Published in final edited form as:

World Neurosurg. 2014 ; 82(0): 239.e13–239.e17. doi:10.1016/j.wneu.2013.01.093.

ORBITAL CONGESTION COMPLICATING TREATMENT OF CEREBRAL VASCULAR ANOMALIES

Marc H. Levin, M.D., Ph.D.¹, Heather E. Moss, M.D., Ph.D.^{1,2}, Stacy L. Pineles, M.D.^{1,2}, Linda J. Bagley, M.D.³, Gregory G. Heuer, M.D., Ph.D.⁴, Eric L. Zager, M.D.⁴, Laura J. Balcer, M.D.^{1,2}, Steven L. Galetta, M.D.^{1,2}, and M. Reza Vagefi, M.D.¹

¹Department of Ophthalmology, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA

²Department of Neurology, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA

³Department of Radiology, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA

⁴Department of Neurosurgery, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA

Abstract

BACKGROUND—The decision between conservative management and invasive treatment of juxtaorbital intracranial vascular anomalies can be challenging. Whereas arteriovenous malformations (AVMs) can lead to vision loss and are potentially life threatening if they rupture, invasive endovascular and surgical procedures also carry risks.

CASE DESCRIPTIONS—Two patients developed blinding orbital congestion soon after successful treatment of cerebral vascular anomalies. The first patient, a 36 year-old man, underwent partial embolization followed by resection of a congenital frontal lobe AVM. The second patient, a 62 year-old woman, underwent embolization of a peri-orbital/skull base dural arteriovenous malformation. Following intervention, both patients developed unilateral vision loss proptosis, chemosis, complete ophthalmoplegia, and elevated intraocular pressure. The first patient suffered from acute orbital compartment syndrome in the absence of any acute localized hemorrhage or thrombosis. The second patient experienced refractory acute glaucoma from orbital congestion, secondary hyphema and angle closure due to superior orbital vein and cavernous sinus thromboses.

CONCLUSIONS—These cases highlight the potential orbital congestion to worsen acutely after invasive treatment of juxtaorbital cerebral vascular anomalies, and suggest diverse mechanisms of resultant visual and orbital compromise. The first case represents the first report of orbital compartment syndrome following resection of a congenital AVM.

To whom correspondence should be addressed: Marc H. Levin, M.D., Ph.D., Department of Ophthalmology, University of Pennsylvania, 51 North 39th Street, Philadelphia, PA, 19104, marc.levin@uphs.upenn.edu.
Dr. Moss is now with the Department of Ophthalmology and Visual Sciences at the University of Illinois, Chicago, IL, USA
Dr. Pineles is now with the Department of Ophthalmology at the University of California, Los Angeles, USA
Dr. Vagefi is now with the Department of Ophthalmology at the University of California, San Francisco, USA

Keywords

arteriovenous malformation; dural arteriovenous fistula; orbital compartment syndrome

Abnormal cerebral arteriovenous networks that alter orbital blood flow frequently result in neurologic and ocular signs and symptoms that prompt treatment (1, 4). Such lesions are often amenable to transarterial or transvenous endovascular embolization procedures utilizing a variety of agents. These procedures are often performed in conjunction with microsurgical resection and/or stereotactic radiosurgery.

Dural arteriovenous fistulas (DAVFs) are indirect, low-flow shunts supplied by vessels arising from internal and/or external carotid arteries. These malformations may develop spontaneously, possibly secondary to venous thromboses, and may also regress spontaneously (1, 4). Though DAVFs usually cause subtle and chronic orbital signs, invasive treatment is considered in patients with progressive vision loss, ocular hypertension, headache, intractable self-audible bruits, or proptosis leading to corneal exposure. Carotid cavernous fistulas (CCFs) are direct, high-flow communications between the internal carotid artery and cavernous sinus that typically arise following trauma. CCFs typically manifest with rapidly progressive chemosis, proptosis, and decreased vision, often necessitating urgent therapy.

DAVFs and CCFs are acquired arteriovenous malformations (AVMs) without capillary vessels. In contrast, congenital AVMs involve arterial-capillary-venous networks. Congenital cerebral AVMs are often asymptomatic, but can cause headache, seizure, or other focal neurological symptoms, and carry a risk of life-threatening rupture and hemorrhage. Rarely, congenital AVMs involve orbital drainage and can result in orbital congestion or even compressive or ischemic optic neuropathy (13).

