Introduction

Intraocular pressure (IOP) is dependent on the rate of aqueous production, facility of outflow, and episcleral venous pressure. Increased IOP and visual field loss may result from an elevation of episcleral venous pressure. Most cases of dilated episcleral vessels with increased IOP can be attributed to carotid-cavernous-sinus fistula, cavernous sinus thrombosis, dural arteriovenous shunt, superior vena cava syndrome, Sturge–Weber syndrome, thyroid ophthalmopathy, orbital obstructive lesions or orbital varices. There are rare cases of open-angle glaucoma and arterialized episcleral vessels without an underlying cause. This condition is known as Idiopathic Elevated Episcleral Venous Pressure (IEEVP). Nearly 40 cases have been reported till now. We report this case because of its rarity of presentation.

Case Report

A 63 year old female presented with history of defective vision. She reported redness of eye lasting for 10 years. She had a history of fall 12 years back. There was no proptosis, tinnitus or other ocular illness. She had no other previous medical problems and was not taking any medications. Her best corrected visual acuity was 6/12 OD and 6/36 OS. A prominent left APD was present. IOPs measured 22mm Hg OD and 28 mm Hg OS. Ocular motility was full. There was no visible proptosis or chemosis and retropulsion was normal. Neurological exam showed no focal deficits. No carotid or ocular bruits were heard

Slit lamp examination revealed cork screwing and dilatation of episcleral vessels both eye. (OS >OD).(figure-1,2) Gonioscopy revealed bilaterally open angles for 360 degrees with blood in the canals of Schlemm in both eyes, greatest within the inferior 90 degrees, but without evidence of neovascularisation or angle recession.(figure-3)

Dilated fundus examination revealed optic disc pallor in the left with cupping, 0.7x0.5. Right eye showed cupping of 0.6 x 0.5 and NRR was normal.(figure-4) Visual field examination showed supranasal scotomas OD and severely depressed field OS.(figure-5) OCT showed loss of double hump pattern left eye with marked nerve fibre loss (figure-6)

With these clinical findings, all the relevant investigations like thyroid function test, chest X-ray,USG B scan and CT head were done to find out causes for elevated episcleral venous pressure, which turned out to be normal. Digital subtraction angiography was also done which showed that the superior ophthalmic veins were of normal caliber and there was no evidence of reversal of flow in either orbit.(figure-7).

All these investigations revealed no intraorbital or intracranial causes for elevated episcleral venous pressure. So we came to a final diagnosis of idiopathic elevated episcleral venous pressure.

Topical anti glaucoma medications- timolol, dorzolamide and brimonidine were given. Even with maximum medications IOP remained high. So trabeculectomy was done. IOP reduced to 16mmHg(OD) 18mmHg (OD).

Discussion

Intraocular pressure (IOP) is dependent on the rate of aqueous production, facility of outflow, and episcleral venous pressure (EVP). Increased IOP and visual field loss may result from an elevation of EVP. Normal EVP is 8 to 10 mm Hg. Arteriovenous fistula is the most frequent cause of elevated episcleral venous pressure. Other causes include cavernous sinus thrombosis, dural arteriovenous shunt, superior vena cava syndrome, Sturge–Weber syndrome, thyroid ophthalmopathy, orbital obstructive lesions or orbital varices.

Clinical features of these cases depend on the cause, but the consistent features are dilated and tortuous episcleral vessels and elevated IOP. The ocular injection results from engorged, arterialized conjunctival vessels.

There is, however, an infrequent syndrome of ocular injection and increased IOP which is idiopathic. The diagnosis of IEEVP is one of exclusion after intraorbital and intracranial pathology has been excluded. Although originally described by Minas and Podos, the same entity is termed the Radius–Maumenee syndrome in the German literature.

The diagnosis is based on the clinical findings of arterialized episcleral veins, elevated IOP causing characteristic optic nerve and visual field changes typical of glaucoma, and an open angle on gonioscopy. Diagnostic evaluation should include a complete ophthalmic examination, radiological imaging, such as MRI, and vascular imaging such as DSA or carotid Doppler to exclude a cerebrovascular disorder. Blood in the Schlemm canal is a general sign of elevated EVP but is not present in all cases of IEEVP.

The diagnosis of glaucoma secondary to IEEVP is clinical. Measurement of EVP is not practical. Familial and sporadic cases have been reported. Both unilateral and bilateral cases have been reported.
No clear aetiology has been identified for IEEVP. Various postulations include localised venous obstruction at the region of extraocular muscles and congenital anomaly of the vasculature.

Treatment is mainly by reducing IOP with aqueous suppressants. Drugs which affect the outflow pathway are not effective. Laser trabeculoplasty is not helpful due to normal facility of outflow. Patients who do not respond to medical management will need trabeculectomy. Other surgeries include non penetrating surgeries like microsurgical sinusotomy to reduce pressure gradient from schlemms canal to episcleral vessels. Certain complications are more likely to occur in the setting of elevated episcleral venous pressure like choroidal effusion, expulsive hemorrhage and flat anterior chamber. Preoperative mannitol is mandatory. Other precautions like slow decompression, usage of intracameral viscoelastics and preplaced sclera flap sutures should be considered.

Summary

It is important for clinicians to be aware of IEEVP so that the patients presenting with dilated ESV can be screened appropriately for serious underlying conditions. Catheter angiography should be done in patients with the clinical features of this syndrome, who have normal orbital colour doppler to avoid delay in the treatment of elevated IOP. Patients generally require filtering surgery and clinicians should be cautious about the development of choroidal effusions in the postoperative period occurring at IOPs not typically thought of as being hypotonous.
References