Orbital Arteriovenous Malformations

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Objective: To present the clinical features, management, and outcomes in a series of patients with orbital arteriovenous malformations (AVMs).

Methods: Clinical records of patients with orbital AVMs confirmed using angiography were reviewed as a retrospective, noncomparative, interventional case series.

Results: Eight patients (3 women and 5 men) with unilateral AVMs and a mean age of 39 years (median, 36.5 years; range, 26-70 years) were reviewed. Findings existed for an average of 11.2 years before diagnosis and included periocular mass (7 patients, 88%); periocular edema, pulsation/bruit, proptosis, episcleral congestion, and previous trauma (4 patients each, 50%); elevated intraocular pressure (3 patients, 38%); pain and reduced visual acuity (2 patients each, 25%); and restriction of extraocular movements, and diplopia (1 patient each, 12%). All of the patients except 1 underwent surgical resection, with 3 (38%) receiving preoperative embolization of feeder vessels; all of the patients had initial resolution of manifestations after treatment.

Conclusions: Angiography is essential for diagnosis and for planning the management of orbital AVMs. Treatment depends on patient-specific features and includes observation, embolization, and surgical excision or combined preoperative embolization/excision. Given their vascular nature, the main cause of poor management outcomes is perioperative hemorrhage. Outcomes after a multidisciplinary approach are good, with few recurrences reported at follow-up.

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Intraorbital arteriovenous malformations (AVMs) are rare lesions that are thought to be congenital in origin. They are derived embryologically from the arterial system, the venous system, or both. They are high-flow, progressively enlarging communications between arteries and veins that bypass normal capillary beds, usually with multiple feeder arteries, a central nidus, and numerous dilated draining veins. The diagnosis of an orbital AVM is based on angiographic and histologic findings. Findings on selective angiography include an engorged, rapidly filling, proximal arterial system; the malformation; and the distal venous outflow. Histologic analysis reveals thick-walled, irregular arterial and venous channels with and without stromal hemorrhage.

Appearance on initial examination is variable, as although the lesions are often congenital, they may not cause symptoms in childhood. Stimuli for growth include menarche, pregnancy, and trauma. Common findings include periocular pain, dilated corkscrew vessels on the globe extending to the limbus, proptosis, pulsation, bruit, and raised intraocular pressure. We report the largest case series, to our knowledge, of symptomatic orbital AVMs, with angiographic evidence and outcomes after intervention. A review of the literature to better define the features and management of orbital AVMs was also undertaken.

Methods

This is a multicenter, retrospective study of all patients with orbital AVMs who were seen in 2 orbital units: South Australian Institute of Ophthalmology, January 1, 2004, to January 1, 2007 (2 patients), and Royal Brisbane Hospital, January 1, 2001, to January 1, 2006 (6 patients). The inclusion criterion was an angiographically proved AVM involving the orbit. Medical records were reviewed for the following data: age; sex; ethnicity; duration of signs and symptoms; clinical presentation; site involved; imaging findings on angiography, computed tomography, or magnetic resonance imaging angiography; treatment modalities; and outcomes. Response to treatment was evaluated by resolution of the signs and symptoms noted at the initial examination and patient satisfaction.

Results

Case Series

This study included 8 patients (3 women and 5 men) with a mean age of...
Table 1. Initial Clinical Signs and Symptoms in 8 Patients With Orbital Arteriovenous Malformations

<table>
<thead>
<tr>
<th>Patients, No. (%)</th>
<th>Periocular mass</th>
<th>Periocular swelling</th>
<th>Pulsation/bruit</th>
<th>Proptosis</th>
<th>Previous trauma</th>
<th>Edema/congestion of episclera/conjunctiva</th>
<th>Elevated intraocular pressure</th>
<th>Pain</th>
<th>Reduced visual acuity</th>
<th>Restriction of extraocular movement</th>
<th>Diplopia</th>
</tr>
</thead>
<tbody>
<tr>
<td>7 (88%)</td>
<td>4 (50%)</td>
<td>4 (50%)</td>
<td>4 (50%)</td>
<td>4 (50%)</td>
<td>4 (50%)</td>
<td>4 (50%)</td>
<td>3 (38%)</td>
<td>2 (25)%</td>
<td>2 (25%)</td>
<td>1 (12%)</td>
<td>1 (12%)</td>
</tr>
</tbody>
</table>

*Intermittent swelling was described in 1 patient.