CASE REPORTS

Patient 1

A 36 year-old man underwent a left fronto-temporal craniotomy for resection of a large, symptomatic congenital left frontal lobe AVM (Spetzler-Martin grade 4) that was diagnosed 8 years prior during an evaluation for seizures (Figure 1A). Neurologic and psychiatric assessments attributed progressive memory loss and migraines in part to the AVM and resultant venous hypertension.

Initial cerebral angiography demonstrated a $6.0 \times 4.5 \times 5.0$ cm AVM that was supplied mainly by anterior and middle cerebral artery lenticulostriate branches (Figure 1B) with venous drainage primarily into the superior sagittal sinus (Figure 1C). A segmental anterior superior sagittal sinus occlusion with collateral reconstitution of the mid-superior sagittal sinus was also noted on serial angiography. Enlargement and ectasia of anterior inferior left frontal veins suggested cortical venous hypertension. There was no supply to the AVM from the ophthalmic artery nor was there orbital venous drainage from it. Gamma knife radiation at another institution did not alter the size of the lesion. Partial arterial Onyx particle embolization substantially decreased arteriovenous shunting in preparation for surgical

resection. After the procedure, the AVM drainage still did not involve the orbit. Pre-operative ocular exams documented a visual acuity of 20/20 with each eye, chronic bilateral color vision loss and mild chronic papilledema.

The AVM was resected via a left fronto-temporal craniotomy. The AVM nidus extended to the frontal horn of the left lateral ventricle and to the left Sylvian fissure. There were no intraoperative complications. The blood pressure remained in a normal range throughout. Estimated blood loss was 1250 mL and serial hematocrits nadired at 34%. The patient was in a supine position with his head turned approximately 30 degrees to the right for the duration of the 12-hour procedure. Intraoperative angiogram demonstrated complete AVM resection with elimination of arteriovenous shunting and a significant reduction in size of multiple cortical veins, indicating improvement of cortical venous hypertension. A total of 11 liters of fluid were administered with a net positive load of 7 liters. High dose dexamethasone dosing was begun intraoperatively.

Following extubation, the patient described a complete lack of vision with the left eye. Computed tomography demonstrated proptosis of the left globe with posterior tenting and a taut optic nerve (Figure 1D). No retrobulbar or intracranial hemorrhage was present. Intracranial pressure, monitored by external ventricular drain, remained normal.

On the first post-operative day ophthalmic examination demonstrated facial and periorbital edema worse on the left side, with serous fluid draining from the craniotomy incision. Vision was 20/20 in the right eye and no light perception (NLP) in the left eye. The right pupil was reactive while the left was fixed in mid-dilated position. Intraocular pressure (IOP) was 30 mmHg in the right eye and 90 mmHg in the left eye. There was non-pulsatile proptosis with significant chemosis and complete ophthalmoplegia on the left. Sensation in the distribution of ophthalmic and maxillary branches of the trigeminal nerve were absent and diminished, respectively. Corneal haze, a flat anterior chamber, and a cataract were present on the left side. Fundus examination of the left eye revealed hemorrhagic disc edema with large folds emanating radially, and diffuse retinal ischemia with edema consistent with a central retinal artery occlusion (Figure 1E). These findings were diagnostic of orbital compartment syndrome (OCS). A lateral canthotomy with superior and inferior cantholysis was urgently performed. IOP immediately decreased to 18 mmHg OS.

Angiography confirmed AVM resection with no residual arteriovenous shunting, an unchanged segmental superior sagittal sinus occlusion with intact collateral flow, and normal ophthalmic artery filling. No flow abnormality of the cavernous sinus was noted. Subsequent magnetic resonance imaging (MRI) of the brain and orbits with venography demonstrated left ophthalmic vein engorgement, numerous additional smaller flow voids, and extraocular muscle swelling (Figure 1F), without evidence of acute arterial, venous, or cavernous sinus thrombosis. MRI and serial B-scan ultrasound examinations done over the next several days showed persistent tenting of the posterior globe with scleral thickening. At 2-year follow up, the left eye was blind, phthisical, and enophthalmic with an IOP of 2 mmHg. There was normal extraocular motility and trigeminal nerve function.

