Carotid Cavernous Fistula: A Case Study and Review

laswarya Ganapathiraju, OMS-IV¹

Douglas T Summerfield, MD²

Melissa M Summerfield, MD²

¹Des Moines University College of Osteopathic Medicine

Des Moines, IA USA

²Mercy Medical Center North Iowa and North Iowa Eye Clinic

Mason City, IA USA

Abstract

Carotid cavernous fistulas are rare complications of craniofacial trauma, resulting in abnormal connections between the arterial and venous systems of the cranium. The diagnosis of carotid cavernous fistulas and other injuries as a result of trauma can be confounded by the traumatized patient's inability to communicate their symptoms to their physician. The following case study demonstrates the importance of a thorough physical exam in caring for such patients and serves to remind physicians to have a low threshold for consultation when managing numerous injuries following trauma.

Introduction

Carotid cavernous fistulas (CCFs) are aberrant connections between the carotid arterial system and the cavernous sinus, which form as complications of craniofacial trauma, or are congenital or spontaneous in nature (1). They occur in up to 3.8% of patients with basilar skull fractures and are more common with middle fossa fracture (2). Prompt diagnosis and treatment of CCF is necessary as approximately 20 – 30% of carotid cavernous fistulas lead to vision loss if not addressed appropriately (3). The following is a case study of a patient who presented with multiple traumatic injuries including CCF with subsequent discussion of the typical presentation, diagnosis, and treatment of direct CCF.

Case Presentation

A 64-year-old woman with a therapeutic INR on Coumadin for atrial fibrillation sustained a fall down a flight of stairs. She was found unresponsive the next day by her relatives and was subsequently brought to the emergency department for evaluation. A maxillofacial CT showed a nondisplaced right maxillary wall fracture and nondisplaced zygomatic arch fracture, as well as a subtle inferotemporal orbital fracture, none of which was determined to require immediate treatment by the otolaryngology service. Further imaging included a CT of the head which revealed a large subdural hematoma,

a superotemporal hematoma, and subfalcine herniation. She was taken to the OR for emergent craniotomy and evacuation of the hematoma before transfer to the critical care unit. In the CCU, she remained intubated and sedated but her condition improved until extubation on hospital day 3. She continued to have swelling surrounding both eyes during this time, but physical exam showed pupils which were equal, round, and reactive to light.

On day 6 of her stay, the patient was noted to have waxing and waning confusion and slightly increased oxygen requirement. Thus, she was re-intubated and sedated for “agitation” and “hypoxic respiratory failure.” Physical exam on the next day was notable for pupillary anisocoria with the right pupil at 1 mm diameter and left at 2.5 mm. There was a poor pupillary light reaction bilaterally. Neurology was consulted and recommended repeat imaging and EEG. Repeat CT and MRI of the brain showed no evidence of herniation, and EEG was negative for seizure-like activity. The anisocoria was thought to be from mass effect of the temporal lobe on cranial nerve III. The patient’s condition continued to deteriorate; physical exam elicited grimace to painful stimuli and the patient was able to open her eyes but did not track movement or follow commands. She was subsequently noted to have a left orbit that became harder to compress with ballottement test compared to the right, so Ophthalmology was consulted.

An ophthalmologic exam showed extensive chemosis of the left eye compared to the right with conjunctival hemorrhage in bilateral eyes (Figure 1).

Figure 1. Ophthalmologic exam revealed chemosis, exophthalmos, and a mid-dilated, fixed pupil of left eye compared to right.

Ocular tonometry revealed a pressure of 14 mmHg in the right eye and 53 mmHg in the left. There was a mid-dilated, fixed pupil on the left. The differential at this point included traumatic acute angle closure glaucoma versus a retroorbital process. The patient was started on timolol, pilocarpine, and dorzolamide eye drops for intraocular pressure control. An orbital CT was obtained, which showed an engorged superior ophthalmic

vein on the left with a new 4 mm proptosis of the left eye (Figure 2) when compared to previous imaging.

Figure 2. A: CT scan showed proptosis of 4 mm of left eye compared to right eye. B: Enlarged left ophthalmic vein also noted on CT scan (arrow).

This raised concern for traumatic carotid cavernous fistula. A CTA obtained the following morning confirmed this suspicion (Figure 3).

Figure 3. A: Reconstructed coronal CT coronal angiogram showing enlarged left cavernous sinus, confirming diagnosis of carotid cavernous fistula. B-E: Static coronal images from CT angiogram with major arteries labeled.

The patient was transferred to an outside facility for surgical management, which consisted of angiography and embolization via coiling of her CCF.

Discussion

Carotid cavernous fistulas are abnormal connections that form between the cavernous sinus and the internal or external carotid arteries, or branches of the internal or external carotid arteries. They are divided into direct and indirect variants per Barrow classification (Table 1, Figure 4).

Type	Direct vs. Indirect	Description
A	Direct	Direct connection between ICA and cavernous sinus.
B	Indirect	Connection between dural branches of ICA and cavernous sinus.
C	Indirect	Connection between dural branches of ECA and cavernous sinus.
D	Indirect	Connection between dural branches of both ICA and ECA and the cavernous sinus.

ICA = Internal carotid artery, ECA = External carotid artery

Figure 4. A: The normal eye: superior ophthalmic vein draining into cavernous sinus and internal and external carotid arteries traversing the cavernous sinus. B: Barrow Classifications for types of carotid cavernous fistulas: Type A: direct connection between internal carotid artery and cavernous sinus. Type B: connection between dural branches of internal carotid artery and cavernous sinus. Type C: connection between dural branches of external carotid artery and cavernous sinus. Type D: connection between dural branches of both internal carotid artery and external carotid artery and the cavernous sinus.

