

Access this article online

Quick Response Code:



Website:

http://www.braincirculation.org

DOI:

10.4103/bc.bc_50_21

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

Ammad A. Baig^{1,2}, Audrey L. Lazar², Muhammad Waqas^{1,2}, Rimal H. Dossani^{1,2}, Justin M. Cappuzzo^{1,2}, Elad I. Levy^{1,2,3,4,5}, Adnan H. Siddiqui^{1,2,3,4,5}

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] 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.^[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 with no 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

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Baig AA, Lazar AL, Waqas M, Dossani RH, Cappuzzo JM, Levy EI, *et al.* Spontaneous resolution of nontraumatic bilateral Barrow Type D indirect carotid-cavernous fistulas: A case report. *Brain Circ* 2021;7:289-93.

¹Department of Neurosurgery, Jacobs School of Medicine and Biomedical Sciences, University at Buffalo,

²Department of

Neurosurgery, Gates Vascular Institute at Kaleida Health,

³Department of Radiology, Jacobs School of

Medicine and Biomedical Sciences, University at Buffalo, ⁴Canon Stroke and Vascular Research Center, University at

Buffalo, ⁵Department of Neurosurgery, Jacobs Institute, Buffalo, New York, USA

Address for correspondence:

Dr. Adnan H. Siddiqui, University at Buffalo Neurosurgery, 100 High Street, Suite B4, Buffalo, NY 14203, USA. E-mail: asiddiqui@ubns.com

Submission: 28-06-2021

Revised: 28-08-2021

Accepted: 26-09-2021

Published: 21-12-2021

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

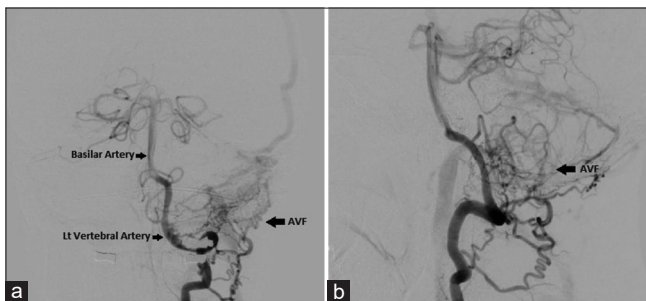


Figure 1: Occipital Cognard Type IIB dural arteriovenous fistula noted on digital subtraction angiogram communicating with the superior sagittal sinus on anterior-posterior (a) and lateral views (b)

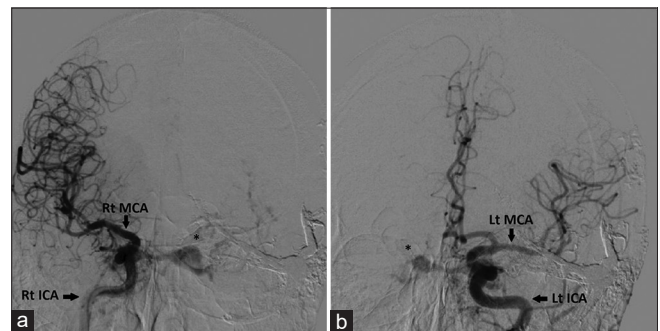


Figure 2: Barrow Type D indirect carotid-cavernous fistulas on the right (a) and left (b) sides are seen, indicated by the asterisk* on the initial angiogram on anterior-posterior view

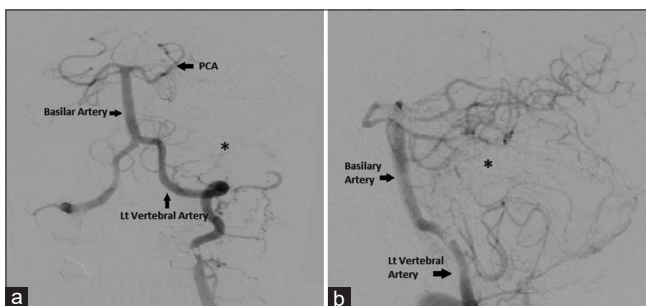


Figure 3: Complete resolution of the occipital dural arteriovenous fistula noted on anterior-posterior (a) and lateral views (b) with no residual filling of the dural fistula post combined treatment approach with transvenous embolization and open surgical resection. Of note, the left vertebral artery,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