Patient 2

A 62 year-old woman underwent embolization of a transverse sinus DAVF centered about the right jugular fossa. This had been diagnosed during an evaluation for pulsatile tinnitus and had been treated with arterial embolization 4 and 7 years prior, with only temporary resolution of a mastoid bruit after each procedure. In the preceding month, she noted increased right eye prominence, redness, and blurring.

Visual acuity was 20/25 with each eye without visual field loss or dyschromatopsia. IOP was 18 mmHg in the right eye and 16 mmHg in the left eye. On the right side there was 5 mm of relative proptosis and 2 mm of blepharoptosis. There was an 8 prism diopter exodeviation in primary gaze on the right side. Arterialization of the episcleral vessels were noted, but the retinal vasculature was normal on the right side. The left eye was normal, as was the neurological exam. MRI revealed prominence of the right superior ophthalmic vein (Figure 2A).

Cerebral angiography demonstrated a DAVF with multiple arterial feeders from the right and left ascending pharyngeal arteries, as well as a smaller contribution from the right internal carotid artery (Figure 2B). Venous drainage was retrograde into the inferior petrosal sinus, to the ipsilateral cavernous sinus, and to the right superior ophthalmic vein. There was also some shunting into the clival venous plexus but no retrograde cortical venous drainage. Selective embolization of the feeding branches was performed using polyvinyl alcohol particles. The post-embolization arteriogram demonstrated reduced but not complete elimination of fistula shunting and no significant drainage into the orbital region (Figure 2C).

Several hours after the procedure, the patient complained of diplopia associated with decreased abduction and infraduction of the right eye. Visual acuity and IOP were normal. The next day, right eye visual acuity was reduced to 3'/200E, IOP was 48 mm Hg, 8 mm of proptosis was noted, and the pupil was fixed in mid-dilation. Head CT revealed proptosis of the right globe (Figure 2D) with enlarged and attenuated superior ophthalmic vein (Figure 2E) and attenuated cavernous sinus, consistent with venous thromboses. There was no evidence of retrobulbar hemorrhage, and a heparin drip was started. Vision on the right side declined further to light perception, the anterior chamber angle was closed on gonioscopy, and a hyphema had developed. She was treated with lateral canthotomy and inferior cantholysis, topical pressure lowering agents, high dose intravenous corticosteroids, and continued systemic anticoagulation. The next day, the right eye vision deteriorated to NLP. Severe ocular hypertension did not resolve for another 4 days. The right fundus was never clearly visible due to corneal opacity and hyphema. An MRI obtained nine days post-embolization demonstrated a large choroidal detachment flattening the anterior chamber in the right eye (Figure 2F). Enhancing orbital congestion, proptosis, and an engorged, thrombosed superior ophthalmic vein were redemonstrated.

Over the next 5 months, right eye ocular motility normalized. Eleven years after the procedure, the vision remained NLP in the right eye with band keratopathy and a white cataract. Ectropion secondary to inferior cantholysis and ptosis on the right have been surgically repaired.

DISCUSSION

Patient 1 highlights the potential for OCS to occur immediately following surgical resection of a congenital cerebral AVM. OCS is an ophthalmic emergency characterized by an acute rise in orbital pressure from a rapid increase of volume within the confined space of the orbit (6). Markedly elevated orbital pressure can compromise blood flow to the optic nerve, retina, and choroid, and cause direct nerve compression (14), all of which can result in irreversible blindness if not addressed emergently. While radiologic studies may demonstrate orbital hemorrhage, emphysema, or a tented posterior globe, the diagnosis of OCS is clinical and should not be delayed for imaging or diagnostic testing (3). A history of acute vision loss accompanied by proptosis, periorbital edema, ophthalmoplegia, and elevated IOP should prompt urgent canthotomy and cantholysis.

