

JAMA Ophthalmology Clinical Challenge

Red Eye and Choroidal Detachment in an Older Woman

Sourabh Jadhav, MBBS, MS, DNB; Krishnakumar Padmalakshmi, DNB; Muna Bhende, MS

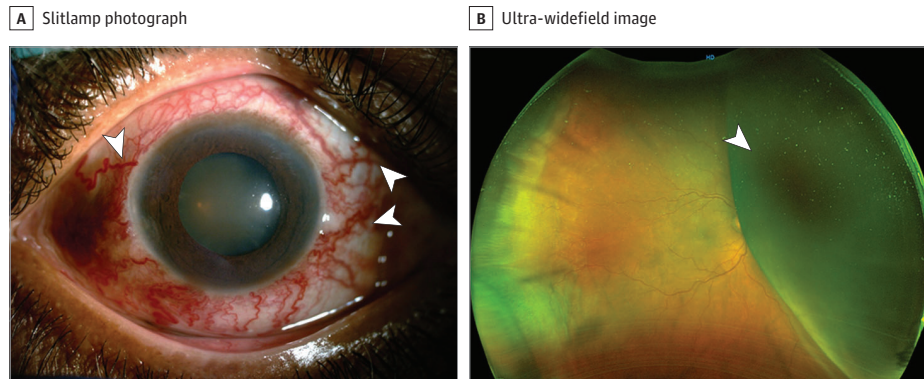


Figure 1. Slitlamp photograph of the right eye showing corkscrew episcleral vessels (A) and ultra-wide field imaging of the right eye showing a large choroidal detachment nasally and shallow detachment all around (B).

A 64-year-old woman presented with diminution of vision, progressive redness, and dull aching pain in the right eye for 8 months. There was no history of trauma. Her medical history was unremarkable.

On examination, her best-corrected visual acuity measured 20/60 OD and 20/25 OS. The anterior segment showed dilated corkscrew conjunctival vessels (Figure 1), a shallow anterior chamber in the right eye, and nuclear cataract in both eyes. Pupillary reaction and color vision were normal in both eyes. Intraocular pressure was 16 mm Hg by Goldman applanation tonometry in both eyes. Gonioscopy revealed an occludable angle in the right eye without evidence of blood in the Schlemm canal. Both eyes' extraocular movements were full and free in all directions of gaze. Dilated fundus examination showed a clear vitreous cavity in the right eye, 360° serous choroidal detachment partly obscuring the optic disc nasally (Figure 1), and the fundus in the left eye was unremarkable.

WHAT WOULD YOU DO NEXT?

- A. Ultrasound biomicroscopy
- B. Uveitis workup and steroids
- C. Thyroid profile
- D. Magnetic resonance imaging with magnetic resonance angiography or digital subtraction angiography

+ CME Quiz at jamacmelookup.com

Diagnosis

Low-flow carotid cavernous fistula

What to Do Next

D. Magnetic resonance imaging with magnetic resonance angiography or digital subtraction angiography

Discussion

Carotid cavernous fistula (CCF) is an abnormal communication between the carotid artery and its branches and the cavernous sinus (CS), causing high-pressure arterial blood flow within the low-pressure veins, sinuses, and cavernous sinus.¹ Direct CCF (high-flow fistula) is a direct connection between the intracavernous segment of the internal carotid artery and CS, commonly caused by head injury. Presentations include blurred vision, pulsatile proptosis, orbital bruit, chemosis, corkscrew conjunctival vessels, raised intraocular pressure, and nerve palsies.^{1,2}

Indirect CCF (low-flow fistula) involves 1 or more meningeal branches of the internal carotid artery, external carotid artery, or both, communicating with CS. It can be spontaneous, associated with hypertension, older age, and female gender. Often asymptomatic, the most common presentation is conjunctival congestion misdiagnosed as conjunctivitis.^{1,3} This patient had corkscrew vessels with serous choroidal detachment (CD) and normal intraocular pressure.^{4,5} This may occur due to increased orbital venous pressure and intracapillary pressure in the choroid, resulting in transudation into the suprachoroidal space.⁵

When CCF is suspected clinically, the patient requires neuroimaging (magnetic resonance imaging of the brain and orbit/magnetic resonance angiography). Digital subtraction angiography is the gold standard for diagnosis^{1,6} (choice D). Magnetic resonance angiography in this case did not reveal dilatation of the superior ophthalmic vein or abnormal flow in CS. Due to strong clinical suspicion, the patient underwent digital subtraction angi-

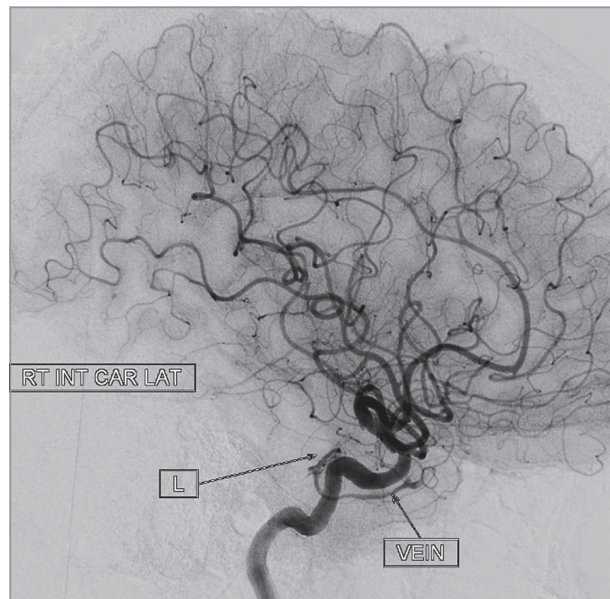


Figure 2. Digital subtraction angiographic image after transfemoral catheterization showing right-sided low flow type B carotid cavernous fistula (L marks the fistula). RT indicates right; INT, internal; CAR, carotid artery; LAT, lateral view.

