Case report

Orbital arteriovenous fistula presenting with choroidal pulsations on optical coherence tomography

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ABSTRACT

Purpose: To describe a case of orbital arteriovenous fistula diagnosed based on choroidal pulsations on optical coherence tomography (OCT).

Observations: A 69-year-old female originally referred for evaluation of macular degeneration. During acquisition of OCT images, choroidal pulsations of the right eye were noted on the B-scan on the instrument display. The pulsations were not noted on gross or funduscopic examination. Fluorescein angiography was unremarkable. Indocyanine green angiography revealed engorged choroidal vasculature in the right eye. OCT angiography revealed relative tortuosity and dilation of the superficial and deep vascular complexes respectively. B-scan ultrasonography revealed orbital pulsations on the right. MRI imaging of the brain and orbits was unremarkable. Findings were attributed to a low-flow orbital arteriovenous fistula. The patient subsequently developed mild stasis retinopathy for which anti-VEGF therapy was initiated.

Conclusions and importance: Arteriovenous fistulas of the brain and orbit classically present with gross pulsatile proptosis among other clinical features. Low flow orbital fistulas may present with subtle choroidal pulsations only detectable on OCT.

1. Introduction

Arteriovenous fistulas (AVF) of the brain and orbit can result in a number of ocular findings. While carotid-cavernous fistulas (CCF) and dural cavernous fistulas are the most common types of such AVFs, a purely orbital AVF can rarely occur. The most common ocular findings in cases of AVF formation of the brain and orbit include pulsatile proptosis, arterIALIZATION OF conjunctival veins and retro-orbital pressure elevation. Other findings including eyelid swelling, chemosis, cranial nerve palsies, choroidal effusions, serious retinal detachment and retinal hemorrhages can also occur.1 A purely orbital AVF can also lead to orbital varix formation with associated thrombosis.1 Herein we present a case of an orbital AVF which was diagnosed based on findings on optical coherence tomography (OCT).

2. Case presentation

A 69-year-old woman was referred for evaluation of macular degeneration. Visual acuity was 20/20 in both eyes and intraocular pressure was normal. Blood pressure was 132/72 mm Hg. Anterior segment examination was unremarkable. Funduscopic evaluation revealed intermediate drusen and pigment mottling in the macula of both eyes (Fig. 1). During acquisition of spectral-domain OCT (SD-OCT) (Spectralis HRA + OCT, Heidelberg Engineering, Heidelberg, Germany) images, dramatic choroidal pulsations in the macula of the right eye were noted on the B-scan on the image acquisition display (Video 1). The pulsations were noted to dampen with gentle pressure on the eye and corresponded to the patient’s heartbeat. SD-OCT revealed greater subfoveal choroidal thickness in the right compared to the left eye during systole which thinned during diastole (Fig. 2). Fluorescein angiography was unremarkable. Indocyanine green angiography revealed engorgement of the choroidal vasculature of the right eye. Investigational OCT angiography (OCTA) (Spectralis HRA + OCT, Heidelberg Engineering, Heidelberg, Germany) revealed subtle tortuosity of the superficial vascular complex and subtle engorgement of the deep muscular complex in the right eye compared to the left (Fig. 3). B-scan ultrasonography revealed subtle orbital pulsations of the right orbit but no clear mass (Video 2). The patient denied any history of trauma or orbital surgery. The patient underwent magnetic resonance imaging (MRI), venography (MRV) and angiography (MRA) which were un-revealing apart from slight enhancement of the right orbital soft tissue. Specifically, there was no abnormality or difference in the caliber of the superior ophthalmic vein draining the right or left eye (Fig. 3). Repeat careful examination of the globe and fundus did not reveal any

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detectable pulsations though they persisted on OCT. After conferring with radiology, a low-flow orbital AVF was suspected though a small orbital arteriovenous malformation (AVM) could not be ruled out. Subsequent examination revealed a macular intraretinal hemorrhage and mild intraretinal fluid. Repeat FA and OCTA did not show evidence of choroidal neovascularization. These findings were suspected to be related to early stasis retinopathy and anti-VEGF therapy was initiated.

Supplementary video related to this article can be found at https://doi.org/10.1016/j.ajoc.2018.08.004.

3. Discussion

Few entities can cause unilateral ocular pulsations including AVFs, AVMs and a defect in the orbital roof causing pulsations of the brain to be translated to the orbit. In our case, MRI/MRA/MRV ruled out an obvious CCF, AVM, dural cavernous fistula or orbital wall defect. Computed tomography angiography (CTA) or MRA are the typical imaging modalities employed for detection of low-flow fistulas. Catheter angiography, however, remains the gold standard for detecting low-flow fistulas, but was not felt to be necessary in our patient given their minimal symptoms. Other orbital vascular malformations such as a cavernous hemangioma was not considered on the differential diagnosis as these lesions typically cause a mass effect resulting in proptosis without pulsations.

Orbital AVFs occur between the branches of the external or internal carotid arteries and venous tributaries of the superior or inferior ophthalmic vein. Similar to dural cavernous fistulas, orbital AVFs tend to be spontaneous and low flow with slow progression of symptoms. If, however, ophthalmic vein thrombosis occurs, the progression of symptoms could turn rapid. ICG angiography of the right eye showed dilated choroidal vasculature supporting the diagnosis of an AVF. AVFs can cause a number of vision threatening ocular complications including glaucoma, orbital congestion/compartments syndrome, stasis retinopathy and even central retinal vein occlusion and so prompt recognition is critical. While gross retinal vascular tortuosity was not evident on funduscopy in our patient, OCTA detected subtle retinal vascular tortuosity and engorgement in the affected compared to the uninvolved eye.

Management of AVFs are typically through endovascular techniques though spontaneous closure may occur. As our patient was asymptomatic, only management of the stasis retinopathy was undertaken. If, however, progressive stasis retinopathy with worsening macular edema were to occur, endovascular treatment with interventional neuroradiology may be necessary.

4. Conclusions

Low-flow orbital AVFs may present with choroidal pulsations on OCT. Given the potential for heralding a potentially life-threatening condition such as a CCF, care should be taken to ensure that patients with choroidal pulsations noted on OCT undergo appropriate neuroimaging.

Patient consent

As this case report did not present any unique patient identifiers, consent to publish this case was not obtained.

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Conflicts of interest

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.
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