Spontaneous resolution of nontraumatic bilateral Barrow Type D indirect carotid-cavernous fistulas: A case report

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Abstract:
A Caucasian man in his 60s with a history of Cognard Type IIB dural arteriovenous fistula presented to the emergency room with right eye proptosis, chemosis, hyperemia, epiphora, diplopia, and blurred vision. Magnetic resonance imaging and magnetic resonance angiography revealed spontaneous, bilateral Barrow Type D carotid-cavernous fistulas (CCFs) that were later confirmed through cerebral angiography. The patient had no history of head or ocular trauma. Given the acute nature of presentation and worsening diplopia, the patient was scheduled for transvenous embolization. However, during the preprocedure angiogram, spontaneous resolution of the bilateral CCFs was observed. Complete resolution of all symptoms was noticed during follow-up. Given the rare nature of bilateral, indirect CCFs, our case stands out as the only reported instance whereby resolution of bilateral, indirect CCFs occurred spontaneously without any intervention.

Keywords:
Bilateral carotid-cavernous fistulas, case report, fistula, spontaneous resolution

Introduction
Carotid-cavernous fistulas (CCFs) are abnormal shunts between the carotid arteries and the cavernous sinus.1,2 CCFs are classified by hemodynamics (high flow or low flow), anatomy (direct or indirect), or etiology (traumatic or spontaneous).2 Type A CCFs are high-flow connections between the internal carotid artery (ICA) and the cavernous sinus. In contrast, Types B, C, and D, are indirect, low-flow communications that result from connections between the dural branches of the ICA, external carotid artery (ECA), or both.2,3 Solitary indirect CCFs have been known to resolve spontaneously and are indolent in nature. However, to the best of our knowledge, no case of spontaneous resolution of bilateral, indirect CCFs has been reported in the literature. Herein, we report the unique case of a patient who presented to us with spontaneous, bilateral, indirect Barrow Type D CCFs that resolved spontaneously without any medical or surgical intervention.

Case Report
A Caucasian man in his 60s presented with the chief complaint of whooshing in his left ear, with associated soreness and pain on palpation, for a duration of 1 month in January 2020. The patient had a past medical history of hypertension, glaucoma, and obstructive sleep apnea. Notably, he did not have any history of a head or ocular injury. An initial visit to his primary care doctor...
was unremarkable. The patient’s symptoms worsened in 1 month time, and he developed acute onset of diplopia that was worse on near vision. Magnetic resonance imaging (MRI) was subsequently performed, and interpreted as negative with no significant intracranial pathologies seen such as cavernous sinus thrombosis or orbital inflammatory disorders. Due to persistent diplopia, he was referred to the neurology department where a “left eye nerve palsy” was suspected.

With symptoms worsening, magnetic resonance angiography (MRA) was performed, which revealed multiple arteriovenous fistulas including an occipital dural arteriovenous fistula (dAVF) involving the left transverse sigmoid sinus and bilateral CCFs. A diagnostic digital subtraction angiogram (DSA) confirmed the presence of an occipital Cognard Type IIB dAVF [Figure 1a and b] and bilateral CCFs [Figure 2a and b]. The patient underwent successful treatment for the left transverse sigmoid junction dAVF through a combined open and endovascular approach (clip and retrograde embolization) in a hybrid suite [Figure 3a and b]. The bilateral CCFs were again noted in the postoperative angiogram. The fistulas were categorized as bilateral Barrow Type D indirect CCFs, with the sphenopalatine artery feeding the left fistula and the ascending pharyngeal artery supplying the right [Figure 4a and b]. Owing to the secondary nature of this CCF, it was decided to wait and observe, especially because the patient had recently undergone the combination procedure.

Six months after the procedure, the patient presented to the emergency room with right eye proptosis, blurred vision, and transient diplopia that was more pronounced on lateral gaze, in conjunction with chemosis, swelling, hyperemia, and epiphora. High intraocular pressure (IOP) in the right eye (28 mmHg) was recorded on ophthalmologic assessment. The patient underwent imaging (MRI and MRA) that confirmed persistent bilateral CCFs. During his stay at the hospital, the patient suffered several seizure episodes that were managed and treated by the neurology team. Once stable, he underwent a cerebral angiogram that again confirmed the presence of Type D CCFs draining into the cavernous sinus. The patient was discharged on acetaminophen (1,000 mg), amlodipine (10 mg), dexamethasone (2 mg), famotidine (20 mg), injection insulin regular (3–12 unit, q6 h), latanoprost ophthalmic (1 drop, both eyes), levetiracetam (1,000 mg), and rosuvastatin (10 mg). On discharge, the patient’s clotting

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**Figure 1:** Occipital Cognard Type IIB dural arteriovenous fistula noted on digital subtraction angiogram communicating with the superior sagittal sinus on anterior–posterior (a) and lateral views (b)

**Figure 2:** Barrow Type D indirect carotid-cavernous fistulas on the right (a) and left (b) sides are seen, indicated by the asterisk* on the initial angiogram on anterior–posterior view

**Figure 3:** Complete resolution of the occipital dural arteriovenous fistula noted on anterior–posterior (a) and lateral views (b) with no residual filling of the dural fistula post combined treatment approach with transvenous embolization and open surgical resection. Of note, the left vertebral artery, basilar artery, posterior inferior cerebellar artery, and the posterior cerebral artery can be appreciated on the anterior–posterior (a) lateral run (b)

**Figure 4:** Angiogram showing feeding arteries of the carotid-cavernous fistula on right (a) and left (b) sides on the anterior–posterior view. The right carotid-cavernous fistula (a) can be seen filling from the multiple smaller branches of internal maxillary artery, and the left (b) from the sphenopalatine artery. Asterisk* indicates that bilateral carotid-cavernous fistulas
profile was noted to be within normal limits (prothrombin time: 13.8 s; international normalized ratio: 1.08; and activated partial thromboplastin time: 25.6 s).

