

Iridocorneal endothelial syndrome: Cogan -Reese syndrome. A case report

Kumar K¹, Ingle R²

¹Dr Kavita Kumar, Associate professor, Department of Ophthalmology, ² Dr Rashmi Ingle, PG Resident. Both affiliated with Regional Institute of Ophthalmology Gandhi Medical College, Bhopal, MP, India

Address for Correspondence: Dr Kavita Kumar, Email: kavita.kumar43@gmail.com

Abstract

ICE syndrome is a rare progressive unilateral disease occurring in young middle aged females. Corneal edema with uveal involvement is key feature. Secondary glaucoma is main sight threatening complication. Management depends on severity of ocular findings and is targeted to control corneal edema and secondary glaucoma medically and surgically.

Introduction

ICE (iridocorneal endothelial) syndrome is congenital condition, usually unilateral, seen in middle aged females, not associated with systemic disease¹. Patient complains of blurred vision ocular pain.

Signs include varying amount of corneal oedema, iris heterochromia and distorted pupil¹. ICE syndrome consists of 3 similar syndromes: **Cogan Reese Syndrome**-Iris Nevus, **Chandlers syndrome** -mild iris thinning, greater corneal edema, **Essential Iris atrophy**-progressive thinning, holes, corectopia¹.

Case report

We report an 18 year old patient presented to our clinic with blurred vision and pain persisting for 1 month in the left eye.

Patient was apparently all right 1 month back when she had episode of pain redness followed by progressive diminution of vision and whitening of cornea. Family and personal history were not contributory.

No history of trauma or any major systemic illness like tuberculosis, leprosy was available. Stigmata of neurofibromas /Stills disease / other congenital anomalies were absent.

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Examination-

Vision in right eye was normal. Slit lamp examination of right eye was normal. Left eyeball was prominent (exophthalmos) and exophoria with moderate ptosis. Vision was decreased to perception of light with defective projection in superior and nasal quadrant.

Left eye had following additional findings:

1. Unilateral Severe Corneal edema.
2. Iris Nevus was present on the anterior surface of iris, from 12 to 4 O'clock position extending up to pupillary margin.
3. Ectropion uveae.matted appearance of iris stroma, loss of iris crypts in temporal quadrant.
4. Left eye unilateral secondary glaucoma (IOP 42 mm of Hg schiotz).

On investigation by Fundus and gonioscopy Right Eye was normal and could not be performed on LE due to severe corneal odema.

B scan of RE was normal and LE showed Macrophthalmia with axial length 25 mm. Other investigations like X-ray skull, bilateral knee, USG abdomen, paediatric, cardiology and orthopaedic examinations were within normal limits.

All these findings were in favor of *Iris nevus (Cogan-Reese) syndrome*.



Fig1: Left eye showing exophthalmus, exophoria & ptosis

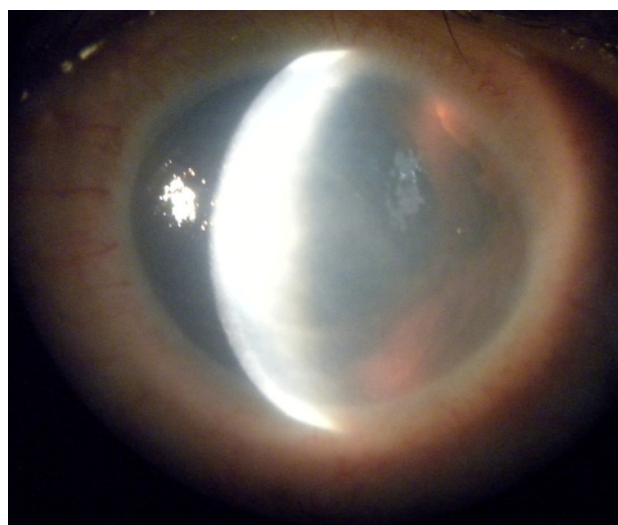


Fig 2: Severe corneal edema with Iris nevus on anterior surface of eyes from 12 to 4 O'clock position.

In our patient medical management was done with local and systemic anti glaucoma drugs and intraocular pressure came down to 14.6 mm of Hg in left eye after 11 days with symptomatic relief. Surgical intervention for glaucoma and corneal edema was not considered owing to poor visual prognosis. Our patient is being followed up for secondary glaucoma.

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, corneal edema, secondary glaucoma, heterochromia and distorted pupil¹ ICE syndrome consists of 3 similar syndromes².

1. Cogan Reese Syndrome- Iris Nevus
2. Chandlers syndrome -mild iris thinning, greater corneal edema.
3. Essential Iris atrophy-progressive thinning, holes, corectopia.

Etiology is unknown. Epstein Barr virus and Herpes simplex viruses have been found serologically in patients of ICE syndrome³. These viruses are suggested to play a

role in cell necrosis and in transformation of endothelial cells. In ICE syndrome the corneal endothelium undergoes an epithelioid metaplasia that migrates in a membrane form². If it migrates over the anterior chamber angle, contraction of this membrane pulls the iris toward the cornea causing a synechial closure of the angle leading to secondary glaucoma⁴. When the abnormal corneal endothelium spreads on to the iris surface, the contraction causes atrophy and corectopia. Pigmented iris nodules are also produced by this contracting abnormal endothelial membrane. Corneal edema occurs due to the subnormal endothelial pump function.

Cogan -Reese syndrome named after David Glendenning Cogan and Aligernon Reese, is an extremely rare eye

disorder seen in young and middle aged females, usually affecting one eye, insidious in onset, characterized by a matted or smudged appearance of the surface of iris, development of modular iris nevi, ectropion uveae, peripheral anterior synechiae, and/or glaucoma. Secondary glaucoma may lead to vision loss.

This disorder most frequently appears in young and middle-aged females, usually affecting one eye (unilateral) and developing slowly over time¹. The differential diagnosis of multiple pigmented iris nodules include Neurofibromatosis, melanomas, inflammatory nodules of leprosy, syphilis, tuberculosis and sarcoidosis⁵

Chandlers syndrome -This variant shows minimum or no iris atrophy, mild corectopia may be present. The corneal edema may be more pronounced, giving pleated silver appearance of posterior corneal surface.

Essential Iris Atrophy-This variant is characterized by severe iris atrophy resulting in heterochromia, marked corectopia, ectropion uveae, pseudopolycoria & iris hole formation are hallmark of essential iris atrophy³

Treatment- Main concern of management is secondary glaucoma which occurs in about 50% of ICE syndrome patients and is major cause of visual loss⁶. Management options are specific to each case and should be tailored to degree and severity of secondary glaucoma and corneal edema.

Conclusion

ICE is a rare, progressive, congenital, unilateral disease, of unknown etiology occurring in middle aged females.

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Case Report

Main clinical feature is ocular pain, blurred vision. Secondary glaucoma is a sight threatening complication. Management is targeted to reduce corneal edema, and control secondary glaucoma medically. In severe cases surgical correction of glaucoma and penetrating keratoplasty is required to achieve and maintain good vision.

References

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