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Case Report

IRIDOCORNEAL ENDOTHELIAL SYNDROME: A CASE REPORT

Sunil Verma[1*], J S Bhalla[2], Rakesh Verma[3]

[1]Department of Ophthalmology, BPS Government Medical College for Women, Khanpur Kalan, Haryana, India.
[2]Department of Ophthalmology, Deen Dayal Upadhyaya Hospital, New Delhi, India.
[3]Department of Ophthalmology, RIO (PGIMS) Rohtak, Haryana, India.

ABSTRACT

Iridocorneal endothelial syndrome is progressive disorder rarely encountered in routine practice. We are presenting here such rare cause of glaucoma because of its diagnostic and therapeutic challenges. A 32 year old female presented with diminision of vision, intolerance and pain in her left eye. The patient was completely evaluated for the complaints. Intraocular pressure was 20mmHg in the right eye and 28mmHg in the left eye. Specular microscopy of the left eye showed pleomorphism and polymegathism. Patient underwent trabeculectomy with Mitomycin-C (MMC) with a well-functioning bleb to reduce the intraocular pressures following failure of topical anti-glaucoma medications.

Key words: Iridocorneal endothelial syndrome, Iris naevus, Cogan Reese, Trabeculectomy.

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INTRODUCTION

Iridocorneal endothelial syndrome (ICE-S) is a rare disease with a prevalence of less than one per two lakh population. It generally affects Caucasian and persons between 20 and 50 years of age with female predilection. Rarely, ICE-S may develop in children and teenagers [1]. There are no recognized systemic or genetic associations with this syndrome [2].Yanoff described three rare overlapping conditions associated with this syndrome– Essential (progressive) iris atrophy, Iris naevus (Cogan-Reese syndrome) and Chandler's syndrome [3].Common features of iris naevus (Cogan-Reese) syndrome include corneal proliferative endotheliopathy secondary to corneal edema, peripheral anterior synechiae and abnormal iris stroma including stromal matting with effacement of normal surface pattern, ectopionuveae, pigmented iris nodules and naevi of the anterior iris surface [4].These pathogenetic mechanisms are due to the abnormal proliferation of the corneal endothelial cells[5]. We are presenting this case for its rarity and management is discussed.

Corresponding Author

Sunil Verma
Department of Ophthalmology, BPS Government Medical College for Women, Khanpur Kalan, Haryana, India.
Email: **verma.sunil4june@gmail.com**

CASE REPORT

A 32 year old previously healthy woman presented to outpatient department with three years history of diminision of vision in left eye (painful), intolerance to light,glare and coloured halos. The patient was systemically stable with no history of known drug allergy. Family and personal history was not contributory. Congenital abnormities were absent.

Ocular Examination

At the time of presentation best corrected visual acuity was 6/6 (P) in the right eye and 6/18 in the left eye with no improvement with a pin hole. Slit lamp examination of right eye was normal. There was mild corneal endothelial haze (+), irregular excrescences over endothelium, sensation- Intact as Comparable to RE shown in figure 1.

Iris and pupil of RE was normal. But in LE iris has atrophic patches (+), pigmented nodules, holes, Irido-Corneal adhesion (+) inferiorly.
Pupil has Corectopia (+), polycoria (+), peaked inferiorly as shown in figure 2.

On fundus examination, the right eye was normal. In the left eye vertical C: D (Cup: Disc) Ratio was 0.9:1. NRR (Neuroretinal rim) thinned in superior and inferior quadrants. A: V (Arteriovenous) Ratio was 2:3. Foveal reflex- Dull. No Haemorrhages/ Exudates.

At gonioscopic examination, the angle was open at all quadrants in the right eye. In the left eye, there was appositional angle closure (>180 degrees). Inferior quadrant showed broad tent like Irido-Corneal adhesion as shown in figure 3.

Specular Microscopy

Specular microscopy of the left eye revealed reduced endothelial cell count with pleomorphism and polymegathism, multiple guttatea and dark cells with light central spot shown in figure 5.

Pachymetry CCT (Central corneal thickness):
528 microns RE, 543 microns LE
OCT Macula: Both eyes found to be normal
Fields charting: (RE) (LE)

After complete evaluation, diagnosis of ICE-S was made. Topical anti glaucoma medications-timolol and brimonidine were given. Even with topical and systemic medications IOP remained high and finally patient underwent trabeculectomy with mitomycin C (.01%) shown in figure 7.

Post Operatively

- BCVA- 6/6 (RE) with -0.5DS, 6/12 (LE) with -1.5DS/ -1.0DC/ 90°
- IOP: Well controlled 12-14 mm Hg.
- Filtering Bleb: Healthy, 100% diffusion area, moderately elevated, normal vascularity.

