Short communication

Churg–Strauss syndrome associated with antiphospholipid antibodies in a patient with retinal vasculitis


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ABSTRACT

Case report: We present the case of a 69-year-old woman with unilateral retinal vasculitis. Investigations showed asthma, rhinosinusitis, nasal polyposis, peripheral blood eosinophilia, increased sedimentation rate, proteinuria, and antiphospholipid antibodies. Anti-neutrophil cytoplasmatic antibodies (ANCA) were negative.

Discussion: Although her anti-neutrophil cytoplasmatic antibody (ANCA) status was negative, taking into account the other clinical and laboratory features, retinal vasculitis was thought to be an ocular manifestation of Churg-Strauss syndrome. Treatment was started with high-dose corticosteroids and anticoagulant therapy.

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Síndrome de Churg-Strauss asociado con anticuerpos antifosfolipídicos en un paciente con vasculitis retiniana

RESUMEN

Caso clínico: Presentamos el caso clínico de una mujer de 69 años, con una vasculitis retiniana unilateral. El estudio del caso mostró la presencia de asma, sinusitis con pólipos nasales, eosinofilia en sangre periférica, aumento de la velocidad de sedimentación, proteinuria y anticuerpos antifosfolipídicos. Los anticuerpos antineutrófilos citoplasmáticos fueron negativos.


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Introduction

The Churg-Strauss syndrome (CSS) is a systemic vasculitis affecting small and medium-sized vessels, characterized by the presence of eosinophilia in peripheral blood and the formation of extravascular granulomas, with ocular involvement being very infrequent.1

CSS is a multisystemic disease characterized by asthma, peripheral eosinophilia, peripheral neuropathy, lung infiltrates and paranasal sinus alterations.2

The case of a patient with CSS and retinal venous vasculitis, negative antineutrophil cytoplasmic antibodies (ANCAs) and positive antiphospholipid antibodies is described. Several months after retinal occlusion, the patient developed other expressions of the disease such as multiple mononeuritis and ischemic colitis.

The presence of said antiphospholipid antibodies is not a frequent finding in association with CSS. Very few published cases have been found.

Clinic case report

Female, 68, referred by the emergency Dept. due to loss of visual acuity (VA) in the right eye (RE) with several weeks evolution. Relevant personal antecedents comprised breast carcinoma treated 10 years earlier, arterial hypertension in treatment, late onset asthma and nasal polyposis.

Discusión: A pesar de la ausencia de anticuerpos antineutrófilos citoplasmáticos, se consideró el diagnóstico de un síndrome de Churg-Strauss, dadas las otras manifestaciones clínicas y de laboratorio, iniciándose un tratamiento con altas dosis de corticoides y anticoagulantes.

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Fig. 1 – RE retinograph showing inferior temporal venous occlusion with large exudative and hemorrhagic component.

VA in RE was of 0.08 without improvement, while the left eye (LE) VA was of 0.4, improving up to 1.0. Biomicroscopy did not show alterations and intraocular pressure (IOP) was of 16 mmHg in both eyes (BE).

Ocular fundus (OF) revealed an occlusion which seemed to involve the inferior temporal venous branch with large exudative and hemorrhagic components in the macula (Fig. 1), corresponding to edema and retina thickening in optic coherence tomography (OCT-SD) (Topcon 3D OCT-1000, Tokyo, Japan) (Fig. 2). Fluorescein angiography (FA) evidenced the

Fig. 2 – RE OCT.
existence of an inferior temporal vein occlusion of vasculitic type that respected the peripheral sections of the vessel, associated to intense ischemia (Fig. 3).

The lab tests reported peripheral blood hyper-eosinophilia of 18.5% (normal up to 5%), with absolute values of $1.28 \times 10^9/l$ (reference values: $0.05-0.5$); erythrocyte sedimentation rate (ESR) 64 mm/h; negative ANCA and ANA; positive anti-cardiolipin antibodies and proteinuria. The serological study produced normal results.

After considering CSS diagnostic, it was decided to carry out photoocoagulation of the macular area involved by the edema secondary to occlusion (Figs. 4 and 5) and the patient was referred to the collagenosis unit, where she was treated with 60 mg of deflazacort orally, intravenous boluses of cyclophosphamide and Sintrom®. In addition, the patient was referred to ENT where the polypoid chronic rhinosinusitis diagnostic was confirmed.

Four months after the first visit, the patient exhibited a condition compatible with multiple mononeuritis. Sural nerve biopsy confirmed the existence of necrotizing vasculitis in a perineural artery. Approximately 4 weeks later and as a consequence of the digestive hemorrhage, endoscopy and biopsy were performed, observing distant colitis with ischemic alterations. Finally, after macular photoocoagulation and immunosuppressant treatment, RE VA was of 0.3 (the patient referred the RE as the amblyopia eye).

**Discussion**

CSS is a form of primary vasculitis characterized by allergy and angitis, its most characteristic expressions being the presence of bronchial asthma, tendency to cardiac and peripheral nervous system involvement and marked eosinophilia in peripheral blood and tissue.³

The present case exhibited 4 out of 6 diagnostic criteria of the American College of Rheumatology, i.e., asthma, eosinophilia above 10%, paranasal sinus involvement and multiple mononeuropathy. Says number is sufficient to consider the diagnostic, including biopsy with necrotizing vasculitis in a perineural artery.⁴

The present patient did not exhibit positive ANCAs. However, these only appear in 40–60% of patients with CSS.⁵ Even though some authors have attempted to classify CSS on the basis of positive or negative ANCAs, the presence or absence of said antibodies does not affect the prognosis or therapeutic approach.⁶

The association between CSS and the presence of antiphospholipid antibodies is an infrequent occurrence. Very few published cases have been found in the literature, and none reporting ophthalmological involvement and said association. The simultaneous existence of both entities could have facilitated the ischemic alterations of the patent, adding severity to the disease. For this reason it became important to look for the presence of antiphospholipid antibodies in CSS and establish anticoagulant prophylaxis to prevent thrombotic episodes.²

CSS is associated to a large variety of ocular expressions that include granulomatous conjunctivitis, retinal vascular occlusions and optic neuropathies which on some occasions could be the first expressions of the disease. Therefore, it is important to take into account said disease in ocular vasculitis cases.

Treatment should be initiated as soon as possible to prevent other systemic expressions of the disease, as the prognostic thereof depends on the stage and the grade of involvement of the disease.⁷ Due to the extension of retinal

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**Fig. 3 – RE fluorescein angiography. (a) Ischemic occlusion of the inferior temporal vein and macular edema and (b) vasculitic occlusion respecting peripheral sections.**
ischemia, early argon laser photocoagulation was performed in the present patient in order to prevent the appearance of neovessels, without waiting for the corticoid treatment to have effects on the vasculitic condition.

**Conflict of interests**

No conflict of interests was declared by the authors.

**REFERENCES**


