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.

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

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 intravitrea 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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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 1g 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.

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 mycophenolate 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

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

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subcapsular cataracts in both the eyes and intraocular pressure of 17/14 mmHg. The right eye fundus was normal, but the left exhibited extensive peripapillary and dotted flame hemorrhages in posterior pole, some cotton-like exudates in proximal vascular arches, venous tortuosity and optic nerve congestion (Fig. 5). OCT revealed no signs of macular edema. FA did not detect retinal ischemia or signs of vasculitis (Fig. 6). The case was approached as visual loss due to cataracts with concomitant left retinal central venous occlusion of non-ischemic onset. Acenocoumarol anticoagulants were prescribed. After 5 months follow-up the patient's visual equity is stable.

Discussion

Retinal occlusive vasculitis is a severe expression of SLE which could bring about irreversible visual loss and significant systemic sequels (association with brain vasculitis). Any unexplained visual loss in patients affected by SLE should be investigated. It is important to differentiate between occlusive arterial or venous vasculitis due to inflammation and venous or arterial vascular occlusion without inflammation. To this end, detailed ocular examination and SA are crucial. Non-vasculitis occlusion is associated to patients affected by antiphospholipid syndrome and other cardiovascular risks such as arterial hypertension (advanced nephropathy cases), and its management does not require increase of immunosuppression. However, these patients have a tendency to develop thrombosis and anti-aggregates or anticoagulants are advised.

Occlusive vasculitis management involves strict control of inflammation. Rituximab, the anti-CD20 monoclonal antibody (lymphocyte-B specific), is initially used in lympho proliferative disorders but with extension for example for uveitis due to Behçet disease. Its application in SLE is

Fig. 3 – Right macular optic coherence tomography, showing macular edema and foveal hemorrhage.

Fig. 4 – Right eye fundus at 2 months of onset, showing the resolution of cotton-like exudates and smaller hemorrhage components, but development of optic disk neovascularization.

Fig. 5 – Left eye fundus at onset, showing posterior visualization due to opacity of posterior subcapsular environment.
infrequent and recent. It is reserved for patients with symptoms which cannot be controlled with standard immunosuppressant therapy, for example, patient 1. In this case, intravenous rituximab was able to control her vasculitis in the right eye and prevent the involvement of the other eye. However, more studies are required for an in-depth knowledge of the role and hierarchy of rituximab in the therapeutic scale of SLE.

Fig. 6 – Left fluorescein angiography: no signs of vasculitis or ischemia can be observed.

Conflict of interest

No conflict of interest has been declared by the authors.

REFERENCES