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

L. Fernando Gonzalez, MD
Thomas Jefferson University

Jurij R. Bilyk, MD
Wills Eye Institute

Pascal Jabbour, MD
Thomas Jefferson University

Stavropoula Tjoumakaris, MD
Thomas Jefferson University

Aaron S. Dumont, MD
Thomas Jefferson University

Follow this and additional works at: https://jdc.jefferson.edu/jhnj

Let us know how access to this document benefits you

Recommended Citation
Gonzalez, MD, L. Fernando; Bilyk, MD, Jurij R.; Jabbour, MD, Pascal; Tjoumakaris, MD, Stavropoula; Dumont, MD, Aaron S.; Chalouhi, MD, Nohra; and Rosenwasser MD, Robert H. (2012) "Paradoxical Worsening of Ocular Symptoms after Spontaneous Closure of a Carotid Cavernous Fistula: Case Report," JHN Journal: Vol. 7 : Iss. 1 , Article 3.
DOI: https://doi.org/10.29046/JHNJ.007.1.006
Available at: https://jdc.jefferson.edu/jhnj/vol7/iss1/3

This Article is brought to you for free and open access by the Jefferson Digital Commons. The Jefferson Digital Commons is a service of Thomas Jefferson University's Center for Teaching and Learning (CTL). The Commons is a showcase for Jefferson books and journals, peer-reviewed scholarly publications, unique historical collections from the University archives, and teaching tools. The Jefferson Digital Commons allows researchers and interested readers anywhere in the world to learn about and keep up to date with Jefferson scholarship. This article has been accepted for inclusion in JHN Journal by an authorized administrator of the Jefferson Digital Commons. For more information, please contact: JeffersonDigitalCommons@jefferson.edu.
Paradoxical Worsening of Ocular Symptoms after Spontaneous Closure of a Carotid Cavernous Fistula: Case Report

Authors
L. Fernando Gonzalez, MD; Jurij R. Bilyk, MD; Pascal Jabbour, MD; Stavropoula Tjoumakaris, MD; Aaron S. Dumont, MD; Nohra Chalouhi, MD; and Robert H. Rosenwasser MD

This case report is available in JHN Journal: https://jdc.jefferson.edu/jhnj/vol7/iss1/3
Paradoxical Worsening of Ocular Symptoms after Spontaneous Closure of a Carotid Cavernous Fistula: Case Report

L. Fernando Gonzalez, MD; Juríj R. Bilyk, MD; Pascal Jabbour, MD; Stavropoula Tjoumakaris, MD; Aaron S. Dumont, MD; Nohra Chalouhi, MD; Robert H. Rosenwasser, MD

1Neurosurgery Department, Thomas Jefferson University, Philadelphia, Pennsylvania

Symptoms after Spontaneous Closure of Paradoxical Worsening of Ocular Symptoms

Note the straightening of the vessel with stent placement.

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 superior ophthalmic vein, (D) left internal carotid artery anterior-posterior view showing the fistula, and (E) left external carotid artery lateral view showing a prominent superior orbital vein.

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 our institution with conjunctival injection associated with elevated IOP.

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 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 orbital signs and symptoms. A DSA showed no evidence of arteriovenous fistula, and a documented CCF on initial cerebral angiography. 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.

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 severe symptoms. In our case, the apparent papillary defect resolved over the next 24 hours. The external ophthalmoplegia and conjunctival injection persisted, but there was a marked improvement of exophthalmos and periscopic pain. On subsequent follow-up two weeks later, a subtle left arcuate defect was noted on automated perimetry, but the IOP had decreased to 22mm 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 severe symptoms. In our case, the apparent papillary defect resolved over the next 24 hours. The external ophthalmoplegia and conjunctival injection persisted, but there was a marked improvement of exophthalmos and periscopic pain. On subsequent follow-up two weeks later, a subtle left arcuate defect was noted on automated perimetry, but the IOP had decreased to 22mm Hg.

In general, treatment of CCF is reserved for severe symptoms. In our case, the apparent papillary defect resolved over the next 24 hours. The external ophthalmoplegia and conjunctival injection persisted, but there was a marked improvement of exophthalmos and periscopic pain. On subsequent follow-up two weeks later, a subtle left arcuate defect was noted on automated perimetry, but the IOP had decreased to 22mm Hg.
Resolution of CCFS has been reported after angiography, where a clot developed during the procedure in the internal carotid artery to 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.

Bajwa 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. Sargent 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 resolu-
tion of the CCF. Since the SOV provides the major and in many cases only venous outflow for the orbit, sudden worsening of orbital congestion manifestations as an orbital compartment syndrome (OCS).10 In addition, since the orbital veins are valved, some orbital drainage may occur in an antegrade 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 results in stagnation of abnormal blood flow within the cavernous sinus, precipitating the occlusion of the CCF, slow flow through 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 concomitant 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, mark-
ably improving within 48 hours.10 The goal of OCS therapy in such situations is to “buy time” until orbital congestion resolves. Presumably, orbital venous outflow forms alternate drain-
age pathways during this time. Initially, topical anti-glaucoma medications are instituted along with intravenous munitsin. 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 symp-
toms does not always represent persistence or progression of the arterio-venous fistula, as in this case Illustrates. In cases of presumed sponta-
naneous 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 time and may be difficult to inter-
pret 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 intervention and the management is focused on managing the acute orbitopathy and raised intraocular pressure.

References

Sandra Ho, BS; Yinn Chor Ooi, MD; Muhamed Adil Sheikh, MBBS; Mitchell Maltenfort, PhD; Jack Jallo MD, PhD
1. Jefferson Medical College, Philadelphia, Pennsylvania
2. Neurosurgery Department, UCLA, Los Angeles, California
3. Doce Medical College, Karachi, Pakistan
4. Thomas Jefferson University, Philadelphia, Pennsylvania

Introduction
Paradoxical worsening of intracranial pressure (ICP), if untreated, may lead to brain ischemia or lack of brain oxygen and even brain death.1-5,9,10 When standard treatments for elevated ICP are exhausted without any signs of improvement, decompressive craniectomy can be an effective alternative solution.6-9

Decompresive 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.1,2 Several studies over the years have demonstrated the efficacy of this procedure.7,9,10 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.7,10,11

There are a limited number of studies that have looked at complications secondary to the procedure itself.12-15 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) as discharge, time period between craniotomy and cra-

Table 1. Complications following Decompressive Craniectomy

<table>
<thead>
<tr>
<th>Complication</th>
<th>Number of Patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hydrocephalus</td>
<td>15 (8.4)</td>
<td>23%</td>
</tr>
<tr>
<td>Vasospasm</td>
<td>10 (5.2)</td>
<td>16%</td>
</tr>
<tr>
<td>Subdural hygroma</td>
<td>8 (4.5)</td>
<td>12%</td>
</tr>
<tr>
<td>Atelectasis</td>
<td>4 (2.2)</td>
<td>6%</td>
</tr>
<tr>
<td>Pneumonia post craniectomy</td>
<td>1 (0.5)</td>
<td>1.6%</td>
</tr>
</tbody>
</table>

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 dis-
charged 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 3-9. 5 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±1.49 (mean±SD) above preoperative GCS scores. Patients with higher pre-op GCS scores or older age tended to have higher GCS upon discharge (p<0.01). Female patients and patients that had one or more complic-a-
tions had lower GCS scores upon discharge (p<0.01). Neither gender nor age was associated with either incidence or total number of complications. Patients that had a decent necrotic brain tissue.