CASE REPORT

Iridocorneal Endothelial Syndrome

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ABSTRACT

A 19 years boy with a 2 years history of reduced and fluctuating vision along with change in pupillary shape and iris colour in his left eye presented to the glaucoma clinic. Ocular examination revealed distinct unilateral stretch holes, iris architecture changes and localized iris atrophy. Intraocular pressure was 16 mmHg in the right and 36 mmHg in the left eye. Gonioscopy of the left angle revealed broad based peripheral anterior synechiae at approximately 2 O'clock to 10 O'clock hours. The optic disc of the left eye had a vertical cup of 0.9 which manifest as superior and inferior arcuate scotoma. Specular microscopy showed unilateral abnormal endothelium with irregular cells of variable shape and size. To our knowledge, this is only the second reported case of iridocorneal endothelial syndrome in a male teenager.

Key Words: *Iridocorneal endothelial syndrome. Essential iris atrophy. Unilateral glaucoma.*

INTRODUCTION

Iridocorneal endothelial syndrome (ICE-S) is a peculiar ocular disorder. The term was proposed by Yanoff to describe three rare overlapping conditions - essential (progressive) iris atrophy, iris naevus (Cogan-Reese syndrome) and Chandler's syndrome.[1] Patient of ICE-S is typically a young adult female. It generally affects Caucasian and persons between 20 and 50 years of age. There are no recognized systemic or genetic associations.[2] Males have been known to develop ICE-S though uncommonly.[3] Even though associated with distinctive clinical features more than half of the cases are originally misdiagnosed.[4]

We are presenting this case because of its rarity, diagnostic intricacy and therapeutic challenge.

CASE REPORT

A 19 years healthy male presented with a 2 years history of gradually decreasing and fluctuating vision in his left eye. His father had noticed that his left iris had been changing shape and colour over the past one year. Furthermore, the patient complained of an increasing pain and photophobia in the preceding 4 months. There was no history of diplopia, haloes or trauma associated with his presenting symptoms. His past medical history was unremarkable.

On presentation, best corrected visual acuity was 6/6 in the right eye (OD) and 6/9 in the left eye (OS), [-1.50 DS/ - 2.50 x 70°]. External examination was normal for both eyes. Pupils were round, reactive to light and accommodating with no relative afferent pupillary defect. Slit lamp examination revealed quiet conjunctiva and clear corneas without posterior embryotoxon in both eyes. Both anterior chambers were deep and quiet. The left iris was notable for effacement of the iris architecture; stretch holes superiorly and temporally with patches of iris atrophy nasally (Figure 1a).

Intraocular pressures (IOP) by Goldmann applanation tonometry were 16 mmHg OD and 36 mmHg OS. On gonioscopy OD the angle was grade 3, with iris processes. The left angle had broad based peripheral anterior synechiae (PAS) extending upto and just anterior to the Schwalbe's line from approximately 2 O'clock to 10 O'clock.

Both lenses were clear with a normal vitreous cavity. Fundus examination revealed healthy macula and vessels with an intact retina in both eyes. The cup to disc ratio (CDR) OD was 0.6. The optic disc OS had a notable elongated vertical cup with a CDR of 0.9 (Figure 1b). Visual fields 30-2 showed a dense superior arcuate scotoma and an early inferior arcuate scotoma associated with temporal sparing OS. Fellow eye had normal visual fields.

Central corneal thickness was 568 mm OD and 498 mm OS. Endothelial cell counts were 2303 cells/mm^2 OD and 1038 cells/mm^2 OS. Specular microscopy showed unilateral polymegathism and dark cells with a lighter periphery OS (Figure 1c). Optical coherence tomography showed superior, nasal and inferior field depression on TSNIT graph and loss of double hump pattern OS (Figure 2). The average retinal nerve fibre layer thickness OS was reduced on OCT, 44.39 microns compared to 99.88 microns OD.

With these findings, a diagnosis of essential iris atrophy was made. The patient was initially prescribed topical pressure lowering drugs OS. Combination of dorzolamide 2% and 0.5% timolol eye drops twice daily, brimonidine eye drops thrice daily and latanoprost eye

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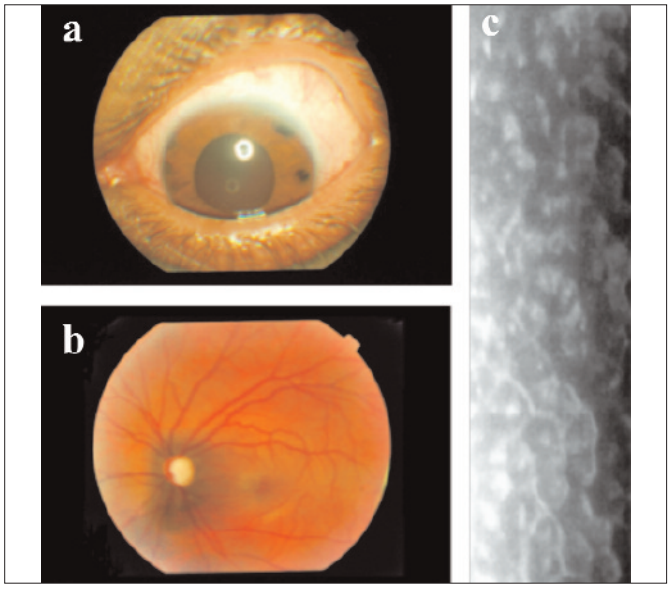

Figure 1 (a,b,c): (a) Stretch holes and iris atrophy. **(b)** Elongated vertical cup OS. **(c)** Specular Microscopy OS.