Types B through D are commonly termed 'indirect' or 'dural' fistulas. These can develop spontaneously as a result of hypertension and are the more common presentation of CCF. More specifically, type B is a connection between the dural branches of the ICA and the cavernous sinus, type C is a connection between the dural branches of the external carotid artery (ECA) and the cavernous sinus, and type D connects the dural supply of both the ICA and ECA and the cavernous sinus (1). Type A, or a 'direct' CCF, is a connection between the intracavernous internal carotid artery (ICA) and the cavernous sinus. Direct CCF is a rare ocular complication that forms most commonly as a result of craniofacial trauma, but can also be due to aneurysmal rupture or spontaneous development. This is also the most dramatic presentation of CCF and was the case in our patient.

Prompt identification and management of CCF is necessary to prevent associated morbidity and mortality. The presentation of CCF depends mainly on the drainage of the fistula. Anterior-drainage of fistulas through the superior ophthalmic vein produces symptoms of exophthalmos, proptosis, acute chemosis or swelling/edema of conjunctiva, and headache, all of which are more common in direct CCFs. The backup of drainage can result in a secondary angle closure with extremely high intraocular pressure. Posterior-drainage of fistulas into the superior and inferior petrosal sinuses tend to lack the aforementioned features of orbital congestion, but can produce painful cranial neuropathy of the trigeminal, facial, or ocular motor nerves. Failure to identify and appropriately treat posterior-draining fistulas can lead to eventual reversal of flow and development of anterior drainage (4).

The signs of CCF are not visible on neuroimaging at a patient's presentation and generally develop over the first week a patient is admitted. Clinical signs which may prompt further investigation and repeat imaging include chemosis, increasing exophthalmos, pain, and increased intraocular pressure. Often, the tools for checking intraocular pressure are not available in an ICU setting. In the absence of signs of a ruptured globe, an intensivist could palpate the orbit over a closed eye (as occurred in this case). If there is asymmetry in resistance to palpation, this should incite an ophthalmologic consult to consider a retro-orbital process.

Repeat neuroimaging is likely to be done in these cases, but it is important to order the right test. Radiologic signs of CCF include proptosis and asymmetric enlargement of a cavernous sinus or superior ophthalmic vein and would be noted on an orbital or maxillofacial CT. A head CT might miss these signs, so it is important to obtain imaging dedicated to examining the retro-orbital space. To confirm the diagnosis of CCF, one must then obtain a CT angiogram, which will show the aberrant connections between the intracranial vessels. Upon confirming a diagnosis of CCF, the preferred mode of

management is endovascular obliteration using an arterial or venous approach as it has been shown to be safe and effective, and confers long-term cure in most cases (5).

A previous review of 16 cases of carotid cavernous fistulas treated with transarterial embolization with detachable balloon show satisfactory results, defined as resolution of CCF without residual disability, in 11 cases and resolution but with residual disability in 5 cases. The most common of the disabilities in these cases was vision impairment, as seen in 4 out of the 5 cases. In addition, 14 out of the 16 cases resolved with preserved internal carotid artery flow (1). As a result, transarterial embolization with detachable balloon (TAEDB) has been established as the preferred method of treatment for carotid cavernous fistulas (6). Other options for treatment include neurosurgery and stereotactic radiosurgery when endovascular approach is not feasible.

Our patient presented with several traumatic injuries following a fall down a flight of stairs and was unable to contribute to history-taking. Detection and treatment of the CCF that she later developed was complicated by several factors. The true exophthalmos of the affected eye was partially masked by the fact that she had an inferotemporal orbital fracture of the opposite eye, which was incorrectly thought to be enophthalmic. Additionally, her altered mental status and subsequent re-intubation limited her ability to vocalize the pain which would have been present in her affected eye due to tremendously increased intraocular pressure.

From a critical care physician perspective, part of the key to her diagnosis was her re-intubation. The patient developed severe agitation requiring sedation without other more typical reasons for intubation such as hypoxia, tachypnea, or dyssynchronous breathing. We suspect this agitation was likely secondary to pain from the rapidly increasing pressure in her affected eye which became symptomatic just prior to her worsening mental status. Her physical exam was ultimately crucial to the detection of her CCF, specifically chemosis, exophthalmos, and increased intraocular pressure in the affected eye. These signs led to the subsequent ophthalmologic consultation, imaging, and eventually the diagnosis of CCF.

An important lesson learned from this patient's management is having a low threshold for consultation when the clinical picture does not match diagnostic workup. In our case, the patient's clinical condition changed but repeat workup including EEG and MRI of the head was negative. Previous imaging had revealed right-sided facial fractures, yet her new findings, including increased resistance to palpation of the orbit and chemosis, were largely left-sided. In situations when the cause of a patient's deteriorating condition is unclear and there is incongruity between the physical exam and diagnostic workup, it is imperative to obtain further consultation. In our case, the ophthalmic exam gave the clues for further workup and the ultimate diagnosis.

In conclusion, this patient's case is a good study in the classic presentation of direct CCF in association with craniofacial trauma, and also illuminates the difficulty in detection of orbital injuries in a trauma patient who cannot vocalize the symptoms they are experiencing. The lesson learned from her presentation is to have a low threshold for ophthalmologic consultation for unexplained changes in ophthalmic condition and discrepancies between clinical presentation and diagnostic findings.

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