Sclerodema Adultorum of Buschke Treated With Radiation

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Background: Sclerodema adultorum of Buschke is an uncommon skin condition that may accompany diabetes mellitus. Current therapeutic options are poor, with only case reports and small series supporting their use.

Observation: We describe 3 patients with severe restrictive scleroderma associated with type 2 diabetes mellitus who responded to radiation therapy after other treatments had failed. Angle of shoulder abduction was used as an objective measurement of improvement, while the patients rated several subjective areas, including arm range of motion, ease of performing daily tasks, ease of breathing, and neck thickening. Objective improvement tended to plateau over time but remained well above baseline. The duration of subjective improvement was variable, lasting from a few months to the end of the observation time of 2 1/2 years.

Conclusion: Though not consistently durable in its effect, radiation therapy appears to be a viable therapeutic option in extreme cases of this difficult-to-treat disease.

Arch Dermatol. 2003;139:780-784

Considered a rare disorder, scleroderma adultorum of Buschke presents as a thickening and tightening of the skin of the neck and back that may extend peripherally and to the face. This thickening results from increased mucin deposition in the reticular dermis as well as broadening of collagen bundles. Three variants are recognized: the first is accompanied by an acute febrile illness and characterized by resolution over several months; the second follows a slow progressive course with no apparent underlying disease; and the third is associated with diabetes mellitus. This third type tends to persist and is reported by some authors to be related to glucose control and by others to be unrelated. Multiple treatment modalities for scleroderma have been published as case reports or small series, including bath psoralen UV-A, cyclosporine, low-dose methotrexate, extracorporeal photopheresis, and high-dose penicillin. No treatment has been shown to be consistently effective in large numbers of patients. Radiation therapy as a successful treatment for scleroderma has been reported in a patient with associated monoclonal hypergammaglobulinemia and in a patient with long-standing type 1 diabetes mellitus. Lee et al reported subjective improvement in 3 patients with diabetes-associated scleroderma who were observed for up to 7 months after electron-beam radiation therapy. We present 3 cases of scleroderma associated with type 2 diabetes mellitus treated with radiation therapy and evaluated both subjectively and objectively. Our patients were not helped by other treatments and were subsequently treated with radiation therapy, either electron-beam or a combination of electron-beam and photon irradiation. All 3 of our patients experienced objective improvement lasting from 1 to 2 1/2 years. In a disease with such few therapeutic options, this small series suggests that radiation therapy may be of some benefit in severe cases of restrictive diabetic scleroderma.

METHODS

Each patient had a 2- to 30-year history of type 2 diabetes mellitus and a 2- to 15-year history of slowly progressive neck thickening, decreased range of motion in the shoulders, and in the case of patient 1, restrictive lung disease. All 3 patients were diagnosed with scleroderma adultorum of Buschke based on clini-
Presentation and skin biopsy findings. The results of all biopsies confirmed an expanded dermis with enlarged collagen bundles, large interbundle spaces, and occasional visible mucin.

Electron-beam radiation therapy was the primary type of radiation therapy used to treat our patients. A computed tomographic scan of each patient was used to determine the thickness of the disease. Electron energy was chosen to deliver 100% of the dose from the skin surface to the maximum thickness of the affected area. In some cases, a bolus (superflab) was added to achieve 100% of the radiation dose at the skin surface. The maximum field size for an electron treatment was \(25 \times 25\) cm.

Multiple matching electron fields were necessary to encompass all sites of disease. All patients were treated with an initial course of radiation therapy to the primary area of restriction, which included the neck, shoulders, and upper torso. Patient 3 underwent a second course of electron-beam radiation therapy to the primary area of restriction 12 months after the initial treatment. Two patients (1 and 3) received additional radiation treatment to the lower torso, including the buttocks and abdomen, while patient 2 received additional radiation treatment to the forearms.

Shoulder abduction was evaluated before treatment (baseline) and periodically through the follow-up period of 12 to 30 months as an objective measure of improvement. In addition to arm abduction, subjective parameters were evaluated, including range of motion, ease of performing daily tasks, ease of breathing, and neck thickness. All patients scored each of these areas from 1 to 10, with 1 being no impairment and 10 being severe restriction. Each patient evaluated these parameters at baseline, at the time of greatest improvement (1 week after treatment for patients 1 and 3; 3-5 months after treatment for patient 2), and 12 to 30 months after initiation of treatment. These values were converted into percentages of improvement (positive values) or worsening (negative values) from baseline. The study was approved by the institutional review board of the University of Utah.

### RESULTS

Table 1 summarizes the clinical presentations and courses for each of our patients. The severe neck thickening is seen in the clinical and histologic photographs of patient 1 (Figure 1). All 3 patients showed significant initial objective responses in shoulder abduction. The initial improvement diminished in patient 3, but improvement returned after a second course of electron-beam radiation therapy to the neck, shoulders, and upper torso. Patient 1 showed initial improvement in shoulder abduction for 6 to 7 months, but then his abduction diminished. Nevertheless, he did not return to his pretreatment restriction. Patient 2 was observed for twice as long as patients 1 and 3 and showed the best overall improvement (Figure 2).

