

# An Arteriovenous Malformation Masquerading as a Carotid-Cavernous Sinus Fistula

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**PURPOSE:** To describe a case of an occipital arteriovenous malformation (AVM) presenting with unilateral signs mimicking a carotid-cavernous sinus fistula (CCF).

**DESIGN:** Interventional case report.

**METHODS:** A 67-year-old normotensive male presented with loss of vision, pain, and proptosis of the right eye. Best-corrected visual acuity was right eye 6/36 and left eye 6/5. Examination of the right eye revealed orbital congestion with arteriolization of the episcleral vessels and an intra-ocular pressure of 44 mm Hg which was refractory to medical treatment.

**RESULTS:** Computed tomography (CT) angiogram and cerebral angiography found an occipital AVM draining into the right sphenoparietal sinus and thereafter the right ophthalmic vein, bypassing the cavernous sinus. Embolization followed by excision of the AVM resulted in recovery of vision, reversal of proptosis, and normalization of intraocular pressure.

**CONCLUSIONS:** Rapid diagnosis and immediate intervention resulted in a rewarding visual recovery despite a persisting left homonymous hemianopia. (*Am J Ophthalmol* 2005;140:548–550. © 2005 by Elsevier Inc. All rights reserved.)

POSTERIORLY LOCATED INTRA-CRANIAL ARTERIOVENOUS malformations (AVMs) may drain anteriorly into the orbital venous system and present with signs masquerading as a carotid-cavernous sinus fistula (CCF).<sup>1</sup> A search using MEDLINE reveals that this is exceedingly rare.<sup>1,2</sup> Here, we describe a patient presenting with unilateral signs suggestive of a CCF who was subsequently found to have an occipital AVM without cavernous sinus involvement.

• **CASE REPORT:** A 67-year-old normotensive male presented with sudden loss of vision in the right eye associated

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with severe pain and proptosis. He had no history of recent trauma but gave a 2-year history of progressive redness and prominence of the right eye.

His best-corrected visual acuities were right eye 6/36 and left eye 6/5. Examination of the right eye revealed eyelid edema, associated with a non-pulsatile proptosis of 7 mm. There was an afferent pupil defect, reduced color vision discrimination, and an almost complete ophthalmoplegia. Slit-lamp biomicroscopy revealed prolapsing conjunctival chemosis, arteriolized episcleral vessels, mild corneal stromal edema, dilated iris vessels, and a semidilated pupil (Figure 1, top). The intraocular pressure (IOP) was 44 mm Hg (left IOP 20 mm Hg). Fundoscopy revealed tortuous retinal vessels, retinal edema, and a cup:disk ratio of 0.6 (left cup:disk ratio 0.2). Gonioscopy revealed an angle open 360 degrees Schaffer grade 3, with no neovascularization. Examination of the left eye was normal. No bruit of the globe or scalp was found.

A computed tomography and computed tomography angiogram of the brain and orbit revealed an occipital AVM and a markedly distended right ophthalmic vein (Figure 2). Cerebral angiography revealed that the occipital AVM was supplied mainly by the right posterior cerebral artery with a contribution from the right occipital and posterior branches of the right middle meningeal artery, but none from the internal carotid artery (Figure 3). Drainage was predominantly through a superficial cortical vein into the right sphenoparietal sinus and, thereafter, the right ophthalmic vein, bypassing the cavernous sinus.

Intravenous acetazolamide and mannitol, and topical anti-glaucoma medications failed to normalize the right IOP. Embolization of the AVM, within 24 hours of presentation, resulted in improved ocular motility, a 3-mm reduction in proptosis, and an IOP of 21 mm Hg. Excision of the AVM by means of an open craniotomy was performed the following day.

One month post-operatively, the right best-corrected visual acuity was 6/6. Examination of the right eye revealed a fixed semidilated pupil, glaucomflecken of the anterior subcapsular lens, full ocular motility, a residual proptosis of 3 mm, and an almost complete reversal of venous arteriolization (Figure 1, bottom). The right IOP was 14 mm Hg off all treatment. Automated perimetry revealed a left homonymous hemianopia without macular sparing.

AVMs are classified according to their arterial supply; dural, supplied by meningeal branches; pial, from cerebral and cerebellar arteries; and mixed dural-pial, with contributions from both sources.<sup>2</sup> This case illustrates a mixed dural-pial AVM.

