Spontaneous Hemorrhage in an Intraorbital Arteriovenous Malformation

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Objective: To report the clinical findings and management of spontaneous hemorrhage in an unsuspected intraorbital arteriovenous malformation.

Design: Interventional case report.

Methods: Review of clinical findings, radiologic studies, and treatment of the patient.

Results: A 75-year-old woman sought treatment for the rapid onset of severe proptosis. Orbital exploration of a “mass” imaged on computed tomography scan and magnetic resonance imaging resulted in massive intraoperative hemorrhage. Subsequent arteriographic and histopathologic analysis confirmed an underlying orbital arteriovenous malformation.


Intraorbital arteriovenous malformations (AVMs) are a rare cause of proptosis. There has been a limited number of previous reports of angiographically documenting orbital AVMs. 1–12 These lesions are believed to be congenital but may enlarge over time. They pose a significant challenge in diagnosis and treatment. They are commonly treated by embolization, surgery, or both. Rapid progression of proptosis resulting from venous thrombosis in an orbital AVM has been documented. Although brain AVMs frequently become apparent as a result of a sudden hemorrhage, spontaneous bleeding in an orbital AVM causing severe proptosis has not been previously reported. We present a patient who experienced rapid onset of massive proptosis that was caused by hemorrhage from an unsuspected orbital AVM.

Patient Report

A 75-year-old woman was referred to our office with a several day history of painless, progressive, proptosis of the right eye with rapid exacerbation during the last 24 hours. She had been initially evaluated by her referring ophthalmologist 7 days before she sought treatment at our office. A computed tomography scan at that time had demonstrated a retrobulbar mass, and a presumptive diagnosis of “orbital pseudotumor” was entertained. She was started on 120 mg of prednisone daily. She had reported minimal discharge and tearing over the last several weeks, but 2 days after initiation of prednisone she noted the onset of diplopia and a marked increase in proptosis. She reported no significant discomfort except for some tenderness to touch. She also reported no conjunctival injection erythema before the sudden onset of chemosis. There was no previous history of ocular trauma or proptosis. She had a history of bilateral cataract extractions, bilateral open-angle glaucoma treated with bilateral filtering procedures, and retinal membrane peeling in the right eye. Past medical history was positive for hypertension, hypercholesterolemia, and previous myocardial infarction.

On presentation to our office, her visual acuity was 20/30...
in the right eye and 20/20 in the left eye. Automated visual fields from 6 months previously demonstrated glaucomatous visual field defects, with the left eye being worse than the right eye. Pupillary examination revealed a relative afferent pupillary defect in the left eye resulting from the more severe glaucomatous damage. Corneal sensation, color vision tested with Ishihara color plates, and intraocular pressure were within normal limits. Thirteen millimeters of proptosis were measured on the right, with prolapsing conjunctival chemosis causing 5 to 6 mm of lagophthalmos (Fig 1). Extraocular movements of the right eye were restricted in all fields of gaze. Fundus examination revealed bilateral

Figure 2. A, B, axial and coronal computed tomography scans with contrast show a 2.5 × 1.8 × 2.0-cm intraconal mass located above the optic nerve with a central low density along the superior aspect on coronal image. This low density was shown to represent subacute hemorrhage on subsequent magnetic resonance imaging.

Figure 3. A, T1 axial precontrast image shows a well-defined mass with an area of T1 hyperintensity compatible with subacute hemorrhage corresponding to the low-density seen on computed tomography. B, a "honeycomb" heterogeneous mass represents the arteriovenous malformation. A feeding or draining vessel is seen prominently at the orbital apex (arrow).
glaucomatous cupping and questionable venous engorge-
ment on the right side relative to the contralateral eye.

The computed tomography scan obtained before her re-
ferral showed a multicystic intraconal mass above and lat-
eral to the optic nerve with variable enhancement (Fig 2).
There was no bony abnormality. Urgent magnetic resonance
imaging was performed that showed the mass to be signif-
ically larger than on the previous computed tomography
scan (Fig 3). The radiologic differential diagnosis was re-
ported as “right orbital lymphangioma or cavernous hem-
angioma with intrinsic bleed versus hemorrhagic metastas-
sis.” The clinical picture was believed to be most consistent
with hemorrhage from a previously existing lymphangioma.
Several adjacent prominent areas of flow void were noted
retrospectively, but not commented on in the initial radio-
logic report.

Over the next 12 hours, a very tight orbit with increasing
pain, reduction of visual acuity to counting fingers, and
reversal of her previous left afferent pupillary defect devel-
oped in the patient. Because of the acute progression and
decreasing vision, an urgent lateral orbitotomy with re-
moval and debulking of the mass was planned. On initial
surgical exposure, a large clot was evacuated from the
superior orbit. This was immediately followed by brisk
hemorrhage with a systolic pressure pulse and rapid loss of
2 units of blood. The surgeon was forced to pack off the
orbit; a tarsorrhaphy was performed and a tight pressure
patch was applied.

