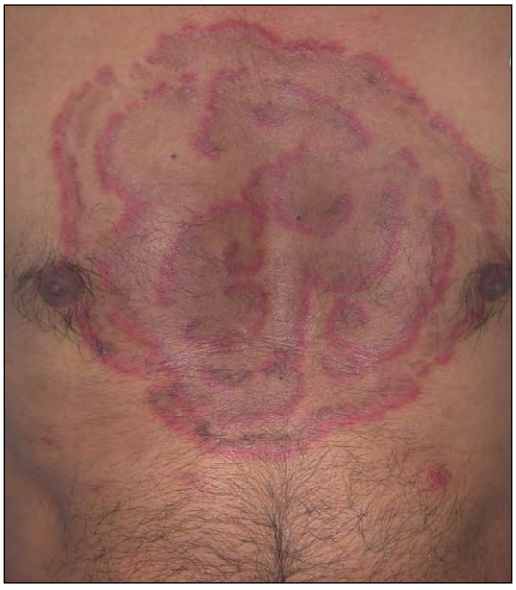
What Is Your Diagnosis?



A 41-year-old man from Bangladesh was referred to our dermatology clinic for evaluation of a mildly pruritic rash spanning the central portion of the chest. According to the patient, the rash had been present and unchanged for approximately 9 years. Prior treatments included oral cephalexin, topical antifungal agents, and topical steroid therapies without substantial improvement. The patient had no other notable medical history and was not taking prescription or over-the-counter medications. Review of systems was noncontributory. Physical examination revealed large, annular, erythematous, scaly plaques with areas of hyperpigmentation forming concentric lesions on the mid chest wall. Potassium hydroxide preparation of lesional scale did not reveal hyphal elements and fungal cultures were negative for organisms. A 3-mm punch biopsy specimen was obtained.

PLEASE TURN TO PAGE 183 FOR DISCUSSION

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The authors report no conflict of interest.
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The Diagnosis: Subacute Cutaneous Lupus Erythematosus

A skin biopsy of the lesion on the mid chest wall (Figure 1) revealed vacuolar changes along the dermoepidermal junction with necrotic keratinocytes accompanied by a perivascular lymphocytic infiltrate in the superficial and deep dermis (Figure 2). Periodic acid–Schiff stain was negative for hyphae. Serologic testing revealed a positive antinuclear antigen assay (quantitative titer of 1:1280 speckled pattern), anti-Ro (Sjögren syndrome antigen A) at a titer of greater than 258.5 EU/mL, and anti-La (Sjögren syndrome antigen B) at a titer of 232.5 EU/mL (reference range, <16 EU/mL [for both anti-Ro and anti-La]). Anti–Smith antibody test, anti–ribonuclear protein test, C3 and C4 levels, complete blood cell count, comprehensive metabolic panel, and urinalysis were within reference range. These findings were consistent with a diagnosis of subacute cutaneous lupus erythematosus (SCLE). Sun protection was advised and treatment with an antimalarial agent was recommended. The patient refused systemic therapy and was lost to follow-up.

Subacute cutaneous lupus erythematosus is a recurring, superficial, nonscarring form of cutaneous lupus erythematosus.[1] The eruption of SCLE is characteristically photodistributed. Therefore, commonly involved regions include the sides of the face, the *v* of the neck, and the extensor aspect of the upper extremities.[2] Lesions are scaly erythematous plaques that may be psoriasiform or may expand and merge to form annular polycyclic configurations. In general, the eruption is asymptomatic, though some patients report associated pruritus.[2]

Subacute cutaneous lupus erythematosus occurs in genetically predisposed individuals; is more common in whites; and affects adults aged 15 to 70 years, with a female to male ratio of 4 to 1.[3] Disease exacerbation usually follows UV light exposure. Therefore, patients often present with flares in the early spring. The majority of the affected individuals, ranging from 60% to 100%, exhibit autoantibodies to the Ro (Sjögren syndrome antigen A) ribonucleoprotein.[4] Antinuclear antibodies also are commonly found, while antibodies to double-stranded DNA are rare and may be an indication of systemic rather than cutaneous lupus erythematosus.[3] In some cases of SCLE, medications, particularly hydrochlorothiazide and nonsteroidal anti-inflammatory drugs, are implicated in the etiology.[5] Prognosis is variable and depends on the extent of internal involvement. Approximately 10% to 15% of patients with a