The patients’ subjective improvement scores, as percentages of baseline function, are summarized in Table 2 for all patients in the study. Figure 3 represents the sub-
Objective data graphically as an average of the 3 patients’ responses. Positive values represent improvement from baseline as a percentage, while negative values indicate percentage of worsening from baseline. All patients reported improvement in all areas initially but waning effect over the next 1 to 2 years. Patients 2 and 3 reported conditions worse than baseline in select areas.

The pathophysiological characteristics and course of sclerema adultorum of Buschke, whether idiopathic or associated with diabetes mellitus, are poorly understood. Disease associated with diabetes tends to begin insidiously and to persist. Additionally, patients with scleredema are more likely to have brittle diabetes. Rho et al, in reporting a series of 11 patients with diabetes-associated sclerema, noted that sclerema lesions improved partially in 5 patients with well-controlled diabetes. This suggests that poor diabetic control may be an etiologic factor in sclerema. Indeed, other diabetic complications, including diabetic nephropathy and neuropathy, seem to be fibrosing processes influenced by the quality of diabetic control. Specifically, these 2 processes involve the abnormal accumulation of extracellular matrix components. Given the

**Table 2. Percentage of Subjective Improvement From Baseline Experienced After Treatment**

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Range of Motion</th>
<th>Ease of Breathing</th>
<th>Ease of Performing Daily Tasks</th>
<th>Neck Thickness</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>75</td>
<td>33</td>
<td>47</td>
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<tr>
<td>3</td>
<td>80</td>
<td>33</td>
<td>60</td>
<td>0</td>
</tr>
</tbody>
</table>

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increased collagen and mucin production in sclerodema, there may be an analogous process occurring in the skin of patients with sclerodema.

The mechanism of action of radiation therapy in sclerodema is unknown. It is unlikely to be related to radiation-induced atrophy, since this adverse effect is usually seen only with much higher doses (5000-6000 rad [50-60 Gy]) than the dose used in the present cases and takes years to become evident. Radiation therapy has been used to treat benign conditions such as keloids and scleromyxedema, where there is excess deposition of connective tissue and mucin, respectively. Keloids and scleromyxedema are both associated with proliferation of fibroblasts, cells known to produce extracellular matrix components such as collagen and mucin. Radiotherapy may suppress or down-regulate, possibly through its acute anti-inflammatory effects, the production of fibroblasts or other cells involved with connective tissue or mucin production.

Though proliferation of fibroblasts is not apparent in histologic sections of sclerodematous skin, increased expression of extracellular matrix proteins has been demonstrated in the fibroblasts of a patient with idiopathic sclerodema adultorum. In addition, Tasanen et al have used quantitative polymerase chain reaction to show increased type I collagen gene expression in lesional sclerodematous skin.

Electron-beam radiation therapy is used to treat a number of cutaneous neoplasms, including cutaneous T- and B-cell lymphomas and basal and squamous cell carcinoma. One mechanism of action of radiation therapy in malignancy is the induction of apoptosis. The key to radiation therapy’s effect in sclerodema may lie in its ability to induce apoptosis in abnormal dermal fibroblasts or to otherwise interfere with their cell signaling, thereby decreasing collagen and mucin production. Given the possible mechanisms of action of radiation therapy,

**Figure 3.** Average subjective improvement. For each subjective measure the percentage of improvement of the 3 patients was averaged for the time of best improvement (1 week after treatment for patients 1 and 3; 3-5 months after treatment for patient 2) and improvement 12 to 18 months after treatment. Error bars indicate SD.
treatment earlier in the course of the disease would seem justified on theoretical grounds. However, since most patients with diabetic scleredema do not have debilitating disease, the adverse effects and cost of radiation therapy do not make early treatment a practical option.

Gauging improvement in scleredema adultorum of Buschke is inherently difficult because of the paucity of objective measures. Nevertheless, by evaluating the angle of arm abduction and patient self-reports, we found that all 3 of the patients described herein experienced varying degrees of improvement after radiation therapy. The treatment was well tolerated with the exception of patient 2, who received a combination of penetrating photons and electron-beam therapy and who complained of a temporary sore throat, headache, and fatigue. All of our patients experienced objective improvement that lasted from 12 to 30 months after initiation of treatment. Though the objective improvement measurement for patients 2 and 3 remained well above pretreatment levels, both patients reported worsening in 2 or 3 subjective areas each. This disparity between objective and subjective measurements may be due to progression of disease, or it could reflect the difficulty in assessing relative functioning over a 1- to 2-year follow-up. This finding highlights the importance of developing additional means of objective assessment in the study of scleredema, such as neck perimeter measurements and the use of computed tomography or ultrasound to measure skin thickness.

Electron-beam radiotherapy is an ideal choice for the treatment of cutaneous diseases such as skin cancers. This type of radiation treatment delivers the target dose to the skin surface with no penetration into deep tissues or organs, thus minimizing radiation exposure to normal tissues. For this reason, electron-beam radiation was primarily used in our patients. The main limitation in the use of electron-beam therapy is the smaller field size of treatment. Photon irradiation can usually accommodate a much larger volume and was used in combination with an electron beam to treat patient 2. To overcome the problem of treating large field sizes with electron-beam radiation, multiple matching fields must be used to encompass the large volumes affected by scleredema adultorum of Buschke. Based on our observation of 3 patients with diabetes and severe restrictive forms of scleredema, treatment with radiation, particularly electron-beam radiation, appears to be a viable option for patients with scleredema who have limited treatment options.

Accepted for publication October 26, 2002.
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REFERENCES