

Orbital arteriovenous malformation mimicking cavernous sinus dural arteriovenous malformation

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Abstract

Aims—Orbital arteriovenous malformations (OAVM) are rare, mostly described with high flow characteristics. Two cases are reported with an OAVM of distinct haemodynamic abnormality. The clinical, angiographic features, and the management considerations are discussed.

Methods—Case review of two patients with dural AVM (DAVM) who presented to referral neuro-ophthalmology and endovascular services because of clinical symptoms and signs consistent with a cavernous sinus dural AVM.

Results—In each patient, superselective angiography revealed a small slow flow intraorbital shunt supplied by the ophthalmic artery. The transarterial and transvenous endovascular approaches to treat the malformation were partially successful. Although, the abnormal flow was reduced, complete closure of the DAVM could not be accomplished without significant risk of iatrogenic injury. Neither patient's vision improved after intervention.

Conclusion—A DAVM in the orbit can cause similar clinical symptoms and signs to those associated with a cavernous sinus DAVM. Even with high resolution magnetic resonance imaging, only superselective angiography can identify this small intraorbital slow flow shunt. The location in the orbital apex and the small size precludes a surgical option for treatment. The transarterial and transvenous embolisation options are limited.

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Arteriovenous malformations (AVM) located within the orbit are rare. Wright reported that among 627 patients with orbital disease only three had arteriovenous shunts.¹ Among the reported intraorbital AVMs, many were high flow shunts located in the anterior orbit which caused proptosis, arterialisated conjunctival vessels, chemosis, and orbital swelling, and were amenable to intravascular embolisation or surgery or a combination of approaches.²⁻¹⁰ When an AVM involves the orbit, it is most often related to an intracranial or maxillofacial arteriovenous shunt.² Orbital dysfunction typically develops as a result of secondary venous hypertension in the ophthalmic veins from AV shunt outside of the orbit, such as with a carotid cavernous fistula or a dural AVM in the cavernous sinus region. We describe two patients, each with an orbital AVM and the rare clinical pres-

entation suggestive of a slow flow dural AVM of cavernous sinus region.

Case reports

CASE 1

A 66 year old Afro-American man presented with symptoms of the sudden onset of left eye pain, redness, and protrusion beginning 3 weeks before his evaluation. He also was bothered by intermittent pulsatile sounds when reclining. His ocular history was significant for open angle glaucoma poorly controlled with oral methazolamide and topical timolol 0.5% in each eye. The visual acuity in the left eye was decreased because of glaucoma for several years before this new problem. His previous medical history was significant for systemic hypertension and diabetes mellitus. He had a renal transplant after renal failure of unknown aetiology and a subtotal parathyroidectomy 1 year later.

The visual acuity was 20/40 in his right eye and hand movements in the left eye. He saw 11 of 15 colour plates with the right eye and he could not see any test or the Ishihara control plate with his left eye. By applanation, the intraocular pressure was 26 mm Hg in his right eye and 38 mm Hg in his left eye. In the left orbit, there was 3 mm of proptosis, upper lid ptosis, periorbital oedema, marked injection, and arterialisation of the conjunctival and episcleral vessels. No bruits were noted on auscultation. Ocular movements of the right eye were full while those of the left eye were mildly limited in all directions. A relative afferent pupillary defect was noted in the left eye. Gonioscopy of both eyes revealed a three plus open angle in all quadrants with no blood in Schlemm's canal. Ophthalmoscopy of the right eye showed a cup to disc ratio of 0.8 and a macular epiretinal membrane. In the left eye, the cup to disc ratio was 0.9 and the macula appeared normal. Threshold perimetry in the right eye demonstrated a dense loss of almost the entire superior field with mild diffuse depression in the remaining field and a mean deviation score of -19.34 dB.

Magnetic resonance imaging (MRI) of the brain and orbits revealed bilaterally enlarged superior ophthalmic veins but no abnormality of either cavernous sinus. The clinical impression was that the patient suffered from a cavernous sinus DAVM that drained anteriorly to both superior ophthalmic veins. An angiogram was performed with the intention of locating and closing the shunt by endovascular techniques. The left internal carotid angiogram revealed a small, slow flow DAVM at the apex of the orbit (Fig 1) supplied by the

