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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Introduction
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 woman presented to our institution with conjunctival injection associated with elevated intraocular pressures (IOP) 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 who referred the patient to a glaucoma specialist for unilateral IOP elevation who then spontaneously obliterated the CCF with paradoxical worsening of her symptoms.

Case Report
A 59-year-old female with a type-D lesion.3 Endovascular treatment algorithms have been recommended, but are beyond the scope of this report. 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 (AVMs) is extremely rare, with just a few case reports in the literature. Most had a hemorrhagic presentation.1,5 In Abdulrauf’s6 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.7

Discussion
We present an interesting case of a type-D carotid cavernous fistula that closed spontaneously with a paradoxical worsening of the symptoms due to thrombosis of the superior ophthalmic vein (SOV). The authors also give directives for the management of these extremely rare cases.

Computerized tomography showing a prominent, hyperdense left SOV suggesting the presence of thrombus within the 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.1 Endovascular treatment was offered but the patient declined. Fifteen months later, the patient presented with an acute exacerbation of her scleral injection, 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. Computers, tomography showed a prominent, hyperdense SOV on the left side suggesting the presence of thrombus within the vein (Figure 2). A DSA, including both internal, external, and vertebral arteries, showed no evidence of arteriovenous fistula and no visualization of the SOV (Figure 3). Brain MRI on gradient echo (GRE) (Figure 4) sequence demonstrated a mixed signal with hyperintensity along the SOV on the left side consistent with intravenous thrombosis. Following DSA, IOP progressively decreased from 45 to 18 mm Hg without any changes to the topical glaucoma regimen and the afferent pupillary defect resolved over the next 24 hours. The external ophthalmoplegia and conjunctival injection persisted, but there was a marked improvement of exophthalmos and periorbital pain. On subsequent follow-up two weeks later, a subtle left afferent defect was noted on automated perimeter, but the IOP had decreased to 22mm Hg.

Figure 1
Digital subtraction angiography showing (A) right internal carotid artery injection lateral view, (B) right internal carotid artery anterior-posterior view showing the fistula, (C) left internal carotid artery injection AP view showing the fistula, (D) left external carotid artery lateral view showing a prominent superior orbital vein, and (E) left internal carotid artery injection showing the terminal branch of the SOV was visible beneath the superomedial eyelid skin. Funduscopic examination, demonstrated increased cupping in the superomedial eyelid skin. Funduscopic examination, demonstrated increased cupping in the superomedial eyelid skin. Note the straightening of the vessel with stent placement.

Figure 2
Computerized tomography showing a prominent, hyperdense left SOV suggesting the presence of thrombus within 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. DSA is the gold standard of diagnosis and management is directed toward decreasing IOP.
Resolution of CCSs 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.

Bukus et al.18 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. Sorgor and colleagues19 reported 2 patients with CCF that developed spontaneous thrombus of the SOV with an acute worsening of symptoms. In contrast to our case, however, thrombus 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 thrombus of the SOV and an angiographically documented complete cure of the CCF. Acute thrombosis of SOV with probable extension proximally into the cavernous sinuses accounts for the resolution of the CCF. Since the SOV provides the major and in some cases only venous outflow for the orbit, sudden worsening of orbital congestion manifests as an orbital compartment syndrome (OCS).16 In addition, since the orbital veins are valvesless, some orbital drainage may occur in an autotransfusion fashion from the SOV to the facial venous system and inferorly through connections with the pterygoplatine 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 sinuses, precipitating the occlusion of the CCF. This phenomenon is known as OCS, a direct consequence of thrombosis of the SOV. In anterior circulation, such thrombosis may temporarily block off the alternate cellular venous return, especially in patients with severe cardiac disease or a stroke that involves brain tissue. The clinical presentation may range from transient visual loss to frank visual acuity loss as a result of thrombosis. Complications of decompressive cranectomy (DC) include intracranial hypertension, edema, ischemia, and seizures.


Figure 4

MRI Gradient Echo sequence showing arrow demonstrates SOV compatible with thrombus within

thrombus of the SOV. Although DSA is the gold standard for diagnosis, there is no role for endovascular therapy and the management is focused on managing the acute ophthalmic hypertension and raised intracranial pressure.

References


