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 woman presented to our institution with conjunctival injection associated with elevated intraocular pressures (IOP) in the left eye. A digital subtraction angiography (DSA) revealed a 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. DSA is the gold standard of diagnosis and management is directed toward decreasing IOP.

Introduction

We present an interesting case of a type-D carotid cavernous fistula (CCF) 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.

Case Report

A 59-year-old female presented 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. On the exam, she was noted to have limited abduction and supraduction of the left eye with an elevated IOP of 45 mm Hg, a left afferent pupillary defect, mild ptosis, external ophthalmoplegia, and upper eyelid edema with minimal exophthalmos. Computerized tomography showed a prominent, hyperdense left SOV. The authors also noted the straightening of the vessel with stent placement and a left abducens nerve paresis. An enlarged cavernous sinus was visible beneath the superomedial eyelid skin. Funduscopic examination demonstrated increased cupping of the superomedial optic disc fiber layer with no evidence of arteriovenous fistula. The large SOV previously seen was not identified at this point.

Figure 1

Computerized tomography showing a prominent, hyperdense left SOV suggesting the presence of thrombus within the superomedial eyelid skin. Funduscopic examination demonstrated increased cupping of the superomedial optic disc fiber layer with no evidence of arteriovenous fistula. The large SOV previously seen was not identified at this point.

Figure 2

Computerized tomography showing a prominent, hyperdense left SOV suggesting the presence of thrombus within the superomedial eyelid skin.

Figure 3

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 external carotid artery injection lateral view showing the SOV. (D) Left internal carotid artery anterior-posterior view showing the fistula, and (E) left internal carotid artery anterior-posterior view showing the fistula. (F) Left external carotid artery lateral view showing the SOV. (G) Left internal carotid artery injection lateral view showing the fistula. (H) Left external carotid artery lateral view showing a prominent superior orbital vein. (I) Left external carotid artery injection lateral view showing the fistula, (J) left internal carotid artery injection lateral view showing the fistula, (K) left external carotid artery injection lateral view showing the fistula, (L) left external carotid artery injection anterior-posterior view showing the fistula, (M) left external carotid artery injection lateral view showing the fistula, (N) left external carotid artery injection lateral view showing the fistula, (O) left external carotid artery injection anterior-posterior view showing the fistula, (P) left external carotid artery injection lateral view showing the fistula, and (Q) left internal carotid artery injection lateral view showing the fistula. (R) Left internal carotid artery injection lateral view showing the fistula, (S) left external carotid artery injection lateral view showing the fistula, and (T) left internal carotid artery injection lateral view showing the fistula. (U) Left internal carotid artery injection lateral view showing the fistula, (V) left external carotid artery injection lateral view showing the fistula, and (W) left internal carotid artery injection anterior-posterior view showing the fistula. (X) Left internal carotid artery injection lateral view showing the fistula, (Y) left external carotid artery injection lateral view showing the fistula, and (Z) left internal carotid artery injection anterior-posterior view showing the fistula.
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 manner 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.

Bujak et al 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. Sergott and colleagues 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. One 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 accounts for the resolution of the CCF. Since the SOV provides the major anastomotic pathway, even a causally 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 antegrade fashion from the SOV to the facial venous system and inferiorly through connections with the pterygopalatine venous plexus, 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 results in stagnation of abnormal blood flow within the cavernous sinus, precipitating the occlusion of the CCF, slow flow to 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 conjunctival 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. 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 SOV 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 arteriovenous fistula, as in this case Illustrates. In cases of presumed spontaneous SOV thrombosis, the use of DSA has been questioned, 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 quandry 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 of ocular 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 therapy and the management is focused on managing the acute orbitopathy and raised intracranial pressure.

References