OCS occurs most commonly in the settings of traumatic retrobulbar hemorrhage (6). It may also be seen as a complication of orbital or sinus surgery, retrobulbar injection, hemorrhage from vascular anomalies, endovascular procedures, blood dyscrasias, orbital infection, inflammation, tumor, or foreign bodies (3, 14). Cases have also been attributed to operative prone positioning, direct pressure from neurosurgical scalp flaps, and massive fluid resuscitation following burn injury (5, 10, 12). A similar clinical presentation can be seen in patients with orbital infarction syndrome after intracranial aneurysm clipping, due to hypoperfusion of the ophthalmic artery and its branches (15).

To our knowledge, patient 1 is the first report of OCS following resection of a congenital AVM. This case is unique due to the absence of an acute thrombotic or hemorrhagic event, and the lack of direct communication between the orbit and AVM. In retrospect, several factors promoted orbital venous congestion in this patient. The preexisting superior sagittal sinus occlusion likely contributed to impaired cerebral venous outflow and, as a result, delayed orbital venous drainage. Bilateral optic nerve elevation as noted pre-operatively, suggests that there may have been chronically elevated ICP due to elevated venous pressures. This presumed papilledema may have placed the optic nerve at greater risk for acute compromise. These factors were compounded by a long surgical time with administration of large volumes of fluid during the procedure. In addition, the proximity of the large AVM to the left orbit could have altered local venous flow patterns subsequent to the resection, leading to congestion. Direct pressure from the bifrontal scalp flap on the orbit may also have played a role in the development of OCS in this patient.

Patient 2 had pre-operative orbital congestion that worsened due to venous thrombosis. Secondary arterial, venous, and cavernous sinus thromboses have all been reported to cause or worsen cranial neuropathies and cerebral or ocular ischemia after DAVF embolization (1, 2, 4, 7–9, 11). Unlike with patient 1, ocular hypertension and decreased vision developed sub-acutely and gradually over 1 day, rather than immediately post-intervention. Also distinct from patient 1, severe post-operative ocular hypertension did not normalize with canthotomy and cantholysis. In this case it is most likely that venous and ciliary body congestion, with resultant hyphema and anterior displacement of the iris and lens diaphragm, were responsible for the refractory acute glaucoma and permanent vision loss.

CONCLUSIONS

In patients undergoing neurosurgical and neuro-interventional procedures, the diagnosis of vision threatening orbital congestion can be challenging given associated cognitive deficits, prolonged wake-up times, and post-operative pain and facial edema. These cases, both of which resulted in blindness, demonstrate the importance of recognizing the constellation of clinical exam findings signifying orbital and ocular complications. Prompt diagnosis and treatment of acute post-procedural orbital compartment syndrome and evolving orbital congestion with resultant acute glaucoma may salvage ocular function.