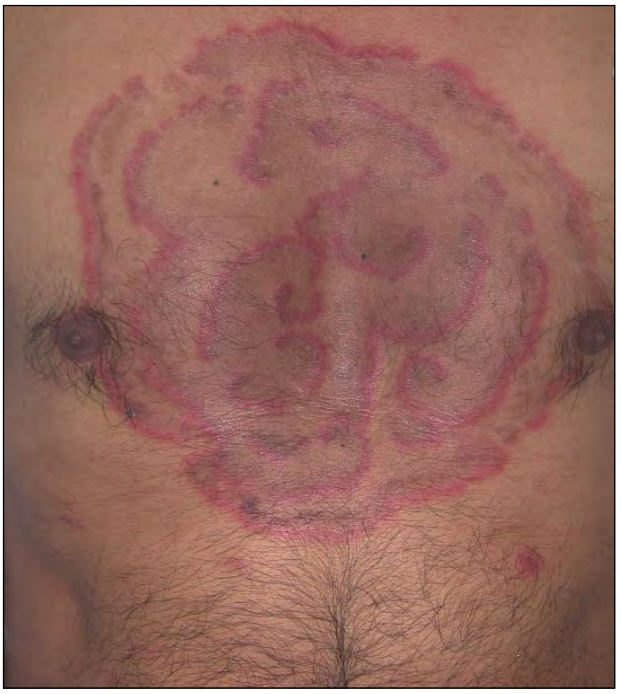

Figure 1. Annular, erythematous, scaly plaques with areas of hyperpigmentation formed concentric lesions on the mid chest wall.

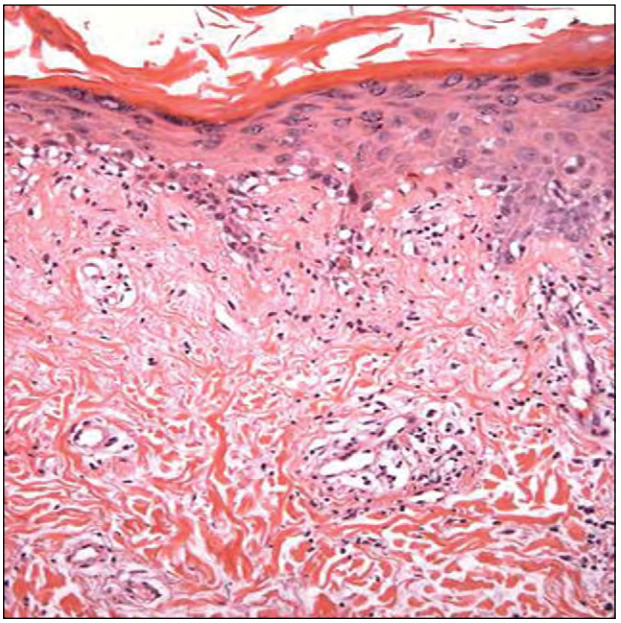

Figure 2. Histopathologic section showed vacuolar changes along the dermoepidermal junction with necrotic keratinocytes accompanied by a perivascular lymphocytic infiltrate in the superficial and deep dermis (H&E, original magnification ×20).

diagnosis of SCLE will develop systemic disease, including nephritis.[6,7]

In patients with cutaneous lesions suggestive of SCLE, the presence of anti-Ro autoantibodies and characteristic histopathologic findings confirm the diagnosis. Classically, biopsy of lesional skin shows necrotic keratinocytes and atrophy of the epidermis, hydropic degeneration in the basal layer, and a sparse interface and perivascular lymphohistiocytic infiltrate. In contrast to discoid lupus erythematosus, there is little or no hyperkeratosis, follicular plugging, perifollicular inflammation, basement membrane zone thickening, or scarring.[8,9] The direct immunofluorescence pattern in SCLE is unique and differentiates it from other forms of lupus, showing granular deposits of IgG and IgM in the epidermis rather than along the basement membrane zone.[10] Deposition of IgG and IgM along the basement membrane zone in nonlesional skin, known as the lupus band, indicates a diagnosis of systemic lupus erythematosus.[11]

Treatment of SCLE begins with broad-spectrum photoprotection and topical corticosteroids. More severe disease warrants therapy with antimalarial agents, particularly with hydroxychloroquine sulfate at doses ranging from 200 mg daily to 200 mg twice daily (up to 6.5 mg/kg) based on ideal body weight. For refractory SCLE, options include systemic corticosteroids, oral retinoids, and immunosuppressive agents.[12] In one report, a patient with SCLE who was administered numerous conventional therapies that failed was successfully treated with the monoclonal antibody rituximab.[13]

An important component in the management of patients with SCLE is monitoring for progression to systemic disease. Patients should be counseled regarding symptoms of systemic lupus erythematosus and assessment of historic information; physical examination should be routinely conducted. In addition, it is recommended that laboratory tests, including a complete blood cell count, renal function analysis, and urinalysis, be checked at least once or twice yearly. Any indication of disease progression should be managed with a multidisciplinary team of the appropriate internists and specialists.[3]

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