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

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Symptoms after Spontaneous Closure of Paradoxical Worsening of Ocular Symptoms

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 intracocular 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 and a progressive superior arcuate defect on automated perimetry. The patient 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 left SOV with feeders from both the internal and external carotid arteries was noted (Figure 1), but predominately on the left side, consistent with a type D lesion. Endovascular treatment was offered but the patient declined.

Case Report
A 59-year-old female presented with conjunctival injection associated with elevated intracocular pressures (IOP) of 45 mm Hg on the left side consistent with intravenous drainage, or when ocular symptoms become significant, such as elevated intracranial pressure, decreased visual acuity, optic neuropathy, or external ophthalmoplegia. Multiple treatment algorithms have been developed, 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 hemorragic presentation. In Abdala’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. The authors also give directives for the management of these extremely rare cases.

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 external carotid artery injection lateral view showing the prominent superior orbital vein, (D) left internal carotid artery anterior-posterior view showing the fistula. (E) left internal carotid artery injection with no evidence of fistula.

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 algorithms have been developed, 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 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 accounted for the resolution of the CCF. Since the SOV provides the major pathway during this time. Initially, topical anti-glaucoma medications are instituted along with intravenous nimodipine. If this fails, a lateral canthotomy with cantholysis is performed, but even this may provide only temporary relief, since the SOV will recoc 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 spontaneou s 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 SOV 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 therapy and the management is focused on managing the acute orbitopathy and raised intracranial pressure.

References