Short communication

Early treatment of idiopathic vasculitis, aneurysms and neoretinitis (IRVAN). A case report

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A B S T R A C T

Case report: A 55-year-old woman presented with retinal vasculitis, multiple aneurysms, macular exudation and widespread retinal nonperfusion and was diagnosed with IRVAN. She was treated with panretinal laser photocoagulation. After 3 years of follow-up visual acuity remains stable and there are no complications due to ischemic sequelae.

Discussion: IRVAN syndrome with neovascularization can progress rapidly despite laser treatment. Panretinal laser photocoagulation has to be considered in the early stages as it is effective in stopping the progression of ischemia.


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T r a t a m i e n t e   p r e c o z   d e   v a s c u l i t i s   i d i o p á t i c a ,   a n e u r i s m a s   y   r e t i n i t i s   ( i r v a n ) .
A propósito de un caso

R E S U M E N

Caso clínico: Se presenta el caso de una mujer de 55 años con vasculitis retiniana, aneurismas arteriales múltiples en el polo posterior, exudación macular e hipoperfusión periférica bilateral (IRVAN) tratada precozmente con panfotocoagulación periférica. Tras 3 años de seguimiento la agudeza visual permanece estable y no hay progresión de la isquemia.

Discusión: El síndrome de IRVAN con neovascularización es difícil de controlar y el riesgo de progresión a glaucoma neovascular, a pesar del tratamiento, es alto. La panfotocoagulación en estadios iniciales es eficaz para controlar la isquemia y debe realizarse lo antes posible.

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Introduction

The IRVAN syndrome is a rare vascular disease described by Chang in 1995, characterized by idiopathic retinal vasculitis, posterior pole multiple arterial aneurysms and peripheral capillary occlusion. It expresses in young and healthy individuals, preferably females. Visual prognosis is uncertain and even though some self-limited cases have been described with good visual acuity, peripheral ischemia and neovascularization are aggressive.

Clinic case

Caucasian female, 55, without relevant pathological history, who visited the center in January 2008 due to diminished visual acuity in the left eye. Best corrected visual acuity was of 20/20 in the right eye and 20/60 in the left eye. Anterior segment exploration and tonometry did not show alterations. Ocular fundus examination revealed multiple large and medium-sized aneurysmatic dilatations in arteries and vasculitis in both eyes (Figs. 1 and 2). The left eye exhibited lipidic exudation in the macular area (Fig. 2). Fluorescein angiography revealed large areas of peripheral ischemia without neovascularization and generalized vasculitis (Fig. 3). Optic coherence tomography (OCT) showed macular lipidic deposits and diffuse macular edema.

Detailed systemic study was carried out with hemogram, glycemia, urine analysis, proteingram, ANCA, ANA, FR, VSG, ECA and cranial MR to discard associated pathology, which gave negative results. Relatives of the patient did not refer vascular malformation history.

According to the new classification proposed by Samuel, the patient was diagnosed with IRVAN syndrome in stage 2.

Discussion

An IRVAN syndrome case in initial stage, treated early prior to the development of neovascularization with peripheral panretinal photocoagulation which remains stable with a three-year follow-up period. After applying the treatment, lipidic exudation in the left eye posterior pole diminished and visual acuity improved (Fig. 5). Even though some authors have described aneurysm regression after treatment, the number and size of aneurysm-like dilatations did not exhibit variations.

Samuel et al. defended the need of early ischemia treatment in order to control the progression of the disease. The case presented herein involves favorably after treatment with peripheral panretinal photocoagulation in initial stages, thereby supporting said theory.

In advanced stages of the disease (stages 3, 4 and 5) the visual prognosis is uncertain despite the treatment and there is high risk of developing complications secondary to retinal ischemia. Due to the poor results obtained with standard treatments in these patients, some authors have utilized intravitreal anti-VEGF (ranibizumab and bevacizumab) prior to laser coagulation with good results.
Fig. 3 – Angiofluoresceingraph showing increased distal vascular patency.

Fig. 4 – Control retinograph after 3 years. Diminished lipidic exudation in the left eye macular area and stability of vascular lesions. Confluent laser coagulation impacts can be observed.

Fig. 5 – (A) Optic coherence tomography image of the macular area at diagnostic, showing intraretinal hyper-reflectiveness points corresponding to lipidic deposits and intraretinal edema. (B) Control optic coherence tomography 3 years later. Smaller subfoveal lipidic exudates and thickened posterior hyaloids.
Conflict of interests

No conflict of interests has been declared by the authors.

REFERENCES