Electron Beam Therapy in Patients with Scleredema

Sir,

Scleredema is characterized by diffuse swelling and induration of the skin, which begins in the cervical region and spreads symmetrically over the upper part of the body (1). Three groups of scleredemic patients can be distinguished (2): one with abrupt onset after infection (Buschke), one with insidious onset and one with previous diabetes. Scleredema associated with infection has a self-limiting course but this is not the case when it is associated with diabetes (1). Many different treatments have been proposed for scleredema (1, 3–5), including electron beam therapy in isolated cases. We report here on 3 patients with diabetes-associated persistent scleredema, in whom substantial clinical improvements were noted after electron beam therapy.

CASE REPORTS

Case 1

A 40-year-old man presented with a 7-month history of progressive induration and thickness of the skin of his posterior neck and upper back. He had had non-insulin-dependent diabetes mellitus for 1 year, for which he had been administered oral hypoglycemics. Physical examination revealed extensive erythematous non-pitting edema of the posterior neck and upper back, with concomitant limitation of rotation of the neck. A biopsy specimen showed a thickened dermis with thick collagen bundles separated by clear spaces with alcian blue-staining mucin lying between them. He was treated with localized electron beam therapy to the lesion. He received a total of 24 Gy in 12 fractions (2 Gy per fraction) for 14 days. He tolerated the treatment well without adverse reactions. At 3-month follow-up, clinical improvement was seen with a reduction in the degree of sclerosis of the skin. The improvement was sustained at 5-month follow-up.

Case 2

A 41-year-old man presented with a 6-year history of hard, woody plaque on the posterior neck. He had suffered from diabetes mellitus for 14 years, which had been controlled by oral medication. A biopsy finding was consistent with scleredema. He was treated with 10 doses of localized electron beam therapy for 2 weeks (total 20 Gy in 10 fractions). At 2-month follow-up, clinical improvement was seen with a reduction in the degree of sclerosis of the skin. The improvement was sustained at 5-month follow-up.

Case 3

A 53-year-old man with a 15-year history of non-insulin-dependent diabetes mellitus and neuropathy presented with an 8-year history of painful hard waxy plaque on the posterior neck and shoulders. A biopsy specimen confirmed the diagnosis of scleredema. He received localized electron beam therapy (total 24 Gy in 12 fractions). His symptoms, including pain and stiffness of the lesion, were markedly improved.

DISCUSSION

Scleredema is a rare disorder whose etiology remains unclear. It is characterized by non-pitting edema and hardening of the skin around the neck, shoulder and trunk (1). Evidence of systemic involvement in scleredema is rare, but various extracutaneous manifestations, including involvement of the tongue, muscles, heart, esophagus and lung, have been described (1). Scleredema may be associated with infections, paraproteinemia, multiple myeloma and poorly controlled diabetes mellitus (1, 6). On microscopic examination, the epidermis is usually unaffected. The major alteration noted is a marked thickening of the reticular dermis, possibly 2–3 times that of normal (1). The collagen fibers appear to be thick collagen bundles separated by clear spaces with alcian blue-staining mucin lying between them.

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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). 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.

REFERENCES


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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 plasmocytosis. 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

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