Combined approach to management of periocular arteriovenous malformation by interventional radiology and surgical excision

Bipasha Mukherjee, Vathsalya Vijay, Swatee Halbe[1]

Periorbital arteriovenous malformations (AVMs) are congenital lesions that may cause significant morbidity such as amblyopia, cosmetic disfigurement, or chronic pain. Due to the rarity of these lesions, they are frequently misdiagnosed and treated inappropriately. We managed a 6-year-old girl with preseptal AVM by endovascular embolization followed by complete surgical excision after 2 days. She was previously diagnosed as capillary hemangioma and was being treated with intralesional steroid injections and oral propranolol. Neuroimaging revealed an AVM fed by the branches of both external and internal carotid arteries and drained by the superior ophthalmic vein. Endovascular embolization with glue followed by complete surgical excision of the lesion was done. The resultant cosmetic and functional outcome was gratifying. Diagnosis and management of periorbital AVM remain a challenge requiring a multidisciplinary approach involving interventional radiologist and trained oculoplastic surgeons. Combined endovascular embolization followed by surgical excision proved safe and effective.

Key words: Arteriovenous malformation, endovascular embolization, surgical excision

Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/ijo.IJO_663_17

Orbit, Oculoplasty, Reconstructive and Aesthetic Services, Sankara Nethralaya, Medical Research Foundation, [1]Department of Interventional Radiology, Apollo Specialty Hospitals, Chennai, Tamil Nadu, India

Correspondence to: Dr. Bipasha Mukherjee, Department of Orbit, Oculoplasty, Reconstructive and Aesthetic Services, Sankara Nethralaya, Medical Research Foundation, 18, College Road, Chennai - 600 006, Tamil Nadu, India. E-mail: beas003@yahoo.co.uk

Manuscript received: 10.08.17; Revision accepted: 09.11.17

Arteriovenous malformations (AVMs) of the orbit are rare, hemodynamically active, high-flow vascular anomalies that may cause significant functional morbidity, cosmetic disfigurement, and high-output cardiac failure.[1] Accurate diagnosis and successful management of such anomalies entail appropriate clinical examination and neuroimaging. A multidisciplinary approach yields satisfactory functional and cosmetic outcome. We present an interesting case of a periorbital AVM managed by embolization followed by surgical excision.

Case Report

A 6-year-old girl presented to orbit clinic with a diffuse progressive soft compressible pulsatile mass lesion in the medial aspect of the right upper eyelid since birth, causing total

Cite this article as: Mukherjee B, Vijay V, Halbe S. Combined approach to management of periocular arteriovenous malformation by interventional radiology and surgical excision. Indian J Ophthalmol 2018;66:151-4.

mechanical ptosis [Fig. 1a]. The surface of the lesion showed dilated blood vessels and bruit was heard on auscultation. Valsalva maneuver did not produce any difference in the size of the lesion. There was no prior history of trauma or presence of similar swellings elsewhere in the body. She had been treated elsewhere as capillary hemangioma of infancy (CHI), with multiple intralesional steroid injections and oral propranolol, which had failed to result in any symptomatic relief. Her vision in the right eye was counting fingers at 1 meter, presumably due to deprivation amblyopia. There were no other ocular or systemic abnormalities. Left eye examination was within normal limits.

Computerized tomography (CT) scans revealed a diffuse preseptal isodense mass in the superonasal aspect of the right upper eyelid, which enhanced brightly during the arterial phase of contrast [Fig. 2a-c]. Magnetic resonance imaging (MRI) showed an isointense signal in T1-weighted and mixed signal in T2-weighted images with multiple flow void signals [Fig. 2d and e]. The right superior ophthalmic vein was dilated. Digital subtraction angiography (DSA) disclosed a compact nidus of AVM with feeder vessels from the right ophthalmic, middle meningeal, internal maxillary, superficial temporal, and bilateral facial arteries [Fig. 3a and b]. Eighty percent of the AVM was embolized using 16% glue (N-butyl 2-cyanoacrylate) by superselective cannulation of the right ophthalmic artery feeder through the right femoral artery as well as direct intralesional glue injection under fluoroscopy under general anesthesia [Fig. 3c-f]. Subsequently, surgical excision was performed after 72 h [Fig. 1b]. The mass was excised completely through a superior eyelid crease incision without encountering any troublesome intraoperative bleeding. Histopathological examination revealed numerous tiny and large vascular spaces lined with endothelial cells and filled with blood. The nidus was seen as cellular stroma in between the vessels, confirming the diagnosis of AVM [Fig. 4]. The postoperative period was uneventful. The functional and cosmetic outcome was satisfactory [Fig. 1c]. The patient had been followed up for 2 years after surgery without any signs of recurrence. Best-corrected visual acuity at last follow-up was noted to be counting fingers at 1 meter, and the child was undergoing amblyopia therapy (left eye patching for 6 h a day) with close follow-up.

