Scleromyxedema: Presentation of 2 new cases and review of the literature☆

Escleromixedema, presentación de 2 nuevos casos y revisión de la literatura

Dear Editor,

Scleromyxedema (SM) is a rare, progressive, chronic disease. Diagnosis is based on 4 criteria: (1) a papular sclerodermoid eruption; (2) microscopic evidence of mucin deposition, fibroblast proliferation and fibrosis; (3) monoclonal gammopathy and (4) absence of thyroid involvement. It may present with severe systemic participation, dermato-neuro syndrome may be fatal. Prognosis and treatment are unclear. At present favourable response has been described with intravenous immunoglobulins (IVIG) and haematopoietic stem cell transplant (HSCT).[1–3]

Our aim was to present 2 new cases and to review the patient series published in the medical literature – Pubmed, with SM diagnosis. A systematic search was made with the following terms: scleromyxedema (MeSH) AND paraproteinemias (MeSH).

Case 1: 37-year-old male, smoker for 10 years of one packet per day. He presented with oedema, erythema and induration in face, arms and hands of 6 months onset. Examination revealed: hard sclerodermiform skin with papular appearance, on the face, microstoma (with dysphagia and loss of 20 kg in weight), and involved the pinna, chest, abdomen and arms with sclerodactyly, with a 31 classification score on the Rodnan scale. Analysis was performed with IgG lambda gammopathy. Treatment was prescribed with prednisone and hydroxychloroquine, with no improvement. Skin biopsy confirmed SM.

Case 2: 69-year-old male, with high blood pressure and dyslipidemia. The condition began with a papular eruption in the first finger of his right hand, left hand and face, and treatment was initiated with phototherapy, prednisone and hydroxychloroquine, with no improvement. There was a progression of the skin induration in his face, with microstoma (associated with dysphagia and loss of 7 kg in weight), arms and hands with sclerodactyly, chest, abdomen and lower limbs, and classification of 26 on the Rodnan scale. Analysis was performed with IgG lambda gammopathy. A study of systemic involvement was conducted with diagnosis of sensory polyneuropathy in lower limbs and oesophageal hypomotility. Skin biopsy confirmed SM. In both cases treatment was initiated with IVIg, with partial response and he was referred to the haematological department for autologous stem cell transplant (ASCT), with improvement in skin symptoms, reduction of induration on the Rodnan scale and systemic improvement.

SM is a rare fibromucinosis-type skin disorder, described by Montgomery and Underwood in 1953. Classification was reviewed in 2001 by Rongioletti and Rebora. For its diagnosis, the patient has to have met with 4 criteria.[1–3] It presents within an age range of 28–77 years, with no gender differences. As the disease advances, skin infiltration with sclerosis becomes generalised, leading to significant disability. The extra-cutaneous symptoms include: gastrointestinal dysmotility and peripheral neuropathy. Dermato-neuro syndrome describes the involvement of fever, convulsions and coma, which is an uncommon manifestation, and has a mortality rate of 32%[4] SM is associated with a monoclonal protein and particularly with type IgG lambda. The relationship between skin findings and the paraprotein is unknown.[2–4] Differential diagnosis has to be made with sclerodermia, and nephrogenic systemic fibrosis, among other diseases.[5] Treatment of SM is unspecified.[1] Up until now, a study by Blum et al. is the most extensive series of patients treated with IVIg and with favourable long-term follow-up results. However, response to immunoglobulins was transitory and required maintenance therapy.[1] ASCT may be efficacious and efficient.[2,3,6,7] IN 2001 Feasel et al. described the first case of remission of SM after an ASCT.[6] In a later study in 2016, Chockalingam and Duvic concluded that ASCT appears to be a safe and effective long-term treatment.[3]

SM is a chronic disease with a reserved prognosis. There is no specific definitive treatment.[8–10] Randomised, multicentre studies are needed, and larger patient groups.

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