

Case Report

Iridocorneal endothelial syndrome: iris naevus (Cogan-Reese) syndrome. A case report

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ABSTRACT

The purpose is to present a case of iridocorneal endothelium Syndrome with glaucoma and discuss clinical presentation and management strategies. A 45 year old female presented with redness, pain and diminution of vision in left eye. The patient was completely evaluated for the complaints. Slit lamp biomicroscopy revealed semidilated pupil not reacting to light and variation in iris colour pattern. Intraocular pressure was 14mmHg in the right eye and 46mmHg in the left eye. Gonioscopy of the left eye revealed broad based peripheral anterior synechiae. The optic disc of the left eye had a cup of 0.7. Specular microscopy of the left eye showed pleomorphism and polymegathism with multiple guttatae. This case reports the importance of specular microscopy in the evaluation of ICE syndrome and to plan the appropriate management strategies. We performed a trabeculectomy surgery for this patient with a well-functioning bleb to reduce the intraocular pressures following failure of topical anti-glaucoma medications.

Keywords: Iridocorneal endothelial syndrome, Iris naevus, Cogan Reese, Trabeculectomy, Mitomycin C

INTRODUCTION

The iridocorneal endothelial (ICE) syndrome is a rare disease with a prevalence of less than one per two lakh population.¹ The disease complex, which includes essential iris atrophy, Chandler's syndrome, and iris naevus (Cogan-Reese) syndrome. Common hallmarks 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, ectopion uveae, pigmented iris nodules and naevi of the anterior iris surface.² These pathogenetic mechanisms are due to the abnormal proliferation of the corneal endothelial cells.³ 68% of cases are misdiagnosed initially with 46-82% cases presenting with secondary glaucoma.^{1,4}

We are presenting this case because of its diagnostic and therapeutic challenges.

CASE REPORT

A 42 year old previously healthy woman presented with four years history of blurring of vision and pain in the left eye on and off. There was no history of coloured halos, photophobia or any ocular trauma. The patient was systemically stable with no history of known drug allergy. Family and personal history was not contributory. Congenital abnormalities were absent. Initially she was diagnosed with query traumatic mydriasis in the left eye and kept under observation. Then three years later she was diagnosed with angle closure glaucoma but failed to be diagnosed as ICE syndrome.

At the time of presentation the visual acuity was 6/6 in the right eye and 6/18 in the left eye with no improvement with a pin hole.

Examination

Slitlamp examination the right eye was normal. There was mild corneal edema and showed hammered silver appearance in the left eye. There were pigmented nodules on the anterior surface of the iris. The iris nodules were at the inferio-temporal quadrant, from the 2 to 7 o'clock position. They were located on the periphery of the hypopigmented iris, slightly elevated and heteroform (spindle) in appearance. The pupil was dilated to 5 mm not reacting to light. There was iris atrophy at the 8 o'clock position and there was ectropion uveae localized to this area (Figure 1). The intraocular pressure was 16 mmHg in the right eye and 46 mmHg in the left eye.

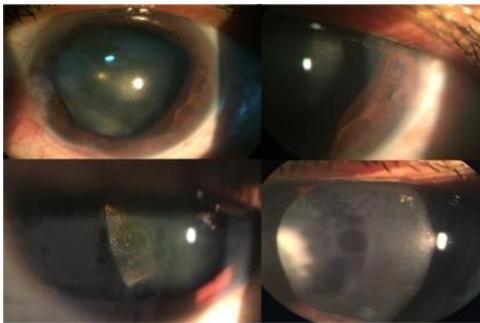


Figure 1: LE anterior segment showing hammered silver appearance and iris nodules.

