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Case Report

Rare case of iridocorneal endothelial syndrome with secondary glaucoma

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ABSTRACT

A rare case of 40 years old female, came with complaints of pain and blurring of vision in the left eye for the past week. The patient had a diminution of vision since birth in her left eye. On examination left eye vision was found to be 6/60 not improving with pinhole, also defective color vision 20/25. The right-eye vision was found to be normal. The left eye showed significant corneal edema, and moderate anterior chamber depth, with iris features of Corectopia, Pseudopolycoria, and iris atrophy. Pressures were found to be 30mm hg in the left eye, gonioscopy revealed high peripheral anterior synechiae extending anteriorly above Schwalbe's line. Left eye fundoscopy was done and it showed increased CDR of 0.8 to 0.9 following which visual field analysis was done, it showed inferior field defect. The patient was started on e/d timolol and hypertonic saline for a week. The patient was reviewed after a week showing reduced pressures of 22mmg in the left eye with reduced corneal edema and pain. Diagnosis of iridocorneal endothelial syndrome-essential iris atrophy type with secondary glaucoma was made and now the patient is being followed up regularly. An iridocorneal endothelial syndrome is a group of disorders with three clinical variants as Chandler syndrome, Essential iris atrophy, and Cogan Reese syndrome. Pathology is the presence of abnormal endothelial cells, which behave as epithelial cells, able to proliferate and migrate. The iridocorneal endothelial syndrome is frequently associated with secondary glaucoma.

1. Introduction

An iridocorneal endothelial syndrome is a rare group of disorder with three clinical variants as

1. Chandler syndrome.
2. Essential/progressive iris atrophy.
3. Cogan reese syndrome.

Viral aetiology has been postulated following the presence of lymphocytes in the corneal endothelium suggesting chronic inflammation. The most common virus involved is herpes simplex and Epstein Barr virus. [1]

Pathology is the presence of abnormal endothelial cells, which behave as epithelial cells, able to proliferate and migrate. [2]

This phenomenon leads to variable degrees of iris atrophy, corneal edema, and secondary angle closure. [1]

2. Case Report

A 40-year-old female came with chief complaints of pain and blurring of vision in her left eye, for the past one week, she also had a diminution of vision present in her left eye since birth.

The patient had no history of any previous ocular treatment. The patient did not seek any specific ocular treatment for defective vision since childhood due to a lack of awareness and education. The low socioeconomic status and financial instability also refrained the patient from seeking any medical advice.

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No associated headache, diplopia, or floaters, and No history of trauma to the eyes.

There is no similar history in the family and there is no history of any consanguinity in the family.

The patient has no known co-morbidities.

Her general examination and vitals were found to be normal.

2.1. Ocular examination

2.1.1. Right eye

Ocular examination of the right eye was found to be normal with visual acuity of 6/6 and normal color vision of 25/25.

Extraocular movements were found to be normal. The right eye anterior segment was found to be normal with normal pupillary size and reaction.

Right eye fundoscopy was found to be normal.

The pressure in the right eye was 21 mmHg.

Indirect gonioscopy done with Zeiss 4 Mirror lens using indentation method showed normal angle structures in the right eye.

2.1.2. Left eye

Left eye ocular examination revealed visual acuity of 6/60 not improving with pinhole and the defective color vision of 20/25.

The extraocular movement was found to be normal.

The cornea showed mild to moderate corneal edema involving the entire cornea.

The anterior chamber was found to be of moderate depth noted as grade II in Van Herrick Grading.

Iris atrophy was noted along with abnormal shape due to hole formations.

The left eye pupil was found to have Corectopia, which is an abnormal pupil position along with Pseudopolycoria which is the false appearance of more than one pupillary opening of the iris. Also, the pupil was sluggishly reacting to light.

Left eye lens examination was found to be normal.

Left eye fundoscopy revealed hazy media and cup disc ratio of 0.8 to 0.9 with superior neuroretinal rim thinning.

Left eye intraocular pressure was found to be 30mmHg in applanation tonometry.[3]

Indirect gonioscopy done with Zeiss 4 mirror lens using indentation method showed high peripheral anterior synechiae extending above the Schwalbe's line in the left eye.