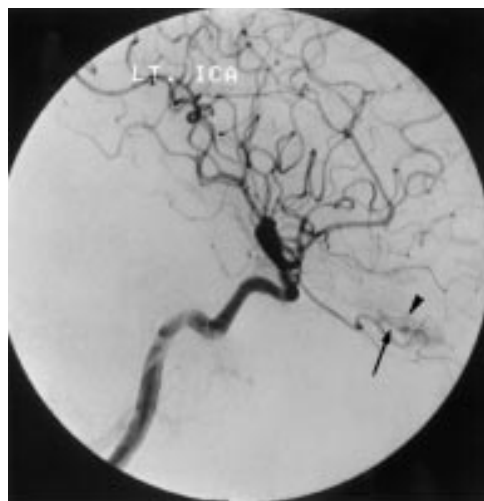


Figure 1 Lateral left internal carotid angiogram demonstrates the ophthalmic artery supply (arrow) to the intraorbital AV shunt with drainage to the ophthalmic vein (arrowhead) in the posterior orbit.

ophthalmic artery (Fig 2). The left internal maxillary artery injection demonstrated only indirect supply to the shunt via the ophthalmic artery (not shown). The shunt drained posteriorly to the cavernous sinus (Fig 2B). Polyvinyl alcohol embolisation of the distal internal maxillary artery was performed in an attempt to reduce the flow in the arteriovenous shunt by blocking ethmoidal and anterior deep temporal artery collateral supply to the ophthalmic artery, but the shunt was not significantly altered.

The following day the patient complained of left orbital pain. Examination of the left eye revealed severe chemosis of the lower conjunctiva, the intraocular pressure was 39 mm Hg, and gonioscopy showed an open angle. Manometric pressure was intermittently applied to the left orbit in order to try and produce thrombosis in the remaining orbital shunt. The intraocular pressure decreased to 25–30 mm Hg. Two days after the end of the intermittent manometric pressure the left eye demonstrated no light perception and the intraocular pressure was 46 mm Hg.

A transvenous embolisation through the left superior ophthalmic vein was attempted. The patient underwent anterior orbitotomy but the ophthalmic venous system posterior to the globe could not be properly cannulated. During the attempt to enlarge the orbitotomy an extensive haemorrhage developed and the procedure was aborted. A repeat angiogram showed that the arteriovenous communication was still present with diminished flow anteriorly. After the procedure the vision was unchanged, the left eye intraocular pressure was 33 mm Hg, there was 7 mm of proptosis, upper lid oedema, significant injection, and prolapse of the left eye conjunctiva. Intermittent manometric pressure was again applied to induce thrombosis in the remaining shunt for 2 days without obvious clinical change.

After 2 years of follow up, the visual acuity in his left eye was hand movements in the temporal field, the intraocular pressure hovered in the

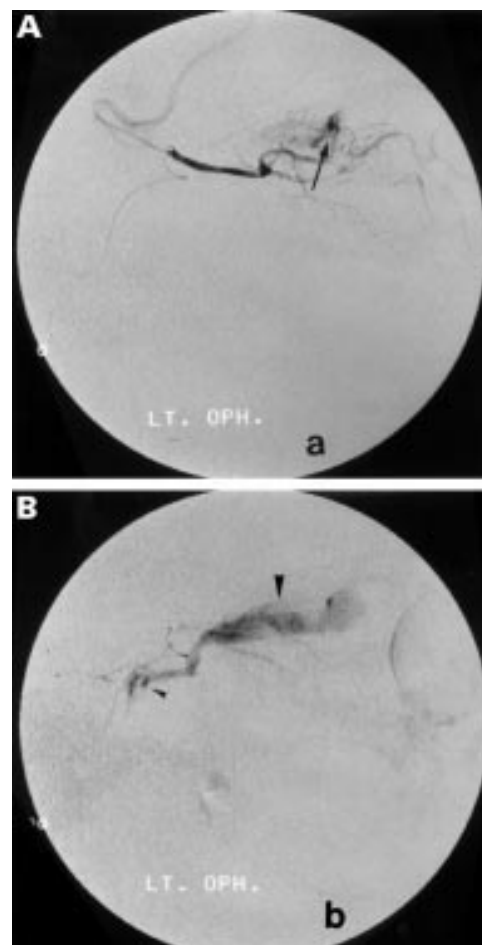


Figure 2 (a) Lateral early selective injection of the left ophthalmic artery shows the AV network (arrow). (b) The venous phase of the same angiogram demonstrates the enlarged superior division of the ophthalmic vein (large arrowhead) with outflow restriction anteriorly and some draining posteriorly to the cavernous sinus (small arrowhead).

mid-20s with glaucoma medications, there was 4 mm of proptosis, and no conjunctival chemosis. MRI with gadolinium still showed enlarged superior ophthalmic veins.