Neuro-ophthalmic manifestations of cerebral AVMs include anterior visual pathway compression,<sup>2</sup> visual field defects,<sup>2,3</sup> oculomotor nerve palsies,<sup>4,5</sup> and raised intracranial pressure.<sup>3</sup>

Orbital drainage from cerebral AVMs is rare.<sup>2</sup> Major re-routing of venous blood may occur in a retrograde direc-



FIGURE 1. Photographs showing ocular appearance before and after excision of the right-sided occipital arteriovenous malformation (AVM). (Top) Photograph illustrates pre-operative conjunctival chemosis, proptosis, and semidilated pupil. (Bottom) Photograph shows post-operative reversal of arteriolization, chemosis, and proptosis.

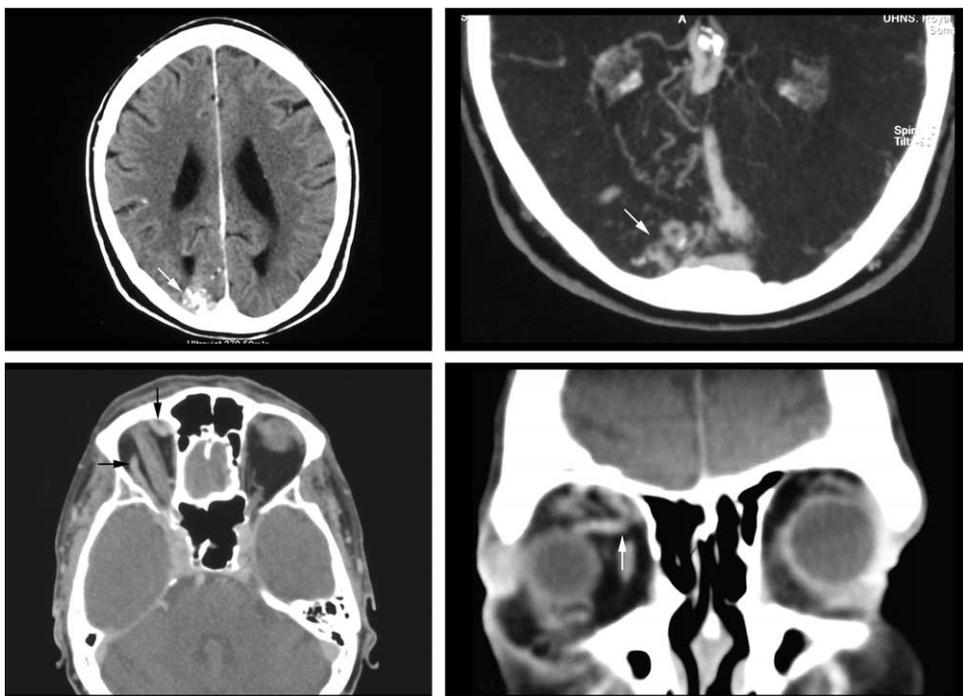


FIGURE 2. A computed tomography and computed tomography angiogram of the brain (top right and left) reveal a right occipital lobe AVM (white arrows). A computed tomography of the orbits (bottom right and left) reveals a dilated right superior ophthalmic vein (bottom left, black arrows; bottom right, white arrow) resulting from drainage of the occipital AVM into the right sphenoparietal sinus and then the orbital circulation.

