Complete Resolution of Traumatic Indirect Carotid-Cavernous Fistula Two Years After Inciting Event

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ABSTRACT

Indirect carotid cavernous fistulas (CCFs) are most often spontaneous, but can rarely be caused by trauma. With traumatic etiology, the timeline for the development of symptoms varies significantly and can be difficult to predict. In this report, we discuss the case of a patient found to have an indirect CCF who presented for acutely worsening ocular symptoms and a history of pulsatile tinnitus that began two years prior after a suspected inciting head injury. To our knowledge, no cases have described a traumatic indirect CCF with a similarly extensive indolent course who demonstrated full symptomatic recovery following treatment.

KEYWORDS: carotid-cavernous fistula, indirect, trauma, endovascular

CASE PRESENTATION

An 82-year-old female with hypothyroidism and no significant ocular history presented to the clinic with three months of left eye pain, redness, decreased vision, and photophobia (**Figure 1A**). She also endorsed persistent left-side facial pain and an intermittent "whooshing" noise in the ipsilateral ear, which began after a fall with loss-of-consciousness two years prior.

Exam on presentation was significant for best-corrected visual acuity (BCVA) of 20/30 OD and 20/800 OS, intraocular pressure (IOP) of 16 mmHg OD and 19 mmHg OS, 2+ afferent pupillary defect (APD) OS, and diffuse visual field deficits on the left with partial sparing of the superior nasal field. External exam showed proptosis OS with exophthalmometry (15mm OD and 20mm OS). Slit lamp examination was significant for prominent episcleral corkscrew vessels OS and no clear posterior findings.

Computed tomography angiogram (CTA) of the head and neck demonstrated asymmetric enlargement of the left superior ophthalmic vein, and asymmetric enhancement of the left cavernous sinus overall suggestive of an indirect (Barrow's type B, C, or D) carotid cavernous fistula (CCF) (**Figure 2A, 2B**).

The patient was hesitant to undergo an endovascular procedure at her initial visit, but at subsequent evaluation

Figure 1. Patient photograph at initial presentation to the clinic (**A**), at one-week post-operative follow-up (**B**), and at one-month post-operative follow-up (**C**).

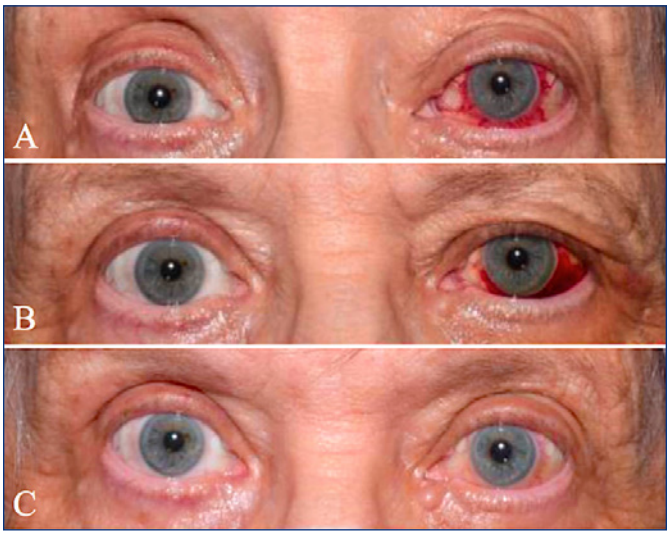
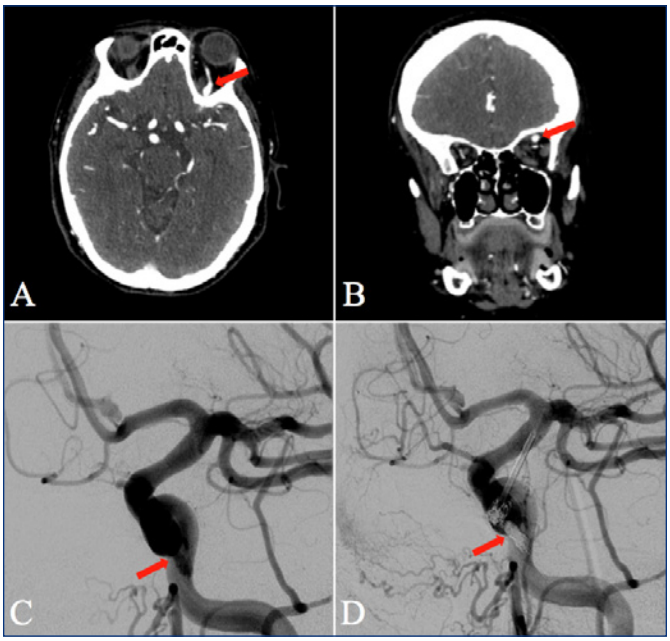


Figure 2. Computed Tomography Angiography (CTA) with coronal (**A**) and axial (**B**) views of the dilated left superior ophthalmic vein. Interventional angiograms with arrows demonstrating the patent fistula (**C**) and the resolved fistula following coil embolization (**D**).



decided to proceed with treatment secondary to persistent ocular and facial pain. She was admitted to the hospital and, after an initial failed endovascular approach via peripheral venous access that identified thrombosis of the distal ophthalmic veins, underwent successful coil embolization of the fistulous pouch by direct transorbital puncture of the cavernous sinus via the superior ophthalmic vein (**Figure 2C, 2D**). She was discharged on the first postoperative day and asked to return to the clinic for follow-up in one week.

