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CASE REPORT

Good Response of Scleromyxedema and Dermato-Neuro Syndrome to Treatment With Intravenous Immunoglobulins☆

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KEYWORDS
Scleromyxedema;
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Abstract Scleromyxedema is a potentially serious disease that can have various systemic complications. One of the most frequent forms of central nervous system involvement is dermato-neuro syndrome. High-dose intravenous immunoglobulins are among the drug treatments that have been used for this syndrome. We describe 2 patients with scleromyxedema, one of whom developed dermato-neuro syndrome. Both patients responded well to treatment with high-dose intravenous immunoglobulins. We suggest this therapy as a suitable first-line treatment for scleromyxedema and for its neurological complications.

PALABRAS CLAVE
Escleromixedema;
Complicaciones neurológicas;
Síndrome dermato-neuro;
Inmunoglobulinas endovenosas

Escleromixedema y síndrome dermato-neuro: buena respuesta al tratamiento con glucocorticoides e inmunoglobulinas endovenosas

Resumen El escleromixedema es una enfermedad potencialmente grave que puede asociarse a complicaciones sistémicas diversas, entre las que se encuentran las que involucran al sistema nervioso central, siendo el síndrome dermato-neuro una de las más infrecuentes. Se han utilizado para su tratamiento fármacos variados, entre ellos, altas dosis de inmunoglobulinas endovenosas. Se describen dos casos de escleromixedema, uno de los cuales desarrolló un síndrome dermato-neuro, ambos con una excelente respuesta al tratamiento con inmunoglobulinas endovenosas. El uso de altas dosis de inmunoglobulinas endovenosas en el tratamiento de esta enfermedad y sus complicaciones neurológicas se perfila como un tratamiento de primera línea.

Introduction

Scleromyxedema is an uncommon disease characterized by mucin deposits, fibrosis, and proliferation of fibroblasts in the dermis. It is often associated with paraproteinemia and very varied systemic manifestations,[1-3] the most notable of which—because of its rarity—is the so-called dermato-neuro syndrome.[4] Response to the treatment of this disease and its complications is often disappointing, although favorable

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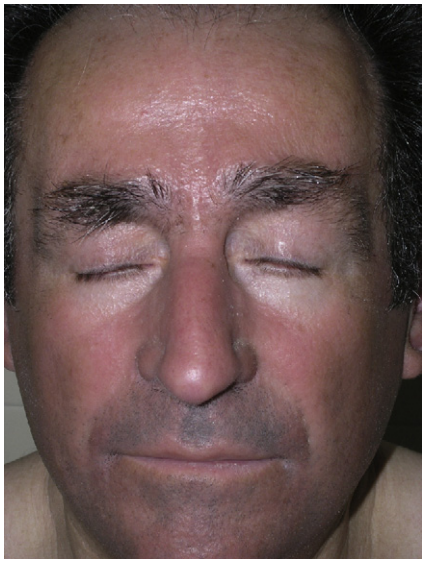

Figure 1 Minute skin-colored papules that are waxy to the touch are visible on the forehead. The wrinkles around the eyes and mouth have disappeared as a consequence of mucin deposits in the skin.

results have been achieved in recent years with intravenous immunoglobulin (IVIG).

We report on 2 patients with scleromyxedema, one of whom developed neurocutaneous syndrome. Both patients had an excellent response to IVIG.

Case Descriptions

Patient 1

A 72-year-old man with a diagnosis of scleromyxedema was referred to our department in 2005 for follow-up and treatment. Physical examination revealed firm, waxy papules (2-3 mm) arranged close together on the auricle of the ear, the face, and the dorsum of the hands. In addition, the skin was hard and thick to the touch at these sites and on the neck, chest, and arms (Figs. 1 and 2). Microscopy revealed an irregularly distributed proliferation of fibroblasts in the dermis, together with increased collagen fibers and diffuse mucin deposits in the middle

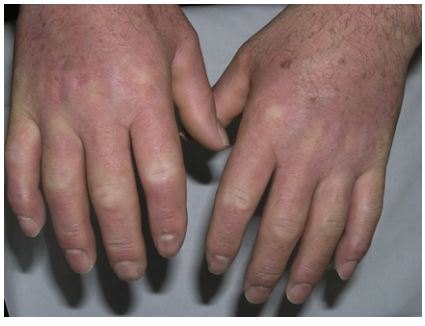

Figure 2 Rigid appearance of the hands and depression over the proximal interphalangeal joint surrounded by redundant skin forming the characteristic donut sign.

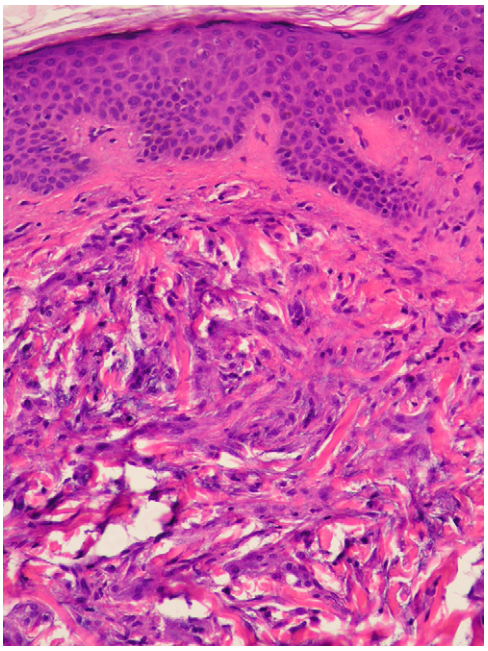

Figure 3 Irregular proliferation of fibroblasts with mucin deposits in the superficial and middle dermis (hematoxylin-eosin, original magnification ×20).

reticular and papillary dermis (Fig. 3). Protein electrophoresis revealed a monoclonal band of immunoglobulin (Ig) Gλ; bone marrow aspirate was normal. Echocardiography revealed mild asymptomatic pulmonary hypertension. The patient received various treatments over the following years, including melphalan, retinoids, corticosteroids, thalidomide, and photochemotherapy; however, the response was poor. In March 2008, the patient began treatment with IVIG (2 g/kg/mo) and initially experienced a notable improvement (skin less thickened, greater mobility of the limbs, and increased mouth opening) that did not continue during the following months. IVIG was, therefore, stopped after 7 cycles. The patient then received chlorambucil for 4 months, with no improvement. In April 2009, following placement of a partially tunneled central catheter, extracorporeal photopheresis was started. After the second session of the first cycle, the patient went to the emergency department complaining of fever (38 °C) that had started 24 hours earlier with no accompanying symptoms. The laboratory workup showed neutrophilia (90%) with no leukocytosis. Blood cultures were positive for *Streptococcus lugdunensis*, although this entity was not detected in the catheter culture. The patient's initial progress with antibiogram-guided antibiotic therapy was good; however, on the fifth day of admission he presented 2 episodes of tonic-clonic seizures preceded by high fever (39 °C) and hypertension in less than 24 hours. The second episode was followed by diminished level of consciousness and stupor. The patient went into coma and was intubated for mechanical ventilation. His condition did not improve despite therapy with anticonvulsants. Two new blood cultures and a catheter tip culture were negative. No abnormal findings were detected in electroencephalography, lumbar puncture, cranial computed tomography, magnetic resonance imaging, or magnetic resonance angiography.