An unusual presentation of lupus tumidus mimicking cutaneous lymphoma

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Introduction

Tumidus lupus erythematosus (TLE) is an uncommon subtype of cutaneous lupus erythematous. It is usually confined to the skin and presents as erythematous, edematous plaques which leave no scars after regression. Usually, TLE has no systemic involvement compared to other forms of cutaneous lupus erythematosus¹. Autoimmune workup is often negative. Only a few case reports have been reported in the literature. Herein we report an unusual presentation of lupus tumidus.

Case history

A 78-year-old patient with hypertension and dyslipidemia, who had undergone common iliac artery embolectomy two months ago, presented with multiple asymptomatic skin lesions over face, scalp, both upper limbs and upper trunk for four weeks. Skin lesions initially appeared on the face, and scalp and then progressed to involve both the anterior and posterior trunk and the upper limbs. There was no history of constitutional symptoms. She denied any history of inflammatory-type joint pains, oral ulcers, photosensitivity and other features suggestive of connective tissue diseases. The rest of the systemic inquiry was negative. The drug history was unremarkable.

On examination, there were multiple well-defined erythematous nontender papules, nodules, and plaques distributed over both sun-exposed and sunprotected areas including the face, neck, chest abdomen, back of the chest, and extensor aspects of upper limbs (Figure 1-5). Lesions over the back of the chest had slight scaling. The rest of the physical and general examination was normal.

Differential diagnoses included cutaneous lymphoma, cutaneous deposits of secondary malignancy,

leukaemia cutis, sarcoidosis, and lupus tumidus. Investigations including full blood count, blood picture, renal function tests, urinalysis, liver function tests, serum calcium, chest x-ray, and ultrasound abdomen were all normal. Antinuclear antibody (ANA) was positive (1:100) and the extractable nuclear antigen-antibody panel was negative except for the weakly positive anti-U1RNP antibody. The antiphospholipid antibody panel was negative.



Figure 1.

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Figure 2.



Figure 3.



Figure 4.



Figure 5.

Incisional skin biopsy revealed mild epidermal hyperkeratosis, atrophy, follicular plugging, attenuated rete ridges, and basement membrane thickening. Vacuolar degeneration with apoptotic keratinocytes was seen in the dermo-epidermal junction. The dermis had increased mucin and moderate superficial perivascular chronic inflammation. There was no evidence of epidermotropism or atypical lymphocytes (Figure 10-12).

A diagnosis of lupus tumidus was made.

The patient clinically improved with topical and systemic corticosteroids, oral hydroxychloroquine, and photoprotection (Figure 6-9).



Figure 6.

Figure 7.



Figure 8.

Figure 9.

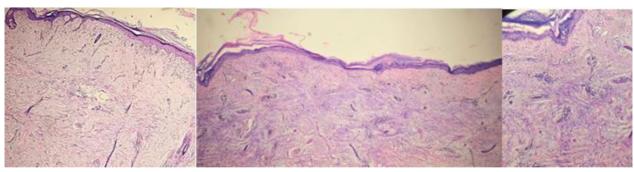


Figure 10.

Figure 11.

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Discussion

Lupus tumidus may have polymorphic presentations that mimic other clinical conditions leading to a diagnostic dilemma. Diagnosis is made based on clinicopathological correlation. Males and females are equally affected. The age of onset is around 30 to 40 years^{1,2}. TLE is characterized by erythematous, succulent, urticariform, non-desquamative plaques mainly over sun-exposed areas and healing without dispigmentation¹. Some patients can have annular lesions imitating subacute cutaneous lupus erythematosus. Approximately 70% have photosensitivity. It is mainly a skin-limited disease without systemic involvement.

TLE is typically characterized by a low titre of ANA and negative other autoantibody panels^{1,3}. Classical histological findings include a superficial and deep dense lymphocytic infiltrate in the perivascular and periadenexial regions and diffuse mucin deposition⁴.

Although TLE follows a chronic relapsing course it responds well to photoprotection, topical corticosteroids, and oral antimalarials which are the firstline treatment. Second-line treatment includes methotrexate and mycophenolate. Options for refractory disease include thalidomide⁵. It generally has a better prognosis compared to other variants of cutaneous lupus.

Conclusion

This case highlights the difficulty in distinguishing TLE from other neoplastic and inflammatory skin conditions and the importance of clinicopathological correlation.

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