Fig 1. Showing findings on slit lamp examination (LE)

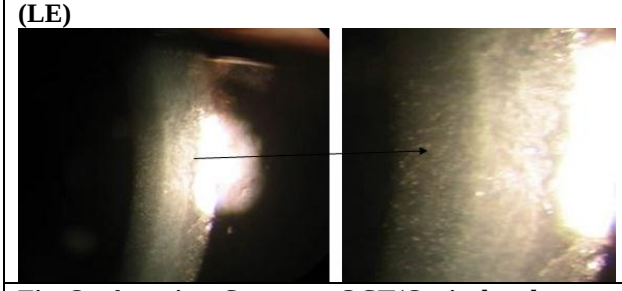


Fig 2. Showing findings on slit lamp examination

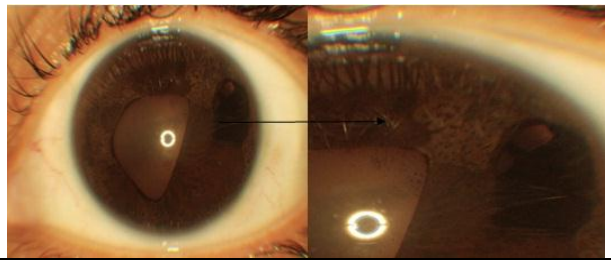


Fig 3. Anterior Segment OCT(Optical coherence tomography)

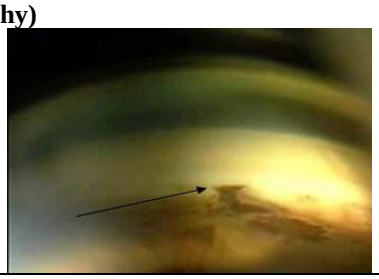


Fig 4. Showing PAS (Peripheral anterior synechiae) in LE on OCT examination

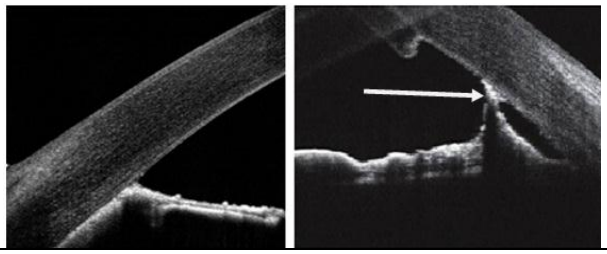


Fig 5. Showing findings on specular microscopy examination

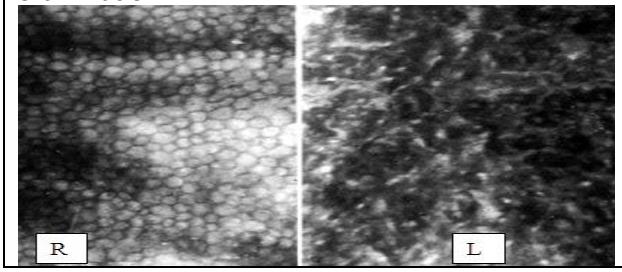


Fig 6. Showing glaucomatous findings on field charting

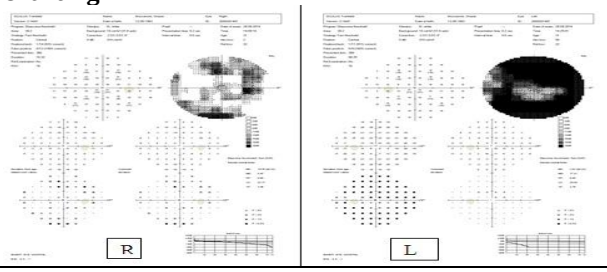
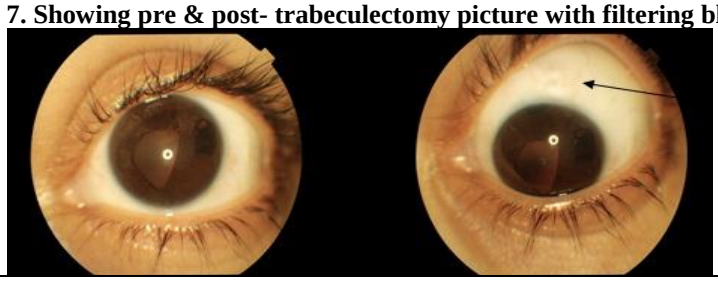


Fig 7. Showing pre & post- trabeculectomy picture with filtering bleb.



DISCUSSION

Iridocorneal endothelial syndrome is a congenital condition, usually unilateral, seen in middle aged females, not associated with systemic disease. Exact aetiology is unknown but viral (Herpes) aetiology is the most commonly accepted theory. Three main variants are progressive iris atrophy, Cogan Reese syndrome and Chandler's syndrome. Cogan Reese is the commonest type in Orientals [6]. All subtypes show aberrations of the cornea, anterior chamber angle and iris. The term "primary proliferative endothelial degeneration "emphasizes the pathogenic origin (Campbell Membrane Theory). Contraction of a sheet of ICE cells can distort the pupil, thin the iris and create holes in it. Subsequent contraction causes progressive synechial closure of theangle. Patients with essential iris atrophy and iris naevus syndrome typically have more significant glaucomatous optic atrophy, visual field loss and elevated IOP [2]. Confocal microscopy shows pleomorphic epithelioid cells with hyper-reflective nuclei alternating with uniform cells having darker nuclei. An abnormal endothelium gives a `fine hammered silver' appearance. The differentials to be considered in ICE-S include posterior polymorphous corneal dystrophy, Fuchs endothelial dystrophy, Axenfeld-Rieger anomaly and iridoschisis. Management of ICE-S depends on degree of corneal oedema and severity of secondary glaucoma. Filtration surgery is eventually required in most cases with variable success rate.

CONCLUSION

ICE syndrome is a progressive disorder rarely encountered in routine practice in middle aged patients who presented with blurred vision, ocular pain and corneal edema. Patient should be examined carefully for any associated iris and angle abnormalities. Specular microscopy is an important investigation which helps to reach the diagnosis of this rare syndrome. Management can be medical or surgical with variable success and failure rates. We presented this case for its rarity and management difficulties.

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CONFLICT OF INTEREST

No financial interest or any conflict of interest.

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