CASE 2

A 71 year old man presented with a swollen red right eye over a month's duration, without visual disturbance or headache. The medical history was significant for hypertension, nephrectomy for renal cell carcinoma, and partial thyroidectomy for a benign nodule.

The neuro-ophthalmic examination revealed visual acuity of 20/30 in the right eye and 20/20 in the left eye, full colour vision using Ishihara test plates, normal tangent perimetry with 2 mm white test object performed at 1 metre, proptosis of 4 mm in the right eye, marked limbal loop arterialised vessels in the right eye conjunctiva, and episclera. Both pupils were round and reactive to light without a relative afferent pupillary defect. Intraocular pressure was 18 mm Hg in the right eye and 10 mm Hg in the left eye. There was a mild shallowing of the anterior chamber in the right eye. Gonioscopy demonstrated the right eye had a one plus open angle in all quadrants and there was no

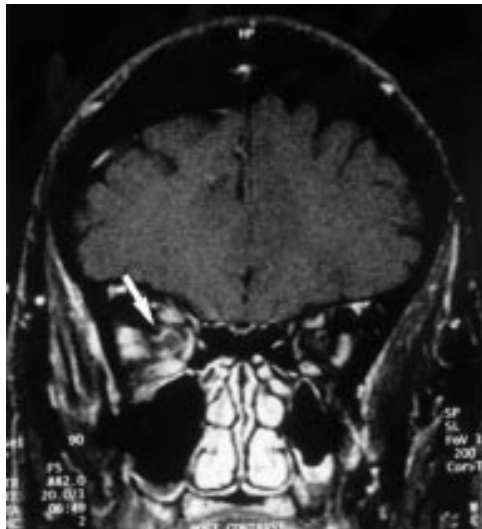


Figure 3 Coronal, fat suppressed, gadolinium enhanced magnetic resonance image of the orbit shows increased enhancement of the right optic nerve sheath (arrow).

blood in the Schlemm's canal. The right eye had a normal optic disc, 0.3 cup to disc ratio, dilated and tortuous retinal veins, mild macular oedema, and dot and blot small haemorrhages throughout the entire retina.

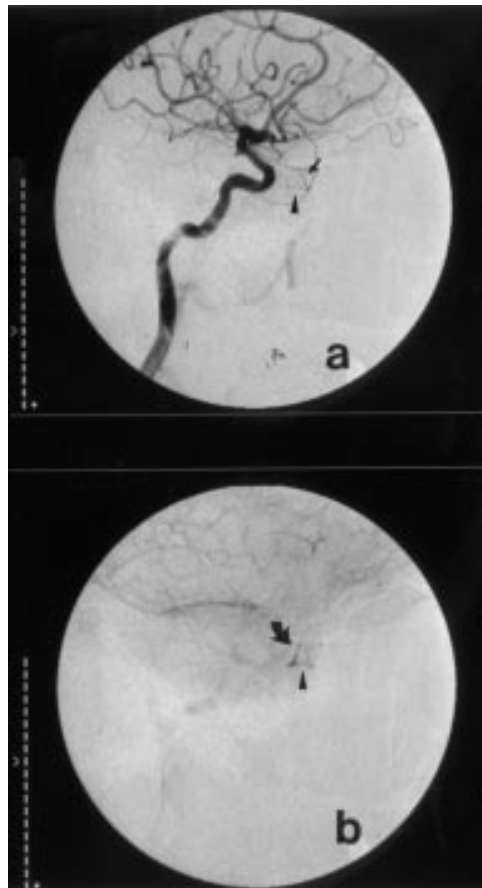


Figure 4 (a) Early lateral right internal carotid angiogram demonstrates a branch (arrow) from the ophthalmic artery supply to the AV shunt. The shunt venous drainage (arrowhead) curves posteriorly to the superior division of the ophthalmic vein. (b) The venous phase of the same angiogram shows the venous side of the shunt (arrowhead) emptying into the ophthalmic vein (curved arrow) with stagnant blood flow suggesting partial thrombosis.

Ophthalmoscopy of the left eye was normal with a 0.3 cup to disc ratio. No bruits of the orbit or posterior auricular regions were noted by auscultation.

