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Case Series

Ophthalmology Section

Trabeculectomy with Mitomycin-C in Patients with Iridocorneal Endothelial Syndrome: A Case Series

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ABSTRACT

Iridocorneal Endothelial syndrome (ICE) is a rare ocular disorder characterized by abnormal endothelization of angle structure and iris producing characteristic manifestations including secondary angle closure glaucoma. We describe the clinical course of three patients with secondary glaucoma due to ICE syndrome who underwent trabeculectomy with mitomycin-C. At last follow-up, all patients had controlled IOP (<18 mm Hg) off drug. Early postoperative period was accompanied by shallowing of anterior chamber diffuse vascularisation of bleb frequent episodes of high IOP which were managed successfully. Trabeculectomy with MMC offers a good treatment choice for the management of secondary glaucoma associated with ICE syndrome.

Keywords: ICE syndrome, Secondary glaucoma, Trabeculectomy with MMC

CASE SERIES

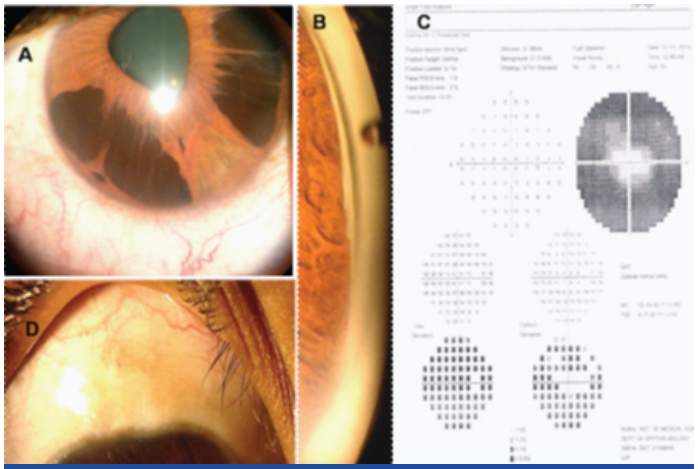
Case 1

A 43-year-old male presented with redness, pain and diminution of vision in the right eye (OD) for 3 months. There was no history of similar complaints in the left eye (OS). Past clinical history and systemic evaluation did not reveal any abnormality. There was no family history for any ocular disease. Patient underwent comprehensive ophthalmic examination at our Institution, which is a rural based tertiary health care centre. His un-corrected visual acuity (UCVA) was 6/9 OD and 6/6 OS using Snellen's visual acuity chart. With +0.25 DS, right eye Best Corrected Visual Acuity (BCVA) was 6/6. Intra-ocular pressure (IOP) with Goldmann Applanation Tonometry (GAT) at the first clinical presentation was 56 mmHg OD, and 10 mmHg OS. Mild bulbar conjunctival congestion was noticed OD. On further slit lamp biomicroscopic examination, patient revealed mild corneal haze, beaten silver appearance of posterior corneal surface associated with iris pigments over the corneal endothelial surface. Anterior chamber was shallower as compared to other eye. Pupil was updrawn superotempoarlly revealing corectopia, pseudopolycoria [Table/Fig-1a]. Iris thinning was noticeable due to stretching of iris over the inferior area. Lens was transparent with few iris pigments depositions. Anterior segment was within-normal limits OS. Gonioscopic examination revealed 360° synechial angle closure (Grade 0) OD, and open angles (Grade IV) OS [Table/Fig-1b]. On fundus evaluation, vertical cup disc ratio of 0.8 with diffuse thinning of neuroretinal rim was seen OD, whereas fundus OS was with-in normal limits.

Based on clinical features and examination, the diagnosis of OD ICE syndrome with secondary angle closure glaucoma was made. Patient was managed on in-patient basis with intravenous mannitol (20%) injections, oral acetazolamide (250 mg QID), topical timolol (0.5%) and brimonidine (0.2%). Although, patient was symptomatically better 4-6 hours later after starting treatment but IOP was still in higher range (30 mmHg). Visual field (Humphrey Field Analyser 30-2) of right eye showed the advanced visual field defect [Table/Fig-1c], whereas left eye was with-in normal limits. The target IOP was not achieved despite using maximal anti-glaucoma medications; therefore it was decided to go for glaucoma filtration surgery. Patient was counselled regarding the trabeculectomy with mitomycin C (MMC).

