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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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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 AP view showing the fistula, (D) left external carotid artery lateral view showing a prominent superior orbital vein, (E) left internal carotid artery injection showing the fistula and a large superior ophthalmic vein treatment with a 3-stents telescoping technique. 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

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, raised intraocular 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 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

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

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 and a progressive superior arcuate 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 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

on the left side and thinning of the retinal nerve fiber layer was confirmed on ocular coherence tomography.

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.³ 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. Computerized 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 periocular pain. On subsequent follow-up two weeks later, a subtle left arcuate defect was noted on automated perimetry, but the IOP had decreased to 20mm 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.^{1,5} In Abdulrauf's¹ 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

Resolution of CCFs has been reported after angiography, where a clot developed during the procedure in the internal carotid artery,⁹ possibly occluding the arteriovenous connection in a similar mechanism as just described. Similar events have been described soon after gamma knife radiotherapy,⁸ also potentially secondary to a thromboembolic event from the angiogram used during the treatment planning, and not from an acute radiation effect.

Bujak et al⁴ 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 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 in many cases only venous outflow for the orbit, sudden worsening of orbital congestion manifests as an orbital compartment syndrome (OCS).² 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 triggers 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 orbitopathy, 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.¹⁰ The goal of

Figure 4
MRI Gradient Echo sequence showing (arrow) a hypointense SOV compatible with thrombosis within

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,¹⁰ since the diagnosis of SOV thrombosis can be made with MRI. However, the MRI signal characteristic of thrombosis evolve over time⁶ 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 intraocular pressure.

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Complications of Decompressive Craniectomy

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Introduction

Persistent elevation of intracranial pressure (ICP), if untreated, may lead to brain ischemia or lack of brain oxygen and even brain death.^{1-6,10} When standard treatments for elevated ICP are exhausted without any signs of improvement, decompressive craniectomy can be an effective alternative solution.^{7,19}

Decompressive craniectomies (DC) have been used as a method of controlling intracranial pressure in patients with cerebral edema secondary to cerebral ischemia, subarachnoid hemorrhage (SAH), and traumatic brain injury (TBI), among others.⁸⁻¹⁰ Several studies over the years have demonstrated the efficacy of this procedure.^{7-9,11,35,36} However, consensus is still lacking in the utility of DC as an effective first tier treatment for intractable intracranial pressure due to the rudimentary neurological outcome assessments, and the many complications associated with this procedure.^{11,12,59}

There are a limited number of studies that have looked at complications secondary to the procedure itself.¹³⁻¹⁸ The majority of these studies only investigated the impact of this procedure in patients with traumatic brain injury. The purpose of this study is to investigate the rates of various complications associated with the decompressive craniectomy procedure in patients that did not suffer from traumatic brain injury, and to determine whether the same associations between preoperative parameters and development of complications can be made.

Methods

A retrospective review of a prospectively collected data set of patients who had a decompressive craniectomy done at our institution between January 2003 and January 2010 was performed. Electronic charts were reviewed to obtain the following data: patient age, gender, diagnosis, type of decompressive craniectomy, any complications following the procedure, patient outcome as measured by Glasgow coma scale (GCS) at discharge, time period between craniectomy and cranioplasty and type of flap used for cranioplasty. Rates of various complications were tabulated and we investigated the association of several patient parameters with patient outcome, and rates of the various complications. These factors included age, gender and preoperative GCS.

Appropriate statistical tests were used to determine the strength of associations; Spearman's p, Student's t-test and multivariate regression were performed using the JMP statistical package (version 7.02; SAS Institute, Cary NC).

Results

191 patients were identified, including 99 females, 91 males. The mean age was 50 years old (range 17-85). The mean preoperative GCS score was 8 (range 3-15). 70 patients had intracerebral hemorrhage (36.6%), 60 had ruptured aneurysm (31.4%), 21 had brain edema secondary to a prior elective brain surgery (11%), 15 had stroke (7.8%), 11 had closed head trauma (5.7%), 4 had thrombosed aneurysm (2.1%), 3 had ruptured arteriovenous malformation (AVM) (1.6%), 2 had penetrating trauma (1.4%), 1 had tumor (0.5%), and 3 were unreported (1.6%). A bifrontal craniectomy was performed on 4 cases (2.1%) and 187 were unilateral craniectomies (97.9%). The incidences of complications are summarized in Table 1.

101 of the 191 patients (53%) had at least one complication. 42 patients died despite the procedure. Of the survivors (n = 149), a significant number were discharged to rehabilitation (n=121), 8 were discharged to full time nursing facilities, 2 remained in the hospital, 1 was discharged to hospice, and the rest returned home (n = 13). Three cases did not report discharge destination. There was no correlation between age and mortality.

19 patients had a preoperative GCS score ranging from^{3-5, 49} patients ranged from 6-9 and 33 patients were greater than 9. The mean preoperative score was 8. Twelve patients had a postoperative GCS score of 6 or less, 40 were between 6-9 and 68 patients had scores greater than 9. Mean postoperative GCS scores were 3.87±0.49 (mean±SE) above preoperative GCS scores. Patients with higher pre-op GCS scores or older age tended to have higher GCS upon discharge (p<0.091). Female patients and patients that had one or more complications had lower GCS scores upon discharge (p<0.037,p<0.016). Neither gender nor age was associated with either incidence or total number of complications. Patients that had a

Table 1. Complications following Decompressive Craniectomy	
Complication	N (%)
Hydrocephalus	55 (28.7)
VP shunt	37 (19.4)
Herniation	40 (20.9)
Vasospasm	10 (5.2)
Subdural hygroma	18 (9.4)
Seizures	2 (1)
Sunken flap	2 (1)
Flap resorption	0
Increased ICP	9 (4.7)
Infection*	42 (21.9)
*Pneumonia was the commonest infection in this study	