Case Report

Rare case of unilateral iridocorneal endothelial syndrome with secondary glaucoma

Vinit Rewanwar[1], B S Joshi[2*], V H Karambelkar[3], D K Sindal[4]

[1]Junior Resident, [2]Associate Professor, [3]Professor, [4]Professor and HOD, Department of Ophthalmology, Krishna Institute of Medical Sciences, Karad, Maharashtra, INDIA.
Email: v.rewanvar@gmail.com

Abstract

Iridocorneal endothelial syndrome (ICE) is a rare disease with a prevalence of less than one per two lakh population. These conditions are predominantly sporadic, almost always unilateral and generally arise in early adulthood, usually in women. Male to female ratio varies from 1:2 to 1:5. Although associated with distinctive clinical features, 68% cases are misdiagnosed initially. Prevalence of glaucoma associated with ICE ranges from 46-82% and all are accompanied by iridocorneal adhesions (peripheral anterior synechiea) This report discusses a case of 45 year old female of Right eye Iridocorneal Endothelial Syndrome with secondary glaucoma.

Key Words: unilateral iridocorneal endothelial syndrome.

[*]**Address for Correspondence:**
Dr. B S Joshi, Associate Professor, Department of Ophthalmology, Krishna Institute of Medical Sciences, Karad, Maharashtra, INDIA.
Email: v.rewanvar@gmail.com

Received Date: 24/04/2017 Revised Date: 11/05/2017 Accepted Date: 02/07/2017
DOI: https://doi.org/10.26611/1009317

Access this article online	
Quick Response Code:	Website: www.medpulse.in
	Accessed Date: 14 July 2017

INTRODUCTION

Iridocorneal endothelial syndrome (ICE) is a rare disease with a prevalence of less than one per two lakh population. These conditions are predominantly sporadic, almost always unilateral and generally arise in early adulthood, usually in women.[1] Male to female ratio varies from 1:2 to 1:5. Although associated with distinctive clinical features, 68% cases are misdiagnosed initially. Prevalence of glaucoma associated with ICE ranges from 46-82% and all are accompanied by iridocorneal adhesions (peripheral anterior synechiea)[2]. Following is a case report of Right eye Iridocorneal Endothelial Syndrome with secondary glaucoma. A 45 year old female presented with history of gradual painless diminution of vision for distance and near in right eye since one year. There was no history of trauma or redness. Patient was not a known case of any systemic illnesses. No similar complaints in the past in left eye. No history of intra-ocular surgery. At the time of presentation her best corrected visual acuity for distance was 6/24 OD and 6/6 OS and for near N8 OD and N6 OS. Anterior Segment Slit lamp examination of Right eye showed mild corneal oedema. Few hyperpigmented iris nodules were seen superonasally. On specular reflection, hammered silver appearance of corneal endothelium was seen. Iris showed full thickness atrophic patch infero-nasal to the pupil. Anterior segment of Left eye was normal.

Figure 1: Anterior segment of Right eye

How to cite this article: Vinit Rewanwar, B S Joshi, V H Karambelkar, D K Sindal. Rare case of unilateral iridocorneal endothelial syndrome with secondary glaucoma. *MedPulse International Journal of Ophthalmology.* July 2017; 3(1): 26-28.
https://www.medpulse.in/Ophthlmology/

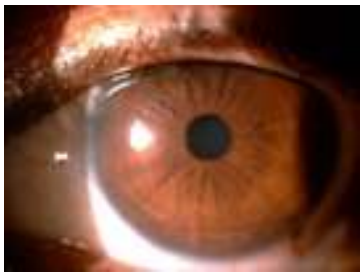

Figure 2: Anterior segment of Left eye

Intraocular pressure (IOP) with Goldmann Applanation Tonometer of Right eye was 40mmHg and Left eye 20mmHg. Fundus examination of Right eye showed Optic disc with Cup to Disc ratio of 0.5. Rest fundus was within normal limits. Left eye fundus was normal with Cup to Disc ratio of 0.3.

Figure 3: Fundus Photograph of Right eye

Gonioscopy with Four mirror gonioscope, showed peripheral anterior synechiae (PAS) in the right eye from 10- 5'o clock with Grade 2 angles according to Schaffer's grading, in all other quadrants. Left eye gonioscopy was normal. Visual field charting of both eyes was done on Octopus perimeter and did not show any significant visual field defects. She was given oral acetazolamide 250mgand started on topical combination of Brimonidine 0.2% and Timolol 0.5% 2 times a day. On follow up after one week, her right eye IOP reduced to 30mm Hg and visual acuity improved to 6/12. Patient was advised to undergo specular microscopy, Anterior segment OCT and surgery but unfortunately, she did not follow up. The clinical presentation and investigations led to the diagnosis of Right eye Iridocorneal endothelial syndrome with secondary glaucoma.

