Scleredema of Buschke Successfully Treated With Electron Beam Therapy

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The Cutting Edge: Challenges in Medical and Surgical Therapeutics

REPORT OF A CASE

A 58-year-old white woman with an 11-year history of type 1 diabetes mellitus presented in April 1995 with a complaint of flushing of the neck and the upper part of her trunk. She had had no antecedent febrile illness. The flushing resolved within a few days, but approximately 1 month later, the patient developed thickening and induration of the skin of the posterior section of her neck (Figure 1). She described restriction of neck movement, tenderness of the involved area, and a rash on the back of her neck. The results of her physical examination showed that she had several violaceous, indurated plaques involving the posterior section of her neck and the upper part of her back.

The diagnosis of scleredema was confirmed with a skin biopsy specimen of the upper part of her back that showed thick bundles of collagen in the middle and lower dermis that were separated by abundant mucin (Figure 2). The results of serum protein electrophoresis were normal on 2 separate occasions.

The plaques were treated with intralesional and topical corticosteroid therapy for 2 months without improvement. Oral corticosteroids were initially avoided given the patient’s history of diabetes mellitus, but when the induration spread to the lateral and anterior part of her neck and shoulders, oral corticosteroid therapy was initiated. This approach produced no improvement, and the patient developed cushingoid features. Prednisone therapy was therefore tapered and discontinued. In November 1995, she began to complain of tightness of her chest and face, with difficulty opening her eyes. Pulmonary function tests performed in December 1995 showed the following results: forced vital capacity, 57% of predicted; forced expiratory volume in 1 second, 53% of predicted; vital capacity, 57% of predicted; and total lung capacity, 64% of predicted. The restrictive component of these test results was thought to be secondary to the presence of scleredema on the patient’s trunk.

THERAPEUTIC CHALLENGE

The treatment of a progressive case of scleredema not responding to topical, intralesional, or systemic corticosteroid therapy.

SOLUTION

Electron beam therapy was begun in November 1995. The patient received a total of 20 Gy in 10 fractions (2 Gy per fraction) twice weekly for 36 days. The treatment required 6 abutted fields that were localized to the affected regions of her neck and torso. The superior extent of the radiation field was at the level of the external acoustic meatus. The largest field, encompassing the pos-
The results from a clinical examination showed a marked improvement with decreased induration of the affected areas of her skin. On follow-up, 3 months after completing electron beam therapy, she reported total resolution of her symptoms, and no clinical evidence of sclerema was noted on her physical examination. Six months after treatment, pulmonary function tests were repeated and the results showed improvement of all values: forced vital capacity, 71% of predicted; forced expiratory volume in 1 second, 84% of predicted; vital capacity, 75% of predicted; and total lung capacity, 73% of predicted.

The patient has been seen for further follow-up at 3- to 6-month intervals and has remained free of disease. Photographs taken in November 1997, 2 years following treatment, showed no clinical evidence of sclerema (Figure 3). A repeated skin biopsy sample, also taken in November 1997, shows no significant mucin deposition on histological examination (Figure 4).

Scleredema of Buschke is an uncommon disorder characterized by induration of the skin, usually of the neck and upper part of the trunk. This disease was first described by Curzio in 1752.1 In 1902, Buschke gave the condition the name “scleroedem” and added the descriptor “adultorum” to distinguish it from scleredema (sclerema) neonatorum.2 Currently, the disease is simply referred to as sclerema, or scleredema of Buschke, because many afflicted individuals are younger than 20 years. The cause of sclerema is currently unknown, but there is an apparent association with diabetes mellitus.3,4 The link between these 2 conditions is unclear, but it is known that diabetic patients with sclerema usually have difficulty controlling their diabetes, as well as diabetic complications.5

Sclerema produces findings that include a non-pitting hardening of the skin around the neck, shoulders, and trunk. This induration may be preceded by a transient erythematous eruption, as occurred in our patient.6 Thickening usually begins on the neck and later spreads to the shoulders, upper part of the trunk, and sometimes the face. Patients may complain of difficulty in opening their eyes and occasionally restrictive pulmonary disease may result, as it did in our patient. Other systemic symptoms include dysphagia, dysrhythmias, and pericardial or pleural effusions.7 Sclerema can be associated with a monoclonal gammopathy that may progress to multiple myeloma.8 Rarely is sclerema fatal.9