References

1. Aletich V, Misra M, Shownkeen H, Camras LR, Debrun G. Evaluation and endovascular treatment of juxtaorbital vascular anomalies. *Radiol Clin North Am.* 1999; 37:123–133. [PubMed: 10026733]
2. Chen Z, Zhu G, Feng H, Liu Z. Worsening of cavernous sinus dural arteriovenous fistula with incomplete superior ophthalmic thrombosis after palliative transarterial embolization. *Neurol India.* 2007; 55:390–392. [PubMed: 18040115]
3. Dalley RW, Robertson WD, Rootman J. Globe tenting: a sign of increased orbital tension. *AJNR.* 1989; 10:181–186. [PubMed: 2492721]
4. De Keizer RJW. Carotid-cavernous and orbital arteriovenous fistulas: ocular features, diagnostic and hemodynamic considerations in relation to visual impairment and morbidity. *Orbit.* 2003; 22:121–142. [PubMed: 12789591]
5. Leibovitch I, Casson R, Laforest C, Selva D. Ischemic orbital compartment syndrome as a complication of spinal surgery in the prone position. *Ophthalmology.* 2006; 113:105–108. [PubMed: 16389105]
6. Lima V, Burt B, Leibovitch I, Prabhakaran V, Goldberg RA, Selva D. Orbital compartment syndrome: the ophthalmic surgical emergency. *Surv Ophthalmol.* 2009; 54:441–449. [PubMed: 19539832]
7. Nagy ZZ, Nemeth J, Süveges I, Lanyi F. A case of paradoxical worsening of dural-sinus arteriovenous malformation syndrome after neurosurgery. *Eur J Ophthalmol.* 1995; 5:265–270. [PubMed: 8963165]
8. Schmidt D, Schumacher M. Central vein occlusion as a sequel of spontaneous arteriovenous fistula of the carotid artery to the cavernous sinus. *Fortschr Ophthalmol.* 1991; 88:683–689. [PubMed: 1794790]
9. Sergott RC, Grossman RI, Savino PJ, Bosley TM, Schatz NJ. The syndrome of paradoxical worsening of dural-cavernous sinus arteriovenous malformations. *Ophthalmology.* 1987; 94:205–212. [PubMed: 3587894]
10. Singh CN, Klein MB, Sullivan SR, Sires BS, Hutter CM, Rice K, Jian-Amadi A. Orbital compartment syndrome in burn patients. *Ophthal Plast Reconstr Surg.* 2008; 24:102–106.
11. Swann KW, Heros RC, Debrun GM, Nelson C. Inadvertent middle cerebral artery embolism by a detachable balloon: Management by embolectomy. *J Neurosurg.* 1986; 64:309–312. [PubMed: 3944642]
12. Takahashi Y, Kakizaki H, Selva D, Leibovitch I. Bilateral orbital compartment syndrome and blindness after cerebral aneurysm repair surgery. *Ophthal Plast Reconstr Surg.* 2010; 26:299–301.
13. Volpe NJ, Sharma MC, Galetta SL, Langer DJ, Liu GT, Hurst RW, Flamm ES. Orbital drainage from cerebral arteriovenous malformations. *Neurosurgery.* 2000; 46:820–824. [PubMed: 10764254]
14. Wladis EJ, Peebles TR, Weinberg DA. Management of acute orbital hemorrhage with obstruction of the ophthalmic artery during attempted coil embolization of a dural arteriovenous fistula of the cavernous sinus. *Ophthal Plast Reconstr Surg.* 2007; 23:57–59.

15. Zimmerman CF, Van Patten PD, Golnik KC, Kopitnik TA Jr, Anand R. Orbital infarction syndrome after surgery for intracranial aneurysms. *Ophthalmology*. 1995; 102:594–598. [PubMed: 7724176]

Patient 1.

A: MRI T1-weighted gadolinium enhanced axial image demonstrating proximity of the AVM to left orbit. **B:** Arterial phase of pre-operative, left internal carotid artery angiogram showing the large AVM fed by branches of the left anterior and middle cerebral arteries. **C:** Angiogram late venous phase revealing superior drainage into the mid-superior sagittal sinus, with a stable segmental anterior superior sagittal sinus occlusion (see arrow). No direct arterial or venous communication between the left orbit and the AVM is noted. **D:** Axial image from a non-contrast CT scan of the head performed several hours following AVM resection demonstrating a markedly proptotic left globe with posterior tenting and a taut optic nerve, as well as diffuse engorgement of the extraocular muscles. **E:** Left eye fundus appearance two days after lateral canthotomy depicting acute optic nerve swelling and ischemic retinal thickening with large radial folds. **F:** T1-weighted gadolinium enhanced fat saturated axial image from MRI image performed on the third post-operative day revealing left-sided pre-septal swelling and enhancement, posterior globe tenting, extraocular muscle thickening, and optic nerve enlargement.

Patient 2.

A: Pre-procedure MRI T1-weighted contrast enhanced fat saturated axial image showing right eye proptosis and right superior ophthalmic vein prominence. **B:** Arterial phase of pre-embolization, right internal carotid artery angiogram demonstrating the DAVF (see arrow) fed by the ascending pharyngeal arteries. **C:** Arterial phase of post-embolization angiogram showing no significant filling of the DAVF (see arrow). **D:** Axial image from a non-contrast CT scan of the head performed Day 1 post-AVM embolization demonstrating a markedly proptotic right globe with posterior tenting and a taut optic nerve. **E:** Axial image from the same CT shown in (D) showing an engorged and an hyperattenuated superior ophthalmic vein from thrombosis. **F:** MRI T1-weighted gadolinium enhanced fat saturated axial image obtained 9 days post-embolization showing right eye proptosis, enhancing orbital fat, and a large choroidal detachment displacing the iris anteriorly (see arrow).