39 years (median, 36.5 years; range, 26-70 years). Seven patients were white and 1 was Malay. All of the cases were unilateral. The mean duration from initial clinical signs and symptoms to diagnosis and treatment was 11.2 years (median, 8 years; range, 3 months to 30 years). The presenting signs and symptoms are summarized in Table 1. The most common finding was a periorbital mass in 7 patients (88%), with periorbicular swelling and edema, pulsation/bruit, proptosis, previous trauma, and episcleral congestion in 4 patients each (50%).

Computed tomography or magnetic resonance imaging localized the AVM to the superomedial quadrant of the orbit in 3 patients, the superolateral quadrant in 2, and the entire medial orbit in 1. The latter patient had a giant facial AVM that had a significant orbital component. Aside from this giant lesion, all of the AVMs were located in the anterior orbit. Angiography was available for all 8 patients. Feeder arteries were noted on the right temporal conjunctiva nonaxial proptosis of the right eye. Dilated, abnormal vessels were noted on the right temporal conjunctiva and sclera together with enlargement of the palpebral lobe of the lacrimal gland (Figure 1). Ocular movements were not restricted, and no distensibility was noted when using the Valsalva maneuver. The remainder of the ocular examination, including the fundus, was unremarkable. Magnetic resonance imaging of the orbits revealed a vascular malformation in the supero-temporal orbit (Figure 2). Angiography showed an AVM in the right orbit with feeder vessels from the ophthalmic artery and multiple branches arising from the superficial temporal and maxillary branches of the external carotid artery. After discussion with the patient, it was decided that preoperative embolization of the malformation would not include the feeder vessels from the ophthalmic artery because of the risk of blindness. Hence, preoperative embolization using polyvinyl alcohol particles occluded the external carotid artery feeder vessels, but residual flow was evident from the ophthalmic artery branches (Figure 3). The lesion was debulked via a lateral orbitotomy after the largest feeder vessel from the lacrimal artery was exposed and clipped (Figure 4). Histopathologic examination confirmed an AVM (Figure 5). The patient was asymptomatic for 8 months before developing recurrent symptoms of persistent pain and discomfort. Further debulking surgery was performed, with initial symptom resolution, but the patient remains symptomatic at this stage. She is considering superselective catheterization of the ophthalmic artery preoperatively.

ILLUSTRATIVE CASE

A 47-year-old woman complained of persistent pain, discomfort, and increasing redness and size of a right periorbital lesion, which was present since childhood and had been debulked at least 20 times before review. She had episodes of severe pain associated with recurrent intralesional hemorrhages. Her best-corrected visual acuity was 6/6 in both eyes. She had 2.5 mm of nonaxial proptosis of the right eye. Dilated, abnormal vessels were noted on the right temporal conjunctiva and sclera together with enlargement of the palpebral lobe of the lacrimal gland (Figure 1). Ocular movements were not restricted, and no distensibility was noted when using the Valsalva maneuver. The remainder of the ocular examination, including the fundus, was unremarkable. Magnetic resonance imaging of the orbits revealed a vascular malformation in the supero-temporal orbit (Figure 2). Angiography showed an AVM in the right orbit with feeder vessels from the ophthalmic artery and multiple branches arising from the superficial temporal and maxillary branches of the external carotid artery. After discussion with the patient, it was decided that preoperative embolization of the malformation would not include the feeder vessels from the ophthalmic artery because of the risk of blindness. Hence, preoperative embolization using polyvinyl alcohol particles occluded the external carotid artery feeder vessels, but residual flow was evident from the ophthalmic artery branches (Figure 3). The lesion was debulked via a lateral orbitotomy after the largest feeder vessel from the lacrimal artery was exposed and clipped (Figure 4). Histopathologic examination confirmed an AVM (Figure 5). The patient was asymptomatic for 8 months before developing recurrent symptoms of persistent pain and discomfort. Further debulking surgery was performed, with initial symptom resolution, but the patient remains symptomatic at this stage. She is considering superselective catheterization of the ophthalmic artery preoperatively.

