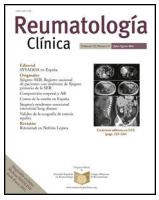
Reumatol Clin. 2020;16(6):497–498

Sociedad Española de Reumatología - Colegio Mexicano de Reumatología

Reumatología Clínica

www.reumatologiaclinica.org



Case Report

Chronic anterior uveitis in a patient with CREST syndrome☆

Sara Fuente Cosío,[a,*] Beatriz Arca Barca,[b] Paula Martínez García,[c] Antonio Sampedro López[d]

[a] *Medicina Interna, Hospital Comarcal de Jarrio, Coaña, Asturias, Spain*
[b] *Reumatología, Hospital Universitario San Agustín, Avilés, Asturias, Spain*
[c] *Medicina Interna, Hospital Universitario San Agustín, Avilés, Asturias, Spain*
[d] *Oftalmología, Hospital Universitario San Agustín, Avilés, Asturias, Spain*

ARTICLE INFO

Article history:
Received 11 July 2018
Accepted 18 October 2018

Keywords:
CREST syndrome
Systemic sclerosis
Uveitis

ABSTRACT

Systemic sclerosis is a connective tissue pathology with very heterogeneous clinical manifestations, associated in a small percentage with inflammatory eye diseases. In the specific case of uveitis, only isolated cases have been reported in the literature, especially in relation to the CREST syndrome. We present the case of a 53-year-old woman with CREST syndrome and chronic anterior uveitis, which we consider of clinical relevance given its low prevalence.

Uveítis anterior crónica en paciente con síndrome de CREST

Palabras clave:
Síndrome de CREST
Esclerosis sistémica
Uveítis

RESUMEN

La esclerosis sistémica es una conectivopatía con unas manifestaciones clínicas muy heterogéneas, relacionándose en un pequeño porcentaje con enfermedades oculares inflamatorias. En el caso concreto de las uveítis, únicamente se han descrito casos aislados en la literatura, sobre todo con relación al síndrome de CREST. Presentamos el caso de una mujer de 53 años, con síndrome de CREST y uveítis anterior crónica, que consideramos de relevancia clínica dada su baja prevalencia.

Introduction

Systemic sclerosis (SSc) is a connective tissue disease characterised by 3 pathological processes: fibrosis (essentially cutaneous and pulmonary), small vessel vasculopathy and specific autoantibodies.[1] It can be classified as diffuse and limited (lcSSc) depending on the extent and distribution of the skin manifestations. LcSSc is frequently associated with CREST syndrome (Calcinosis cutis, Raynaud's phenomenon, Esophageal dysmotility, Sclerodactyly and Telangiectasia).[2] In this syndrome, ocular involvement is infrequent, the case presented below is the fourth described in the literature on uveitis with CREST syndrome.[3–5]

Description of the case

A 53-year-old female, housewife, no family history of interest, no known drug allergies or toxic habits, with a history of Hashimoto's thryoiditis and multinodular goiter. Followed up by rheumatology for CREST syndrome and secondary Sjögren's syndrome, under treatment with nifedipine, esomeprazole, cinitapride and levothyroxine.

Complementary studies showed titre positive ANA (1/2560) centromere pattern and capillaroscopy with sclerodermal pattern. No anomalies were observed on chest X-rays, functional, respiratory and diffusion tests or transthoracic echocardiograph.

The patient attended ophthalmology consultations for repeated episodes of red eye, pain and loss of right eye vision. On examination, she presented a non-hyperaemic eye with some isolated cells inside the anterior chamber (Tyndall 1^{+} and negative flare),[6] synechias and pigment deposits in anterior crystalloids, together with cystoid macular oedema with visual acuity (VA) .5 in the right eye (RE); and was diagnosed with chronic anterior uveitis

☆ Please cite this article as: Fuente Cosío S, Arca Barca B, Martínez García P, Sampedro López A. Uveítis anterior crónica en paciente con síndrome de CREST. Reumatol Clin. 2020;16:497-498.