DISCUSSION

ICE is a spectrum of disease characterized by primary corneal endothelial abnormality. Typically a unilateral condition although sub clinical abnormalities may be seen in other eye[1]. Usually it is seen in middle aged adult and have a female predilection. There is no systemic or genetic association. Three main variants are Progressive Iris Atrophy, Cogan Reese syndrome and Chandler's. Cogan Reese is the commonest type in Orientals[7]. Clinically, the corneal endothelium has been described to have a "hammered silver" or "beaten bronze" appearance in ICE syndrome patients, similar to corneal guttae seen in Fuchs corneal endothelial dystrophy.[3,4] Pathologically, the normal endothelial cells are 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, tonofilaments, and microvilli.[5] The altered endothelium migrates posteriorly, moving beyond Schwalbe's line, onto the trabecular meshwork, and at times, onto peripheral iris. Contraction of this tissue within the angle and on the iris results in the peripheral anterior synechiae (PAS) 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.[3] As a result, patients may initially present with what appears to be open-angle glaucoma because the fibrovascular membrane obstructing aqueous flow can be difficult to visualize with gonioscopy. The corneal edema found in ICE syndrome patients is thought to be secondary to both elevated intraocular pressure (IOP) from secondary angle-closure glaucoma, and from subnormal pump function from the altered corneal endothelial cells.[6] Specular microscopy will show characteristic ICE cells. [Dark cells except for a light central spot and light peripheral zone]. Clear hexagonal margins will be lost. Associated features are pleomorphism and decreased cell count[1]. Corneal changes are predominantly seen in Chandler's. In Chandler's corneal oedema can persist even after controlling IOP. Iris changes are more in progressive iris atrophy. Iris atrophy is more on the side opposite to pupillary distortion. Corectopia is always towards a prominent PAS. There will be ectropion uvea and iris holes [stretch holes or melting holes]. In Cogan Reese pedunculated iris nodules are seen. These nodules are normal iris tissue pinched by contracting endothelial membrane. Aetiology is unknown. Viral [Herpes] aetiology is the most commonly accepted theory. Campbell and associates proposed a membrane theory according to which abnormality of corneal endothelium is the primary defect[1]. Primary aim of the treatment is to reduce corneal oedema and glaucoma. Control of corneal oedema can be achieved by lowering IOP and by using hypertonic saline and soft contact lens. Persistence of corneal oedema can lead to corneal decompensation and keratoplasty. Routine evaluation for glaucoma in these patients should be done by measuring intraocular pressure and evaluating the angle for PAS with gonioscopy.

Glaucoma in early stages can be controlled by aqueous suppressants. Miotics are not effective due to obstruction of trabecular meshwork. Long term medical management is usually ineffective[2]. When no longer controlled medically, surgery is indicated, of which better options are augmented trabeculectomy and shunt surgeries. Surgical procedures have variable success rate. Late failure can occur due to obstruction of fistula by synechiae or by endothelialization .Failure can occur due to inflammatory response also. Failure rate is high in young individuals. Trabeculectomy is the surgery of choice for ICE syndrome. Shields *et al*. have reported a 69% success rate in a study conducted in 1978 on 33 eyes [8], while Yanoff reported a 64% success rate 1 year postoperatively and a 36% at 3 years[9]. When the Trabeculectomy proves to be ineffective, the reason is usually excessive subconjunctival scarring[10] (a frequent occurrence in patients with ICE syndrome, given their young age). ICE-specific phenomena that lead to failure are bleb and/ or filtering ostium endothelialization[11] and PAS formation that obstruct the drainage pathway.

SUMMARY AND CONCLUSIONS

Though uncommon in routine practice, ICE syndrome has attracted much attention both for the enigma in its pathogenesis and challenges in its diagnosis and treatment. Failure rate of medical treatment is more than 70%. So it is better to intervene surgically as early as possible. But there is a high chance of failure even after surgery. We present this case for its rarity and management difficulties.

REFERENCES

1. Shield's textbook of Glaucoma, 4th edition, 226-236
2. Laganowsky H, Kerr-Muir, Hitchings KA. Glaucoma and Iridocorneal endothelial syndrome. Arch Ophthalmol. 1992; 110:346- 350.
3. Glaucoma. Basic and Clinical Science Course (BCSC). American Academy of Ophthalmology, 2010-2011; pp142-144.
4. Doan A, Alward W: Iridocorneal Endothelial Syndrome (ICE) - essential iris atrophy : 63 year-old female with PAS, "iris mass", corectopia, and increased IOP OS.February21,2005; Available from: http://www.EyeRounds.org/cases/case14.htm.
5. Levy SG, Kirkness CM, Moss J, Ficker L, McCartney AC. The histopathology of the iridocorneal-endothelial syndrome. Cornea. 1996 Jan; 15(1):46-54.
6. External Disease and Cornea. Basic and Clinical Science Course (BCSC). American Academy of Ophthalmology, 2010-2011; pp344-345.
7. Teekhasaenee C, Ritch R. Iridocorneal endothelial syndrome in Thai patients. Arch Ophthalmol. 2000; 18:187-192.
8. Bruce Shields M, Campbell DG, Simmons RJ. The Essential Iris Atrophies. Am. J. Ophthalmol. 1978; 85, 749–759.
9. Yanoff M, Duker JS. Ophthalmology. 2004
10. Hong B. Intracameral injection of tissue plasminogen activator to treat severe post-operative fibrinous reaction in iridocorneal endothelial syndrome. Digit. J. Ophthalmol. 2013; 90033.
11. Yanoff M, Scheie HG, Allman MI. Endothelialization of Filtering Bleb in Iris Nevus Syndrome. Arch. Ophthalmol. 1976; 94, 1933–1936.

Source of Support: None Declared
Conflict of Interest: None Declared