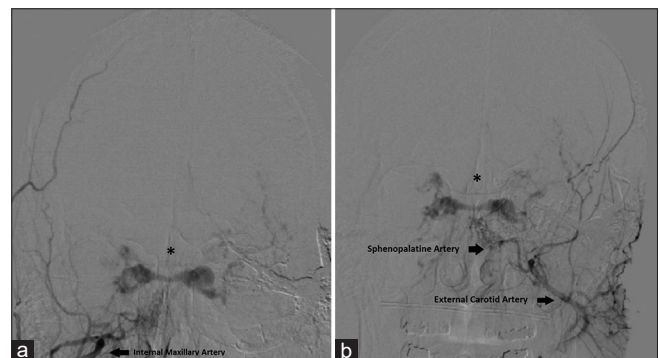


Figure 4: Angiogram showing feeding arteries of the carotid-cavernous fistula on right (a) and left (b) sides on the anterior-posterior view. The right carotid-cavernous fistula (a) can be seen filling from the multiple smaller branches of internal maxillary artery, and the left (b) from the sphenopalatine 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 DCCFs 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

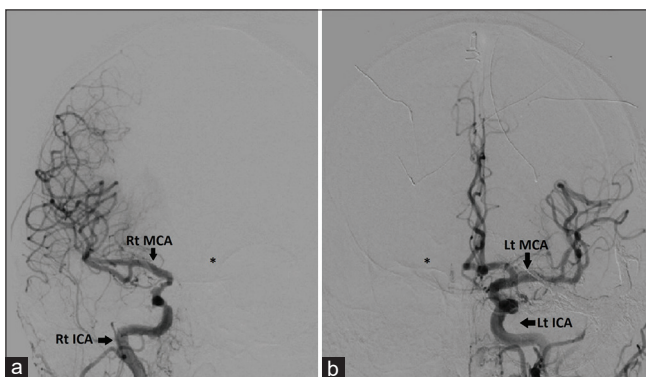


Figure 5: Spontaneous resolution of fistulas on the right (a) and left (b) sides are seen on this angiogram on anterior–posterior view. Asterisk* indicates the previous location of the fistulas. Major intracranial blood vessels are also appreciated

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.^[2,15] 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.

Financial support and sponsorship

Nil.

Conflicts of interest

Disclosure of relationships/Potential conflicts of interest.

AAB, ALL, MW, RHD, JMC: None.

EIL: Shareholder/Ownership interests: NeXtGen Biologics, RAPID Medical, Claret Medical, Cognition Medical, Imperative Care (formerly the Stroke Project), Rebound Therapeutics, StimMed, Three Rivers Medical; National Principal Investigator/Steering Committees: Medtronic (merged with Covidien Neurovascular) SWIFT Prime and SWIFT Direct Trials; Honoraria: Medtronic (training and lectures); Consultant: Claret Medical, GLG Consulting, Guidepoint Global, Imperative Care, Medtronic, Rebound, StimMed; Advisory Board: Stryker (AIS Clinical Advisory Board), NeXtGen Biologics, MEDX, Cognition Medical, Endostream Medical;

Site Principal Investigator: CONFIDENCE study (MicroVention), STRATIS Study—Sub I (Medtronic).

AHS: Financial interest/investor/stock options/ownership: Adona Medical, Inc, Amnis Therapeutics, (Purchased by Boston Scientific October 2017), Blink TBI Inc., Buffalo Technology Partners Inc., Cerebrotech Medical Systems, Inc., Cognition Medical, Endostream Medical Ltd., Imperative Care, International Medical Distribution Partners, Neurovascular Diagnostics Inc., Q'Apel Medical Inc, Rebound Therapeutics Corp. (Purchased 2019 by Integra Lifesciences, Corp), Rist Neurovascular Inc., Sense Diagnostics, Inc., Serenity Medical Inc., Silk Road Medical, Spinnaker Medical, Inc., StimMed, Synchron, Three Rivers Medical Inc., Vastrax, LLC, VICIS, Inc., Viseon Inc; Consultant/advisory board: Amnis Therapeutics, Boston Scientific, Canon Medical Systems USA Inc., Cerebrotech Medical Systems Inc., Cerenovus, Corindus Inc., Endostream Medical Ltd., Imperative Care, Inc. Integra LifeSciences Corp., Medtronic, MicroVention, Minnetronix Neuro, Inc., Northwest University—DSMB Chair for HEAT Trial, Penumbra, Q'Apel Medical Inc., Rapid Medical, Rebound Therapeutics Corp.(Purchased by Integra LifeSciences Corp.), Serenity Medical Inc., Silk Road Medical, StimMed, Stryker, Three Rivers Medical, Inc., VasSol, W.L. Gore & Associates; Principal investigator/steering committee for the following trials: Cerenovus NAPA and ARISE II; Medtronic SWIFT PRIME and SWIFT DIRECT; MicroVention FRED & CONFIDENCE; MUSC POSITIVE; and Penumbra 3D Separator, COMPASS, INVEST, TIGER.