Figure 1: (a) Clinical picture of patient at presentation. (b) Intraoperative photograph showing removal of the lesion with minimal bleeding. (c) Postoperative appearance of the patient

Discussion

AVMs are developmental hamartomas consisting of an abnormal nidus of dysplastic vascular channels with feeding arteries and draining veins and absence of normal intervening capillary network.[2] Mutation of RASA 1 genes may be associated with the development of AVMs.[3] These high-flow lesions undergo hemodynamic changes and worsen during puberty and pregnancy, suggesting possible hormonal influence on these lesions.[2] In a case series reported

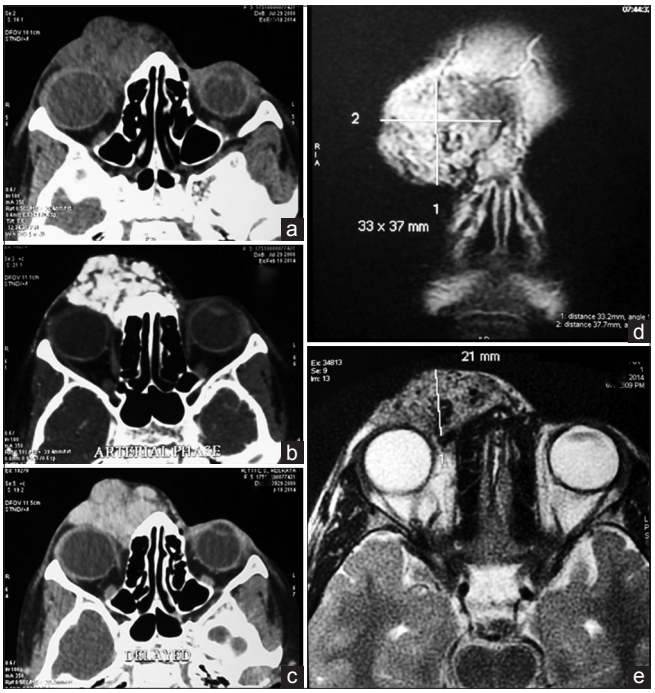

Figure 2: (a) Axial plain computerized tomography scan of the patient. (b) Contrast-enhanced computerized tomography shows intense enhancement in the arterial phase. (c) Rapid washout in the venous phase confirming diagnosis of arteriovenous malformation. (d) Coronal magnetic resonance imaging scan showing soft-tissue lesion in the right orbital region extending up to midline. (e) Axial magnetic resonance imaging showing multiple flow voids within the lesion

by Warrier *et al.*,[2] trauma was found to be associated in 50% of the patients. Orbital AVMs may be associated with similar lesions elsewhere in Wyburn-Mason syndrome and hereditary hemorrhagic telangiectasia or Rendu–Osler–Weber syndrome.[4] A classification system by Schobinger describes the clinical characteristics of superficial AVM.[5] [Table 1]. Our patient falls under Grade II of Schobinger's classification of superficial AVMs and simple sporadic AVM (Type IV of simple vascular malformation) of the International Society for the Study of Vascular Anomalies Classification.

These patients usually present with periocular pain, proptosis, ptosis, periocular swelling, reduced visual acuity, diplopia, discoloration, tinnitus, and high-output cardiac failure.[1,2] An orbital AVM may cause sudden mass effect due to thrombosis or intralesional hemorrhage.[6] On examination, dilated corkscrew vessels, pulsation, bruit, compressibility, and raised intraocular pressure may be found. AVMs are difficult to differentiate clinically from other vascular anomalies such as caroticocavernous fistulas, orbital arteriovenous fistulas (AVFs), and cerebral AVMs with drainage into orbital veins. Differentiating congenital vascular malformations from CHI is crucial as both the entities represent different pathologies and treatment approaches vary greatly. The imaging characteristics and histopathological findings confirm the diagnosis.[7] Orbital AVFs and AVMs have similar appearances on angiographic study, and hence, demonstration of the nidus is crucial in ascertaining AVMs, which is typically absent in an AVF.[2] The other differential diagnosis is orbital varix, which typically increases in size on Valsalva maneuver. Imaging by various modalities such as dynamic arterial and Valsalva-augmented venous phase, multidetector CT angiography (DP-MDCTA), dynamic MRI, magnetic resonance angiography, and Doppler and selective angiography aids in the diagnosis of an AVM.[7] DSA is the gold standard for AVMs, which involves injection of contrast directly into one of its feeder vessels and digital image acquisition. This selective angiography maps the feeder vessels as well as draining veins.[6] Current management options for AVM are based on their hemodynamic characteristics, vascular anatomy, and location of these lesions. Asymptomatic lesions may be observed. However, AVMs tend to recruit new feeder vessels and continue to progressively enlarge in size. Surgical excision of orbital AVMs is challenging as they carry a significant risk of intraoperative bleeding and surgical morbidity. Preoperative embolization closes off surgically inaccessible arterial feeders,

