weekly for the next 4 months, and patient showed a complete clinical response. During this time, she also received several intralungal Kenalog injections to the larger nodules of the face and groin.

**Discussion** | There have been a variety of treatment techniques used for CRDD, including cryotherapy, surgical excision, irradiation, oral corticosteroids, dapsone, thalidomide, and isotretinoin. To our knowledge, the use of methotrexate alone or in combination with other agents has been reported in 9 cases of systemic Rosai-Dorfman, and a complete to partial response was reported in most cases. By contrast, methotrexate therapy has been reported in only 3 cases of CRDD, and partial or no improvement was reported. However, in all of these cases, the eruption had already been present for well over a year. In our patient, a lack of response to prednisone, preexisting diabetes, and significant disease burden prompted the choice of low-dose methotrexate, to which a complete clinical response was seen over 11 months.

Though the exact cause of CRDD remains unknown, the presence of Epstein-Barr virus, human herpesvirus 6 by polymerase chain reaction, and reported eruption after vaccination with spontaneous remission over months to years suggests that CRDD is a benign reactive process involving a particular pattern of immune dysregulation. Early diagnosis remains a challenge in CRDD owing to its nonspecific clinical manifestations, including variable numbers of papules, nodules, plaques, or tumors. Timely diagnosis and initiation of methotrexate therapy may be key to effecting a rapid clinical remission while this disease remains in its active phase.

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**Published Online:** April 30, 2014. doi:10.1001/jamadermatol.2013.8679.

**Conflict of Interest Disclosures:** None reported.


**Paraproteinemia-Associated Scleredema Treated Successfully With Intravenous Immunoglobulin**

Scleredema is a fibromucinous connective tissue disease characterized by symmetric, nonpitting edema and induration of the face, neck, and trunk. Although the pathogenesis remains elusive, associations with infection, diabetes mellitus, and paraproteinemia have been established. Paraproteinemia-associated scleredema is typically characterized by a progressive course, for which no standard therapeutic protocol exists. To our knowledge, the patient described herein represents the first reported case of scleredema with paraproteinemia successfully treated with intravenous immunoglobulin (IVIG).

**Report of a Case** | A woman in her 40s presented with a 2-year history of progressive erythema and induration of the face, neck, and upper trunk. She denied a history of preceding infection or diabetes mellitus. Physical examination revealed erythema, brawny edema, and induration of the face, neck, upper trunk, and upper arms. There was significant limitation of neck flexion, extension, and lateral rotation as well as shoulder abduction and internal rotation (Figure A). The patient denied difficulty swallowing or restricted breathing.

A skin biopsy from the upper back revealed dermal thickening with separation of enlarged collagen bundles by Alcian blue–positive mucin deposition, consistent with a diagnosis of scleredema. Findings from a complete metabolic panel, complete blood cell count, and assays for fasting glucose and glycosylated hemoglobin levels were within normal limits. Serum protein electrophoresis revealed an abnormal globulin peak. Immunofixation confirmed an IgA κ-band, and serum IgA level was elevated (933 mg/dL; normal range, 140-260 mg/dL). Investigation for multiple myeloma was negative, with normal findings on skeletal survey, bone marrow biopsy, and urine
protein electrophoresis. A diagnosis of sclerema associated with IgA-κ paraproteinaemia was established.

Initial treatment with thalidomide was unsuccessful. The patient then completed a 1-year regimen of weekly to bi-monthly extracorporeal photopheresis (ECP) with initial success. However, therapeutic response to ECP plateaued, and eventually skin induration worsened and neck and shoulder range of motion (ROM) became more restricted. Interestingly, the patient subsequently reported improvement in symptoms after a flulike illness. For this reason, a 6-week course of pegylated-interferon was administered by her hematologist, but this resulted in no benefit.

Based on 2 recent publications demonstrating the efficacy of IVIG for sclerema associated with diabetes and streptococcal infection, monthly IVIG treatment was initiated at a dose of 2 g/kg over 2 consecutive days. After just 2 cycles, the patient noted significant improvement. After 5 cycles, her ROM continued to improve, as evident on clinical examination and careful evaluation of high-quality photographs of shoulder and neck ROM before and after IVIG therapy (Figure). Shoulder abduction improved from approximately 140 to 170 degrees over this time period. Serum IgA level after 6 monthly cycles of IVIG therapy remained elevated, though less than the initial measurement (841 mg/dL). At the patient’s most recent follow-up evaluation, the patient continued to receive monthly IVIG treatments with ongoing improvement in skin induration and stabilized ROM.

Discussion | Although the efficacy of IVIG has been established in a variety of inflammatory diseases, its mechanism of action remains elusive. It may exert immunomodulatory and anti-inflammatory effects by suppression of proinflammatory cytokines, downregulation of adhesion molecules and chemokine receptors, and neutralization of superantigens.

Treatment of sclerema, regardless of subtype, remains a challenge. Variable improvement has been reported with phototherapy, systemic corticosteroids, methotrexate, cyclosporine, ECP, and radiation. IVIG has had reported success in diabetes-associated and streptococcal-associated sclerema, in 1 case each. The rapid response to IVIG in this patient with IgA-κ paraproteinemia further supports the potential efficacy of IVIG treatment for sclerema. Despite marked clinical improvement, the patient’s IgA levels did not significantly decrease in concordant fashion with IVIG treatment. This finding suggests that the mechanism of action of IVIG resulting in therapeutic benefit in paraproteinaemia-associated sclerema may be similar to that seen in sclerema associated with other causes rather than from directly lowering the paraprotein level. Additional investigation is necessary to further define the role of IVIG in the treatment of sclerema.

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Conflict of Interest Disclosures: None reported.

Funding/Support: Dr Vleugels’ career has been supported by a Medical Dermatology Career Development Award from the Dermatology Foundation.

Role of the Sponsor: The Dermatology Foundation had no role in the design and conduct of the study; collection, management, analysis, and interpretation of the data; preparation, review, or approval of the manuscript; and decision to submit the manuscript for publication.

Additional Information: Dr Femia is now with the Department of Dermatology, New York University Medical Center, New York.