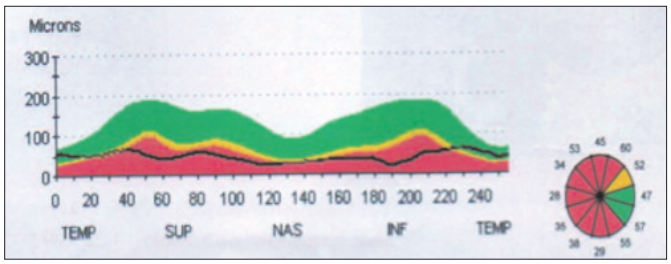

Figure 2: OCT showing loss of double hump pattern OS.

drops at night. IOP after 6 weeks of topical treatment was 26 mm of Hg.

Surgical intervention was decided and accordingly offered to the patient. Trabeculectomy with adjunctive 5-fluorouracil chemotherapy was eventually performed. Subsequently, IOP has been controlled in the low teens with last measurement of 14 mmHg on prednisolone eye drops thrice daily; with a well formed bleb.

DISCUSSION

The exact etiology of ICE-S is uncertain with researchers implicating Herpes simplex and Epstein Barr viruses. Role of a low grade chronic inflammation has also been suggested in the pathogenesis. All subtypes show aberrations of the cornea, anterior chamber angle and iris. The term "primary proliferative endothelial degeneration" emphasizes the pathogenic origin. It seems that the endothelial cells undergo a metaplastic transformation into “epithelial-like” cells that migrate in a membrane like form over the anterior chamber angle on to the iris. 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 the angle. The extent of PAS does not correlate with the degree of reduction in aqueous outflow. Raised IOP occurs because an unobserved retrocorneal ICE membrane covers the angle.

Confocal microscopy shows pleomorphic epithelioid cells with hyper-reflective nuclei alternating with uniform cells having darker nuclei. An abnormal endothelium gives an appearance of `fine hammered silver'. Abnormal epithelium along with ensuing corneal oedema causes visual reduction in ICE patients. Cystoid macular oedema can also cause visual deterioration. Ultrasound biomicroscopy is a valuable diagnostic aid to assess anterior segment and angle features especially when corneal oedema hinders view.

In essential iris atrophy there is progressive angle closure as PAS form. As the iris stroma atrophies, a full thickness iris hole forms opposite the PAS. This causes the pupil to be displaced towards the area of PAS. Iris naevus shows a diffuse solid mass covering the anterior iris, replacing the stroma locally. These nodules are normal iris tissue pinched by a contracting ICE membrane. Naevus close to the angle can cause closure with PAS. Chandler's syndrome is characterized by corneal oedema, monocular glaucoma and PAS.

Clinically, the condition is sporadic and unilateral, with subclinical irregularities of the corneal endothelium usually noted in the fellow eye. Bilateral cases have been documented.[5] Chandler's syndrome is the most common clinical variant. Glaucoma is found in approximately half of all cases of ICE-S. Patients with essential iris atrophy and iris naevus syndrome typically have more significant glaucomatous optic atrophy, visual field loss and elevated IOP.[2] Rarely, ICE-S may develop in children and teenagers.[6]

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 should be dictated by the degree of corneal oedema and severity of secondary glaucoma. Agents decreasing aqueous production are more effective than those increasing outflow or miotics. Laser trabeculoplasty and iriodotomy are ineffective for the same reason.

Filtration surgery is eventually required in most cases. The success rate of trabeculectomy in ICE-S is comparable to that for primary open-angle glaucoma (POAG). Failures have been attributed to progressive propagation of abnormal endothelium into the filtering bleb and to the young age of the patients. In these cases, another filtering procedure can be done at a different site. A YAG membranectomy can be attempted if failure occurs due to endothelialization of the fistula. Adjunctive antimetabolite use is highly recommended.[7]

Glaucoma drainage implants have slightly improved long-term surgical outcomes. However, most implants require revision due to tube blockage by iris, an ICE membrane or tube migration secondary to contraction of an ICE membrane. YAG membranectomy to the tube tip can be attempted in blockage. In pseudophakic eyes or eyes having high PAS the tube can be placed through the pars plana. Combined glaucoma surgery has shown promising potential.[8]

Patients with corneal oedema may benefit from lowering IOP even if it is within normal limits. However, cornea may remain oedematous even at the lowest attainable IOP so filtering surgery cannot be recommended exclusively for resolving oedema. Hypertonic saline drops and soft contact lenses are helpful in mild oedema. Iris defects can be corrected by reconstruction with multipiece endocapsular prosthesis. Femtosecond assisted keratopigmentation has been attempted for cosmesis and diplopia in an ICE-S patient with satisfactory results.[9]

If visually significant corneal oedema persists then keratoplasty is indicated. Provided intraocular pressure remains controlled, the prognosis of keratoplasty is primarily good. One report states that all cases of penetrating keratoplasty for ICE-S fail within 2 years.[10] A better surgical option is deep lamellar endothelial keratoplasty (DLEK). It provides patients rapid visual recuperation with minimal refractive changes. Additionally, replacement of the dysfunctional endothelium through descemet stripping with endothelial keratoplasty (DSEK) can successfully treat corneal oedema and associated visual loss.

Progression in ICE-S is variable and unpredictable. Due to its unilaterality, patients rarely become visually disabled. Medical therapy has a high failure rate so early surgical intervention is recommended. A meticulous anterior segment exam is indispensable in any patient presenting with monocular glaucoma.

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