* Corresponding author.
E-mail address: sarafuentecosio@gmail.com (S. Fuente Cosío).

with secondary macular oedema. She was prescribed cycloplegic and topical corticosteroids (dexamethasone eye drops and subtenon's injection of triamcinolone acetonide) with progressive improvement. The patient continued to experience new flare-ups, coinciding with decrease and/or withdrawal of the corticosteroid; and therefore in approximately one year, oral methotrexate was started (15 mg/week). After 5 months it was necessary to increase the dose (20 mg/week/sc) and then discontinue it due to elevated transaminases after 2 months of treatment. She has remained stable since that time, 5 further flare-ups presented after that, and she started oral cyclosporine (50 mg/12 h) with improvement and clinical stabilisation until now (VA .7 in RE and .8 in LE).

Discussion of the case

Uveitis consists of inflammation of the uvea, the middle portion of the eye, comprising the iris and the ciliary body (anterior part) and choroid (posterior part); although, in general terms, it includes any eye inflammation. Its principal aetiologies are: infections, immune-mediated systemic diseases (40%), syndromes confined to the eye and idiopathic (up to 30%). Most notable among the immune-mediated aetiologies are spondyloarthritis, sarcoidosis, Behçet's syndrome and inflammatory bowel disease, among others.

As for the ocular manifestations of SSc, the most notable are the palpebral alterations due to fibrotic changes, which occur more frequently in young patients, with greater skin involvement and earlier onset of the disease. Other ocular manifestations frequently related to this disease are: pinguecula, keratoconjunctivitis sicca, cataracts and blepharitis. Inflammatory ocular manifestations have been described, although much less frequently: of interest is the recent description of 2 cases of CREST syndrome with retinal vasculitis.[7] We should also mention that in the study by Gomes et al.[8] only 4.4% (2 patients) episcleritis was found and no cases of uveitis were described. In fact, this combination (uveitis and SSc) is very rare and only isolated cases have been described in the literature: 3 in patients with CREST syndrome,[3–5] one in a patient with SSc[9] and another in a patient with SSc without sclerodermia.[10] although the frequency of this association is low, we believe that it should be taken into consideration when managing these situations in order to deliver multidisciplinary treatment in which ophthalmologists would play an important role.

Conflict of interest

The authors have no conflict of interest to declare.

References

1. Korn JH. Pathogenesis of systemic sclerosis. In: Koopman WJ, editor. Arthritis and allied conditions. 14th ed. Philadelphia: Lippincott Williams & Wilkins; 2001. p. 1643–54.
2. Varga J. Overview of the clinical manifestations of systemic sclerosis (scleroderma) in adults. UpToDate; 2018. Available from: http://www.uptodate.com [accessed 01 May].
3. Akman A. Granulomatous anterior uveitis in a patient with CREST syndrome. Ocul Immunol Inflamm. 2000;8:201–3.
4. Santos PS. Granulomatous uveitis, CREST syndrome and primary biliary cirrosis. Br J Ophthalmol. 2000;84:548–9.
5. Courtade M, Gicquel JJ, Mercie M, Vabres B, Dighiero P. Granulomatous uveitis and CREST syndrome: a case study. J Fr Ophtalmol. 2004;27:918–20.
6. Jabs DA, Nussenblatt RB, Rosenbaum JT, Standardization of Uveitis Nomenclature (SUN) Working Group. Standardization of uveitis nomenclature for reporting clinical data. Results of the first international workshop. Am J Ophthalmol. 2005;140:509–16.
7. Yang S, Kopplin LJ, Rosenbaum JT. Retinal vasculitis associated with CREST syndrome. Am J Ophthalmol Case Rep. 2018;10:185–8.
8. Gomes BAF, Santhiago MR, Magalhães P, Kara-Junior N, Azevedo MNL, Moraes HV Jr. Ocular findings in patients with systemic sclerosis. Clinics. 2011;66:379–85.
9. Bolad W. Anterior uveitis in a patient with scleroderma: a case report. Ocul Immunol Inflamm. 2013;21:11–2.
10. Borges T, Vilaça J, Ferreira S, Chora I, Silva S, Dias C. Systemic sclerosis sine scleroderma: a case report of anterior uveitis. Reumatismo. 2015;67:21–5.