Table 1: Schobinger's classification of superficial arteriovenous malformations

Grade	Description
Grade I	Quiescent lesion, pink or blue cutaneous stain, warm lesion, AV shunt on Doppler sonography
Grade II	Expansive lesion, pulsatile lesion, thrill; dilated draining veins
Grade III	Grade II + necrosis, infection, hemorrhage, or pain
Grade IV	Grade III + cardiac failure

AV: Arteriovenous

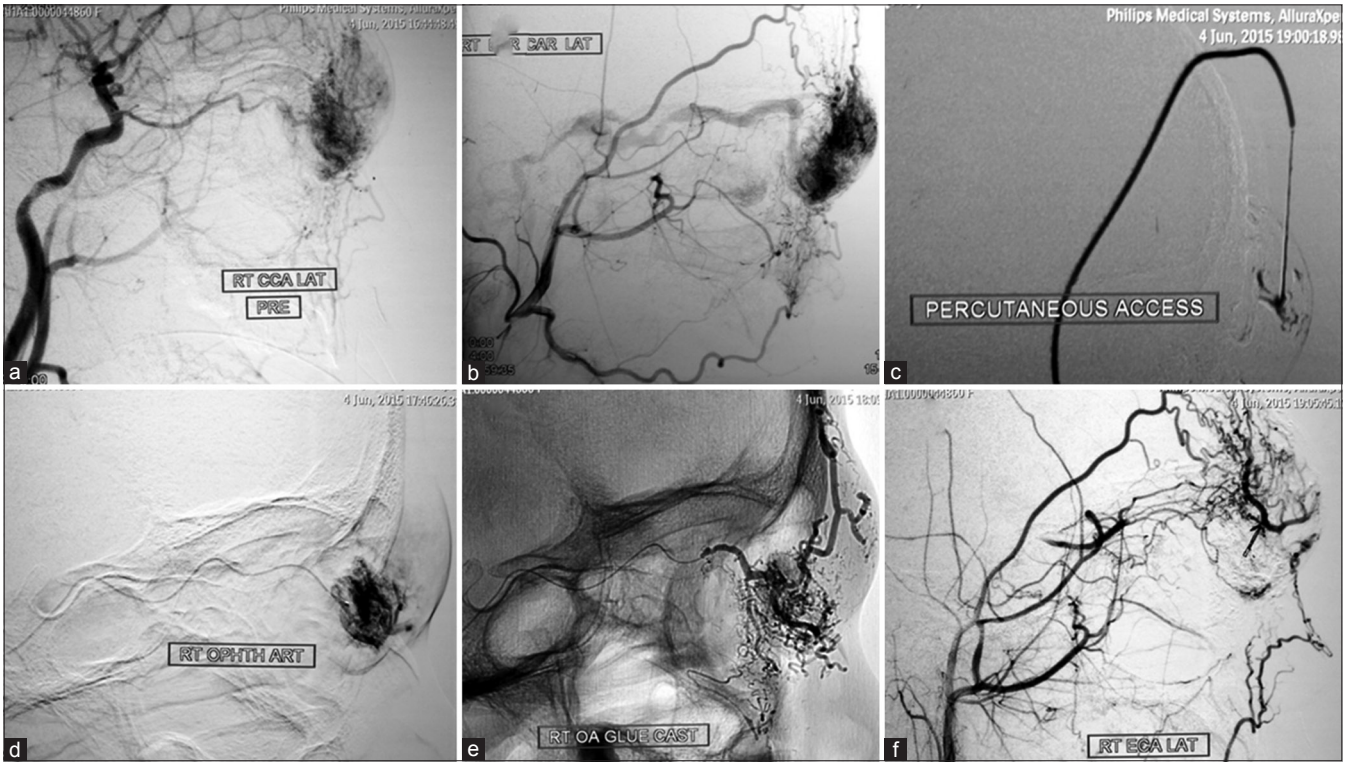

Figure 3: (a) Right common carotid angiogram showing nidus of the right orbital arteriovenous malformation supplied by the right ophthalmic artery as well as external carotid artery branches. (b) Right external carotid angiogram showing nidus supplied by branches of the right external carotid artery, namely, superficial temporal, internal maxillary, and facial artery. (c) Direct puncture of nidus by 22-g needle and injection of glue. (d) Microcatheter in the right ophthalmic artery and embolization of nidus with glue. (e) Lateral view of the skull showing cast of glue after embolization. (f) Postembolization right external carotid angiogram showing residual nidus

