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Short communication

Sclerodermatomyositis, ocular manifestations☆

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ABSTRACT

Background: Sclerodermatomyositis is an overlap syndrome of myositis and scleroderma, with dermatological, muscular and joint involvement, but may also present with ocular manifestations.
Clinical case: A 57 year-old woman presented with ophthalmological manifestations, including scleral thinning 360°, and the presence of cells in the anterior and posterior chamber. Oriented physical examination and laboratory studies led to the diagnosis, with the need for systemic treatment.
Conclusion: Sclerodermatomyositis is a rare disease. Its diagnosis needs thorough clinical and laboratory studies, and its management should be multidisciplinary when inflammatory ocular manifestations may be present.

Esclerodermatomiositis con manifestaciones oculares predominantes

Palabras clave:
Esclerodermatomiositis
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Anticuerpos antinucleares
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RESUMEN

Antecedentes: La esclerodermatomiositis es un síndrome de superposición que tiene manifestaciones dermatológicas, musculares y articulares, y que puede presentar afección ocular.
Caso clínico: Se presenta el caso de una mujer de 57 años en quien la exploración oftalmológica hizo evidente adelgazamiento escleral 360 grados, celularidad anterior y vítrea. La exploración física orientada y los estudios de laboratorio permitieron sustentar el diagnóstico de esclerodermatomiositis, por lo que se trató de forma sistémica.
Conclusión: La esclerodermatomiositis es una enfermedad rara, cuyo diagnóstico implica estudio clínico y de laboratorio, y su manejo debe ser multidisciplinario, donde las manifestaciones oculares inflamatorias pueden estar presentes.

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Introduction

Sclerodermatomyositis is an "overlap" syndrome, defined as having the concurrent presence of diseases that fulfill defined clinical data and diagnostic criteria independently, occasionally accompanied by specific serological tests.[1–4] The etiopathogeny of dermatomyositis has been linked to the deposit of immune complexes in vessels and is regarded as a supplement-regulated vasculopathy.[1,2]

Clinical expressions include symmetric proximal weakness, myalgia, myositis, Gottron sign (erythematous or purple plates appearing in the dorsal area of the interphalanx, metacarpian-phalanx, elbows and knee joints) and calcinosis.[1]

Some ocular expressions have been described including heliotrope erythema (hyperpigmentation, scaling and palpebral edema), avascular areas in the conjunctiva, uveitis, scleritis and glaucoma, as well as retinopathy with cotton-like lesions, retinal hemorrhages, retinal edema and optic nerve paleness.[5–8]

Diagnostic is based on clinical findings, data compatible with inflammatory myopathy and antibody pattern. The auto-antibodies reported in this conditions include anti-ribonucleoprotein (anti-RNP) which are associated with this overlap in variable percentages (generally under 30%) whereas nearly 90% of patients with scleroderma are positive to antinuclear antibodies (AAN), and 35% are positive for rheumatoid factor.[2,9,10] Treatment includes systemic glycocorticoids although generally an immunosuppressant agent is also required.

Clinic case report

Female, 57, who consulted due to poor vision and intermittent episodes of throbbing pain in both eyes during 20 years. She was diagnosed in another institution with idiopathic panuveitis and treated with topical steroids and midriatics with apparent clinical improvement. Two years prior to consulting at the authors' practice, the patient debuted with pain in proximal and distal inter-phalanx, metacarpo-phalanx and shoulder joints without morning stiffness and with proximal myalgia in thoracic limbs. In addition, she exhibited dyspnea with median effort and dysphagia against solids as well as lesions in the forehead which increased with exposure to the sun. Pathological personal history included hypertension with 7 years evolution.

Ophthalmological examination produced visual acuity in the right eye (RE) of 20/200 that improved to 20/80 and 20/100 in the left eye (LE) that improved to 20/80. Biomicroscopy revealed 360° scleral thinning, cornea with discrete peripheral thinning, anterior chamber with cellularity 2+ and clear lens in the RE (Fig. 1). The LE showed sclera without alterations, anterior chamber with cellularity 2+, posterior synechiae and clear lens (Fig. 2). RE ocular fundus did not show alterations whereas the LE ocular fundus identified 2+ vitreous cellularity (Fig. 3).

General physical examination found Gottron's papules, heliotrope erythema, changes suggesting Raynaud in the distal phalanx of hands, increased skin consistency in those areas, Heberden and Bouchard nodules and abnormal capillaroscopy (Fig. 4).

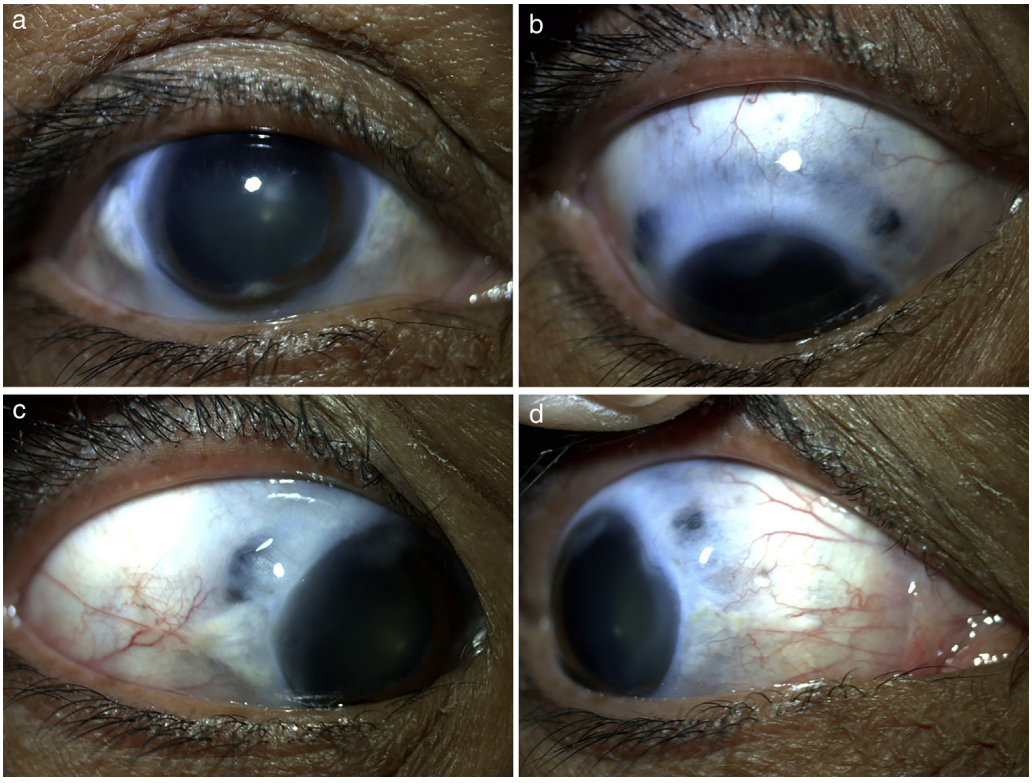

Fig. 1 – Clinic images of the right eye, showing scleral thinning.