We describe herein a series of 8 patients with angiographically and histologically proved orbital AVMs, which, to our knowledge, is the largest series in the literature. All of the patients except 1 underwent surgery, with or without previous embolization, generally with good results.

Vascular malformations of the orbit are complex and varied lesions. Rootman1 used hemodynamic concepts to classify them into subtypes. Type 1 encompasses lymphangiomas and combined venous–lymphatic system mal-
Table 2. Patient Demographics and Diagnosis and Management of Orbital AVMs

<table>
<thead>
<tr>
<th>Patient No./Sex/Age, y</th>
<th>Duration of Symptoms</th>
<th>Symptoms</th>
<th>Side</th>
<th>Angiography</th>
<th>Treatment</th>
<th>Progress</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/M/27</td>
<td>6 y</td>
<td>Periocular mass, slowly increasing size, pulsation positive</td>
<td>L</td>
<td>AVM anterior superolateral orbit; multiple ICA feeder vessels</td>
<td>Refused treatment</td>
<td>Lost to follow-up after 3 mo</td>
<td>Boxing trauma to left side</td>
</tr>
<tr>
<td>2/M/26</td>
<td>4 y</td>
<td>Periocular mass, intermittent eyelid swelling, pulsation positive</td>
<td>L</td>
<td>AVM anterior superolateral orbit; feeder vessels from STA, facial artery, ophthalmic artery</td>
<td>Surgical excision without embolization owing to numerous feeder vessels</td>
<td>Mild and regressing swelling postoperatively; follow-up, 11 mo</td>
<td>Trauma causing left orbital fracture</td>
</tr>
<tr>
<td>3/F/41</td>
<td>3 y</td>
<td>Pain, eyelid lesion, and bulbar conjunctival hyperemia</td>
<td>R</td>
<td>AVM anterior superomedial orbit; feeder vessels from ophthalmic artery and ethmoidal artery</td>
<td>Surgical excision without embolization</td>
<td>Decreased pain only on bending; follow-up, 37 mo</td>
<td>Encephalocoele surgically managed in past</td>
</tr>
<tr>
<td>4/M/43</td>
<td>20 y</td>
<td>Periocular mass and swelling, diplopia, 6-mm proptosis, pulsation positive</td>
<td>L</td>
<td>AVM anterior superomedial orbit; feeder vessels from ophthalmic artery</td>
<td>Preoperative embolization using platinum microcoils; surgical excision</td>
<td>Horizontal diplopia that required strabismic correction; other symptoms resolved; follow-up, 48 mo</td>
<td>Trauma to left orbit 20 y before initial examination</td>
</tr>
<tr>
<td>5/M/28</td>
<td>16 y</td>
<td>Periocular mass, 4-mm proptosis, reduced VA</td>
<td>L</td>
<td>Giant AVM filling entire medial orbit; feeder vessels identified from ethmoidal artery</td>
<td>Surgical excision</td>
<td>Resolution of symptoms; mild diplopia in extreme up and down gaze; follow-up, 10 mo</td>
<td>None</td>
</tr>
<tr>
<td>6/F/70</td>
<td>3 mo</td>
<td>Periocular mass, pulsation positive</td>
<td>R</td>
<td>AVM anterior superomedial orbit; multiple feeder vessels from supraorbital vessels</td>
<td>Surgical excision</td>
<td>Resolution of symptoms; temporary supraorbital nerve hypoesthesia; follow-up, 48 mo</td>
<td>None</td>
</tr>
<tr>
<td>7/M/32</td>
<td>10 y</td>
<td>Periocular mass and swelling, 5-mm proptosis, decreased VA, increased IOP, edema, and congestion of episclera</td>
<td>R</td>
<td>AVM anterior inferolateral orbit; feeder vessels from maxillary, facial, lingual, and ophthalmic arteries</td>
<td>Preoperative embolization using PVA particles; surgical excision</td>
<td>Resolution of signs and symptoms; follow-up, 12 mo</td>
<td>None</td>
</tr>
<tr>
<td>8/F/46</td>
<td>30 y</td>
<td>Periocular mass and swelling, pain, increased IOP, 2.5-mm proptosis, edema, and congestion of episclera</td>
<td>R</td>
<td>AVM anterior supertemporal orbit; feeder vessels from STA and maxillary and ophthalmic arteries</td>
<td>Preoperative embolization of ECA feeder vessels using PVA particles; surgical debulking</td>
<td>Resolution of symptoms for 8 mo before recurrence; follow-up, 12 mo</td>
<td>Considering preoperative embolization of ophthalmic artery feeder vessels</td>
</tr>
</tbody>
</table>

