OBSERVATION

Complete Remission of Scleromyxedema Following Autologous Stem Cell Transplantation

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Scleromyxedema is characterized by dermal fibroblast proliferation and mucin deposition, associated with plasma cell dyscrasia. Therapy for systemic progression is often not effective, and the disease is potentially fatal.[1] We describe a man with rapidly progressive scleromyxedema in whom multiple treatments had failed before complete remission was achieved with treatment with high-dose pulse dexamethasone, high-dose melphalan, and autologous stem cell transplantation.

REPORT OF A CASE

A 46-year-old white man was referred to the M. D. Anderson Cancer Center, Houston, Tex, to undergo photopheresis for scleromyxedema. Six years earlier, pruritic papules appeared on the dorsal part of his hands but resolved with oral and topical corticosteroid therapy. Three years later, the lesions returned as waxy plaques, thickened skin, and orbital swelling. A skin biopsy specimen showed dermal mucinosis and fibrosis, consistent with scleromyxedema. IgG λ light chain plasma cell dyscrasia was noted on serum electrophoresis. The disease progressed rapidly during the next 2 years, despite therapy with prednisone, hydroxychloroquine sulfate, and azathioprine. As shown in **Figure 1**, sclerosis with pink waxy papules gave a cobblestone appearance to the central face, and bilateral ectropion prevented complete closure of the eyelids. Marked limitation in movement, with significant sclerodactyly and flexion contractures of both hands, was present (**Figure 2**).

The patient was unable to tolerate treatment with etanercept (Enbrel) or interferon alfa-2b. He experienced transient decreased skin tightness and fatigue after initiation of photopheresis, but the improvement was not sustained.[2] He could not eat because his incisor-to-incisor distance measured only 0.6 mm. Dysphagia with reflux resulted in significant weight loss. Aspiration pneumonia necessitated percutaneous endoscopic gastrostomy. Pulmonary function test results showed a restrictive pattern.

Because of his downhill course, transplantation was considered to augment chemotherapy. High-dose pulse dexamethasone was given first to reduce the plasma cell dyscrasia and to improve functional status. Two months later, he received filgrastim (granulocyte colony-stimulating factor) and underwent stem cell mobilization with high-dose melphalan.[3] This was followed by autologous stem cell transplantation. The only complications were neutropenia and thrombocytopenia on day 5, requiring platelet transfusions. He did well and was discharged from the hospital on day 21.

At the 3-month follow-up, there was significant improvement in functional status and skin examination results, showing marked skin softening and increased mobility. The cobblestone appearance of the glabella had cleared, and the patient was able to close his eyes and open his mouth to an incisor distance of 2.2 cm. He had increased quality of life and physical endurance and was able to swallow, resulting in a weight gain of 11.8 kg. By 6 months posttransplantation, the patient

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Figure 1. Facial skin sclerosis with confluence of waxy lichenoid papules on the forehead and cheeks and bilateral ectropion 3 months before transplantation.

demonstrated continued skin softening and new hair growth on the trunk and arms. Complaints of dyspnea had resolved, and pulmonary function test results showed markedly improved respiratory function. Serum electrophoresis no longer showed the monoclonal γ-globulin spike. He is in complete remission at 10 months post-transplantation and is doing well.

COMMENT

Scleromyxedema is similar to scleroderma, but has predominantly mucin deposition and is usually associated with a serum IgG monoclonal paraprotein. There has been controversy over the causative effect of this paraprotein. In one similar patient, the IgG paraprotein isolated from his serum did increase the glucosaminoglycan production by skin fibrobasts over control IgG (M.D. and

Figure 2. Marked sclerodactyly with flexion contractures of the digits.

Gerald Lazarus, MD, unpublished data, 1979). Treatment of scleromyxedema has been largely unsatisfactory, with varying results. Chemotherapy targeting the plasma cell dyscrasia has been reported to improve the disease and further supports the paraprotein's role in causing sclerosis.[4] High-dose melphalan has been reported to be effective, but may be toxic. Autologous stem cell transplantation allowed our patient to receive high-dose therapy and recover. He achieved a sustained complete remission of his skin lesions, dysphagia, and dyspnea, allowing him to return to normal activities. To our knowledge, this is the first report of treating scleromyxedema with transplantation, and additional investigations would be helpful for determining the response rate for progressive scleromyxedema.

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