3. Discussion

The diagnosis of Irido corneal endothelial syndrome – Essential Iris atrophy with secondary glaucoma was done based on the anterior segment, tonometry, and gonioscopy findings.[4]

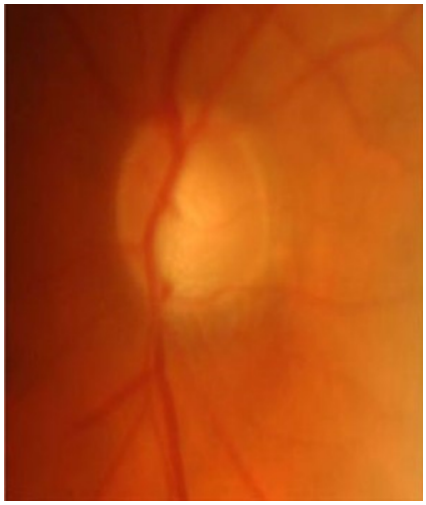

Fig. 1: Left eye disc – CDR 0.8 to 0.9 cupping with superior NRR thinning

Fig. 2: Left eye – Corneal haze due to corneal edema

An important feature noted in the case of essential iris atrophy is the atrophied iris. Iris atrophy occurs due to the contraction of the endothelial cells in the iris resulting in distortion of the iris.[2] Pseudopolycoria is a hallmark finding in the case of essential iris atrophy, which is a hole formation commonly noted as a false pupil. Corneal edema was initially present which can occur secondary to either abnormal endothelium or elevated intra-ocular pressures.[1] Low vision in the left eye since birth was due to corneal edema and elevated pressures, which were not evaluated. In this case, there was the clearing of corneal edema and

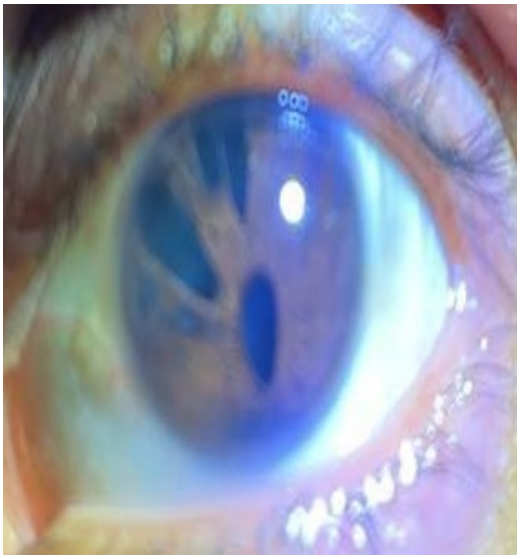

Fig. 3: Left eye – IRIS atropy with pseudopolycoria and corectopia

Fig. 4: Left eye gonioscopy – High peripheral anterior synechiae extending above schwalbes line

improvement of vision to 6/12 in the left eye after medical management of glaucoma.[4] Tonometry evaluation showed an elevated IOP of 30mmhg in the left eye after management IOP was reduced to 22mmhg in the left eye.

Gonioscopy revealed – high peripheral anterior synechiae reaching above Schwalbe's line.[5] Fundus examination of the left eye shows CDR -0.8-0.9 cupping with superior NRR thinning. Visual field analysis using HFA 30-2– left eye shows an Inferior field defect.

The abnormality of the endothelium is the primary defect in ICE Syndrome. It leads to corneal edema and proliferation of cellular membrane across the angle of the anterior chamber, associated glaucoma occurs following a secondary angle closure from PAS formation.[1,4]

1. The abnormal corneal endothelium migrates posterior to the Schwalbe's line, onto the trabecular meshwork and iris.[1] PAS formation occurs while these migrated endothelium contract along with collagenous fibrous tissue, Further contraction of these membranes causes progressive angle closure.
2. Electron microscopy has shown, that endothelium varies in thickness, has multiple layers like epithelium, and contains filopodial processes for movement.[6]
3. ICE syndrome is mostly unilateral, occurring in young adult females, Patients present with elevated IOP, decreased vision, and pain (secondary glaucoma).[4]

A varying degree of iris atrophy and corneal involvement helps to narrow down the specific entity of ICE Syndrome. However apparent transition from one form to another has been reported.