After surgery, there was no light perception in the af-
fected eye. The patient experienced increased proptosis over
the ensuing 10 days, with prolapse of hemorrhagic chemo-
sis through the previous tarsorrhaphy. Two weeks after surgery

Figure 4. A, B, C, right lateral common carotid injection showing a
mildly enlarged ophthalmic artery (thick arrow) with a small collection of
blood vessels (thin arrow) and an early draining vein (arrow head).

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we elected to proceed with arterial angiography and attempted embolization of what was presumed to be an orbital AVM. The angiogram revealed an orbital AVM fed by the ophthalmic artery (which was enlarged approximately twice its normal size) and draining through both anterior and posterior venous channels (Fig 4). Despite multiple attempts, it was not possible to cannulate the ophthalmic artery to allow embolization. Options for management, including transcranial orbitotomy to gain control of the proximal ophthalmic artery, were discussed with the patient, but because of her advanced age and poor health, it was decided to proceed with enucleation of the blind eye to allow an anterior approach to excise the underlying AVM. At the time of enucleation, a tangle of large-caliber vessels was encountered in the retrobulbar space. Gross excision of all visibly abnormal tissue and all abnormal vessels was carried out, and an 18-mm silicone implant was inserted. Histologic examination of formalin-fixed, paraffin-embedded tissue revealed numerous abnormal blood vessels of various sizes (Fig 5). The larger vessels had thick, muscular walls consistent with arteries or veins, and these were admixed with thin-walled vessels resembling capillaries or venules. No single, well-defined connection between artery and vein characteristic of an arteriovenous fistula was found. In addition to the vessels of the AVM, an organizing hematoma with abundant fibroblastic proliferation was noted (Fig 6).

Discussion

Arteriovenous malformations are benign hamartomas that have been classified into high and low flow on the arterial side and into distensible and nondistensible on the venous side. Orbital AVMs have been described as being anterior (near lids) or posterior (being intraconal or communicating intracranially). Arteriovenous malformations are congenital lesions that may evolve and present during later life. They can be stimulated to enlarge by menarche, pregnancy, or trauma. Histopathologically, they are composed of many microvascular connections between arteries and veins, with a nidus of cellular stroma interspersed between the vessels. Growth may occur with recruitment of additional arterial feeders and venous enlargement. It is important to differentiate AVMs from arteriovenous fistulas, which are characterized by a single connection between an artery and a vein and which may also subsequently enlarge. Most orbital arteriovenous fistulas occur after an ethmoidal fracture, although one spontaneous case has been reported. Intraocular endothelial papillary hyperplasia can be the cause of an acute or progressive proptosis in patients with varix, cavernous hemangioma, or lymphangioma.

Orbital AVMs are rare. Wright found only three AVMs in a series of 627 patients with proptosis. They have been reported to present at between 8 and 53 years of age. Similar to brain AVMs, it is presumed that orbital AVMs may be present and remain asymptomatic in an unknown larger number of patients. Arteriovenous malformations may evolve and change as a result of high-pressure angiopathy resulting in aneurysmal dilation and vascular proliferation, or they may thrombose and decrease in size. They can cause proptosis (which can be pulsatile or nonpulsatile), compressive optic neuropathy, congestive conjunctival signs, bruit, and limitation of extraocular excursions. A few cases have been reported with rapid progression of signs and symptoms. Hieu et al reported a patient in whom painful proptosis developed over 1 month that was thought to be the result of venous thrombosis. Murali et al reported a similar case of a patient who had experienced two episodes of rapidly progressing proptosis, both of which resolved spontaneously. It is possible that these cases were actually the result of extravasation of blood with subsequent organization and resorption, as suggested in our current patient. Our patient had an initial bleed that was apparent on magnetic resonance imaging. When the clot was dislodged during surgery, rapid hemorrhage ensued.

Our literature review disclosed no previous patient in whom hemorrhage from an orbital AVM has been documented. This is, however, a common presentation of brain AVMs. An unruptured brain AVM carries a 1% to 2% yearly risk of bleeding, with a mortality rate from hemorrhage estimated to be from 6% to 14%. Hemorrhage
usually occurs as a result of rupture of vessel walls weakened by "high flow angiopathy." Hemorrhage is more common in older patients (whose vessels have experienced a longer duration of high flow) and most often occurs at weak points in the vessel wall, occurring from acquired aneurysmal dilation or intraluminal mesenchymal proliferation with vessel wall thinning.

Orbital magnetic resonance imaging and magnetic resonance angiography may be helpful in delineating the true extent and location of vascular lesions, and areas of flow void should alert the surgeon to the possibility of an AVM or shunt. Orbital angiography is most helpful in identifying the feeding and draining channels of the AVM.

Arteriovenous malformations may remain relatively stable, may enlarge progressively, or may even involute, depending on the local and systemic vascular dynamics. Approximately 50% of brain AVMs that have bled will bleed again. Treatment of each lesion is individual, depending on symptomatology and anticipated morbidity. When intervention is elected, it is usually effected by angiographic localization and embolization followed by surgical excision.

Intraorbital hemorrhage into an orbital AVM is extremely rare, but should be considered in the differential diagnosis of rapidly progressive proptosis.

References