References

1. Miller NR. Diagnosis and management of Dural carotid-cavernous sinus fistulas. *Neurosurg Focus* 2007;23:E13.
2. Law G, Docherty G. Dural carotid cavernous fistula following hypertensive emergency. *BMJ Case Rep* 2019;12:e230823.
3. Kohli GS, Patel BC. Carotid Cavernous Fistula, in StatPearls. Treasure Island (FL): StatPearls Publishing Copyright© 2021, StatPearls Publishing LLC; 2021.
4. Barrow DL, Spector RH, Braun IF, Landman JA, Tindall SC, Tindall GT. Classification and treatment of spontaneous carotid-cavernous sinus fistulas. *J Neurosurg* 1985;62:248-56.
5. Alexander MD, Halbach VV, Hallam DK, Cooke DL, Ghodke B, Dowd CF, *et al.* Relationship of clinical presentation and angiographic findings in patients with indirect cavernous carotid fistulae. *J Neurointerv Surg* 2019;11:937-9.
6. Ke L, Yang YN, Yuan J. Bilateral carotid-cavernous fistula with spontaneous resolution: A case report and literature review. *Medicine (Baltimore)* 2017;96:e6869.
7. Khan S, Gibbon C, Johns S. A rare case of bilateral spontaneous indirect carotid-cavernous fistula treated previously as a case of conjunctivitis. *Ther Adv Ophthalmol* 2018;10:2515841418788303.
8. Meyers PM, Halbach VV, Dowd CF, Lempert TE, Malek AM, Phatouros CC, *et al.* Dural carotid cavernous fistula: Definitive endovascular management and long-term follow-up. *Am J Ophthalmol* 2002;134:85-92.
9. Haugen OH, Sletteberg O, Thomassen L, Kråkenes J. Bilateral

- non-traumatic carotid cavernous sinus fistula with spontaneous closure. *Acta Ophthalmol (Copenh)* 1990;68:743-7.
10. Al-Mufti F, Amuluru K, El-Ghanem M, Changa AR, Singh IP, Gandhi CD, *et al.* Spontaneous bilateral carotid-cavernous fistulas secondary to cavernous sinus thrombosis. *Neurosurgery* 2017;80:646-54.
 11. Kurata A, Takano M, Tokiwa K, Miyasaka Y, Yada K, Kan S. Spontaneous carotid cavernous fistula presenting only with cranial nerve palsies. *AJNR Am J Neuroradiol* 1993;14:1097-101.
 12. Lewis AI, Tomsick TA, Tew JM Jr. Management of 100 consecutive direct carotid-cavernous fistulas: Results of treatment with detachable balloons. *Neurosurgery* 1995;36:239-44.
 13. Iampreechakul P, Tirakotai W, Tanpun A, Wattanasen Y, Lertbusayanukul P, Siritwimonmas S. Spontaneous resolution of direct carotid-cavernous fistulas: Case series and literature review. *Interv Neuroradiol* 2019;25:71-89.
 14. Henderson AD, Miller NR. Carotid-cavernous fistula: Current concepts in aetiology, investigation, and management. *Eye (Lond)* 2018;32:164-72.
 15. Chen CC, Chang PC, Shy CG, Chen WS, Hung HC. CT angiography and MR angiography in the evaluation of carotid cavernous sinus fistula prior to embolization: A comparison of techniques. *AJNR Am J Neuroradiol* 2005;26:2349-56.