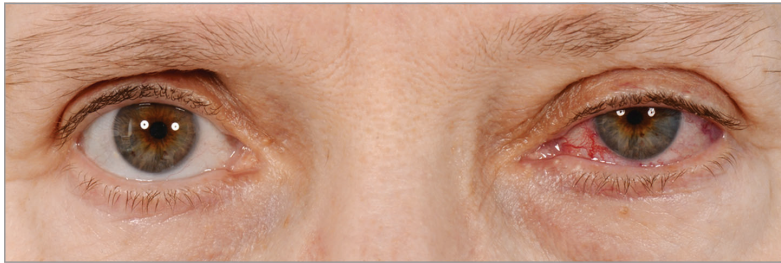


## JAMA Ophthalmology Clinical Challenge

## Persistent Red Eye Unresponsive to Topical Treatment

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A Normal appearance of the right eye



B Dilated tortuous bulbar blood vessels in the left eye



Figure 1. Clinical photographs.

**A 68-year-old woman** presented to the emergency eye clinic with a 3-week history of progressive left eye redness and discomfort unresponsive to topical antibiotics or lubricants, as prescribed by her optometrist. The intraocular pressure (IOP) was raised (26 mm Hg) and was treated with a topical  $\beta$ -blocker. Otherwise, her condition was managed conservatively for presumed viral conjunctivitis.

She self-presented 6 days later with intermittent visual blurring, mild eyelid swelling, and conjunctival chemosis. The IOP remained raised (24 mm Hg). An inflammatory or allergic etiology was suspected; therefore, topical corticosteroids and a prostaglandin analogue were also prescribed.

One week later at planned follow-up, the patient had dilated and tortuous bulbar blood vessels (Figure 1). Further interrogation of the history revealed a left-sided nonspecific dull headache, constant for the previous 2 months, and an annoying "whooshing" noise in the patient's left ear.

## WHAT WOULD YOU DO NEXT?

- A. Suggest supportive strategies (eg, cold compresses, ocular lubricants) and consider increasing the topical corticosteroid treatment or prescribing a topical nonsteroidal anti-inflammatory drug (NSAID)
- B. Prescribe an oral NSAID and/or systemic immunosuppression (oral corticosteroid) and perform chest radiography, urinalysis, blood tests, and other studies
- C. Refer the patient for urgent neuroimaging
- D. Perform a general examination (for a port-wine stain) and dilated ophthalmoscopy

[Quiz at jamacmelookup.com](https://www.jama.com/lookup/quiz)

## Diagnosis

**Carotid-cavernous fistula**

## What to Do Next

**C.** Refer the patient for urgent neuroimaging

## Discussion

Clinically, the most likely diagnosis is carotid-cavernous fistula (CCF)—an abnormal communication between the carotid artery (or its branches) and the cavernous sinus, causing high-pressure arterial blood flow within the low-pressure veins, sinuses, and cavernous sinus.<sup>1,2</sup> Arterialized ("corkscrew") conjunctival blood vessels are shown

in Figure 1. Other features of this case consistent with CCF included raised IOP (due to increased episcleral venous pressure), conjunctival chemosis, and visual blurring. Significant visual loss may occur secondary to traumatic, ischemic, or glaucomatous optic neuropathy.

In 75% of CCF cases there is a direct connection between the intracavernous segment of the internal carotid artery (ICA) and the cavernous sinus, usually due to arterial wall trauma after head injury.<sup>3</sup> Acute high-flow symptoms of direct CCFs (commonly used terms) include visual blurring, motility disorders, proptosis, chemosis, and orbital bruits.