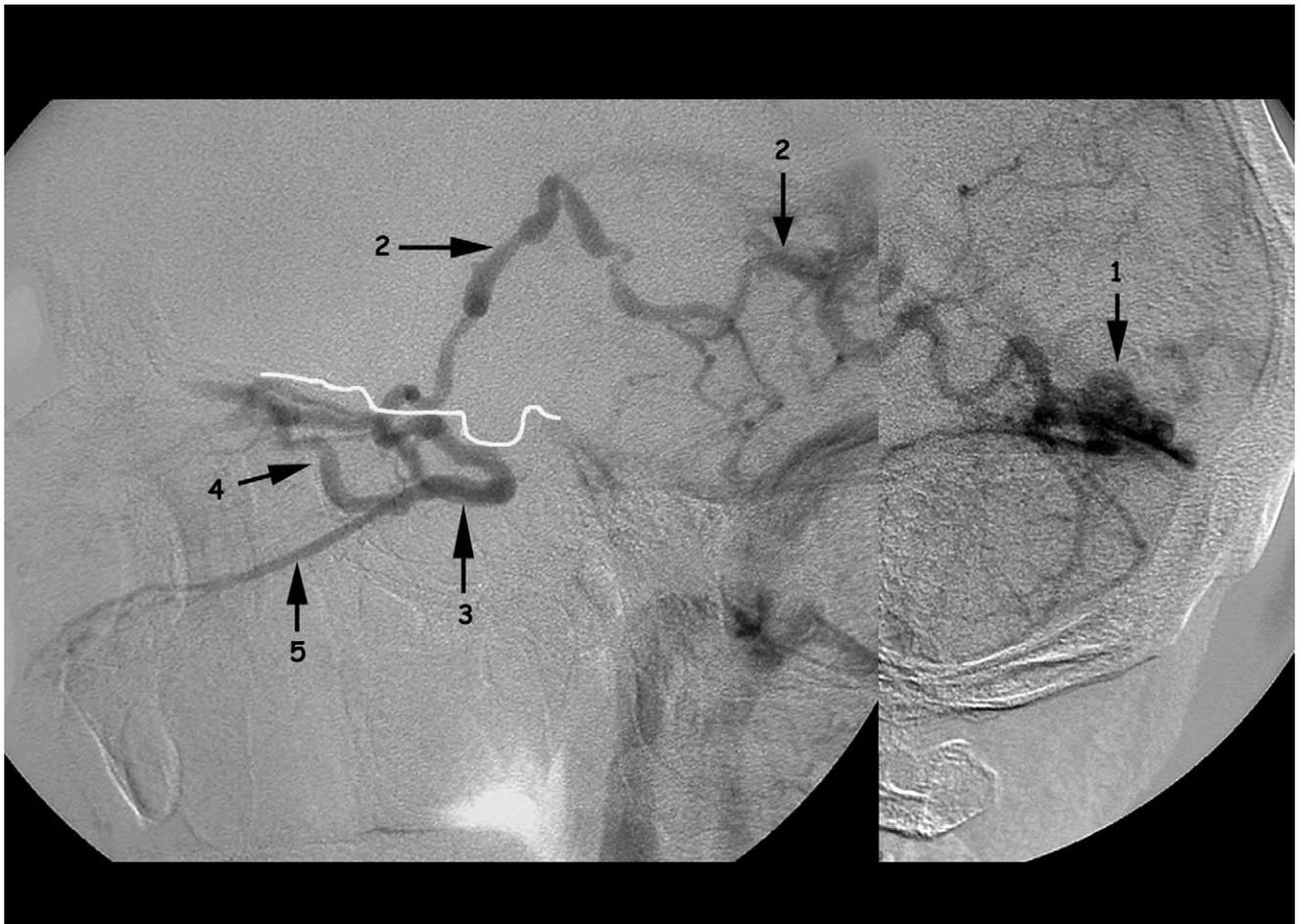


FIGURE 3. A Cerebral angiogram digitally reconstructed using sequential films shows the occipital arteriovenous malformation (AVM) and its drainage. Indicated are <sup>1</sup> AVM, <sup>2</sup> superficial cortical draining vein, <sup>3</sup> sphenoparietal sinus, <sup>4</sup> dilated superior, and <sup>5</sup> inferior ophthalmic veins. The position of the sella turcica is outlined in white.

tion through the basal, cortical, and dural veins toward the sphenoparietal and cavernous sinuses, and subsequently into the orbital venous system.<sup>1</sup> However, not all cases of AVMs that drain orbitally result in the constellation of signs characteristic of CCF; some may have signs of orbital congestion without the presence of proptosis, glaucoma, and episcleral arteriolization.<sup>2</sup> Additionally, bruits over the eye and pulsating exophthalmos, which are common in CCF, are rarely present in dural AVMs,<sup>4</sup> where bruits are more commonly heard over the cranium.<sup>3</sup>

Because the early ocular clinical signs resulting from an intra-cranial AVM are often subtle, many patients are initially treated for non-specific ocular inflammation, resulting in delayed diagnosis and management.<sup>4</sup> Despite the late presentation in this case, rapid diagnosis and immediate intervention, resulting from close cooperation between the ophthalmic and neurosurgical teams, resulted in visual recovery.

#### REFERENCES

1. Buchanan TAS, Harper DG, Hoyt WF. Bilateral proptosis, dilatation of conjunctival veins, and papilloedema: a neuro-ophthalmological syndrome caused by arteriovenous malformation of the torcular Herophili. *Br J Ophthalmol* 1982;66: 186–189.
2. Volpe NJ, Sharma MC, Galetta SL, et al. Orbital drainage from cerebral arteriovenous malformations. *Neurosurgery* 2000;46:820–824.
3. Newton TH, Weidner W, Greitz T. Dural arteriovenous malformation in the posterior fossa. *Radiology* 1968;90:27–35.
4. Moster MR, Kennerdell JS. B-Scan ultrasonic evaluation of a dilated superior ophthalmic vein in orbital and retro-orbital arteriovenous anomalies. *J Clin Neuro-ophthalmol* 1983;3: 105–108.
5. Forman AR, Luessenhop AJ, Limaye SR. Ocular findings in patients with arteriovenous malformations of the head and neck. *Am J Ophthalmol* 1975;79:626–633.