For the bilateral, indirect CCFs, the patient was presented with all treatment options such as medical management, endovascular intervention including embolization and alternatives, as well as no treatment, after which he gave informed consent for transvenous embolization that was scheduled 1 week after the angiogram. The patient’s ocular symptoms persisted with no major improvements recorded during the period between the two angiograms. The patient was prepared and draped for the procedure, and the initial arterial angiographic run demonstrated spontaneous obliterations of both CCFs [Figure 5a and b]. Hence, no embolization was attempted, and the procedure was aborted. The previous dAVF was also noted to be fully resolved. On physical examination at discharge, the patient demonstrated no proptosis, chemosis, or redness with resolution of most visual symptoms except some restricted extraocular motion in the right eye. The patient was otherwise at his baseline level and denied any headache, numbness, tingling, weakness, or facial droop. According to the patient’s ophthalmologist, his vision was 20/25 OU. The patient reported positive changes in vision and improvement in all his symptoms.

**Discussion**

We present here a rare case of bilateral nontraumatic indirect Barrow Type D CCFs that resolved spontaneously without any medical or surgical intervention. Barrow Type D CCFs are classified as indirect, low-flow communications with a blood supply from meningeal branches of both the ICA and ECA.[4] Symptoms and signs, such as proptosis, subjective orbital bruit, diplopia, cranial nerve palsy, chemosis, orbital pain, raised IOP, headache, and loss of visual acuity are most common in CCFs.[5-7] Although unilateral spontaneous, indirect CCFs are not uncommon, bilateral, indirect CCFs are rarely cited in the literature without an underlying etiology.[8] In a literature search conducted by Khan et al., 35 spontaneous, nontraumatic, bilateral CCFs (both direct and indirect) were reported since 1963, of which only 2 were Type C and 7 Type D, thus signifying the uncommon nature and rarity of these cases.[7] It is noteworthy to mention that among all these cases, only 2 were managed nonsurgically. This first case reported by Haugen et al. was a 74-year-old man who had left-sided exophthalmos, chemosis, and dilated episcleral veins.[9] The patient was diagnosed with bilateral Type B CCFs and treated nonsurgically, although manual carotid compression technique for more than 2 weeks was practiced, after which his symptoms resolved. Similarly, Al Mufti et al. reported two cases of bilateral CCFs (Type B and D) secondary to cavernous sinus thrombosis, with 1 treated endovascularly and the other through anticoagulation.[10] Our case stands out as the only reported case whereby resolution of bilateral, indirect CCF occurred without any intervention. Notably, the demographics of our case are also uncommon (Caucasian male), as indirect CCFs are far more commonly reported in postmenopausal women, with some studies reporting a 7:1 female-to-male ratio.[8,11,12]

The mechanism for spontaneous resolution remains unknown but could be attributed to the low-flow nature or small size of the fistula.[13] A few authors have speculated that stagnancy and reduction of blood flow could potentially lead to thrombosis of the feeding arteries and subsequent closure of fistula possibly due to a reduction in pressure gradient between the ICA and the fistula.[13] It is also proposed that navigation of microguidewires and catheters through the small-sized fistulas during neuroangiography and attempted intervention could induce abrupt closure of the fistula (via arterial spasm or dissection), especially those supplied by small feeding arteries.[13] In our case, two prior DSAs may explain the spontaneous resolution, although no vasospasm or dissections were seen intraoperatively.

It is important to note that although we report spontaneous resolution of the CCFs, that is not the case generally. Indications for intervention such as uncontrollable IOP, severe proptosis with corneal exposure, retinal ischemia, unremitting diplopia, and cortical venous drainage from the fistula should be considered during medical decision-making since all of these could potentially lead to loss of vision.[14] This was the case in our patient who was scheduled for an endovascular intervention but was later found to have complete resolution. Notably, the patients’ glaucoma, which was present before the fistula.
diagnosis, continued to persist at its' baseline severity and was managed medically as part of routine care. Although glaucoma secondary to unremitting IOP due to high-flow fistulous connection is reported commonly in literature, this was not the case in our presentation.[10] Furthermore, in cases of unilateral spontaneous regression, changes such as loss of light perception have been reported to persist,[6] but our patient experienced improvement in all his visual symptoms except some residual restricted movement following the resolution.

To confirm the presence of CCFs, imaging techniques such as MRI and MRA should be obtained. Currently, cerebral angiography (digital subtraction angiography) is the “gold standard” modality for the diagnosis of CCFs, regardless of the etiology.[2,13] Once confirmed, CCFs may be treated with endovascular or open surgical approaches. Although CCFs may resolve spontaneously during watchful waiting, if left untreated, they may be considered a threat to vision.[6,7,13]

Ethics
The patient gave informed consent for treatment. Institutional review board approval was deemed unnecessary.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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