

Case Report

Medical and surgical management has a role in control of intraocular pressure in early detected iridocorneal endothelial syndrome: case series

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ABSTRACT

Iridocorneal endothelial syndrome is a rare ocular disorder characterized by abnormal endothelialization of angle structure and iris producing characteristic manifestation including secondary angle closure glaucoma. The purpose is to present four cases of iridocorneal endothelial syndrome with glaucoma and discuss clinical presentation and management strategies. Out of four cases, two cases of Cogan-Reese syndrome who present early have well controlled IOP with trabeculectomy with mitomycin-C. One case who presented late with progressive iris atrophy and secondary angle closure glaucoma went to absolute stage in spite of trabeculectomy with mitomycin-C. One case with Chandler's syndrome has well controlled IOP with topical anti-glaucoma medication.

Keywords: Cogan-Reese syndrome, Chandler's syndrome, Progressive iris atrophy, Trabeculectomy with mitomycin-C

INTRODUCTION

Iridocorneal endothelial (ICE) syndrome is a rare disorder characterized by proliferative and structural abnormalities of the corneal endothelium, progressive obstruction of the iridocorneal angle and iris abnormalities such as atrophy and hole formation.¹

The consequence of these changes are corneal decompensation and secondary glaucoma which represent the most frequent causes of visual function loss in patients with ICE syndrome. The three subtypes of this syndrome are: the progressive iris atrophy, the Cogan-Reese syndrome and the Chandler syndrome.² It is commonly found in women in adult age group. Clinical history and complete eye examination including slitlamp biomicroscopy, tonometry and gonioscopy are necessary to reach a diagnosis. Imaging techniques like confocal microscopy and ultrasound bio-microscopy are used to

confirm the diagnosis by revealing the presence of ICE cells on the corneal endothelium and the structural changes of the anterior angle. An early diagnosis is helpful to better manage the most challenging complication such as secondary glaucoma and corneal edema. Treatment of glaucoma due to ICE syndrome requires glaucoma filtering surgery with antifibrotic agent and glaucoma drainage implants. Visual impairment and pain associated with corneal edema can be managed with endothelial keratoplasty.

CASE REPORT

Case 1

A 42-year-old female presented to our out-door with headache, intermittent pain, blurring of vision-left eye since one year. She was treated irregularly with some topical medication.

No history of similar complaints in the right eye. Her BCVA in right eye was 6/6 and in left eye 6/60. On slit lamp examination of left eye cornea was clear, AC-clear content, PACD= ½ CT, iris atrophic patches, iris holes with numerous small pigmented nodules found inferiorly. Pupil was single but correctopia was seen. Fundus shows glaucomatous cupping with C:D ratio 0.8:1 with NRR thinning. IOP on aplanation tonometry in left eye was 42 mmHg. Gonioscopy of left eye showed 360° peripheral anterior synechiae. Visual field showed inferior arcuate defect. Specular microscopy revealed reduced endothelial cell count with pleomorphism and polymegathism. All ocular examination and finding were normal in right eye with IOP 16 mmHg. With these findings, the patient was diagnosed as Cogan Reese variety of ICE syndrome (Figure 1). The patient was started on Brimonidine Tartarate (0.2%) eye drop- twice daily, Timolol maleate (0.5%) eye drop twice daily and Acetazolamide tablet (250 mg) - thrice daily. As IOP was not controlled within two weeks of topical anti-glaucoma medication, patient underwent trabeculectomy with mitomycin-C (0.04%). First follow-up visit after one week showed a well formed diffuse bleb with IOP of 26 mmHg. In the 2nd week, IOP was 18 mmHg with a diffuse avascular bleb. Visual acuity in left eye was 6/60 and IOP was maintained with 18 mmHg after one month, three months, six months' follow-up.

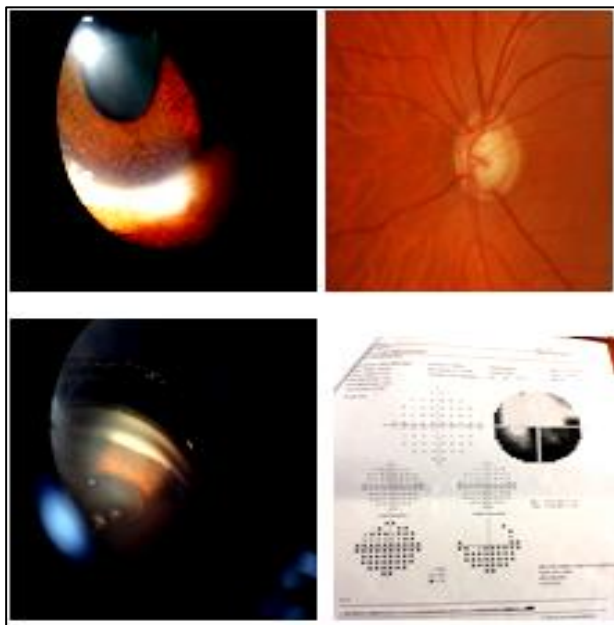


Figure 1: The patient was diagnosed as Cogan Reese variety of ICE syndrome.

