CASE REPORT

Intraorbital Arteriovenous Malformation: Case Report

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ABSTRACT

An intraorbital arteriovenous malformation became symptomatic without exophthalmos but with chemosis and bruit of the left eye. The patient underwent only surgical treatment. The orbital rim was removed through a fronto-orbital approach. The chemosis was corrected and postoperative magnetic resonance imaging and angiography confirmed complete removal of the malformation. Histopathological examination disclosed an arteriovenous malformation in the extrabulbar fatty tissue without invasion of the extraocular muscles or optic nerve. The differences between various types of intraorbital malformations are discussed.

KEYWORDS: Arteriovenous malformation, intraorbital, surgery

Arteriovenous malformations (AVMs) are developmental anomalies that occur when the embryonic vascular network fails to differentiate.¹ Intraorbital AVMs are rare, and their management represents a challenge for multidisciplinary teams. Unilateral exophthalmos can be caused by both intraorbital and extraorbital vascular malformations.² The differential diagnosis includes anterior cranial fossa dural fistulas, cirroid aneurysms of the scalp, traumatic carotid-cavernous fistulas, and congenital vascular malformations like Wyburn-Mason or Bonnet-Dechaunne-Blanc syndrome. Arteriography is essential for confirming the diagnosis and defining the arterial supply and venous drainage pathways.³ We present the preoperative findings

and surgical outcome of a patient with a left intraorbital AVM who underwent successful surgical treatment.

CASE REPORT

An 18-year-old, right-handed man was referred for evaluation of an increasing lesion in the region of the left brow. The lesion had first been noted at the age of 3 years. The patient had no history of trauma. At 18 years he was admitted to a local hospital with a 6-month history of increasing lesion size and ipsilateral chemosis.

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At the time of the patient's initial presentation to our institution, a 6 cm mass in the region of his left brow was painless, soft, and pulsatile, and associated with a bruit to auscultation. There was no displacement of the globe. A temporal ectatic artery feeding the supraciliar lesion from the preauricular area was evident (Fig. 1). He was neurologically intact and exhibited no exophthalmos.

Plain skull radiographs and computed tomography (CT) revealed a well-defined lytic lesion of the frontal bone involving the left brow area. Enhanced CT showed a vascular abnormality in the superior portion of the orbit. Magnetic resonance imaging (MRI) SE-weighted revealed the lesion was homogeneous. It measured 4 cm in its greatest axis and projected intraorbitally. Angiography showed increased vascularity in the area of the left brow. Feeder vessels extended from the left

Figure 1 Intraoperative photograph shows the supraciliar mass, lack of exophthalmos, and superficial vessels from temporal artery.

ophthalmic and middle meningeal arteries. The principal drainage was to the internal jugular vein via the facial veins (Fig. 2).

The lesion was excised completely through a bicoronal approach and a fronto-orbital craniotomy with two-step removal of the orbital rim and roof. Extraorbitally, several feedings vessels from the meningeal artery were ligated and divided. Intraorbitally, the branches of the left ophthalmic artery distal to the central retinal artery were identified using the surgical microscope, then coagulated and divided. The craniotomy was repaired using microplates (Fig. 3).

The specimen, which consisted of multiple pieces of soft tissues with numerous vascular channels, was diagnosed as an AVM in the bulbar fatty and connective tissue (Fig. 4).

At discharge, the patient showed evident cosmetic improvement, and the appearance of his left eye had recovered completely. Postoperative MRI and angiography confirmed total removal of the AVM (Fig. 5). There has been no intra- or extra-orbital recurrence during 4 years of follow up.

DISCUSSION

With the exception of bruit, the most common presenting symptoms of orbital AVMs are related to venous drainage involving the orbit. The elevated pressure in the venous pathways draining the orbit produces orbital venous congestion. The congestion is reflected by dilation of the veins on the surface of the sclera, producing chemosis. The severity of the visual symptoms correlates with the adequacy of the venous drainage pathways more than with the speed or volume of the arteriovenous shunting. Occasionally, a severely congested orbit produces mechanical restriction of ocular motility in all directions without cranial nerve dysfunction. However, the presence of a dilated pupil would suggest a third cranial nerve palsy. The bruit is produced by arterialized turbulent flow entering the venous drainage. Unilateral exophthalmos can be caused

Figure 2 Contrast-enhanced axial computed tomography shows the nidus of the arteriovenous malformation (AVM) (upper left). Sagittal spin-echo magnetic resonance imaging shows extra- and intraorbital components of the AVM (upper right). Lateral carotid angiograms show the AVM fed by the left ophthalmic (lower left) and meningeal arteries (lower right) and maxillary vein drainage (lower right).

by both intraorbital and extraorbital vascular malformations.^{4,5}

Intraorbital AVMs are considered to be progressive congenital lesions, but a history of penetrating or blunt trauma should always be considered. At the age of 3 years, our patient had a small left brow tumor without head trauma that began increasing in the 6 months before treatment. The onset of symptoms was probably caused by progressive thrombosis of a draining vein because the chemosis and bruit were only present for 6 months. In this case, the AVM was probably spontaneous.