Exam at one-week follow-up demonstrated BCVA of 20/40 OS, exophthalmometry of 16.5mm OS, and only trace APD OS. Slit lamp examination revealed improved episcleral corkscrew vessels and a new subconjunctival hemorrhage temporally OS (**Figure 1B**). The patient reported mild frontal headache, but otherwise felt well with no ocular pain. Patient reported near-complete resolution of presenting symptoms after one month (**Figure 1C**).

At five-month follow-up, BCVA was 20/25 OS, exophthalmometry was 15.5mm OS, and no APD was present. Slit lamp exam revealed resolution of episcleral corkscrew vessels and subconjunctival hemorrhage. The patient endorsed some residual left-side facial pain, but otherwise complete resolution of ocular symptoms and the pulsatile tinnitus that has persisted for two years prior.

DISCUSSION

Indirect CCFs, also called "low-flow" fistulas, are abnormal communications between the cavernous sinus (CS) and dural branches of the internal carotid artery (Barrow type B), the external carotid artery (Barrow type C), or both (Barrow type D). In contrast, direct CCFs (Barrow type A) connect the internal carotid artery and CS without intermediary tracts, and thus function under higher arterial pressures. As such, the presentation of indirect fistulas is more indolent as accommodations in venous drainage from the CS may delay the onset of visual loss, proptosis, and redness.

The etiology of indirect CCFs is often unclear. Unlike with direct fistulas, obvious precipitating trauma is relatively rare. It has been postulated that some indirect CCFs may occur secondary to a subclinical CS thrombosis that causes local revascularization[1]. Other risk factors include pregnancy, local surgical instrumentation, and sinusitis.[2]

As the anatomy of indirect CCFs is highly variable, so too is the natural history. Spontaneous closure is relatively common, with estimates varying between 40–90% depending on duration of observation and means of non-invasive treatment.[3,4] Therefore, conservative management may be a consideration depending on patient preference and severity of presenting symptoms. Factors that have demonstrated increased risk for poor prognosis include progressive vision loss, proptosis, and signs of elevated intracranial pressure.[2]

Our patient reported a two-year history of ipsilateral facial pain and pulsatile tinnitus, which began after a fall with loss-of-consciousness, progressing to include ipsilateral ocular pain and decreased vision starting three months prior to presentation. Her history, combined with the cessation of tinnitus following treatment, strongly suggests that the fall two years prior was responsible for the indirect CCF. While a traumatic indirect CCF is a highly uncommon presentation, perhaps more remarkable is this patient's timeline of symptoms.

To our knowledge, no cases of traumatic CCFs have been described with such a lengthy indolent course, particularly given the patient's full recovery following treatment. It is unclear why the current patient was symptomatic, but stable, for two years with a precipitous worsening of symptoms. Possible causes for the worsening of her symptoms may include an acute anatomical change to the fistulous connection resulting in increased inflow to the CS, similar to the acute process responsible for traumatic, direct (Barrow type A) CCFs. Also possible is an acute congestion of the venous outflow tract, such as with a venous thrombosis, leading to decreased drainage of the sinus.

The current case demonstrates that an unusually long symptomatic time to presentation should not steer the clinician away from adding indirect CCF to the differential diagnosis for symptoms including, but not limited to, red eye, chemosis, or ocular pain that is refractory to conservative management. The misdiagnosis of indirect CCF as more common benign conditions can cause delays in treatment and possible permanent ocular damage[5]. If signs of smoldering indirect CCF are correctly identified early by caregivers, episodes of severe exacerbation with threatened vision loss and potential permanent ocular damage may be avoided.

The current case also demonstrates that even after a two-year symptomatic period with an acute clinical decompensation, endovascular therapy can be highly efficacious. The outcomes of various endovascular interventions for patients with traumatic indirect CCFs has been described, but more research is needed to delineate the relationship between symptomatic time-to-presentation and degree of recovery post-intervention.[6,7,8] We demonstrate here full symptomatic resolution of an indirect, traumatic CCF following a time course not previously documented. In future encounters, it may be warranted to pursue consultation for endovascular therapy as soon as an indirect CCF is suspected, irrespective of etiology, degree of vision loss, or extent of symptomatic time course.

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