ography, which showed a small extremely low-flow type B CCF (dural shunts between the meningeal branches of the internal carotid artery and CS) on the right side. (Figure 2) Jamison et al⁷ described

a patient with persistent red eye and glaucoma, later found to have CCF. The current case emphasizes the need to suspect CCF when a patient presents only with CD and red eye. In asymptomatic cases, close observation or ipsilateral manual carotid compression can achieve a success rate of 35%, with resolution occurring between 6 weeks and several months; 20% to 70% of these cases resolve spontaneously.¹ Surgical options for closure include endovascular treatment (transarterial or transvenous) and stereotactic radiosurgery.^{1,6}

Choice A (ultrasound biomicroscopy) is not the best option as there was no mass lesion, no sentinel conjunctival vessels (choroidal melanoma), severe pain, or inflammation (scleritis). Choice B (uveitis workup and steroids) is not an option as there was no evidence of inflammation. Choice C (thyroid profile) was not the best option because though chronic congestion can occur in thyroid eye disease, other features like eyelid retraction, proptosis, impaired extraocular movement, and systemic features of thyroid disease were absent. Low-flow CCF should be considered in cases of chronic red eye and CD. Prompt neuroimaging is essential to initiate appropriate management.

Patient Outcome

The patient was advised to perform ipsilateral carotid compression with the contralateral hand for 30 seconds⁸ with 30-second release 6 to 7 times per hour in a sitting or supine posture, 40 to 50 times per day for 8 weeks. This resulted in resolution of the CD. Follow-up was advised every 4 to 6 weeks. Intermittent manual carotid compression is the initial choice of management for low-flow indirect CCF before more aggressive intervention.

ARTICLE INFORMATION

Author Affiliations: Sri Bhagwan Mahavir, Vitreoretinal Services, Sankara Nethralaya, Chennai, India (Jadhav, Bhende); Department of Neuro-Ophthalmology, Medical Research Foundation, Sankara Nethralaya, Chennai, India (Padmalakshmi).

Corresponding Author: Muna Bhende, MS, Shri Bhagwan Mahavir Vitreoretinal Service, Medical Research Foundation, Nethralaya, 41 College Rd, Nungambakkam, Chennai 600006, Tamil Nadu, India (drmunab@snmail.org).

Published Online: May 18, 2023.
doi:10.1001/jamaophthalmol.2023.1513

Correction: This article was corrected on June 8, 2023, to delete the corresponding author's middle initial.

Conflict of Interest Disclosures: None reported.

Additional Contributions: We thank the patient for granting permission to publish this information.

We also thank Ambika Selvakumar, MD, Department of Neuro-Ophthalmology, Medical Research Foundation, and Swatee Halbe, MD, Apollo Specialty Hospitals.

REFERENCES

- Henderson AD, Miller NR. Carotid-cavernous fistula: current concepts in aetiology, investigation, and management. *Eye (Lond)*. 2018;32(2):164-172. doi:10.1038/eye.2017.240
- 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(2):248-256. doi:10.3171/jns.1985.62.2.0248
- Slusher MM, Lenington BR, Weaver RG, Davis CHJ Jr. Ophthalmic findings in dural arteriovenous shunts. *Ophthalmology*. 1979;86(5):720-731. doi:10.1016/S0161-6420(79)35454-9
- Berk AT, Ada E, Kir E, Saatci AO. Choroidal detachment associated with direct spontaneous carotid-cavernous sinus fistula. *Ophthalmologica*. 1997;211(1):53-55. doi:10.1159/000310875
- Komiyama M, Nishikawa M, Yasui T. Choroidal detachment and dural carotid-cavernous sinus fistula—case report. *Neurol Med Chir (Tokyo)*. 1997;37(6):459-463. doi:10.2176/nmc.37.459
- Texakalidis P, Tzoumas A, Xenos D, Rivet DJ, Reavey-Cantwell J. Carotid cavernous fistula (CCF) treatment approaches: a systematic literature review and meta-analysis of transarterial and transvenous embolization for direct and indirect CCFs. *Clin Neurol Neurosurg*. 2021;204:106601. doi:10.1016/j.clineuro.2021.106601
- Jamison A, Siddiqui A, Lockington D. Persistent red eye unresponsive to topical treatment. *JAMA Ophthalmol*. 2021;139(1):119-120. doi:10.1001/jamaophthalmol.2020.4062
- Gemmete JJ, Chaudhary N, Pandey A, Ansari S. Treatment of carotid cavernous fistulas. *Curr Treat Options Neurol*. 2010;12(1):43-53. doi:10.1007/s11940-009-0051-3