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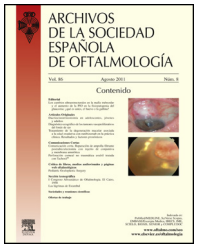

Short communication

Retinal vasculopathy in systemic lupus erythematosus: A case of lupus vasculitis and a case of non-vasculitis venous occlusion☆

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ABSTRACT

Clinical case: Two patients with systemic lupus erythematosus presented with vision loss and were diagnosed with retinal vasculopathy. Patient 1 had occlusive vasculitis with macular edema and retinal ischemia in the right eye. Corticosteroid therapy was increased and intravenous rituximab added. Intravitreal therapy and panretinal photocoagulation were performed. Patient 2 presented with a left central retinal vein occlusion without vasculitis but was on anticoagulation therapy due to having an antiphospholipid syndrome. Both patients maintained a stable visual acuity.

Discussion: Occlusive lupus retinal vasculitis has severe visual and systemic consequences (central nervous system vasculitis). It is crucial to differentiate it from standard vascular occlusion syndromes.

Vasculopatía retiniana en el lupus eritematoso sistémico: caso de vasculitis lúpica y caso de oclusión venosa no vasculítica

Palabras clave:
Lupus
Vasculitis
Oclusión venosa
Rituximab
Síndrome antifosfolípido

RESUMEN

Caso clínico: Dos pacientes con lupus eritematoso sistémico consultaron por pérdida visual y fueron diagnosticadas de vasculopatía retiniana. La paciente 1 presentó vasculitis oclusiva en ojo derecho con edema macular e isquemia retiniana. Se aumentó la inmunosupresión, añadiendo rituximab iv, y realizó terapia intravítrea y panretinofotocoagulación. La paciente 2 presentó oclusión venosa central retiniana izquierda no vasculítica y fue anticoagulada por presentar síndrome antifosfolípido. Ambas pacientes mantuvieron su agudeza visual estable.

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Discusión: La vasculitis oclusiva lúpica retiniana puede ocasionar graves repercusiones visuales y sistémicas (vasculitis cerebral). Es importante diferenciarla de la oclusión vascular común.

Introduction

Systemic lupus erythematosus (SLE) is a chronic disorder of unknown etiology which mainly affects young females. It is based on anomalous self-immune reactions with lymphocyte-B hyperactivity and lymphocyte-T deregulation which form circulating immunocomplexes and histological injury. At the ophthalmological level,[1] SLE can express with orbitopathy, dry syndrome, scleritis, anterior and/or posterior uveitis, optic neuritis and oculomotor palsy, among others. Retinal involvement (up to 10%) includes lupic retinopathy, occlusive vasculitis, central or branch arterial occlusions, central or branch venous occlusions and exudative retina detachment due to lupic choroidopathy. Up to 30% of lupus patients can exhibit anti-cardiolipin antibodies and lupic anticoagulants, although only 50–70% of these develop thrombosis and antiphospholipid syndrome at 20 years. This study reports two cases of patients affected by SLE exhibiting retinal vascular occlusion, analyzing the clinic and management thereof.

Clinical case

Patient 1

A female patient, age 51 years, was diagnosed with SLE 20 years ago with cutaneous onset. Treatment consisted of oral prednisone (25 mg/day), mycophenolate mofetil (720 mg/day) and pentoxifylline (800 mg/day). No antiphospholipid antibodies were present. She visited due to visual loss (finger counting at 1 m) in right eye starting 2 months earlier, with visual acuity of 1 in the left eye. She also presented posterior subcapsular cataracts in both eyes and bilateral intraocular pressure of 15 mmHg. Right eye funduscopy identified multiple flame-blurry intraretinal hemorrhages in posterior pole, macular edema with foveal hemorrhage and cotton-like exudates, as well as arteriole thinning with sheathing (Fig. 1). Fluorescein angiography (FA) revealed macular ischemia, diffuse vasculitis with capillary amputation and peripheral ischemia (Fig. 2). Optic coherence tomography (OCT, Zeiss Cirrus HD-OCT) confirmed cystic macular edema (Fig. 3). The left eye exhibited 3 cotton-like exudates at the level of the inferior vascular arch. The case was approached as lupic retinal occlusive vasculitis,[2] intensifying immunosuppression in order to protect the left eye from a similar condition. In treatment, 50 mg/day prednisone was administered in descending amount as well as 1 g intravenous rituximab (2 administrations separated by 15 days). At 2 months follow-up, the patient developed capillary neovascularization in the right eye which required panretinophotocoagulation for stabilization (Fig. 4). The macular edema was resolved with intravitreal therapy, first with 0.1 ml (4 mg) triamcinolone and subsequently with 0.05 ml (1.25 mg) bevacizumab. At 20 months follow-up visual acuity remains stable.

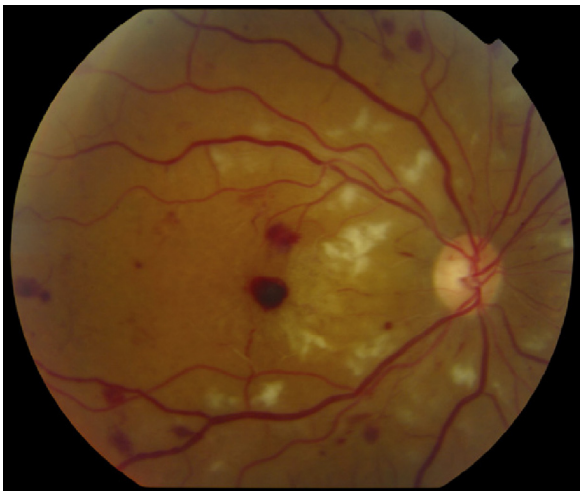

Fig. 1 – Ocular fundus at onset.

Patient 2

A female patient, age 60 years, was diagnosed with SLE 33 years ago with nephropathy and dry syndrome. She also had advanced lupus nephropathy with arterial hypertension. Treatment consisted of 15 mg/day prednisone, 1800 mg/day myofenolate mofetil enalapril and acetyl-salicylic acid. She was in anti-aggregation due to positive analysis for antiphospholipid antibodies. She visited due to visual loss (visual acuity of 0.5 in the right eye and 0.7 in the left eye) with onset 2 weeks earlier. She also presented posterior

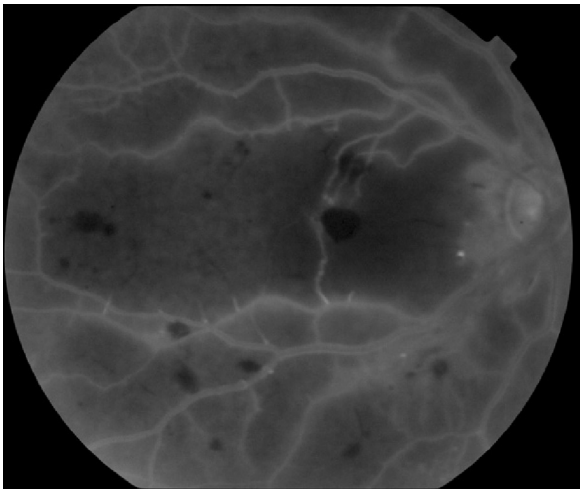

Fig. 2 – Right fluorescein angiography showing diffuse vasculitis with vascular amputation and retinal ischemia.