET was approximately 20%. The other commonly systemic lupus-associated serologies such as positive dsDNA, and low C3 and C4 complement levels, were also rarely observed in LET (2). In our case, the positivity of ANA and dsDNA together with hypocomplementemia might be related to the presence of systemic lupus symptoms.

The prognosis of LET is better than that of most cases of cutaneous lupus erythematosus. Furthermore, the rash can spontaneously resolve without treatment (3). Topical corticosteroids, oral antimalarials, and

photoprotection are the first-line treatments for LET. Daily hydroxychloroquine was considered the first choice of antimalarial drugs. Methotrexate and mycophenolate were used as second-line treatment, whereas thalidomide and lenalidomide were used as third-line treatment.

CONCLUSION

LET has a better prognosis than other cutaneous lupus erythematosus. Patients with LET, especially those with systemic symptoms should be investigated for other autoimmune disease because it can be one of the presenting symptoms of the underlying condition. The uncommon coexistence of LET and SLE highlights diagnostic challenges in disease management. The importance of detailed clinical examination as well as histopathological analysis should be emphasized in the diagnosis of this rare form of cutaneous lupus.

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