At gonioscopic examination, the angle was open at all quadrants in the right eye. In the left eye, there was peripheral anterior synechiae in all quadrants, most prominent at 3 o'clock position (Figure 2). On fundus examination, the right eye was normal. In the left eye, there were glaucomatous changes of the optic disc with a cup/disc ratio of 0.7 in the horizontal and vertical meridians (Figure 3). Humphrey automated perimetry examination revealed normal findings in the right eye and concentric narrowing reaching to the central 15 in the left eye. Anterior segment OCT of the left eye showed peripheral anterior synechiae in all four quadrants. Specular microscopy of the left eye revealed reduced endothelial cell count with pleomorphism and polymegathism, multiple guttata and dark cells with light central spot (Figure 4).

The patient was given the maximum medical treatment (brimonidine tartrate twice daily, timolol maleate 0.5% twice daily, acetazolamide 250 mg thrice daily).

Trabeculectomy with mitomycin C has been performed for the patient as the intraocular pressure could not be controlled with maximum medical treatment. First post-operative follow up visit after 5 days, showed a well formed diffuse bleb with intraocular pressure of 24

mmHg. During the follow up visit in the second week the intraocular pressure was 18 mmHg. Subsequent visits the bleb was diffusely elevated and avascular (Figure 5). The visual acuity in the left eye was 6/18 after 1 month follow up with intraocular pressure less than 18 mmHg without any medication.

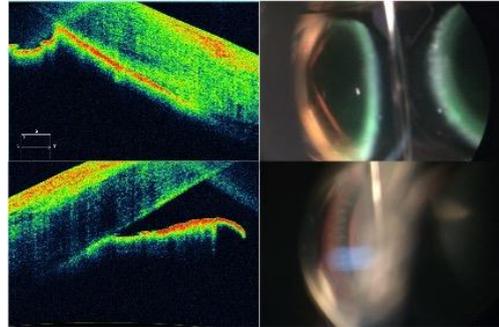


Figure 2: LE AS-OCT and gonioscopy showing peripheral anterior synechiae.

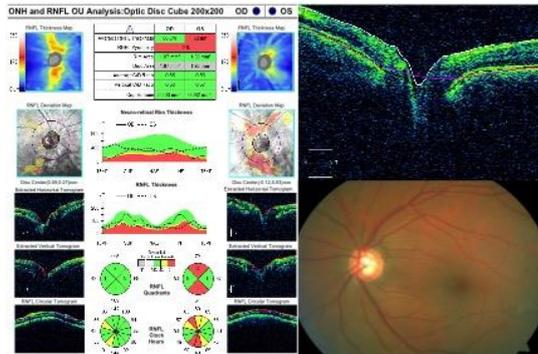


Figure 3: LE Posterior segment glaucomatous changes.

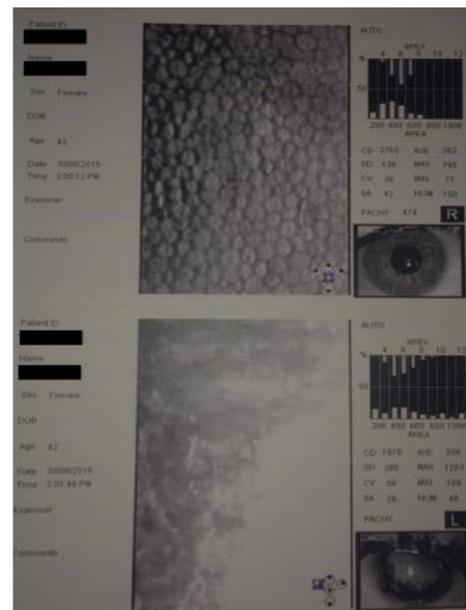


Figure 4: BE specular microscopy.

