Arterial Occlusion Following Embolization of an Orbital Arteriovenous Malformation
A Case Report

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SUMMARY – Orbital arteriovenous malformation is a rare condition which poses a management problem in view of the complexity of the vessels involved, and the potentially blinding and life-threatening complications from the modes of treatment. Treatment requires obliteration of the abnormal vascular communication. Vision may be adversely affected by the condition itself, as well as a result of invasive treatment modalities. This case reports such a situation where the visual outcome was severely affected following embolization of the feeding vessels.

Case Report

A 32-year-old man presented with progressive painless proptosis for two years which had worsened over the past two months. There was also worsening diplopia in all directions of gaze. The proptosis was more obvious when he coughed. There was a vascular mass growing over the left medial canthus and left conjunctival vessels were becoming more prominent. There was no limb weakness or speech abnormalities. There was a history of blunt trauma to the left eye, with traumatic retinal hole a year earlier. Mild proptosis was already noted at that time, however he defaulted follow-up after undergoing laser treatment. Medical history was unremarkable.

On examination, the patient had nasal speech, and left carotid bruit was present. There was an obvious non-pulsatile non-axial proptosis of the left eye. Hertel’s exophthalmometer measured 20 mm for the right eye and 30 mm for the left eye, with a base of 115 mm. The left eye was displaced downward and outwards. There was a vascular conjunctival mass over the medial canthus, with dilated and tortuous left conjunctival vessels. A haemangioma was noted at the tip of the nose. The best corrected visual acuity was OU 6/6 with a vertical diplopia in all directions of gaze. Ocular eye movements were full, and relative afferent pupillary defect was absent.

Magnetic resonance imaging of the orbit showed a heterogeneously-enhancing mass containing a dilated high flow vessel. The mass measured 5.1 cm × 2.1 cm × 3.3 cm in size and was suggestive of a left orbit vascular malformation (figure 1).

A four vessel cerebral angiogram showed an extensive torrential arteriovenous malformation of the left ophthalmic artery as well as left maxillary/nasal region (figure 2). The left ophthalmic artery was grossly dilated with five feeder vessels, receiving multiple blood supply from the left facial, superficial transverse, internal maxillary and ascending pharyngeal arteries. There was no supply from the middle meningeal or internal carotid arteries. The malformation was found to be predominantly extraconal in origin with involvement of the left medial rectus, floor of the left orbit and left ethmoid sinus.

During the first stage, several embolizations were attempted (figures 3 and 4). Ultraselective cannulation with glue embolization was performed on the ophthalmic feeder vessels.
and the superficial transverse artery. The left internal maxillary artery was embolized with polyvinyl alcohol (PVA). Sixty percent of the ophthalmic component was ablated. There was residual supply from the ophthalmic artery and facial and inferior alveolar branches. The left internal carotid artery was preserved post embolization.

Following the first stage of embolization, the visual acuity was good, with OD 6/6 and OS 6/9. The intraocular pressure was 18 mmHg bilaterally, and the optic discs were pink and healthy. Hertel measurements remained unchanged. The second stage embolization was performed one month later. The left facial and left inferior alveolar arteries and branches of the left ophthalmic arteries were embolized with PVA. The left central retinal artery and choroidal arteries were intact immediately post embolization. However, the patient developed left central retinal artery occlusion the next day, complaining of sudden left visual loss to light perception only. This important symptom was only alerted to the attending doctor.
after two hours had passed. Upon examination, there was a marked left relative afferent papillary defect. The left optic disc was pink with a classical cherry red spot at the macula. The peripheral retina was pale. Despite the immediate management which included ocular massage and intravenous acetazolamide, the vision remained poor.

One month later, there was no light perception in the left eye. The left optic disc was pale. The proptosis had decreased by 5 mm, and the small hemangioma at the tip of the nose almost disappeared. However, the patient defaulted further treatment.

Discussion

AVMs are the most common form of intracranial vascular hamartoma. AVMs are usually congenital, but they may become symptomatic at any age. However, 70% of AVMs produce symptoms during the second and the third decades of life. This patient presented with progressive proptosis secondary to an enlarging left orbital AVM in the early fourth decade of life. Fortunately, there was no associated intracranial AVMs which could have predisposed him to more serious complication of intracerebral or subarachnoid haemorrhage.

The management of orbital AVM involves a team of ophthalmologists, neurosurgeons and interventional neuroradiologists. Improvements in microguidewire and microcatheter technology have made it possible to treat previously unreachable and untreatable AVMs. The definitive management would be embolization of the AVM. Materials used to embolize vascular malformations include glue, coil, polyvinyl alcohol, ethanol, cyanoacrylate and microfibrillar collagen. In this case, polyvinyl alcohol was used, and the procedure was carried out in multiple stages. Embolization carries a high risk of retinal artery occlusion, and even intracerebral haemorrhage which could be life-threatening. The patient was carefully counseled regarding the risk of central retinal artery occlusion and bleeding that may follow the embolization procedure. Following the first stage of embolization, the proptosis had decreased slightly.

Following the second stage of embolization one month later, the patient developed central retinal artery occlusion (CRAO). The retina is particularly susceptible to hypoxic injury because of its high rate of oxygen consumption. Hayreh et Al reported irreversible cell injury after 90 minutes of total CRAO in their primate model. Despite performing emergency maneuvers, significant visual loss may persist in the majority of patients with CRAO. This patient most likely had irreversible retinal ischaemia by the time emergency measures were instituted, and thus the measures were not successful in aborting the occlusion event.

Untreated advanced arteriovenous malformation of the orbit can result in serious sequelae. Vision can decrease because of optic nerve or retinal ischaemia, which develops as a result of a combination of venous congestion and increased intraocular pressure. Current management of orbital arteriovenous malformation is based on embolization of feeder vessels followed by surgical excision. In complex lesions, embolization does not completely eradicate the lesion. If surgery is indicated, it should be performed soon after the embolization procedure when the lesion is most quiescent.

Complications of embolization are severe but infrequent, and include post-operative retrobulbar haemorrhage, dislodgement and migration of embolus, and failure to identify and occlude anastomotic vessels. Such was the case with this patient, who presumably developed dislodgement of emboli causing a central retinal artery occlusion (CRAO). Neuroradiologic techniques are highly useful as an adjunct to surgical procedures in the management of vascular lesions, but are not without marked risk of complications. The patient with arteriovenous malformation of the orbit is best treated with a solid understanding of the lesion, careful consideration of the treatment issues, and sound preparation in current endovascular and surgical approaches.

This case illustrates the serious complications that may occur as a result of invasive interventional procedures. Risks of serious complications like blindness or strokes must be clearly explained to all patients scheduled to undergo such procedures.
References


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