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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 (IOP) and mild exophthalmos of her left eye. A digital subtraction angiography (DSA) demonstrated 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 conjunctival injection associated 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 intracranial pressure, decreased visual acuity, optic neuropathy, or external ophthalmoplegia. Multiple treatment approaches have been tried, but are beyond the scope of this report.

Case Report

A 59-year-old female presented with conjunctival injection associated with elevated intracranial 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. 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 a limited abduction on the left eye was noted, consistent with a left abducens nerve paresis. An enlarged terminal branch of the SOV was visible beneath the superomedial eyelid skin. Funduscopic examination, demonstrated increased cupping of the optic nerve head. Based on these findings, the patient was 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 a limited abduction on the left eye was noted, consistent with a left abducens nerve paresis. An enlarged terminal branch of the SOV was visible beneath the superomedial eyelid skin. Funduscopic examination, demonstrated increased cupping of the optic nerve head. Based on these findings, the patient was referred to a neuro-ophthalmologist for further management.

Figure 1

Figure 2

Digital subtraction angiography showing (A) right internal carotid artery injection lateral view, (B) right internal carotid artery injection posterior view showing the fistula, (C) left external carotid artery injection AP view showing the fistula, (D) left external carotid artery lateral view showing a prominent superior ophthalmic vein, (E) left internal carotid artery injection showing the fistula and a large superior ophthalmic vein treated with a 3-stents telescoping technique. Note the straightening of the vessel with stent placement.

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 a large SOV previously seen was not identified at this point.

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 intracranial pressure, decreased visual acuity, optic neuropathy, or external ophthalmoplegia. Multiple treatment approaches have been tried, 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 Abdulrauf’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.

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. MRI of the head showed a dilated left SOV, exophthalmos, and an enlarged cavernous sinus. 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.

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. Computed tomography showed a prominent, hyperdense left SOV suggesting the presence of thrombus within the vessel (Figure 2). A DSA, including both internal, external, and vertebral arteriographs, 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 intravascular thrombus. 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 perisceral pain. On subsequent follow-up two weeks later, a subtle left afferent defect was noted on automated perimetry, but the IOP had decreased to 22mm Hg.

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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 intracranial pressure, decreased visual acuity, optic neuropathy, or external ophthalmoplegia. Multiple treatment approaches have been tried, but are beyond the scope of this report.

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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.

Bukh 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. Sergot 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 accounted for the resolution of the CCF. Since the SOV provides the major and in many cases only venous outflow of the CCF, slow flow triggers the coagulation cascade, manifesting as thrombosis. Based on the CCF; slow flow triggers the coagulation cascade, manifesting as thrombosis. Based on the MRI signal characteristic of thrombosis this case illustrates. In cases of presumed spontaneous SOV thrombosis the use of DSA has been questioned 13 since the diagnosis of SOV thrombosis can be made with MRI. However, the MR signal characteristic of thrombosis evolve over time and may be difficult to interpret accurately in the SOV. The clinician 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 of oculomotor 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