Abbreviations: AVM, arteriovenous malformation; ECA, external carotid artery; ICA, internal carotid artery; IOP, intraocular pressure; L, left; PVA, polyvinyl alcohol; R, right; STA, superficial temporal artery; VA, visual acuity.

Formations with essentially no flow. Type 2 encompasses low-flow lesions, including distensible, directly communicating venous malformations and nondistensible lesions with little venous communication. Both AVMs and cavernous hemangiomas are included in type 3, with antegrade flow from the arterial through to the venous side.

As described at the beginning of this article, AVMs are high-flow communications between arteries and veins, bypassing normal capillary beds. Orbital AVMs are rare lesions, and Wright, in a series of 627 patients with proptosis, found only 3 AVMs. A review of the literature disclosed only 36 reported cases of orbital AVMs. A selection of these cases is summarized, highlighting various management options and outcomes (Table 3). The most common presenting feature was proptosis, followed by a periocular lesion and pain. Most lesions were located in the superior orbit. Spontaneous hemorrhage is uncommon, with only 1 case
being reported. This is in contrast to histologically similar cerebral AVMs, which manifest most commonly with hemorrhage (approximately 50%) and are responsible for 1% of all strokes. There is a tendency for orbital AVMs to expand slowly, with factors such as menarche, pregnancy, and trauma implicated in their growth. Trauma was a feature in 50% of the patients in this series but was previously reported in only 5 patients.

Orbital AVMs are best considered to be congenital hamartomas, with trauma possibly precipitating hemodynamic changes, leading to symptoms. Based on location, they may be classified into 3 types: purely orbital, orbital and periorbital, and orbital with retinal or cerebral AVMs (Wyburn-Mason syndrome). The first 2 groups are more common (33 of 36 reviewed cases [92%]: 26 [72%] purely orbital and 7 [19%] orbital and periorbital). Wyburn-Mason syndrome can include any combination of cutaneous angiomas and retinal, orbital, and cerebral AVMs, with cutaneous manifestations being the least common. Three of 36 orbital AVMs (8%) reviewed for this article could be classified into this category. It is unclear whether the Wyburn-Mason syndrome is a separate disorder or simply a multifocal manifestation of AVM. Note that cerebral AVMs may cause secondary orbital congestion because of atypical venous drainage into the orbital veins. Garrity et al use the term secondary type of orbital AVM to describe this phenomenon, although no AVM of the orbit exists in this condition.

Diagnosis of orbital AVMs is based on angiographic findings highlighting an engorged, rapidly filling proximal arterial system, a malformation, and distal venous outflow. Histologic analysis of these lesions includes irregularity in the thickness of the muscularis layer in the affected arteries and veins and the presence of a partial

Figure 2. Magnetic resonance images (T1, contrast enhanced) of the orbits. A, Axial image showing a vascular malformation in the right superotemporal orbit with a characteristic flow void (arrow). B, Coronal image highlighting a right superotemporal vascular malformation with vessels evident in the muscle cone.

Figure 3. Preoperative angiogram after embolization revealing residual flow via the ophthalmic artery feeding vessels to a right orbital arteriovenous malformation.

Figure 4. Intraoperative photograph of the excision of an orbital arteriovenous malformation with identification of a lacrimal artery feeder vessel via lateral orbitotomy.

Figure 2.
elastica in some vessels. A nidus of cellular stroma is found between the vessels.

