

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, dermatoneuro 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 scleroderma, 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.

Acknowledgements

Our thanks to Daniel González Fernández, Ana M. Pilar González Rodríguez and Pablo Gonzalvo Rodríguez.

References

- Blum M, Wigley FM, Hummers LK. Scleromyxedema: a case series highlighting long-term outcomes of treatment with intravenous immunoglobulin (IVIg). *Medicine (Baltimore)*. 2008;87:10–20.
- Lacy MQ, Hogan WJ, Gertz MA, Dispenzieri A, Rajkumar SV, Hayman S, et al. Successful treatment of scleromyxedema with autologous peripheral blood stem cell transplantation. *Arch Dermatol*. 2005;141:1277–82.
- Chockalingam R, Duvic M. Scleromyxedema: long-term follow-up after high-dose melphalan with autologous stem cell transplantation. *Int J Dermatol*. 2016;55:e539–43.
- Fleming KE, Virmani D, Sutton E, Langley R, Corbin J, Pasternak S, et al. Scleromyxedema and the dermatoneuro syndrome: case report and review of the literature. *J Cutan Pathol*. 2012;39:508–17.
- Wigley F, Friday RP, Shepard JA, Nazarian RM. Case records of the Massachusetts General Hospital. Case 8-2015. A 68-year-old man with multiple myeloma, skin tightness, arthralgias, and edema. *N Engl J Med*. 2015;372:1056–67.
- Feasel AM, Donato ML, Duvic M. Complete remission of scleromyxedema following autologous stem cell transplantation. *Arch Dermatol*. 2001;137:1071–2.
- Donato ML, Feasel AM, Weber DM, Prieto VG, Giralt SA, Champlin RE, et al. Scleromyxedema: role of high-dose melphalan with autologous stem cell transplantation. *Blood*. 2006;107:463–6.
- Dinneen AM, Dicken CH. Scleromyxedema. *J Am Acad Dermatol*. 1995;33:37–43.
- Le Moigne M, Mazereeuw-Hautier J, Bonnetblanc JM, Astudillo L, D'Incan M, Bessis D, et al. Clinical characteristics, outcome of scleromyxedema: a retrospective multicentre study. *Ann Dermatol Venereol*. 2010;137:782–8.
- Rongioletti F, Merlo G, Cinotti E, Fausti V, Cozzani E, Cribier B, et al. Scleromyxedema: a multicenter study of characteristics, comorbidities, course, and therapy in 30 patients. *Am Acad Dermatol*. 2013;69:66–72.

Eva M. Fonseca Aizpuru,^{a,*} Carlos Delgado Verges,^a Esther González García,^b Luis Barcala Vázquez^c

^a Servicio de Medicina Interna, Hospital Universitario de Cabueñes, Gijón, Asturias, Spain

^b Servicio de Hematología, Hospital Universitario de Cabueñes, Gijón, Asturias, Spain

^c Servicio de Dermatología, Hospital Universitario de Cabueñes, Gijón, Asturias, Spain

* Corresponding author.

E-mail address: evamfonseca@yahoo.es (E.M. Fonseca Aizpuru).

2173-5743/ © 2018 Elsevier España, S.L.U. and Sociedad Española de Reumatología y Colegio Mexicano de Reumatología. All rights reserved.

[☆] Please cite this article as: Fonseca Aizpuru EM, Delgado Verges C, González García E, Barcala Vázquez L. Escleromixedema, presentación de 2 nuevos casos y revisión de la literatura. *Reumatol Clin.* 2020;16:430–30.