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Generalized acute cutaneous lupus erythematosus in a young female

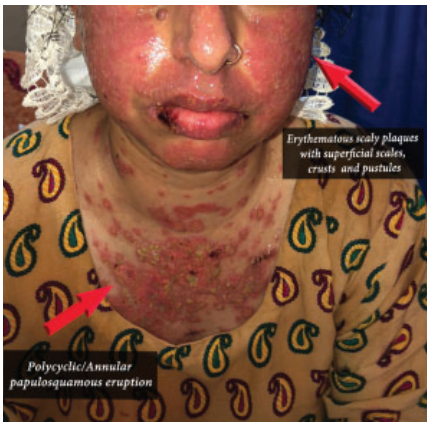

Figure 1. Erythematous scaly plaques with superficial scales, crusts, and pustules on the face, and polycyclic/annular papulosquamous eruption on the chest.

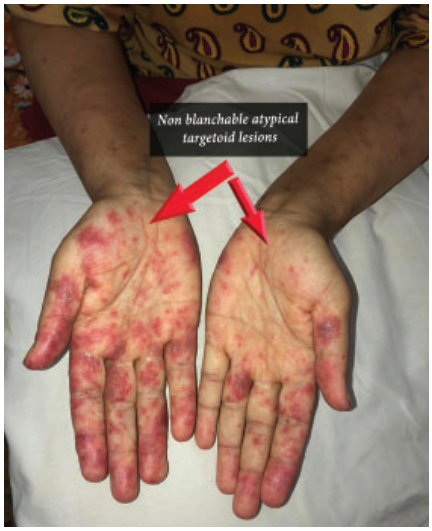

Figure 2. Symmetrical nonblanching atypical targetoid lesions on the palms.

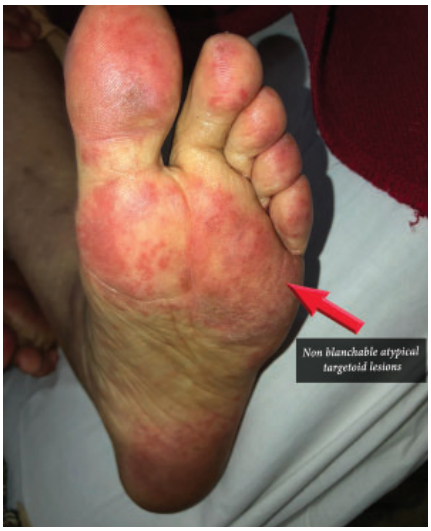

Figure 3. Symmetrical nonblanching atypical targetoid lesions on the soles.

A 28-year-old woman with systemic lupus erythematosus (SLE) presented to the emergency department with erythematous plaques on sun-exposed areas of the body that had developed in the previous 24 to 36 hours after sun exposure, as well as oral ulcers and joint pain. She had been prescribed oral steroids, hydroxychloroquine, and methotrexate 1 year previously when she was diagnosed with SLE but had stopped taking these medications 3 months ago because of herpes zoster infection.

Her temperature was 101°F (38.3°C) and her heart rate was 110 beats per minute. On examination, erythematous scaly plaques with superficial scales, crusts, and pustules were seen on her face with sparing of the nasolabial fold, along with an annular papulosquamous eruption on the chest (**Figure 1**). Symmetrical nonblanching targetoid lesions were seen on the palms and soles (**Figures 2 and 3**). Examination of the dorsum of the hands revealed periungual erythema and erythematous macules on the fingers, hands, and forearms, with sparing of the knuckles (**Figure 4**). Significant laboratory testing results included the following:

- Positive antinuclear antibody titer of 1:1280 (less than 1:160 is considered negative)
- Anti-dsDNA 80 IU/mL (reference range < 35 IU/mL)
- Hemoglobin concentration of 9 g/dL (ref-

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erence range 12–16 g/dL)
- Erythrocyte sedimentation rate 45 mm/hour (reference range 0–29 mm/hour)
- Complement component 3 = 40 mg/dL (reference range 75–175 mg/dL); complement component 4 = 10 mg/dL (reference range 14–40 mg/dL)
- Blood urea nitrogen 16 mmol/L (reference range 3–8 mmol/L)
- Creatinine 160 µmol/L (reference range 60–120 µmol/L)
- Urinalysis: 10–12 red blood cells per high-power field (reference range < 4 cells); 8–10 erythrocyte casts per high-power field (reference range 0–4 casts)
- 24-hour urinary protein 1.2 g/24 hours (reference range <0.45 g/24 hours).