The diagnosis can be aided by clinical history and non-invasive tests, such as flow Doppler studies, and by computed tomography and magnetic resonance imaging to highlight the extent of the lesions. Because these lesions are rare, they must be considered in the differential diagnosis of orbital vascular lesions with similar clinical features, such as carotid-cavernous fistulas, dural-cavernous sinus fistulas, orbital arteriovenous fistulas (AVFs), and cerebral AVMs with drainage into orbital veins. Orbital AVF is the only condition that may be confused with orbital AVM on radiology and angiography. Orbital AVFs may be traumatic or spontaneous and are limited to the orbit, with no connection to the cavernous sinus. These lesions can be differentiated from orbital AVMs on angiography: AVFs demonstrate a direct arteriovenous connection without the intervening nidus that is characteristic of AVMs. Orbital AVFs are rare, with only 10 reported cases; the subject is well reviewed by Yazici and coworkers.

The management of orbital AVMs is based on a multidisciplinary approach. As described earlier, the slow growth and the low incidence of hemorrhage permit observation in many cases. Regression is well documented in cerebral AVMs but has not been reported in orbital lesions. Indications for intervention include visual compromise, patient discomfort related to symptoms, and aesthetic concerns when the lesions extend outside the orbit. The primary treatment for orbital AVMs is surgical excision with or without preoperative embolization. Radiotherapy using newer techniques to focus radiation onto the lesion (linear accelerator, proton beam, or gamma knife) has been used for cerebral AVMs and works by inducing thrombosis; however, this method has not been used for orbital lesions.

Surgery seems to be a safe and effective treatment for orbital lesions, and the predominantly excellent results from this and other case series highlight the importance and effectiveness of preoperative embolization. Goldberg et al in 1993 reported 3 cases of orbital AVM that were successfully managed with combined embolization and surgical resection. One patient complained of persistent supraorbital swelling on bending that, on angiography, was found to represent a varix. Another series consisted of 3 patients with orbital AVMs managed with combined embolization and surgical resection, with little morbidity. Exposure keratopathy developed in 1 patient owing to vertical shortening of the lower eyelid, which was subsequently surgically corrected. Since the advent of superselective angiography and small catheters, it has become possible to locally restrict flow to the lesions, thus reducing their bulk before resection. It also changes the flow property from a dynamic to a static process, thus reducing the risk of hemorrhage perioperatively. In small lesions, with high surgical risk, embolization alone has been successful. This is possible in only a few cases given the propensity of AVMs to form collateral circulation. This form of catheterization is also useful in the diagnosis of small AVMs of the posterior orbit, such as those involving the dura of the optic nerve. Currently, these lesions have few therapeutic options.

Positioning of a catheter when embolizing ophthalmic artery feeder vessels must be distal to the posterior ciliary and central retinal vessels, or larger embolization particles aimed at greater-caliber vessels should be used. This should be accompanied by provocative testing with lidocaine to ensure ocular blood flow. The technique involves the injection of enough lidocaine to replace blood flow for at least 2 seconds so that there is sufficient time for perfusion through the tissue and, hence, provocation. The detection of a scotoma on testing localizes ocular blood flow. Surgical resection should be via an approach specific to the location of the AVM. Rootman et al advocated the use of microvascular clips as opposed to bipolar cautery whenever possible to ensure a more precise effect around delicate tissue.

Review of the literature shows that recurrences were reported when incomplete excision or partial embolization alone was performed, highlighting the tendency of these lesions to recruit new feeder vessels when their supply is partially reduced. This tendency to recurrence was also demonstrated in the illustrative case in this series, in which preoperative embolization of the external carotid feeder vessels but not the internal carotid feeder vessels was performed.

The risk-benefit ratio must be evaluated on a case-by-case basis before interventional management is undertaken in orbital AVMs. Their natural history must be understood and considered, alongside the risks of neuroradiologic and surgical interventions. Visual compromise and persistent or progressive patient discomfort are the main indicators for intervention. We found, in this series of 8 patients and in the literature, that a multidisciplinary approach, when successful, is often curative in these rare lesions.

Orbital AVMs are rare lesions that usually manifest in a chronic manner. They are an important differential diagnosis in any suspected orbital vascular abnormality. Angiography is essential for diagnosis and for planning management. Their management depends on patient-specific features and includes observation, embolization, and surgical excision or combined pre-
operative embolization and surgical excision. Given the vascular nature of orbital AVMs, the main cause of poor management outcomes is perioperative hemorrhage. Outcomes after a multidisciplinary approach are generally good, with few recurrences reported at follow-up.
REFERENCES


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