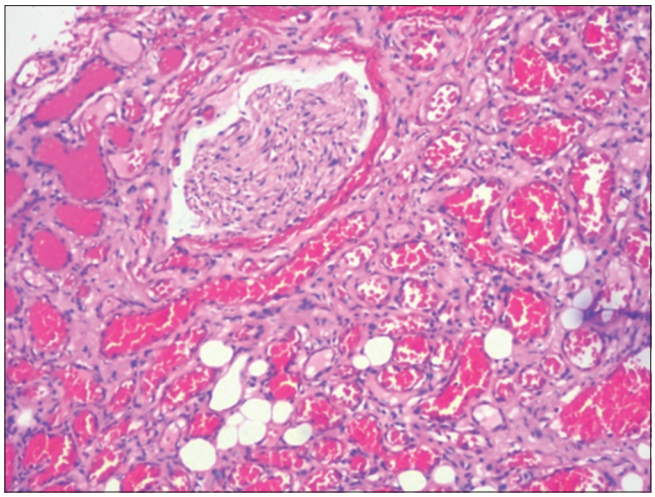

Figure 4: Microphotograph of the excised lesion showing vascular malformation composed of numerous tiny and large vascular spaces lined with endothelial cells and filled with blood (H and E, ×100)

reduces intraoperative blood loss, aids in complete surgical resection, and decreases the chances of recurrence.[6,8,9] The embolization procedure, however, is not without risks and may cause sight/life-threatening complications. Occlusion of vessels due to reflux of liquid embolic material and small particles into adjacent vessels can cause cranial nerve palsies[9] or permanent visual loss from ophthalmic artery occlusion or central retinal artery occlusion.[9] Embolization should be followed by surgical excision to completely remove the nidus, thus preventing recurrence. Ideally, surgical excision should be done within 48 h of embolization.[9] The combined approach of preoperative embolization of the AVM followed by surgical excision of the nidus yields successful outcomes in both function and cosmesis.[6] There are a few reported cases of orbital AVM managed successfully by this combined approach.[10]

Conclusion

Multidisciplinary approach by a competent interventional radiologist and a trained oculoplastic surgeon yields satisfactory functional and cosmetic outcome in the management of periorbital AVM.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Acknowledgments

The authors would like to thank Dr. Olma Veena Noronha, VRR Scans, Chennai.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Starks VS, Gilliland G, Hise J, Thacker I, Layton KF. Effect of resection of an orbital arteriovenous malformation on central venous pressure. Proc (Bayl Univ Med Cent) 2015;28:185-7.
2. Warrier S, Prabhakaran VC, Valenzuela A, Sullivan TJ, Davis G, Selva D. Orbital arteriovenous malformations. Arch Ophthalmol 2008;126:1669-75.
3. Brouillard P, Vikkula M. Genetic causes of vascular malformations. Hum Mol Genet 2007;16:R140-9.
4. Van Went C, Ozanne A, Saliou G, Dethorey G, De Monchy I, Krings T, *et al.* Spontaneous thrombosis of an orbital arteriovenous malformation revealing hereditary haemorrhagic telangiectasia (Rendu-Osler-Weber disease). A case report. Interv Neuroradiol 2011;17:466-71.
5. Schobinger R. Proceedings of International Society for the Study of Vascular Anomalies Congress Rome, Italy; 23-26 June, 1996.
6. Rootman J, Heran MK, Graeb DA. Vascular malformations of the orbit: Classification and the role of imaging in diagnosis and treatment strategies. Ophthal Plast Reconstr Surg 2014;30:91-104.
7. Lee BB, Antignani PL, Baraldini V, Baumgartner I, Berlien P, Blei F, *et al.* ISVI-IUA consensus document diagnostic guidelines of vascular anomalies: Vascular malformations and hemangiomas. Int Angiol 2015;34:333-74.
8. Ernemann U, Kramer U, Miller S, Bisdas S, Rebmann H, Breuninger H, *et al.* Current concepts in the classification, diagnosis and treatment of vascular anomalies. Eur J Radiol 2010;75:2-11.
9. Lazzaro MA, Badruddin A, Zaidat OO, Darkhabani Z, Pandya DJ, Lynch JR. Endovascular embolization of head and neck tumors. Front Neurol 2011;2:64.
10. Wu CY, Kahana A. Immediate reconstruction after combined embolization and resection of orbital arteriovenous malformation. Ophthal Plast Reconstr Surg 2017;33:S140-3.