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Case Report

A rare case of Radius Maumenee syndrome with unilateral open angle glaucoma

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ABSTRACT

Elevated episcleral venous pressure (EEVP) is a rare but important cause of raised intraocular pressure. The resultant glaucoma is often severe and refractory to routine anti glaucoma treatment. Usually it occurs due to arteriovenous fistulas or venous congestion. This entity is important to detect as it may be associated with serious pathologies like retrobulbar tumor, cavernous sinus thrombosis, mediastinal tumor to name a few. EEVP is considered idiopathic after ruling out such disorders. Here, we have described idiopathic EEVP also known as Radius-Maumenee syndrome along with a detailed diagnostic and therapeutic approach of management.

1. Introduction

IOP is regulated by a complex process of generation and drainage of aqueous humour.[1] Elevated episcleral venous pressure (EEVP) is a rare cause of IOP elevation with almost 1 mm rise in IOP for every 1 mm of EEVP.[2] Chronic and persistent elevation of EVP results into secondary open angle glaucoma which is often refractory to routine therapy.[3]

EEVP is often secondary to arteriovenous fistulas or venous congestion.[1] There are numerous causes of EEVP, which include, obstruction at the level of episcleral vein, vortex veins or superior ophthalmic veins due to retrobulbar tumour or cavernous sinus thrombosis, obstruction at the level of jugular vein, internal carotid artery or an obstruction at the level of superior vena cava which may be associated with mediastinal tumour and rarely idiopathic.[2–5]

Idiopathic elevated epi-scleral venous pressure (IEEVP) or Radius-Maumenee syndrome is a rare clinical entity which was first described in 1968 by Minas and Podos.[4] Approximately only 60 cases have been reported in the literature to the best of our knowledge. Hence, here we are reporting a rare case of Radius-Maumenee syndrome. We have described the need of investigations, approach to various treatment and surgical options.

2. Case Report

50 year old male presented with chief complaints of chronic redness of left eye (LE) and gradual diminution of vision with headache. There was no antecedent history of head trauma, systemic illness or family history of ocular disease. Patient was a known case of glaucoma in left eye and he had a history of piles. On torchlight examination, there were no facial or external abnormalities and ocular movements were normal. There was no proptosis, bruit or visible pulsation in eyes. Visual acuity was normal (6/6)

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in the right eye (RE) and 6/12p in the LE with only temporal vision. Anterior and posterior segment of RE was within normal limits. (Figure 1 A) LE had dilated, hyperaemic and tortuous episcleral veins with grade 4 RAPD. (Figure 1 B) Intraocular pressure (IOP) was 42 mm Hg in the LE. Gonioscopy examination showed bilateral open angle. However blood was noted in schlemm's canal in LE suggestive of elevated episcleral venous pressure (EEVP). (Figure 1 C, D) Severe glaucomatous cupping of 0.95-0.99 was also seen in LE. (Figure 1 E, F) Visual field analysis was normal in right with advanced field defect and constriction in LE. (Figure 2 A, B) Magnetic resonance angiography of brain and orbit (Figure 2 C) was performed to rule out intracranial pathologies but was normal. All other investigations (Figure 2 D, E, F) such as x-ray skull, x-ray chest, and B scan were also normal. Thus, after thorough evaluation, diagnosis of Radius Maumenee syndrome

2.1. Management

Patient was started with oral acetazolamide 250 mg four times/day with combination of dorzolamide 2% and timolol 0.5%, along with brimonidine 0.2% eye drops. However, IOP could not be controlled and eventually patient underwent trabeculectomy with mitomycin C. Post-op tablet prednisolone (1 mg/kg/day) was also added to the routine postoperative regimen.

Immediate postoperative course was unremarkable. However, at 2 months of follow up, (figure 1g) patient presented with visual acuity of 1/60 and flat, fibrosing bleb with IOP of 32 mm Hg along with cataractous changes. Bleb revision was performed at this visit and phacoemulsification with intraocular lens implantation was done at 5 months follow up visit. At the last follow up visit, (figure 1h) visual acuity was 6/60 with a functioning bleb and IOP of 12 mm Hg. Patient was on two antiglaucoma medications additionally.

3. Discussion

Radius Maumenee syndrome is a diagnosis of exclusion once other causes of EEVP are ruled out by investigations. Magnetic Resonance angiography (MRA), CT angiography, B scans and colour Doppler must be done to rule out cerebrovascular pathologies.[6–8] Chest x-ray is necessary to rule out mediastinal mass.

Aqueous suppressants are the first line of treatment.[9] Beta blocker and alpha agonist were used in this patient. Newer drugs like natursudil and repasudil can also be used.[7] However, earlier articles have reported inefficiency of medications in Radius maumenee syndrome.[9]

Surgical management is challenging with options like prophylactic deep sclerectomy, viscocanalostomy, trabeculectomy, and shunt surgery.[9,10] In this case, trabeculectomy with MMC was done considering advanced glaucoma and high IOP. Failure rate is more here due to intra-operative bleed during conjunctiva handling, scleral incision, or bleed from exposed ends of schlemm's canal. This also potentially leads to heightened post-operative inflammation and bleb failure.[3] Mitomycin application is necessary to reduce the failure rate.

Several studies reported choroidal effusion without hypotony is frequent in these patients after filtration surgery.[9,10] To combat this, we gave systemic steroid in our case. Choroidal effusion was not seen in this patient intraoperatively or on follow up visits.

