Commentary: Retinal arteriovenous malformations and retinal exudation and/or macular edema

This issue of the Indian Journal of Ophthalmology features an interesting case of macular edema in retinal arteriovenous malformation (AVM, racemose hemangioma, or cirsoid hemangioma of the retina),[1] which responded well to posterior subtenon triamcinolone (PST) after treatment failure of intravitreal bevacizumab.

Archer and colleagues [Am J Ophthalmol. 1973;75(2):224-41] have classified the AVMs into three groups-

Group 1 AVM is characterized by retinal macrovessels with intervening arteriolar or abnormal capillary plexus between retinal artery and vein. Typically, these AVMs do not leak on fundus fluorescein angiogram (FFA). Associations of congenital retinal macrovessels and group 1 AVMs include a reduced diameter of foveal avascular zone/foveal hypoplasia (FAZ),[2,3] macular hemorrhage, conjunctival macrovessels, abnormal vessels in the tongue, and possibly in the brain, though brain involvement is rare.[4]

Group 2 AVM is direct communication between a dilated artery and vein without any intervening capillary or arteriole. On cursory examination, these may appear like the feeding and draining vessels of capillary hemangioblastoma[5] seen in von-Hippel–Lindau disease except the fact that intervening vascular tumor (hemangioblastoma) is not present. Though such AVMs are usually stationary, these may decompensate leading to retinal exudation (hard exudates), edema, and hemorrhages. These AVMs may get occluded and cause macular edema secondary to vascular occlusion and new AVMs may form in a place different from the occluded vessel. Other features include paravascular capillary abnormality and nonperfusion, beading or fusiform dilation of the involved vessel, perivascular scarring/sheathing, and an association with intracranial vascular malformation. Complications include intraretinal or vitreous hemorrhage, and loss of macular photoreceptors (detected with spectral-domain optical coherence tomography, and adaptive optics flood illuminated fundus camera) leading to central scotoma [Ophthalmic Surg Lasers Imaging. 2010; 1-4, PMID: 20337274].

Group 3 AVMs are characterized by more extensive direct arteriovenous communication and artery and vein cannot be differentiated. These are dilated, tortuous, convoluted, intertwined vessels that may appear like a bag of worms. The color is bright red due to the presence of oxygenated arterial blood. There may be sheathing around the AVM due to hyperdynamic stress on the vessel wall. There is a high risk of intracranial AVM (Wyburn–Mason syndrome).[6] Some of these eyes may be blind from birth. Complications of Group 3 AVMs include occlusion of the AVM, retinal arterial occlusion, retinal venous occlusion (RVO), retinal hemorrhage, Valsalva retinopathy, retinal (macular) edema- cystoid macular edema with or without subretinal fluid, retinal exudation, disc edema, arterial macroaneurysm, visual decline due to compromise of the optic nerve, vitreous hemorrhage, retinal ischemia, neovascular glaucoma, and macular hole.[4]

Cause of macular edema and/or hard exudates in AVM include

- Exudation from the AVM especially during pregnancy [J Fr Ophtalmol. 2018; 41(8):e383-e385]. The AVM may form a hairpin loop near the fovea which may leak on FFA. High intraluminal pressure due to increased speed of blood column may lead to leakage from the venous side of the AVM and/or the capillaries [Case Rep Ophthalmol. 2018; 9(3): 504–509]. The changes in the vascular wall due to hyperdynamic stress may play a crucial role in the increased permeability of the AVM. The pressure gradient along the AVM may increase the venous transmural pressure leading 'to reverse flow and excessive pressure in venous capillaries around the anastomosis'[7]
- Exudation from aneurysms at the wall of AVM
- Leak from the adjoining capillary
- RVO which might occur due to [Retin Cases Brief Rep. 2010;4(2):112-5]
  - Steal phenomenon- The blood flow is shunted via the low resistance pathway of AVM and other areas suffer.
  - Compression of the vein by AVM
  - High flow angiopathy- The exposure of veins to rapid arterial blood flow may compromise or damage the endothelium, thus predisposing to thrombus formation as per the Virchow's triad
  - The AVM itself may spontaneously get occluded leading to RVO of the distal draining peripheral retinal veins
- Possible impairment of choroidal circulation [Semin Ophthalmol 2015;30(2):154–6].

Management of macular edema in AVM

Systemic optimization is needed for macular edema in AVM patients with systemic conditions including diabetes, hypertension, and pregnancy. If vision is not compromised, then observation can be done as macular edema or retinal exudation may improve with time spontaneously. Ocular management includes intravitreal anti-vascular endothelial growth factor (VEGF) agents including bevacizumab. VEGF plays an important role in the causation of macular edema in RVO. Also, the role of anti-VEGF agents in reducing vascular permeability and improving the tight junction of the vascular endothelium is well known. A 37-year-old lady has been reported in whom three intravitreal bevacizumab injections were noted to control the macular edema.[8] Occlusion of the AVM was also noted in this patient which could have been spontaneous or triggered by the anti-VEGF injection.[8]

Periocular steroids (PST) may also work well to reduce macular edema in cases with AVM.[1]

Laser 'directly on the leaking shunt segment and over the adjacent area of leaking microvessels' has been used with success.[7] The power used was sufficient enough to induce a visible 'crimping' of the vessel. However, it should be avoided if the leaking shunt is at or very near to the FAZ. In the same report, laser was not applied in the fellow eye which did not show any leak on FFA and no fluid on optical coherence tomography.[7]

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