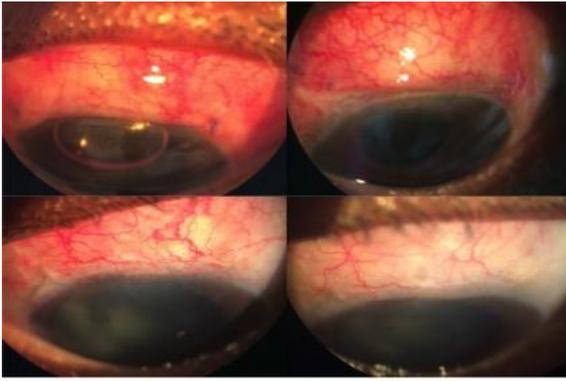


Figure 5: LE post-operative bleb formation.

DISCUSSION

Iridocorneal endothelial syndrome is congenital condition, usually unilateral, seen in middle aged females, not associated with systemic disease. Patient complains of blurred vision, ocular pain.⁵

ICE syndrome consists of 3 similar syndromes.

1. Cogan Reese syndrome - Iris nevus
2. Chandlers syndrome - Mild iris thinning, greater corneal edema.
3. Essential iris atrophy - Progressive thinning, holes, corectopia.

There is silver hammer beaten or plated silver appearance of the posterior corneal surfaces mainly the endothelium. Iris abnormalities vary in severity from mild stromal atrophy to full thickness iris hole.⁴ These changes with associated glaucoma are thought due to proliferation of the corneal endothelium and basal membrane. Histopathological examination revealed, the membrane over the trabecular meshwork and the iris.⁶

The differential diagnosis of ICE syndrome is important as there are many similar ocular pathologies. In a series of 25 cases with ICE syndrome, the correct diagnosis of ICE syndrome was made only in 8 (32%) of the cases at presentation. The false initial diagnoses were Posner Schlossman syndrome, chronic hypertensive uveitis, Fuchs' corneal endothelial dystrophy, Herpes simplex keratouveitis, acute and chronic angle closure glaucoma and Reiger's anomaly.¹

Specular microscopy examination reveals large endothelial cells, with a dark area and a bright central spot, that are located at the areas of the cornea that appear to be hammered silver. These cells are known as the ICE cells, pathognomic for ICE syndrome. This causes corneal edema which causes symptoms of reduced vision and pain. Corneal edema is more associated with

Chandler's syndrome but less with Cogan Reese syndrome and progressive iris atrophy.⁷

Glaucoma was reported in more than 50% of the cases of ICE Syndrome.⁴ Glaucoma was thought to occur as a result of the obstruction of the anterior chamber angle by an abnormal membrane composed of endothelium-like cells, a descemet-like membrane and the peripheral anterior synechia. It was recommended to search for ICE syndrome in young cases with unilateral glaucoma.⁴

The etiological factor for ICE syndrome is unknown. Viral [Herpes] etiology is a commonly accepted theory. Viruses cause cell necrosis and transformation of the endothelial cells. The ICE cells are thought to secrete corneal endothelial modulation factor and an abnormal matrix.^{8,9}

During the initial phases of the ICE syndrome, the glaucoma can be managed by medical treatment. However, in the later phases of the disease, surgical intervention is required in almost all cases. The improvement of glaucoma by conventional trabeculectomy in these cases is poor. Kidd et al. reported the success rates for the 1st, 2nd and the 3rd trabeculectomy operations in a series of 42 cases with ICE syndrome as 64%, 79% and 63%, respectively.¹⁰ Failure of first trabeculectomy surgery occur due to subconjunctival fibrosis and aggressive inflammatory reaction with development of the scar tissue, might be the cause of the insufficiency of the bleb.⁴

We performed trabeculectomy with mitomycin C in our case as the intraocular pressure did not improve with maximum medical treatment. During the post-operative follow up period the intraocular pressure has been well controlled.

CONCLUSION

ICE syndrome is a rare progressive disorder in unilateral middle aged adults presenting with blurred vision, ocular pain and corneal edema which should be examined carefully for any associated iris and angle abnormalities. Specular microscopy helps to clinch the diagnosis of ICE syndrome. In our case, because the diagnosis of ICE syndrome was missed patient had irreversible loss of vision.

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