Short communication

Recurrent neurosensory macular detachment in carotid-cavernous fistula

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ARTICLE INFO

Article history:
Received 1 August 2013
Accepted 18 March 2014
Available online 13 August 2015

Keywords:
Carotid-cavernous sinus fistula
Dural arteriovenous fistula
Neurosensory detachment
Macular detachment
Serous macular detachment

ABSTRACT

Case report: A 46 year-old man was seen in the emergency department complaining of vision loss and exophthalmos in his right eye. He also complained of headache, diplopia of 4 months onset, and neurosensory detachment that resolved spontaneously the month before. The study revealed tortuous conjunctival and episcleral vessels and neurosensory macular detachment in his right eye. A carotid-cavernous fistula was confirmed by computed tomography angiography. The fistula closed spontaneously during the hospitalization. One month later, the neurosensory detachment disappeared again.

Discussion: Carotid-cavernous fistula should be included in the differential diagnosis of neurosensory macular detachments. These neurosensory detachments can resolve spontaneously 11 the fistula is closed.

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Desprendimiento neurosensorial macular recurrente en fistula carótido-cavernosa

RESUMEN

Caso clínico: Varón de 46 años, acudió a Urgencias por disminución de agudeza visual y exoftalmos en ojo derecho. Aquejaba cefalea, diplopia de 4 meses de evolución e historia de desprendimiento neurosensorial (DNS) resuelto espontáneamente un mes antes. Presentaba tortuosidad de vasos conjuntivales y episclerales y nuevo DNS macular derecho. La sospecha de fistula carótido-cavernosa quedó confirmada mediante angiotomografía computarizada (angio-TC). Durante su ingreso la fistula se cerró espontáneamente. Al mes, el DNS había desaparecido.


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Introduction

Carotid-cavernous fistulae (CCF) are abnormal communications between the arterial system and the cavernous sinus. The presentation of this disease is highly variable. Frequently, patients consulted due to headache and symptoms derived from orbital congestion, although they could also exhibit loss of vision.

A case report is presented of a patient with CCF who exhibited recurring macular neurosensory detachments (NSD) which were resolved completely after the spontaneous closure of the fistula.

Case report

Male, 46, with history of vitiligo and recent dental infection who presented with cephalgia, tinnitus and diplopia with one month evolution. Visual acuity (VA) was 1 in both eyes, and biomicroscopy did not reveal alterations. Ocular fundus (OF) showed parafoveal retina pigment epithelium detachment (PED) in the right eye (RE) (Fig. 1). Cranial computerized tomography was requested to discard complicated acute middle ear keratitis (Gradenigo syndrome) with negative results.

One month later, the patient visited due to scotoma in RE and worsening diplopia. VA was 0.2 in RE and 1 in left eye (LE). Anterior segment did not exhibit anomalies, while funduscopy showed macular NSD in RE. Optic coherence tomography and fluorescein angiography were performed (Cirrus-OCT®, Carl-Zeiss Meditec Inc., Dublin, CA) (Fig. 2) which confirmed exploration findings. The patient was diagnosed with central serous chorioretinopathy. Full analyses were requested, including VSG and PCR. At the three-week checkup the patient referred improvement in symptoms, RE VA was of 0.5 while OF showed spontaneous subretinal fluid reduction (Fig. 3).

Two months later, the patient experienced a new worsening of diplopia and vision, exhibiting ptosis, exophthalmos and ophthalmoplegy in the RE. VA was 0.1 in the RE and 1 in the LE. Biomicroscopy revealed medusahead conjunctival vessels while the right OF showed reappearance of PED, NSD and macular intraretinal fluid without peripheral alterations (Fig. 4).

Emergency angio-CT confirmed the presence of right CCF (Fig. 5). During admission for embolization the fistula closed spontaneously, making treatment unnecessary. One month

Fig. 1 – Optic coherence tomography centered on the right eye fovea in the first visit showing three retina pigment epithelium detachments.

Fig. 2 – (A) Right eye fluorescein angiography in late phase, coinciding with the loss of vision in the same eye. Contrast leak can be observed in relation to retina pigment epithelium detachments and neurosensory detachment in the perimacular area. (B) Right eye optic coherence tomography centered on the fovea at the same time, showing neurosensory detachment in the macular area together with subfoveal pigment epithelium detachments.
after diagnostic, VA in the RE was of 0.5 with another spontaneous complete remission of PED and NSD (Fig. 6).

**Discussion**

CCF are anomalous communications established between the carotid system and the cavernous sinus. They are classified according to etiology (traumatic or spontaneous), to hemodynamic properties (high or low flow) and anatomy (direct or indirect).1,2

Typical expressions are periocular pain, conjunctival and episcleral vessel tortuosity, chemosis, proptosis and diplopia.2 In addition, open angle glaucoma has been described secondary to increased episcleral venous pressure3,4 as well as neovascular glaucoma. In addition, retinopathy due to venous stasis has been reported.5 Central retinal vein obstructions5,6 retina detachments,5,7 macular serous detachments,8 choroidal detachments5,5,9 and optic neuropathies.5 The arterialization of the orbital veins which takes place in CCF can cause venous stasis. This could give rise to choriocapillary hypoxia and alteration in the pumping function of the retina pigment epithelium. Retina pigment epithelium dysfunction leads to retina detachments.10 The present case illustrates the reversibility of this situation once hemodynamic stability is restored.

**Fig. 3** – Right eye optic coherence tomography centered on the fovea, taken three weeks after the one shown in Fig. 2. Spontaneous reduction of subretinal fluid and neurosensory detachment can be observed.

**Fig. 4** – Right eye optic coherence tomography centered on the fovea, taken two months after that of Fig. 3, during the recurrence of clinical worsening: recurrence of the neurosensory detachment of pigment epithelium detachments and intraretinal fluid can be observed.

**Fig. 5** – Computerized angiotomography at orbital level, evidencing right side exophthalmos, upper orbital vein thickening, asymmetry of the right cavernous sinus and venous engorgement of the orbital drainage at the level of the cavernous sinus.

**Fig. 6** – Right eye optic coherence tomography centered on the fovea one month after the spontaneous closure of the fistula, showing complete spontaneous resolution of the neurosensory detachment and normal foveal morphology.
The differential diagnostic of macular serous detachments should include central serous chorioretinopathy. This disease was suspected due to the angiographic characteristics of the retinal lesion and to the absence of other typical expressions of the CCF at onset. Other differential diagnostics would include exudative macular degeneration, idiopathic polypoid vasculopathy, optic nerve fossae and uveal effusion syndrome. The Vogt-Koyanagi-Harada disease was also included in the differential diagnostic due to the patient’s vitiligo history, although the unilaterality and the absence of other typical diseases of this process led us to discard it.

The peculiarity of this case is the form of presentation and evolution. The only funduscopic findings in this patient were the presence of PED and NSD, without other retinal alterations. Very few cases of CCF with this form of presentation have been published as the majority associate peripheral retina and choroidal detachments. The second peculiarity is the evolution of the process, involving NSD improvements with diminished symptoms, as well as a recurrence of the process associated to clinical worsening. Finally, complete resolution of the macular serous detachment was observed after the spontaneous closure of the fistula, with VA improvement.

In conclusion, CCF should be included in the differential diagnostic of macular NSD, as it can disappear spontaneously upon closure of the fistula.

**Conflict of interests**

No conflict of interests was declared by the authors.

**REFERENCES**