Histopathologic study of the skin lesions was consistent with generalized acute cutaneous lupus erythematosus (ACLE). The patient's abnormal renal function test results and proteinuria raised suspicion for lupus nephritis. Renal biopsy revealed subendothelial deposits in glomerular capillaries and hematoxylin bodies. Endocapillary proliferation, glomerular tuft necrosis, and thickening of capillary walls were also observed. Hence, a diagnosis of diffuse (class IV) lupus nephritis was made.

The patient was started on topical and intravenous corticosteroids along with hydroxychloroquine 200 mg for generalized ACLE. Optimal wound care and strict sun avoidance were advised. Renal biopsy-proven lupus nephritis was treated with oral prednisolone 60 mg/day along with mycophenolate mofetil 2 g/day. On follow-up at 2, 4, and 8 weeks, the patient's skin lesions had resolved without scarring.

CUTANEOUS LUPUS ERYTHEMATOSUS

Cutaneous lupus erythematosus is common in patients with SLE, but the lesions can often be seen in the absence of SLE.[1]

Our patient was diagnosed with generalized ACLE which, compared with the localized form, is an extremely rare cutaneous manifestation of SLE, occurring in 5% to 10% of SLE patients and having a wide variety of presentations in a photosensitive distribution.[2] In our patient, a history of SLE and discontinuation of medications for 3 months led to increased disease activity, which has previously been associated with generalized ACLE.[1] Scarring is seldom seen once skin lesions resolve. However, dyspigmentation is common.[2]

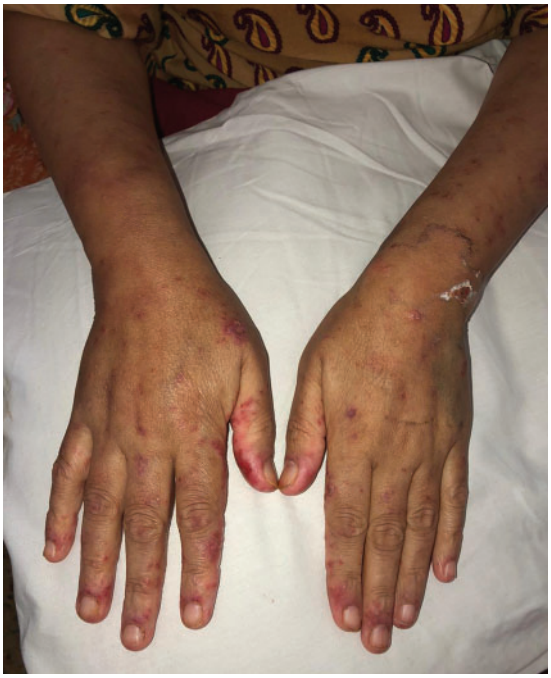

Figure 4. Periungual erythema and erythematous macules on the fingers, hands, and forearms, with sparing of the knuckles.

Diagnosis is made with skin biopsy. The classic histologic findings of localized and generalized ACLE are consistent with interface dermatitis (ie, at the interface between the dermis and epidermis) and include apoptotic keratinocytes, vacuolization of the basal cell layer of the epidermis, lymph histiocytic infiltrate in the superficial dermis, and dermal mucin deposition.[3] Management includes topical and intravenous corticosteroids along with antimalarials such as hydroxychloroquine.[4]

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Considering its rarity, generalized ACLE may be missed or mistakenly diagnosed as one of a number of other conditions:
- Drug-induced photosensitivity, for which a history of initiation of a photosensitizing drug must be present
- Dermatomyositis, which is diagnosed based on the presence of skin findings such as heliotrope rash, Gottron sign and papules, and shawl sign[5]
- Pemphigus erythematosus, which is excluded if there is involvement of any other organ besides the skin

- Atopic dermatitis, which is diagnosed after taking a careful history and performing a physical examination that reveals the presence of chronic lesions and a history of other atopic conditions.[6]

This makes it imperative for clinicians to be aware of such rare disease presentations and familiarize themselves with the essential diagnostic criteria for optimal management.

DISCLOSURES

The authors report no relevant financial relationships which, in the context of their contributions, could be perceived as a potential conflict of interest.

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