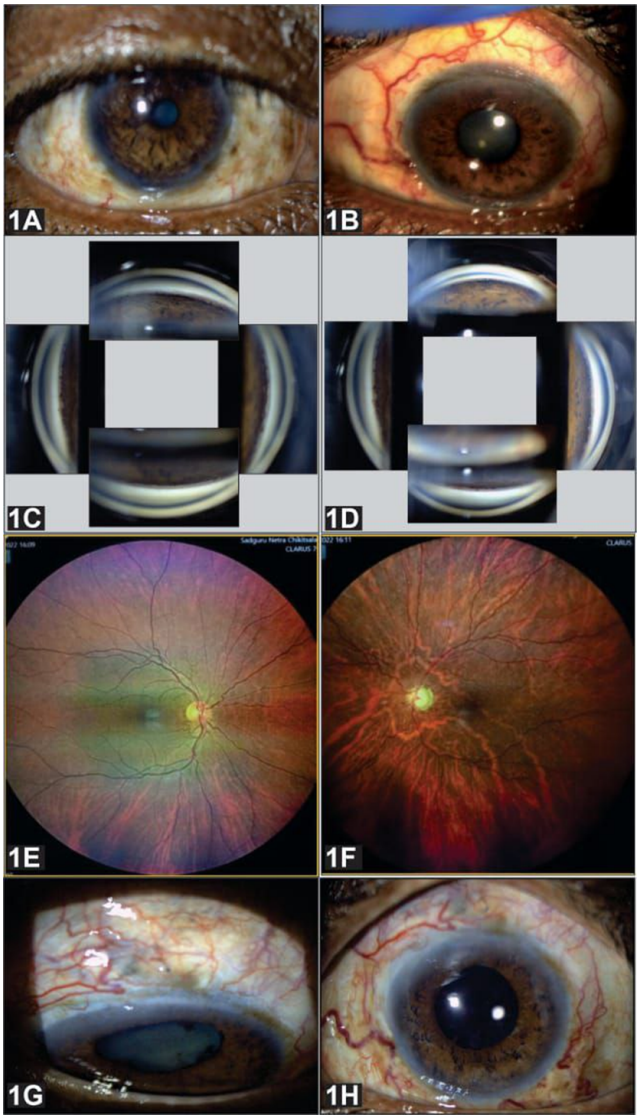

Figure 1: A): Right eye showing early cataractous changes; **B**): Left eye showing dilated tortuous episcleral veins; **C,D**): Shows bilateral open angles with left eye with blood in schlem's canal; **E**): Fundus photo showing right eye normal disc; **F):** Left eye glaucomatous changes; **G**): Follow up showing cataractous changes with flat bleb; **H**): Last follow up showing bleb with PCIOL in bag

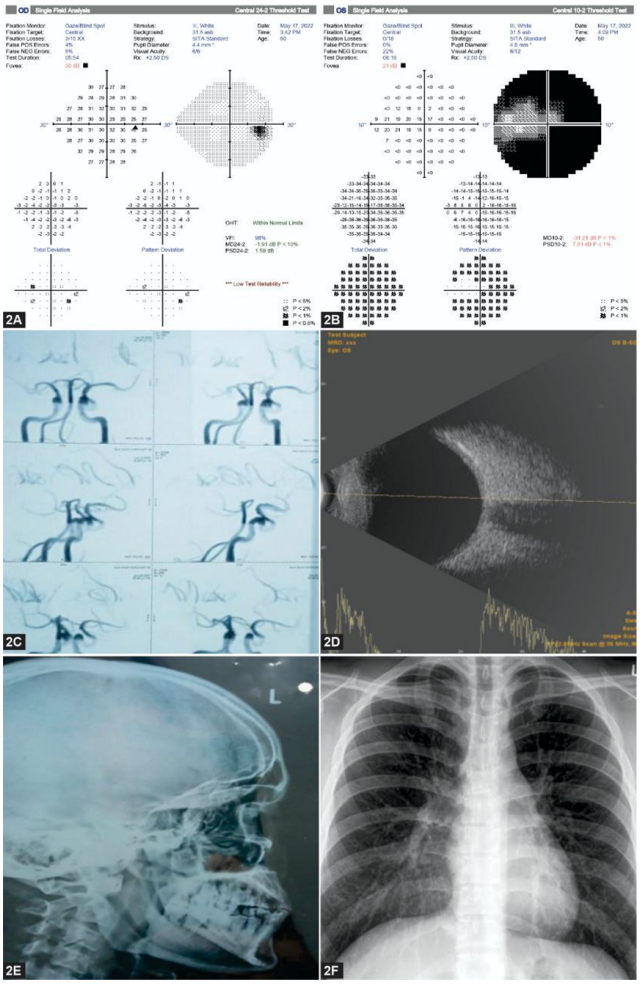

Figure 2: A): Right eye fields are normal; **B**): Left eye fields showing involvement of central 10 degree fields; **C**): Magnetic Resonance Angiography brain and orbit showed no evidence of tumor or arteriovenous malformations; **D,E,F**): B Scan, X-ray skull and x- ray chest were unremarkable

4. Conclusion

Medical management is not sufficient in most of the eyes, needing filtration surgery. IEEVP is a diagnosis of exclusion. So, other systemic causes must be ruled out. These investigations are necessary as they are both sight-saving and lifesaving. Surgical management by trabeculectomy requires the use of antimetabolites, intra-op tight sutures should be applied considering the chances of post op choroidal effusion in the absence of hypotony. Considering high failure in these cases, meticulous close follow up is mandatory.

5. Source of Funding

None.

6. Conflict of Interest

None of the authors have any proprietary interests or conflicts of interest related to this submission.

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