Like dural fistulas, intraorbital AVMs must be evaluated to determine whether they are best

suited to surgical or endovascular management.¹⁷ In many cases, transarterial and transvenous approaches provide minimally invasive access by embolization and occlusion of AVMs and fistulae. Intraorbital AVMs are exceptions to endovascular treatment because embolization of the central retinal artery, a distal branch of the ophthalmic artery, can complicate the procedure and devastate a patient's vision.^{16,18} The small caliber and tortuous course of the ophthalmic artery make selective catheterization difficult. Embolization is possible only in a few extraorbital AVMs.^{2,14}

Cirroid aneurysms of the scalp in the brow region are a possible differential diagnosis.^{19,20} In

Figure 3 Intraoperative photograph after orbital roof has been removed shows a vascular mass identified within the orbital fat (left). Plain anteroposterior postoperative radiograph shows the craniotomy repair (right).

our patient, however, there were feeders from the left ophthalmic artery, and a true intraorbital arteriovenous nidus was demonstrated surgically. These findings exclude this uncommon arteriovenous fistula of the scalp.

True AVMs involving only the orbit are rare.^{3,6-16} In our case, the AVM involved the brow and eyelid tissues by feeders from facial arteries. Often the nidus is supplied by feeders from the external carotid artery (ECA) to the AVM,

Figure 4 Photomicrograph showing multiple vascular channels consistent with arteriovenous malformation adjacent and within fat. Hematoxylin and eosin, original magnification $\times 100$.

Figure 5 Lateral (upper left) and coronal (upper right) T1-weighted magnetic resonance images obtained 6 months after surgery show the normal aspect of the orbit. Follow-up left internal carotid angiograms obtained 9 months after surgery confirm complete removal of the arteriovenous malformation (lower).

moreover by an enlarged ophthalmic artery. The extraorbital component of the AVMs can produce ophthalmological manifestations when draining into the orbital veins. AVMs fed primarily by the ECA system¹⁴ are most amenable to embolization, but the nidus still should be resected.

As in most of the published cases, surgery was the treatment of choice for our patient. The surgical

approach should be large enough to allow complete removal of the lesion. We performed a fronto-orbital craniotomy with a two-step removal of the orbital rim and roof. Access to the posterior area of the orbit was needed to control distal branches to the posterior ciliary and central retinal vessels in the deep orbital tissues. Surgical outcomes are usually satisfactory and associated with minimal morbidity.⁸

CONCLUSION

Intraorbital AVMs are rare, slowly progressive congenital lesions, or as in our case, true traumatic AVMs. Anterior skull base surgery is the treatment of choice to eradicate the lesion completely. Endovascular treatment is appropriate when the AVM is fed primarily by the ECA system. Multidisciplinary teams^{5,21} are likely to offer the optimal options for treatment.

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Commentary

This article describes a unique presentation of a large intraorbital arteriovenous malformation (AVM) and reviews orbital and ocular adnexal AVMs. Although not exhaustive, the concise summary provides enough background to cover the differential diagnoses. The surgical description is a fine review of a cautious approach to dealing with large lateral intraorbital AVMs involving the superior lateral orbit. As alluded to in this article, smaller AVMs may be considered for intravenous embolization. The success rate of embolizing selected smaller AVMs varies from 10 to 40%. These embolization techniques are associated with the same risk profile as surgical approaches to the same lesions. Grade I AVMs are associated with a 5% surgical risk and their surgical success rate is almost 100%. In contrast, embolization techniques are associated, at best, with a 40% chance of therapeutic

control whereas the risk is equivalent to that of surgery. Consequently, most authorities would agree that a surgical approach to orbital AVMs appears to be warranted. The literature and personal experience testify to the efficacy of microembolizing spontaneous and traumatic cavernous-sinus fistulas. Access for embolization can be attained through an upper lid crease cut-down, exposing the superior ophthalmic vein. Gil-Salú, González-Darder, and Vera-Román have presented a case report that highlights a rare but important orbital challenge. They should be congratulated for their endeavors and presentation.

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Commentary

Vascular malformations in and around the brain and skull base are not rare, and many cause neurological and neuro-ophthalmic signs and symptoms. When the orbit is involved, arteriovenous malformations (AVMs) may manifest with primary orbital signs, including proptosis, chemosis, and dystopia. Secondary signs may include problems associated with optic nerve function and morbidity related to orbital venous congestion. Digital subtraction angiography remains the gold standard of diagnosis, but the past 15 years have provided a revolution in neurointerventional therapeutic tech-

niques. Small tracker catheters and more experienced neuroradiologists can maneuver into small feeders, embolizing AVMs transarterially or even transvenously.

Orbital lesions have remained on the frontier of interventional neuroradiology. The extensive connections between the internal and external carotid artery supply as well as the potential for inadvertent embolization of end arteries such as the central retinal artery have limited the role of embolization in AVMs affecting the orbit.

Embolization may be inadequate to cure a lesion permanently unless the nidus is totally obliterated (often requiring placement of glue or some other agent that completely eliminates the primary connection into the venous system). If the nidus is not removed, additional vessels will be recruited. This statement also holds true for orbital lesions. As the authors point out, surgical excision is likely to remain an important part of the treatment of AVMs involving the orbit for the foreseeable future. The use of a multidisciplinary approach to these lesions recognizes the clinical spectrum possible as well as the contributions available from radiologists and ophthalmologists. Not all of these lesions need to be approached. Only when there is evidence of clinical deterioration or interference with function does the risk:benefit ratio for various therapeutic options need to be discussed.

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