

swollen, somewhat fragmented and separated by mucin-containing fenestrations (1).

Many different treatments have been proposed for scleredema, including thyroid hormones, pituitary extracts, systemic corticosteroids, physiotherapy and D-penicillamine, but none has proved to be effective (6). Recently, several cases treated with high-dose penicillin (3), cyclosporine (4), bath psoralen+ultraviolet A (5), electron beam therapy (6, 7) or prostaglandin E₁ (8) have been reported. We used localized electron beam therapy, which has no serious adverse effects and only requires a short duration of treatment (2 weeks). In all our patients, electron beam therapy produced a remarkable clinical improvement of symptoms, including erythema, sclerosis, restriction of movement and pain of the lesions.

The pathogenesis of diabetes mellitus associated scleredema has not been clarified and seems to be heterogeneous. Irreversible glycosylation of collagen and resistance to degradation by collagenase in diabetes mellitus may lead to accumulation of collagen in the dermis (9). Another possible pathogenesis may relate to excess stimulation of insulin, which is one of the growth factors for connective tissue, resulting in over-production of collagen (10). The third hypothesis is that microvascular damage and hypoxia in diabetes mellitus may increase the synthesis of collagen and glycosaminoglycan by fibroblasts (7). The mechanism of electron beam therapy in scleredema is unknown, but it is possible that it may modulate the proliferation of dermal fibroblasts and the production of collagen and glycosaminoglycan.

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Erythema Multiforme-like Subacute Cutaneous Lupus Erythematosus: A New Variety?

Sir,

The recent papers on patients with lupus erythematosus (LE) presenting with features recalling erythema multiforme (EM) (1, 2) prompted us to describe a similar patient. Our experience and a review of the literature suggest that EM-like features are not uncommon in patients with LE. Most of them may be considered another morphological form of subacute cutaneous LE (SCLE) to add to the psoriasiform or annular varieties.

A 76-year-old woman, a former nurse in a department of radiology, had a widespread annular polycyclic rash for 1 month. When lesions first appeared, she was receiving 4 mg/day perindopril for blood hypertension. At the age of 42 years she had been hysterectomized for uterine carcinoma and underwent some cycles of cobalt therapy. Four months before consultation, she had been cholecystectomized for gallstones.

On examination, she exhibited asymptomatic, erythematous/violaceous, oedematous lesions with scaling-crusted

central areas involving the back, chest and abdomen, arms and face (Fig. 1). The oral mucosa was not affected. The patient denied fever, Raynaud's phenomenon, perniosis and hair loss, but she complained of dry mouth, xerophthalmia, malaise and weight loss. Perindopril was stopped with no improvement and 1 week later the lesions spread to the trunk, becoming psoriasiform.

General examination revealed no gross alteration. ESR was elevated (50 mm/h), while liver and renal function tests were normal. There was pancytopenia (haemoglobin 7.3 g/l, RBC 2,350,000/mm³, WBC 1,600/mm³ with 300 lymphocytes, platelets 111,000/mm³). Bone marrow examination revealed hypocellularity with reactive plasmacytosis. Direct Coombs' test was positive up to 2/12, while indirect Coombs' test and antiplatelets auto-antibodies were negative. There were speckle-patterned antinuclear IgG (1/640) and IgM (1/40) with positive anti-La/SSB (1/8) and Ro/SSA (1/4) antibodies. Rheumatoid factor, cryoglobulins and immunocomplexes

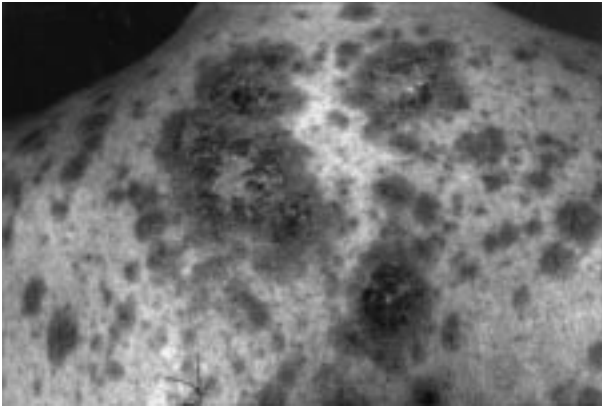


Fig. 1. Erythematous-violaceous, edematous lesions with scaling-crusts central areas involving the back.

were negative or within normal limits despite hypergammaglobulinaemia (23.4 g/dl). C4 (8 mg/dl) and C3 (85 mg/dl) levels were low. Schirmer's test was positive.

Free T4 was 2.55 ng/dl (normal values 0.71–1.85) and TSH was <0.02 UI/ml (normal range 0.38–4.70). Anti-thyroglobulin and anti-thyroid peroxidase antibodies were absent.

Sonography demonstrated a multinodular goitre, defined as hyperactive by scintigraphy. Chest radiography was normal and all tumoral markers were within normal limits. A lesion of the back was biopsied and histopathology showed the presence of many necrotic keratinocytes and subepidermal blisters with a mild perivascular lymphocytic infiltrate. Direct immunofluorescence showed granular deposits of C3 at the dermoepidermal junction.

We diagnosed our patient as having SCLE associated with Sjögren syndrome and toxic multinodular goitre. The patient was given 500 mg/day chloroquine and 25 mg/day prednisone. The lesions cleared in a week, leaving hypopigmented macules and no scarring. The treatment was diminished to 12.5 mg/day prednisone and 250 mg/day chloroquine. Two months later, the patient was still free from lesions. Laboratory examination revealed only mild anaemia (RBC 3,450,000/mm³). ANA and anti-La/SSB and -Ro/SSA antibodies remained positive with the same titres. Six months later, the patient was on 250 mg chloroquine 3 times a week and was still free from lesions.

Lesions of SCLE are typically widespread, non-scarring and asymmetrical in distribution. They frequently involve shoulders, extensor surface of the arms, upper chest, upper back and neck. Psoriasiform and annular-polycyclic forms have also been described.

Histologically, SCLE is characterized by mild hyperkeratosis and focal epidermal atrophy, vacuolar degeneration of the epidermal basal layer and a sparse lymphocytic infiltrate in the upper dermis. Necrotic keratinocytes are not currently included in the diagnostic features. Recently, however, by examining 13 bioptic specimens of SCLE patients, Herrero et al. found necrotic keratinocytes in 6 (54%) (3). By contrast,

Sontheimer, though admitting the existence of early SCLE lesions mimicking EM, relies on histopathology to distinguish the two diseases (4).

Our patient exhibited lesions involving the typical areas affected by SCLE and fulfilled all SCLE diagnostic criteria, but she presented with clinical and histological features closely simulating EM.

In fact, finding EM features in patients with LE is not a novelty. In addition to those of Lyon et al. (1) and Marzano et al. (2), many cases have been described in the past under the title of Rowell's syndrome. According to the original description (5), patients with Rowell's syndrome develop targetoid EM-like lesions, test positive for rheumatoid factor, exhibit speckle-patterned antinuclear antibodies and a circulating antibody direct to an extract of human tissues. Reviewing the literature, we suggested years ago that the overall majority of the cases labelled as Rowell's syndrome were probably only coincidental associations of LE and EM (6). Our point of view has been recently shared by Shteyngarts et al., who went so far as to deny the very existence of Rowell's syndrome (7).

Our patient with EM-like presentation is similar to most of the cases labelled in the past as Rowell's syndrome, but differs from Marzano et al.'s (2) patient in that the latter had not SCLE, but only low-titre Ro-SSA antibodies. We suggest gathering all cases of SCLE with EM-like presentation into a distinct morphological subtype of SCLE.

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