In all three variants, there is abnormal endothelium is seen as a beaten bronze appearance, Corneal diameter has been found to be normal. Corneal edema due to endothelium abnormality even with normal IOP also High PAS (peripheral anterior synechiae) is characteristic in ICE syndrome and almost always extends anteriorly to the Schwalbe line.[3] Iris atrophy is present in varying degrees in different entities of ICE. (1) Glaucoma occurs in 50% of cases of ICE Syndrome.

Among the three entities of ICE syndrome, CHANDLER syndrome is the most common.

Iris atrophy and correctopia are minimal but corneal and angle abnormalities are more significant.

In the case of Cogan Reese syndrome, there are diffuse pigmented lesions or pedunculated nodules but iris atrophy is minimal.

In essential iris atrophy cases, there is severe progressive iris atrophy with hole formation, Corectopia is significantly present in cases of essential iris atrophy. Pseudopolycoria is almost always present. High PAS is commonly present – leading to significant glaucoma.

3.1. Management

The patient was started on a topical anti-glaucoma drug, most commonly an aqueous suppressant beta blocker – Timolol eye drops BD for a week the patient was also started on topical hypertonic saline solution 4 times a day for a week to reduce corneal edema.

The patient was followed up after a week, ocular examination showed decreased intra-ocular pressure of 22mmHg along with clearing of corneal edema

The patient was feeling symptomatically better.

The patient is now being followed up in ophthalmology opd regularly which is monthly once and advised to continue anti-glaucoma medications.

Gonioscopy is a must to identify angular synechiae and identify angle status.[5]

Diagnosis is confirmed with specular or confocal microscopy - showing abnormal endothelial cells as pseudo epithelial cells.

Medical management is mostly ineffective in advanced cases, however, anti-glaucoma medications, usually aqueous suppressants, are given.

If medical management fails, then glaucoma filtration surgery such as trabeculectomy with antifibrotic agents is indicated.[6] However, late failures can occur due to endothelization of the fistula by abnormal corneal endothelial cells causing obstruction. In case of late failures occurring due to endothelization of the fistula, the fistula can be reopened using an ND-YAG laser procedure. The last option will be glaucoma tube shunt procedures. Cases of irreversible corneal edema require penetrating keratoplasty once glaucoma is controlled.

4. Conclusion

Irido corneal endothelial syndrome is frequently associated with secondary glaucoma hence the high level of glaucoma suspicion should be kept in all ICE cases since it can lead to secondary angle closure glaucoma.[1]

Always the specific type of syndrome needs to be identified.

Patient needs to be evaluated regularly even though the patient is asymptomatic.

The patient needs to be aware of the possible complications associated. The long-term prognosis is usually poor.

5. Source of Funding

None.

6. Conflict of Interest

None.

References

1. Levy SG, Kirkness CM, Moss J, Ficker L, Mccartney AC. The histopathology of the iridocorneal-endothelial syndrome. *Cornea*. 1996;15(1):46–54.
2. Alvarado JA, Murphy CG, Maglio M, Hetherington J. Pathogenesis of Chandler's syndrome, essential iris atrophy, and the Cogan-Reese syndrome. I. Alterations of the corneal endothelium. *Invest Ophthalmol Vis Sci*. 1986;27(6):853–72.
3. Shields MB, Campbell DG, Simmons RJ. The essential iris atrophies. *Am J Ophthalmol*. 1978;85(6):749–59.
4. Laganowski HC, Muir MGK, Hitchings RA. Glaucoma and the iridocorneal endothelial syndrome. *Arch Ophthalmol*. 1992;110(3):346–50.
5. Silva L, Najafi A, Suwan Y, Teekhasaenee C, Ritch R. The iridocorneal endothelial syndrome. *Surv Ophthalmol*. 2018;63(5):665–76.
6. Doe EA, Budenz DL, Gedde SJ, Imami NR. Long-term surgical outcomes of patients with glaucoma secondary to the iridocorneal endothelial syndrome. *Ophthalmology*. 2001;108(10):1789–95.

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