Case 2

A 52-year-old female presented with pain and diminution of vision in the right eye (OD) for 1 month. There were no complaints in the left eye (OS). She was treated for raised IOP with antiglaucoma therapy by the local practitioner. Her family history was insignificant.



[Table/Fig-1]: (a) Clinical photograph of a patient with features of iridocorneal endothelial (ICE) syndrome in the right eye at presentation; (b) Gonioscopy of nasal angle showing closed angles with anterior synechiae; (c) Visual field (Humphrey field analyser) demonstrating the advanced visual field defect at presentation; (d) photograph showing diffuse well formed bleb at 1 year.

Her UCVA was 6/18 and 6/6 in OD and OS respectively. IOP measured with GAT were 32 mm Hg (on acetazolamide, brimonidine & timolol) OD and 14 mm Hg OS. On slit lamp examination there were mild corneal haze, iris thinning inferiorly and superior updrawning of pupil revealing corectopia. On gonioscopic examination, 270° anterior synechiae were noticed. Fundus examination revealed glaucomatous cupping of 0.7. Based on the clinical features, the diagnosis of secondary angle closure due to ICE syndrome was reached. As the medical therapy was ineffective, it was planned to go for surgical management in the form of trabeculectomy with MMC.

Case 3

A 38-year-old female presented with mild pain over his right eye since one year. She consulted to the local practitioner and advised antiglaucoma medication. Patients was intolerant to antiglaucoma drugs particularly acetazolamide, so she was referred to the tertiary health care centre for further management. On the first clinical presentation to our centre patient had UCVA 6/12 OD and 6/6/ OS. IOP with GAT was 24 mm Hg OD (on timolol 0.5%, brimonidine 0.2%, latanoprost 0.005%) and 16 mm Hg OS. Also, the patient was intolerant to acetazolamide. Anterior segment

examination revealed diffuse mild corneal haze associated with iris atrophic changes and polycoria. Gonioscopy revealed peripheral anterior synechiae in all 4 quardrants. Fundus evaluation showed advanced glaucomatous cupping of 0.9. Patient was planned for trabeculectomy with MMC due to uncontrolled IOP on maximal medical therapy and intolerance to medications.

Surgical procedure

Trabeculectomy with MMC was performed in all cases by author (VKJ) using fornix based conjunctival flap, making partial thickness scleral flap, application of MMC (0.04%) for 4 minutes and making sclerostomy of around 2x1 mm in size. No significant intraoperative complication was seen in any case.

Postoperative follow-up

All patients received topical moxifloxacin 0.5% and prednisolone 1% 4 times per day and cycloplegic agent once a day for 1 month. Flat anterior chamber with iridocorneal touch without bleb leak was noticed in both case 1 and case 2 in the 1st week which was successfully managed with air injection into the anterior chamber. Case 2 experienced the recurrent episodes of high IOP with low-lying bleb in the first 2 weeks after surgery. It was managed with bleb massage successfully. There were no instances of any significant postoperative complication in case 3. Abnormal vascularisations were noticed in all cases which was more in case 1. It was noted at 2 weeks in case 1, and on 1 week followup in the other two cases. Since abnormal vascularisation is a sign of impending fibrosis so subconjunctival injections of antifibrotic agent 5-fluorouracil (2.5mg / 0.1 ml) were given. At the last follow-up (1 year for case1; and 6 months for case 2 & case 3), all patients had controlled IOP (< 18 mm Hg) without any antiglaucoma medication. The mean IOP was 12± 0.9 mm Hg. All patients maintained vision >6/9 on snellen's chart. Diffuse and well formed blebs were noticed in all cases [Table/Fig-1d].

