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Spontaneous Resolution of Presumed Idiopathic Elevated Episcleral Venous Pressure

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Abstract

Purpose—To report a case of presumed idiopathic elevated episcleral venous pressure initially requiring medical management.

Methods—We present a case report of a 28-year-old Caucasian male presenting with eye redness and soreness as a referral to the glaucoma service. He was diagnosed with IEEVP after systematic workup.

Results—Unilateral elevated intraocular pressure was detected along with dilated episcleral vessels, mild cupping of optic nerve head, and blood in Schlemm's canal, yet negative angiographic imaging and hypercoagulable labs. Topical anti-glaucoma medication normalized intraocular pressure, which then maintained with resolution of episcleral vessel caliber and blood in Schlemm's canal after discontinuation of medication.

Conclusion—This is the first reported case of spontaneous resolution of presumed idiopathic elevated episcleral venous pressure, persisting after discontinuation of medical therapy and without surgical intervention.

Keywords

Episcleral venous pressure; Blood in Schlemm's canal; Radius-Maumenee syndrome

Case Report

A 28 year-old white gentleman was referred to us for unilateral elevated intraocular pressure (IOP). He reported several months of left eye blurriness, redness and pain. History revealed remote, mild, blunt right eye trauma; his father had pulmonary embolism. Best corrected visual acuity was 20/20 right and 20/100 left; IOP was 17 and 51 mm Hg, respectively. Examination demonstrated left “conjunctival injection”. Oral acetazolamide was started with left, topical anti-glaucoma medications. Left IOP decreased the next day to 20 mm Hg. Acetazolamide was stopped and topical treatment continued. Although normalized IOP

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maintained 2 weeks later, the etiology of severely elevated IOP remained unclear, hence referral was made to our institution.

Upon presentation to us, one month after initial examination, the patient reported decreased soreness and blurriness, but persistent redness. Left visual acuity improved to 20/50. IOP was 17 and 19 mm Hg, while central corneal thickness was 534 and 552 microns, right and left respectively. Color vision was full bilaterally. No afferent pupillary defect, proptosis, or facial venous prominence was detected. Extraocular movements were full. Anterior segment examination demonstrated tortuous and dilated episcleral vessels in the left eye, much greater than right, and keratitis (Figure; top left and right). Gonioscopic examination showed open angles, but the left showed blood infero-temporally in Schlemm's canal (SC) (Figure; bottom). Dilated fundus exam showed mild left cupping. No choroidal effusion was detected. Urgent neuro-radiologic consultation was made but magnetic resonance (MR) head/orbits angiography/venography was unremarkable, including normal superior ophthalmic venous and cavernous sinus sizes. Extensive hypercoagulable workup was normal. Brain MR with-and-without contrast, one month later, was also negative. At two-month follow-up, the patient reported improvement; examination revealed complete resolution of SC blood, IOP normalization without medication, and normal episcleral vessel caliber bilaterally.

Discussion

Dilated episcleral vasculature with elevated episcleral venous pressure (EVP) and IOP is most commonly attributable to arteriovenous fistula.¹ Entities include dural arteriovenous shunt, Klippel-Trenaunay syndrome, orbital varix, and Sturge-Weber syndrome. Venous obstruction from superior vena cava blockage, orbital venous obstruction and cavernous sinus thrombosis must also be considered.² A diagnosis of exclusion, idiopathic elevated EVP may be considered when careful history, physical examination, and imaging, as done twice in our patient, fail to identify plausible causes.

Idiopathic elevated EVP with open-angle glaucoma, absent of cranio-orbital pathology, is also known as Radius-Maumenee syndrome, although it was first reported by Minas and Podos.³ Radius and Maumenee described three cases of unilateral-dilated episcleral vessels with elevated IOP and visual field deficit (one with ipsilateral SC blood) devoid of other known vascular or obstructive causes.⁴ These latter cases were also notable for less-prominent episcleral vessel dilation and normal IOP in the contralateral eye. In addition to similar unilateral findings, our case resembles those reported cases with the contralateral eye having subtle dilated episcleral vessels but normal IOP and lack of SC blood. Episcleral venous pressure itself was not measured in our case, just as it was deferred by Radius and Maumenee due to technical limitations.⁴

While Radius-Maumenee syndrome involves poor episcleral venous drainage, its cause remains unknown. Hypotheses include local blood volume and pressure shifts in periocular vasculature.⁵ The evanescent ocular hypertension and venous congestion here is presumably due to transient obstruction or fistula, likely too small to be detected. Such explanation is supported by the resolution, even after repeated radiologic intervention, although we caution

the over-interpretation as low osmolar load and distal location of contrast injection may not be sufficient enough to reverse the obstruction. Though less likely given our patient's young age and male sex,⁶ a low-flow carotid-cavernous fistula may also have been plausible, as previous cases have shown to resolve spontaneously.^{6,7}

Previous Radius-Maumenee syndrome reports typically involved years of "red eye", all having elevated IOP either refractory to medication or necessitating surgery.^{1–5,8} Severe surgical complications are common.⁸ To the authors' knowledge, this is the first spontaneous resolution of Radius-Maumenee syndrome, persisting after discontinuation of medications and without surgery. Provided that the elevated IOP and optic nerve damage are only mild, this case supports careful observation prior to the decision of performing glaucoma surgery in order to allow the spontaneous resolution to occur.

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References

1. Faroozan R, Buono LM, Savino PJ, Sergott RC. Idiopathic dilated episcleral veins and increased intraocular pressure. *BMJ*. May; 2003 327(7418):652–654.
2. Lanzl IM, Welge-Lüssen, Spaeth GL. Unilateral open-angle glaucoma secondary to idiopathic dilated episcleral veins. 1996; 121(5):587–589.
3. Minas TF, Podos SM. Familial glaucoma associated with elevated episcleral venous pressure. *Arch Ophthalmol*. 1968; 80:202–208. [PubMed: 5661886]
4. Radius LR, Maumenee AE. Dilated episcleral vessels and open-angle glaucoma. *Am J Ophthalmol*. 1978; 86:31–35. [PubMed: 677230]
5. Stock RA, Fernandes NL, Pastro NL, Oliveira RSD, Bonamigo EI. Idiopathic dilated episcleral vessels (Radius-Maumenee syndrome): case report. *Arq Bras Oftalmol*. 2013; 76(1):45–7. [PubMed: 23812528]
6. Fernandez N, Murias E, Vega P, Sainz A, Meilan A. Angiographic confirmation of the spontaneous resolution of two low-flow carotid-cavernous fistulae. *Neurologia*. 2010; 25(5):333–6. [PubMed: 20643045]
7. Bujak M, Margolin E, Thompson A, Trobe J. Spontaneous resolution of two dural carotid-cavernous fistulas presenting with optic neuropathy and marked congestive ophthalmopathy. *J Neuroophthalmol*. 2010; 30(3):222–7. [PubMed: 20498622]
8. Rhee DJ, Gupta M, Moncavage MB, Moster ML, Moster MR. Idiopathic elevated episcleral venous pressure and open-angle glaucoma. *Br J Ophthalmol*. 2009; 93(2):231–4. [PubMed: 18567649]

Figure.

Clinical color photographs. External photograph of right eye (top left) demonstrates mildly dilated and tortuous episcleral vessels compared to relatively greater severity and prominence in the left eye (top right). Gonioscopic biomicroscope photograph of left eye (bottom) shows inferotemporal angle filled with blood in Schlemm's canal and an otherwise clear view of structures in an open angle (16x).