Case 2

A 41-year-old female presented with mild pain and diminution of vision in her right eye for past six months. There was no similar complaint in the left eye. Systemic evaluation was normal. Patient underwent complete ophthalmic examination in our hospital. Her BCVA in right eye was 6/18 (with -5 D spherical) and in left eye

6/6 (with -2 D spherical). IOP in right eye was 40mmHg and left eye 14 mmHg. On slit lamp examination of right eye, patient had corneal haze, iris with multiple pigmented nodules at inferior quadrant (Figure 2). Pupil showed correctopia with ectropion uveae. Lens was clear. Fundus examination revealed glaucomatous cupping of 0.7:1. Gonioscopy showed 270° peripheral anterior synechiae. Based on clinical examination, diagnosis of Cogan Reese Syndrome with secondary angle closure glaucoma was made. After putting the patient with topical anti-glaucoma medication i.e., Brimonidine eye drop-twice daily, Timolol eye drop-twice daily and Acetazolamide tablet (250mg) thrice daily; target IOP was not achieved. Then patient underwent Trabeculectomy with mitomycin-C in right eye. Her BCVA was 6/18 in right eye with IOP of 18 mmHg after one, three and six months follow-up.

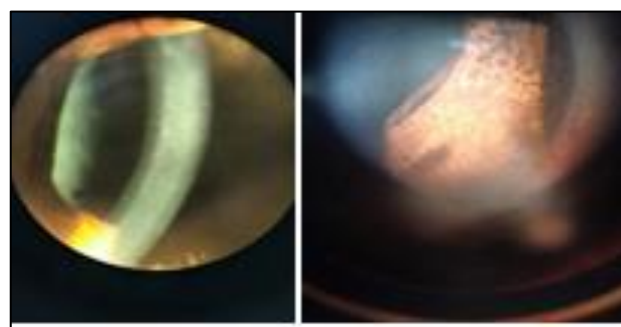


Figure 2: Corneal haze, iris with multiple pigmented nodules at inferior quadrant.

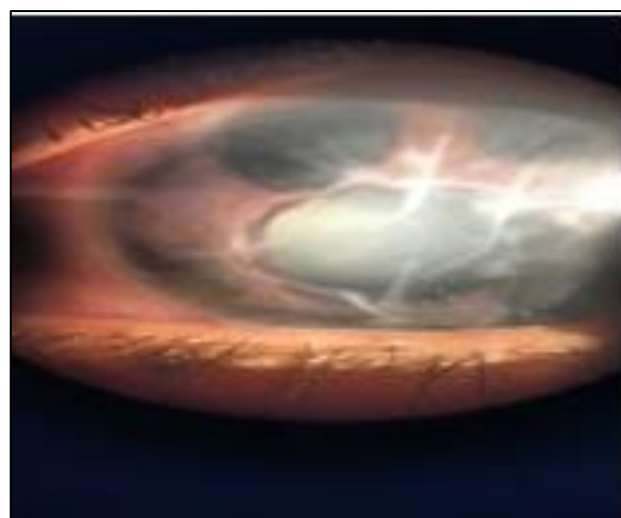


Figure 3: On slit lamp examination of left eye showed iris atrophy, iris holes and ectropion uveae

Case 3

A 48-year-old female presented with pain and gross diminution in vision in left eye for past 3 years. She was diagnosed as primary angle closure glaucoma and under treatment of anti-glaucoma medication by local practitioner. On first clinical examination BCVA in right

eye was 6/6 and in left eye 3/60. On slit lamp examination of left eye, corneal edema was seen. Iris-progressive iris atrophy with iris holes and ectropion uveae (Figure 3). Lens was clear. Fundoscopy revealed glaucomatous cupping of C:D ratio of 0.9:1 with gross NRR thinning. IOP in left eye was 56mmHg. When cornea becomes clear with topical hypertonic saline and topical antiglaucoma medication, gonioscopy revealed 360° synechial angle closure. Perimetry showed advanced visual field defect. All findings were normal in right eye. As IOP was not controlled with topical and oral antiglaucoma medication, in left eye, trabeculectomy with mitomycin-C was done in left eye. Patient had recurrent episodes of high IOP in first one month follow up with a low-lying bleb and vascularisation. Even after bleb massage and topical anti-glaucoma medication, patient went to absolute stage in left eye after one year.

