Paradoxical Worsening of Ocular Symptoms after Spontaneous Closure of a Carotid Cavernous Fistula: Case Report

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Paradoxical Worsening of Ocular Symptoms after Spontaneous Closure of a Carotid Cavernous Fistula: Case Report

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L. Fernando Gonzalez, MD et al.: Paradoxical Worsening of Ocular Symptoms after Spontaneous Closure of a Carotid Cavernous Fistula: Case Report

We report an interesting case of a spontaneous occlusion of a carotid cavernous fistula (CCF) causing a paradoxical worsening of orbital symptoms. A 59-year-old female presented to our institution with conjunctival injection, raised intracranial pressures (ICP) and mild exophthalmos of her left eye. A digital subtraction angiography (DSA) showed a Type-D CCF draining into the left superior ophthalmic vein (SOV). The patient declined endovascular treatment. She presented 15 months later with acute exacerbation of her orbital signs and symptoms. A DSA showed no evidence of arteriovenous fistula, and a brain MRI was consistent with spontaneous thrombosis of the SOV. At her 2-week clinical assessment, the patient showed clinical improvement and her IOP were within normal limits. Spontaneous thrombosis of the SOV can trigger the obliteration of a CCF with possible paradoxical worsening of orbital symptoms. The gold standard of diagnosis and management is directed toward decreasing IOP.

Case Report

A 59-year-old female presented with conjunctival injection associated with elevated intracranial pressures (ICP) in the left eye despite the use of three topical antiglaucoma medications. She initially presented about one year earlier to her local ophthalmologist, who eventually referred the patient to a glaucoma specialist for unilateral IOP elevation and a progressive superior ophthalmic defect on automated perimetry. The patient was then referred to a neuro-ophthalmologist for further management. Visual acuity was 20/25 in each eye and mild left exophthalmos was present. A two prism diopter esotropia with limited abduction and supraduction of the left eye was noted, consistent with a type D lesion. Endovascular treatment was offered but the patient declined.

Fifteen months later, the patient presented with an acute exacerbation of her strabismus, proptosis, and ocular pain, which occurred overnight. On the exam, she was noted to have limited abduction and supraduction of the left eye associated with an elevated IOP of 45 mm Hg, a left afferent pupillary defect, mild ptosis, external ophthalmoplegia, and upper eyelid edema with minimal ecchymosis. Computerized tomography showed a prominent, hyperdense left SOV suggesting the presence of thrombus within the left SOV (Figure 1). A digital subtraction angiography (DSA) revealed a CCF draining into the left SOV with feeders from both the internal and external carotid arteries bilaterally (Figure 1), but predominately on the left side, consistent with a type D lesion. Endovascular treatment was offered but the patient declined.

Follow-up angiography at the time symptoms increased showing (A) left internal carotid artery injection with no evidence of fistula, (B) left external carotid artery injection with no evidence of arteriovenous fistula, and (C) left internal carotid artery injection lateral view showing the fistula. Digital subtraction angiography showing a prominent, hyperdense left SOV suggesting the presence of thrombus within the left SOV. A Type-D CCF draining into the left SOV was noted on automated perimetry, but the IOP had decreased to 20 mm Hg.

Discussion

We present a case of a woman with elevated IOP, conjunctival injection, optic neuropathy, and a documented CCF on initial cerebral angiography who then spontaneously obliterated the CCF with paradoxical worsening of her symptoms. In general, treatment of CCF is reserved for Class A lesions, in the presence of cortical venous drainage, or when ocular symptoms become significant, such as elevated intraocular pressure, decreased visual acuity, optic neuropathy, or external ophthalmoplegia. Multiple treatment algorithms have been recommended, but are beyond the scope of this report.

Spontaneous resolution of arteriovenous malformations (AVM) is extremely rare, with just a few case reports in the literature. Most had a hemorrhagic presentation. In Abdalla’s series a single vein was a common finding in 83% of their patients with spontaneous thrombosis. The proposed mechanism is a thromboembolic event within the AVM itself, although this has not been proven histologically.
Resolution of CCFs has been reported after angiography, where a clot developed during the procedure in the internal carotid artery, possibly occluding the arteriovenous connection in a similar mechanism as just described. Similar events have been described soon after gamma knife radiotherapy, also potentially secondary to a thromboembolic event from the angiogram used during the treatment planning, and not from an acute radiation effect.

Bukul et al.2 reported 2 patients with dural CCF causing severe clinical manifestations that spontaneously resolved before endovascular intervention. Unlike the present case, obliteration of the CCF was associated with a concomitant resolution of orbital signs and symptoms. Seroog and colleagues10 reported 2 patients with CCF that developed spontaneous thrombosis of the SOV with an acute worsening of symptoms. In contrast to our case, however, thrombosis of the SOV in these 2 patients was not associated with an obliteration of the fistula. Our case is therefore unique, since there was an acute worsening in the orbital signs and symptoms caused by a spontaneous thrombosis of the SOV and an angiographically documented complete cure of the CCF. Acute thrombosis of SOV with probable extension proximally into the cavernous sinus accounted for the resolution of the CCF. Since the SOV provides the major and only orbital venous outflow for the orbit, sudden worsening of orbital congestion manifests as an orbital compartment syndrome (OCS). In addition, since the orbital veins are valveless, some orbital drainage may occur in an anterograde fashion from the SOV to the facial venous system and inferriorly through connections with the pterygoid palatine venousplexus, even with an active CCF. Sudden thrombosis of the SOV may temporarily block off these alternate drainage routes.

Thrombosis of the SOV in all likelihood resulted in stagnation of abnormal blood flow within the cavernous sinus, precipitating the occlusion of the CCF, slow flow through the coagulation cascade, manifesting as thrombosis. Based on anatomic studies, the SOV in this particular case was the single major venous drainage for the orbit, resulting in acute orbital, IOP elevation from decreased episcleral venous outflow, and a congestive optic neuropathy. Once there is no visualization of the CCF on DSA, the endovascular options are limited. Despite the presence of severe orbital signs, the management of the OCS may be difficult. In most cases, the OCS is a transient event, markedly improving within 48 hours.7 The goal of OCS therapy in such situations is to “buy time” until orbital congestion resolves. Presumably, orbital venous outflow forms alternate drainage pathways during this time. Initially, topical anti-glaucoma medications are instituted along with intravenous mannitol. If this fails, a lateral canthotomy with cantholysis is performed, but even this may provide only temporary relief, since the OCS will recur as orbital soft tissue congestion fills the decompressed space.

Worsening of the orbital and ocular symptoms does not always represent persistence or progression of the arterio-venous fistula, as in this case Illustrates. In cases of presumed spontaneous SOV thrombosis, the use of DSA has been questioned,3 since the diagnosis of SOV thrombosis can be made with MRI. However, the MRI signal characteristic of thrombosis evolve over time and may be difficult to interpret accurately in the SOV. The clinician is then left in a quandary of “waiting out” a possible thrombosis and delaying DSA or proceeding with timely DSA to confirm thrombosis or treat a worsening CCF. Despite the inherent risks of DSA, we support the use of this modality in all cases of acute worsening of orbital signs, since spontaneous SOV thrombosis is a rare event and delay in definitive care in the face of an acute, severe OCS may result in permanent visual loss.

Conclusions
Paradoxical worsening ofocular symptoms in presence of complete obliteration of a CCF is extremely rare and possibly triggered by thrombosis of the SOV. Although DSA is the gold standard for diagnosis, there is no role for endovascular intervention and the management is focused on managing the acute orbitopathy and raised intracranial pressure.

References