Short communication

Non-arteritic anterior ischemic optic neuropathy as first manifestation of antiphospholipid syndrome in a young patient

M. Serrador-García*, E. Santos-Bueso, F. Sáenz-Francés, J.M. Martínez-de-la-Casa, J. García-Feijoo, J. García-Sánchez

Unidad de Neurooftalmología, Servicio de Oftalmología, Hospital Clínico San Carlos, Madrid, Spain

Article history:
Received 18 February 2013
Accepted 2 July 2013
Available online 26 September 2014

Keywords:
Non-arteritic anterior ischemic optic neuropathy
Antiphospholipid syndrome
Anticardiolipin antibodies
Hypercoagulability
Young patient

Abstract

Case report: We report the case of a young patient with unilateral anterior ischemic optic neuropathy, with no known cardiovascular risk factors and visual acuity, preserved with positive anticardiolipin antibodies as a unique find.

Discussion: Non-arteritic anterior ischemic optic neuropathy in the context of antiphospholipid syndrome is an uncommon finding, but it must be considered in the diagnosis of the atypical anterior ischemic optic neuropathy.

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Neuropatía optica isquémica anterior no arterítica como primera manifestación de síndrome antifosfolípido en un paciente joven

Resumen

Caso clínico: Se presenta el caso clínico de un paciente joven con neuropatía óptica anterior isquémica unilateral sin factores de riesgo cardiovascular conocidos y agudeza visual conservada con anticuerpos anticardiolipina positivos como único hallazgo.

Discusión: El inicio clínico como neuropatía isquémica anterior no arterítica en el síndrome antifosfolípido es un hallazgo infrecuente, pero debe tenerse en cuenta en el diagnóstico de la neuropatía óptica anterior isquémica atípica.

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Introduction

The most frequent cause of unilateral papillary edema in patients over 45 is anterior ischemic optic neuropathy (AION) which represents a multifactor ischemic event of the optic nerve (ON) and produces acute ischemia in the optic disk circulation, mainly in the posterior ciliary arteries. The pathogenicity of non-arteritic (NA) AION has been linked to temporary hypoperfusion of the ON head, which may present in typical or atypical form with different diagnostic approaches.1,2

The antiphospholipid syndrome (APS) is a self-immune disease which produces a state of hypercoagulability in which any organ can be compromised. Ocular involvement can occur in between 8% and 88% of patients and could be the first

Fig. 1 – LE funduscopy: unilateral papilla edema in LE with flame hemorrhage at 3 o’clock.

Fig. 2 – OCT spectrals in axon mode: significant thickening of the retina nervous fiber layer at the overall level as well as sector level in LE.
clinical expression. Early diagnosis of the syndrome by ophthalmologists or clinic physicians could prevent the increase of the systemic and neurological disease.

Clinic case

Male, 49 years, who visited the Emergency Service referring blurred vision in the lower hemifield of the left eye (LE) with 48 h of evolution. The patient did not exhibit relevant ophthalmological antecedents while at the general level he referred having been diagnosed with gastric ulcer treated with proton pump inhibitors. Ophthalmological examination revealed a visual acuity of 1.2 in both eyes and intraocular pressure within normal range. Biomicroscopy did not show significant alterations, without relative afferent pupil defect or pain with extraocular movements. Relevant findings of funduscopy were a unilateral papillary sectional edema in LE with flame hemorrhage at 3 o’clock (Fig. 1). Optic nerve fiber layer was analyzed with the optic coherence tomograph (OCT spectralis in axonal mode, software version 5.3). Heidelberg Engineering, Heidelberg, Germany, which revealed ON edema in LE (Fig. 2), while campimetry (OCTOPUS 1-2-3, Stimuli 3, INTERZEAG AG, Switzerland) revealed inferior altitudinal defect in LE (Fig. 3).

Urgent analysis was requested which did not exhibit alterations in basic hemogram study, coagulation or biochemistry, including acute phase reactants. Subsequently, analytics screening for infections was performed and serology for Borrelia burgdorferi, Rickettsia conorii, human immunodeficiency virus, chickenpox and syphilis was requested. All were negative except IgG for chickenpox. In addition antidiardipin antibody determination was requested, which was positive in title of 79.9 for IgG (1.4–20.0), confirmed with a second determination 12 weeks later. Magnetic angioresonance did not determine significant alterations.

Treatment was initiated with clopidogrel (Plavix®, Sanofi Aventis S.A., Barcelona, Spain) but was voluntarily suspended due to gastric history. After one year of follow-up the patient did not exhibit new episodes, with optic nerve fiber layer OCT revealing reduction in the superior-nasal quadrant of the LE after 3 months (Fig. 4) with persistence of inferior altitudinal defect in campimetry.

Discussion

The diagnosis of APS requires the definition of specific clinical and laboratory criteria. Thromboembolic episode must occur within 5 years of a positive lab analysis, which must be confirmed 12 weeks later. The APS treatment is based on the assessment of the thrombotic risk for the patient, which will be determined by previous thrombosis history, type of thrombotic episode and immunological profile. Initially, treatment consists in reducing modifiable cardiovascular risk factors. Most authors agree that the initial episode must be treated with warfarin for venous thrombosis and antiplatelets for arterial episodes. Numerous retrospective studies recommend prolonged anticoagulation, which becomes indefinite in the case of APS. APS can debut with ocular compromise, with early detection and treatment being the best way to prevent greater systemic morbidity and mortality. Generally, patients present
Fig. 4 – OCT ONFL LE: significant reduction in the mean thickness of the retina nervous fiber layer and of the superior and nasal quadrants after 3 months.

with visual symptoms such as blurred vision, temporary amaurosis or scotoma and visual field defect. Other symptoms pointed out in the literature include reddening and pain.\textsuperscript{4,5} However, the authors have not found in the literature cases similar to those described herein, diagnosed in males with APS of the “primary” type, i.e., without the coexistence of other self-immune diseases, with clinical onset at the
ophthalmological level in the context of AION-NA without neurological clinic, in contrast with the most frequent presentation in other described cases of AION in the context of APS. Described cases referred to females and in some cases included the coexistence of factor V Leyden mutation, treated with oral anticoagulation or corticoids, with evident clinic improvement.\textsuperscript{6,7}

Accordingly, APS must be considered in the differential diagnostic of AION-NA in young patients and in the absence of traditional risk factors, particularly when etiology is uncertain. The patient must be referred to the hematologist for systemic treatment to prevent the progression of the disease, as ocular episodes are associated with an increased risk of brain episodes. In addition, comprehensive ophthalmological follow-up must be carried out in order to immediately detect and treat any neovascular complication.\textsuperscript{1,8}

\textbf{Conflict of interests}

No conflict of interests has been declared by the authors.

\textbf{References}