Case 4

A 45-year-old woman presented with pain in right eye for past four months. There are no similar complaints in left eye. On examination, her BCVA in right eye was 6/9 and in left eye 6/6. On slit lamp examination patient had corneal haze with mild corneal edema with beaten - silver appearance, areas of atrophy of iris, correctopia was seen. Lens was transparent. Left eye anterior segment was normal. IOP in right eye was 30 mmHg and in left eye 12 mmHg. After regression of corneal edema in right eye with hypertonic saline, gonioscopy revealed 180° of synechial angle closure. Fundus shows glaucomatous cupping with C:D ratio of 0.5:1. Perimetry shows early visual field defect. On specular microscopy, there is decrease in endothelial cell count with ICE cells. All findings were normal in left eye. Based on the clinical feature and examination; diagnosis of ICE syndrome with secondary angle closure glaucoma was made in right eye. Patient was started with timolol eye drop (0.5%) twice daily and Brimonidine eye drop (0.2%) twice daily and hypertonic saline thrice daily. After one week of follow up IOP was reduced to 18 mmHg in right eye. IOP is maintained with 14 mmHg in right eye with topical anti glaucoma medication with visual acuity of 6/9 after 3 months and 6 months follow up.

DISCUSSION

ICE syndrome is a rare disorder. It is sporadic in presentation. It is usually unilateral and typically affects more often women in third to fifth decade.

The true etiology of ICE syndrome is not well understood. It has been seen that an underlying viral infection with Herpes simplex virus (HSV) or Epstein-Barr Virus leads to a low-grade inflammation at the level of the corneal endothelium resulting in its unusual epithelial like activity. PCR testing of corneal endothelial cells from ICE syndrome patients shows high percentage of HSV DNA. On a pathological level, it is felt that the normal endothelial cells have been replaced with a more

epithelial like cell with migratory characteristics. Transmission and scanning electron microscopic examination of these cells has demonstrated a population of well differentiated cells with epithelial features such as desmosomes, monofilaments and microvilli. The altered endothelium migrates posteriorly moving beyond Schwalbe line onto the trabecular mesh work and at times onto peripheral iris. Contraction of this tissue within the angle and on the iris results in high peripheral anterior synechiae and iris changes characteristic of ICE syndrome. Secondary angle closure glaucoma is a consequence of high PAS but can at times occur without overt synechiae because the advancing corneal endothelium can functionally close the angle without contraction. So, patients may initially present with what appears to be open angle glaucoma because the fibro vascular membrane obstructing aqueous flow can be difficult to visualize with gonioscopy. The corneal edema found in ICE syndrome patients is felt to be secondary to both elevated intra-ocular pressure from secondary angle-closure glaucoma and from sub-normal pump function from altered corneal endothelial cells.

ICE syndrome should be differentiated from posterior polymorphous corneal dystrophy, Axenfeld-Rieger syndrome, Aniridia (Iris hypoplasia) and malignant melanoma of iris.

Glaucoma was reported in more than 50% cases of ICE syndrome. During the initial phase of ICE syndrome, the glaucoma can be managed by medical treatment. However, in the later phases of the disease surgical intervention is required in all cases. The improvement of glaucoma by conventional trabeculectomy in these cases is poor. Kidd et al reported the success rates for the first, second and third trabeculectomy operation in a series of 42 cases with ICE syndrome is 64%, 79% and 63% respectively.⁶ The use of mitomycin-C has been shown to improve success rate in all glaucoma surgeries. Earlier case series reported different success rate (54% to 80%) of trabeculectomy with antifibrotic agent in patients of ICE syndrome.³⁻⁵ In a latest case series of 16 eyes of the outcome of the primary trabeculectomy with mitomycin-C in ICE syndrome by Chandran P et al showed the moderate surgical success and managing a long term IOP control is still a challenge.⁵

CONCLUSION

Prognosis for patients with ICE syndrome depend on the complications related to the disease like glaucomatous optic neuropathy, corneal edema and decompensation, iris changes and failed glaucoma surgery. This is dependent on the timing of diagnosis within the disease course and success or failure of treatment.

Early diagnosis and management of glaucoma is really a challenge in ICE syndrome. During initial phase of ICE syndrome, glaucoma can be managed by medical treatment. But in later phases surgical intervention is

required. The outcome of trabeculectomy with mitomycin-C may be better and offers a good treatment of choice for patients of secondary glaucoma associated with ICE syndrome. However close follow up should be done in early post-operative period for appropriate management of complication. This will probably ensure a successful long term IOP control.

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