Three types of sclerema have been described. The classic type, described by Buschke, follows a febrile illness, and symptoms usually resolve completely. Another type is not associated with any preceding illness and follows a chronic, slowly progressive course. The third type of sclerema is seen in patients with diabetes. This type also fails to resolve, and its progression is unrelated to control of serum glucose levels.10

The histopathologic features of sclerema are characterized by thickened collagen bundles within the reticular dermis that are separated by mucin-containing fenestrations. The mucin is thought to represent nonsulfated
acid mucopolysaccharides. These thickened collagen bundles are often more prominent within the deeper dermis. This process may extend into the subcutaneous fatty tissue.

Currently, there is no established effective treatment for scleredema. Systemic corticosteroid therapy is typically used, but the patient’s response is usually suboptimal. High-dose penicillin has been used in one report with some improvement of scleredema. However, the patient in that report was only able to tolerate penicillin treatment for a short period. Another report describes 2 patients with scleredema successfully treated with cyclosporine. Both patients had the classic form of scleredema preceded by a febrile illness and would have been expected to recover fully without treatment.

The treatment of our patient was based on a case report that described the use of electron beam therapy to treat a patient with scleredema associated with monoclonal gammopathy. In that report, treatment of the patient’s entire skin was accomplished using 20 Gy in 10 sessions. Our patient was treated with localized electron beam therapy, using the same dose of radiation. In the aforementioned case, electron beam therapy resulted in improvement, although follow-up for an unspecified length of time failed to show complete resolution of symptoms.

Depending on the energy of the electron beam therapy used, its effects may be limited primarily to the epidermis and upper dermis, while largely sparing the deep dermis and subcutaneous tissue. Common acute complications with higher doses, which have been used for conditions such as cutaneous T-cell lymphoma, include erythema, desquamation, and alopecia. Delayed adverse effects consist of temporary loss of fingernails and toenails and transient anhidrosis. Rarely do secondary skin malignancies result from multiple courses of electron beam therapy. Minimal acute or delayed adverse effects would be expected at the low doses of radiation therapy used to treat our patient. In a report on electron beam therapy used to treat scleromyxedema, it was noted that the treatment resulted in inhibition of the proliferation of dermal fibroblasts. We hypothesize that a similar mechanism was responsible for the favorable response in our patient, although the exact reason for the treatment’s effectiveness in this case is unknown.

No report on electron beam therapy as a treatment for scleredema has been described in the American literature. Furthermore, we are not aware of any other report of this treatment in a patient with scleredema and
without associated monoclonal gammopathy. We report a case of scleredema in a patient with diabetes mellitus that responded to electron beam therapy after treatment with topical, intralesional, and systemic corticosteroids had failed. Two years after receiving electron beam therapy, the patient remains free of disease. This treatment should be considered as a therapeutic option in patients with persistent scleredema and associated systemic complications.

REFERENCES


Submissions

Clinicians, local and regional societies, residents, and fellows are invited to submit cases of challenges in management and therapeutics to this section. Cases should follow the established pattern. Submit 4 double-spaced copies of the manuscript with right margins nonjustified and 4 sets of the illustrations. Photomicrographs and illustrations must be clear and submitted as positive color transparencies (35-mm slides) or black-and-white prints. Do not submit color prints unless accompanied by original transparencies. Material should be accompanied by the required copyright transfer statement, as noted in “Instructions for Authors.” Material for this section should be submitted to George J. Hruza, MD, Cutaneous Surgery Center, Suite 16411, 1 Barnes Hospital Plaza, St Louis, MO 63110. Reprints are not available.

Correction

Error in Expansion. In the article titled “Bacteriology of Inflamed and Uninflamed Epidermal Inclusion Cysts” published in the January issue of the ARCHIVES (1998;134:49-51), ANA was mistakenly expanded as “antinuclear antibody” on page 50. Actually, it should have been RapID ANA System.