DISCUSSION

Iridocorneal endothelial (ICE) syndrome is a rare condition caused by an abnormal proliferation and migration of corneal endothelial cells over the angle structures and anterior surface of iris [1]. It comprised of three variants: Chandler syndrome, progressive iris atrophy and Cogan–Reese syndrome [2]. ICE syndrome is usually sporadic and unilateral with middle aged female predilection. Glaucoma associated with ICE syndrome poses a significant challenge to ophthalmologists. It is generally refractive to medical treatment and most often require trabeculectomy with adjunctive antifibrotic agents. Although the success rate of trabeculectomy in ICE syndrome has improved from pre-antimetabolite era [3] to mitomycin C (MMC) era [4,5], but still the chances of failure of surgery are high particularly over the long-term period. We describe a case series of 3 patients of ICE syndrome presented with raised intraocular pressure (IOP), and were managed successfully with trabeculectomy with MMC. Informed consent for reporting of cases was obtained from all patients. Trabeculectomy in a case of secondary glaucoma due to ICE syndrome generally has poor outcome. The failure of trabeculectomy may be due to several factors like younger age, blockage of internal ostium by an abnormal membrane, iris tissue, and most importantly because of higher chances of abnormal bleb fibrosis [3,6]. Since the introduction of antifibrotic agents (5-fluorouracil, MMC) as an adjuvant to trabeculectomy, the success rate has improved especially in secondary glaucoma [4,5]. Trabeculectomy without the use of antifibrotic agents has been reported to be successful in 61-64% of the cases [3,7,8]. The use of mitomycin has been shown to improve success rates in all glaucoma subtypes.

Earlier case series reported different success rates (64% to 80%) of trabeculectomy with antifibrotic agent in patients of ICE syndrome [4,5,9]. In a latest case series of 16 eyes of the outcome of primary trabeculectomy with MMC in ICE syndrome by Chandran P et al., showed the moderate surgical success and maintaining long-term IOP control is still a challenge [9]. However, there are still very few studies reporting the long term success rates of primary trabeculectomy augmented with MMC. Though we had fewer cases and shorter follow-up, author experience in managing these cases add useful information to the literature. In our cases, though the 1st postoperative month was marked by some early postoperative complications like shallow anterior chamber, high IOP, bleb vascularisation, but we succeeded in managing these complications because of close follow-up with early and adequate intervention. The IOP was well controlled and was maintained in lower teens (target IOP). Our cases had shorter follow-up and possibility of failed trabeculectomy could not be ruled out in future. The outcome of trabeculectomy with MMC may be better and offers a good treatment choice for the patients of secondary glaucoma associated with ICE syndrome. However, the early postoperative period needs a close watch for development and appropriate management of complications. This will probably ensure a successful long term IOP control.

REFERENCES

[1] Campbell DG, Shields MB, Smith TR. The corneal endothelium and the spectrum of essential iris atrophy. *Am J Ophthalmol.* 1978;86:317–24.
[2] Eagle RC, Font RL, Yanoff M, et al. Proliferative endotheliopathy with iris abnormalities. The iridocorneal endothelial syndrome. *Arch Ophthalmol*. 1979;97:2104–11.
[3] Laganowski HC, Kerr Muir MG, Hitchings RA. Glaucoma and the iridocorneal endothelial syndrome. *Arch Ophthalmol*. 1992;110:346–50.
[4] Lanzl IM, Wilson RP, Dudley D, et al. Outcome of trabeculectomy with mitomycin-C in the iridocorneal endothelial syndrome. *Ophthalmology.* 2000;107:295–97.
[5] Doe EA, Budenz DL, Gedde SJ, et al. Long-term surgical outcomes of patients with glaucoma secondary to the iridocorneal endothelial syndrome. *Ophthalmology*. 2001;108:1789–95.
[6] Jacobi PC, Dietlein TS, Krieglstein GK. Primary trabeculectomy in young adults: long-term clinical results and factors influencing the outcome. *Ophthalmic Surg Lasers*. 1999;30:637-46.
[7] Kidd M, Hetherington J, Magee S. Surgical results in iridocorneal endothelial syndrome. *Arch Ophthalmol*. 1988;106:199-201.
[8] Shields MB, Campbell DG, Simmons RJ. The essential iris atrophies. *Am J Ophthalmol*. 1978;85:749–59.
[9] Chandran P, Rao HL, Mandal AK, et al. Outcome of primary trabeculectomy with mitomycin-C in glaucoma secondary to Iridocorneal Endothelial Syndrome. *J Glaucoma*. 2015 Apr 24 [Epub ahead of print].

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