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

L. Fernando Gonzalez, MD
*Thomas Jefferson University, Fernando.Gonzalez@jefferson.edu*

Jurij R. Bilyk, MD
*Wills Eye Institute, jbilyk@aol.com*

Pascal Jabbour, MD
*Thomas Jefferson University, Pascal.Jabbour@jefferson.edu*

Stavropoula Tjoumakaris, MD
*Thomas Jefferson University, Stavropoula.Tjoumakaris@jefferson.edu*

Aaron S. Dumont, MD
*Thomas Jefferson University, adumont2@tulane.edu*

*See next page for additional authors*

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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. Rosenwasswer MD

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

L. Fernando Gonzalez, MD1; Jurij R. Bilyk, MD2; Pascal Jabbour, MD1; Stavropoula Tjoumakaris, MD1; Aaron S. Dumont, MD1; Nohra Chalouhi, MD1; Robert H. Rosenwasser, MD1

1Neurosurgery Department, Thomas Jefferson University, Philadelphia, Pennsylvania
2Wills Eye Institute, Philadelphia, Pennsylvania

Symptoms after Spontaneous Closure of Paradoxical Worsening of Ocular Symptoms

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. The patient declined endovascular treatment. She presented 15 months later with acute exacerbation of her symptoms. A digital subtraction angiography (DSA) demonstrated a Type-D CCF draining into the left superior ophthalmic vein (SOV). The authors also gave directives for the management of these extremely rare cases.

Case Report

A 59-year-old female presented with constipation, 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 exophthalmos. Computed tomography showed a prominent, hyperdense left SOV on the left side suggesting the presence of thrombus within 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 constipation, 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 exophthalmos. Computed tomography showed a prominent, hyperdense left SOV on the left side suggesting the presence of thrombus within 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.

Discussion

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 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. 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.

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 ophthalmic vein, (E) left internal carotid artery injection showing the fistula, and a large superior ophthalmic vein treated with a 3-stents telescoping technique. The straightening of the vessel with stent placement is noted in the straightening of the vessel with stent placement.
Resolution of CCs 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.20 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 colleagues21 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 venous drainage to the orbit, resulting in acute orbitopathy, spontaneous SOV thrombosis is a rare event, occurring in patients with cerebral edema secondary to cerebral ischemia, subarachnoid hemorrhage (SAH), and traumatic brain injury (TBI), among others.3-5 Several studies over the years have demonstrated the efficacy of this procedure.7-10 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.3-5,7

Decompensation of the CCF has been described 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.7-10 Several studies over the years have demonstrated the efficacy of this procedure.7-10 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.3-5,7

There are a limited number of studies that have looked at complications secondary to the procedure itself.7-10 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 prognostic variables 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 procedure, 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 prognostic GCS. Additional factors included post-operative complications, including those associated with the surgical procedure, wound infection, and other systemic problems. We also compared the impact of these complications on neurologic outcome. The association between various explanatory variables and the outcome was evaluated using the chi-square test of association and the Fisher exact test. The relationships between survival and the explanatory variables were evaluated using the Kaplan-Meier test, and the log-rank test was used for comparison. The survival was defined as the time from randomization to death, or last follow-up.

Results

191 patients were identified, including 99 females, 91 males. The mean age was 50 years old (range 17-85). The mean GCS scores at baseline (0-15) were 12±5±0.5. The mean postoperative GCS scores were 13±4±0.5. The mean postoperative SOV thrombosis was 1.6% (5/305). The mean postoperative SOV thrombosis was 1.6% (5/305). The mean postoperative SOV thrombosis was 1.6% (5/305). The mean postoperative SOV thrombosis was 1.6% (5/305). The mean postoperative SOV thrombosis was 1.6% (5/305). The mean postoperative SOV thrombosis was 1.6% (5/305).

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. AlthoughDSA is the gold standard for diagnosis, there is no role for endovascular intervention and the management is focused on managing the acute optic neuropathy and raised intracranial pressure.