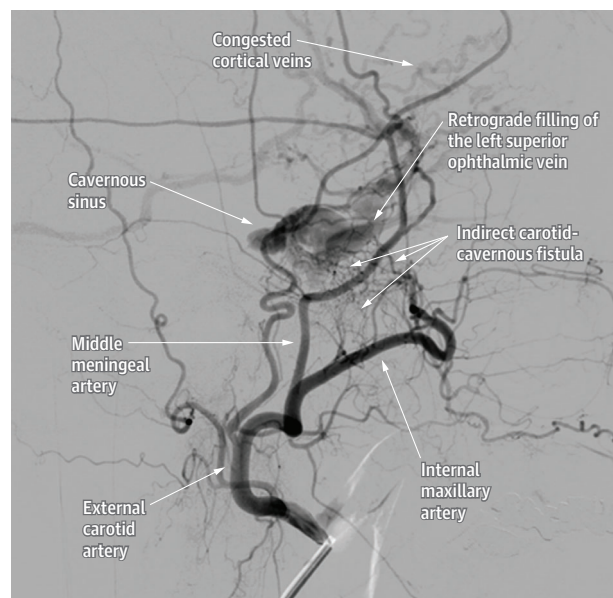
In the remaining 25% of cases, the cavernous sinus communicates with 1 or more meningeal branches of the ICA, external carotid artery, or both. Indirect CCFs (also known as dural CCFs) present with chronic low-flow symptoms, including raised IOP, conjunctival engorgement, and chemosis. Associated systemic conditions, which careful investigation may detect, include venous thrombosis, hypertension, fibromuscular dysplasia, Ehlers-Danlos syndrome type IV, and dissection of the ICA.

When CCF is suspected clinically, the patient's condition should be discussed urgently with interventional neuroradiology and/or neurosurgery specialists regarding appropriate investigations, such as computed tomography (CT) or magnetic resonance angiography and catheter cerebral angiography (choice C).<sup>1</sup> This patient's CT intracranial angiographic image revealed a prominent left cavernous sinus and arterial enhancement of the sinus and left superior ophthalmic vein, consistent with CCF. Figure 2 shows a subsequent digital subtraction angiographic image.

In the absence of high-risk features, such as vision loss or cortical venous drainage, conservative options may be appropriate, including close observation (70% of dural CCFs close spontaneously). Manual intermittent compression of the ipsilateral cervical carotid artery by the patient's contralateral hand is successful in 35% of cases but is contraindicated because of the risk of stroke when carotid atherosclerotic disease exists.<sup>1</sup>

Definitive surgical intervention aims to completely occlude the fistula while maintaining flow within the ICA.<sup>4-5</sup> Endovascular approaches (usually transvenous via the inferior petrosal sinus, but occasionally via the transorbital intraarterial route) use coils or, less commonly, detachable balloons or liquid embolic agents. These approaches successfully occlude the fistula in 90% to 100% of cases, with a mortality rate of less than 1% and a low rate of major complications (notably, cerebral ischemic event, hemiparesis, and permanent ocular motor nerve palsy).<sup>1</sup> Open surgical approaches may be adopted if endovascular intervention fails.

Choice A (supportive strategies) was based on an incorrectly assumed diagnosis of nonspecific (potentially allergic) conjunctivitis, choice B (prescription of an NSAID and/or immunosuppression), on



**Figure 2.** Digital subtraction angiographic image after transfemoral catheterization. A left-sided dural carotid-cavernous fistula is noted, with supply from both the external and internal carotid artery branches, with cortical venous drainage and retrograde filling of the left superior ophthalmic vein.

a diagnosis of scleritis; and choice D (examination to identify a port-wine stain and/or a choroidal hemangioma), on the suspicion of Sturge-Weber syndrome (given the dilated blood vessels, headache, and raised IOP). Sturge-Weber syndrome is one of the phakomatoses, which may feature port-wine stain, seizures, headaches, choroidal hemangioma, and both dilated bulbar vessels and raised IOP or glaucoma (resulting from raised episcleral venous pressure). All 3 differential diagnoses merit consideration, but investigation to exclude or identify CCF must be prioritized because early treatment may avoid substantial morbidity.

Carotid-cavernous fistula should be considered in all cases of persistent red eye, arterialized conjunctival vessels, or chemosis. When clinical suspicion exists, liaising with interventional neuroradiology and/or neurosurgery specialists regarding appropriate urgent neuroimaging, including CT/magnetic resonance angiography or catheter cerebral angiography, should ensure prompt diagnosis and intervention.

### Patient Outcome

This patient underwent successful transvenous coil embolization. Post-treatment digital subtraction angiography showed coils placed within the left cavernous sinus and complete closure of the treated fistula.

### ARTICLE INFORMATION

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