The MRI and magnetic resonance angiography did not demonstrate any abnormalities in the cavernous sinus but the optic nerve sheath of the right orbit demonstrated abnormal enhancement after gadolinium infusion (Fig 3). No enlargement of the superior ophthalmic veins was noted, and there were no abnormalities in the intracranial venous system. The patient was thought to have a cavernous sinus region dural AVM with possible thrombosis in the ophthalmic venous system. Because of the mild degree of ocular dysfunction and marginal functioning kidney (creatinine 2.1 mg/dl) which increased the risk of the contrast agent used during endovascular therapy, expectant follow up was recommended.

Two months later in the right eye, the visual acuity decreased to 20/70 and the colour vision decreased to 60% of the Ishihara plates. The visual field became constricted and the intraocular pressure was elevated to 27 mm Hg. An angiogram was performed with the intention to locate and close the shunt with endovascular techniques. The right internal carotid angiogram demonstrated a small arteriovenous shunt in the posterior orbit supplied by the ophthalmic artery (Fig 4). The external carotid artery supplied the shunt only through indirect supply via collaterals to the right ophthalmic artery (Fig 5). Because the only route to embolise the shunt was through the ophthalmic artery which carried significant risk of causing right eye blindness, endovascular treatment was deferred. The patient was treated using intermittent manual compression of the cervical carotid artery with his left hand.¹¹ During a follow up period of 1 year, his vision deteriorated to 20/200, and the intraocular pressure remained at 22 mm Hg while using twice daily topical timolol 0.5%.

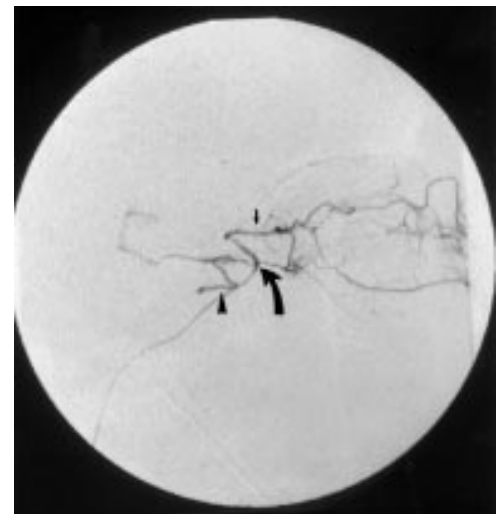


Figure 5 Lateral supraselective left middle meningeal artery injection demonstrates the AV shunt (arrowhead) filled via a middle meningeal artery collateral (curved arrow), probably through Hyrtl's canal to fill the ophthalmic artery (arrow) before supplying the shunt.

Discussion

An isolated intraorbital arteriovenous shunt is a rare vascular disorder of the orbit that arises from a congenital or traumatic cause.¹² Since most of the orbital AVMs reported have high flow characteristics they can be diagnosed non-invasively by MRI and ultrasound techniques. In contrast, our two cases presented with a clinical picture very similar to the one found in patients with cavernous sinus region dural AVMs that drain to the ophthalmic venous system. The sudden onset of symptoms in our cases might have been due to a thrombotic process in the venous drainage of the shunts. A patient who had an orbital dural AVM without visual loss, minimal orbital congestion, and an inferior division third nerve paresis, possibly due to thrombosis in the ophthalmic venous system, has been recently described.¹³

MRI did not demonstrate a definitive arteriovenous malformation in either of our patients. Only superselective angiography determined the exact location and haemodynamic features of each AVM. In both cases, the shunts were supplied by the ophthalmic artery and had very slow flow; and there was compromise of the ophthalmic venous drainage. In contrast to DAVM in the cavernous region,¹² the external carotid arterial system provided no access to treat the shunts. The arteriovenous anomaly appeared to involve the dura around the optic nerve or of the periorbita. The therapeutic options are very limited in any case of AVM in the posterior orbit, particularly when the dura of the optic nerve is involved. Owing to the poor systemic medical condition of our patients, they were not candidates for a craniotomy to unroof the orbit, which would have been needed to expose and resect the AVM. In addition, as suggested in at least one previous report, given the small size and low

flow, such an AVM might not be readily found during a surgical exploration.⁸ The endovascular intervention options were exhausted by attempts to embolise each lesion. Further embolisation attempts appeared to carry a high degree of a risk without assurance of success. In one case similar to our patients, although the AVM was occluded by embolisation and the proptosis decreased, the patient ended up with a blind eye.⁵ We would like to emphasise the unique orbital vascular anomaly, that can mimic the clinical presentation of a cavernous sinus DAVM, requires superselective angiography for diagnosis and, currently, has few therapeutic options.

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