Case Report

A rare case of Iridocorneal Endothelial Syndrome: Case Report

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Abstract

Iridocorneal endothelial (ICE) syndrome includes a group of rare ocular pathologies with unilateral involvement, frequently affecting young women. The disease complex includes essential iris atrophy, Chandler's syndrome, and Cogan–Reese syndrome. We herein report a 46 years old female who presented with diminution of vision in Left Eye since 2 months. Based on characteristic appearance on anterior segment finding, a clinical diagnosis of Iridocorneal Endothelial (ICE) Syndrome with secondary glaucoma was made. Medical management using combination of anti-glaucoma was given to the patient. Trabeculectomy was advised as a long term management for better quality of life to avoid secondary glaucomatous optic atrophy.

Keywords: Essential iris atrophy(ICE), glaucoma, trabeculectomy

Introduction

Iridocorneal Endothelial Syndrome (ICE) syndrome is a rare ophthalmic disorder with a prevalence of less than one per two lakh population, in which there occurs an abnormal corneal endothelium that leads to varying degrees of corneal edema, atrophy of iris and secondary angle-closure glaucoma.[1] This ailment, typically affects young women unilaterally with no family history.[2] It comprises of three clinical variants: Chandler Syndrome, Essential (Progressive) Iris Atrophy and Cogan-Reese Syndrome (Iris Nevus Syndrome). It is considered to be an acquired disorder probably due to Herpes simplex virus and Epstein–Barr virus etiology. [2,8]

The ICE cell is pathognomonic of this syndrome. These corneal endothelial cells are abnormally large and show increased pleomorphism and show epithelial-like characteristics. The abnormal endothelial cells have the tendency to migrate posteriorly thus forming a membrane covering the adjacent structures, iris and trabecular meshwork. This membrane when contracted leads to characteristic changes of iris, iridotrabecular synechiae and corectopia.

Case

A 46 years old female presented with diminution of vision in LE since 2 months. Patient's BCVA (best corrected visual acuity) was 6/6 (RE) 6/36 (LE). NV N18 (RE) & <N36 (LE). On slit lamp examination RE appeared normal whereas LE showed microcytic corneal edema, asymmetrical shallow AC with VH grade I-II, essential iris atrophy with corectopia [Figure 1], ectropion uveae & peripheral anterior synechae (PAS). Gonioscopy finding showed PAS extending anterior to schwalbe's line. The intraocular pressure by Goldmann applanation tonometry were RE 14mmHg and LE 26mmHg at 10 am in the morning.

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Fundus examination showed LE clear media with cup-disc ratio 0.9 with nasalisation of vessels and peri-papillary atrophy [Figure 2].

Anterior segment, gonioscopy and fundus examination of RE were within normal limits.

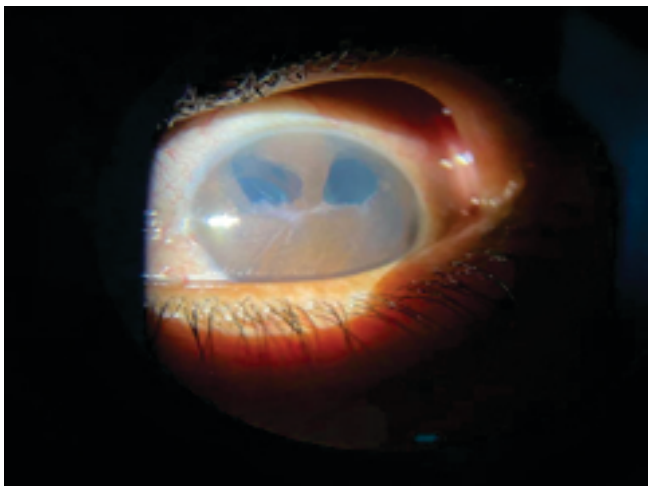

Figure 1: Slit lamp examination showing essential iris atrophy with corectopia

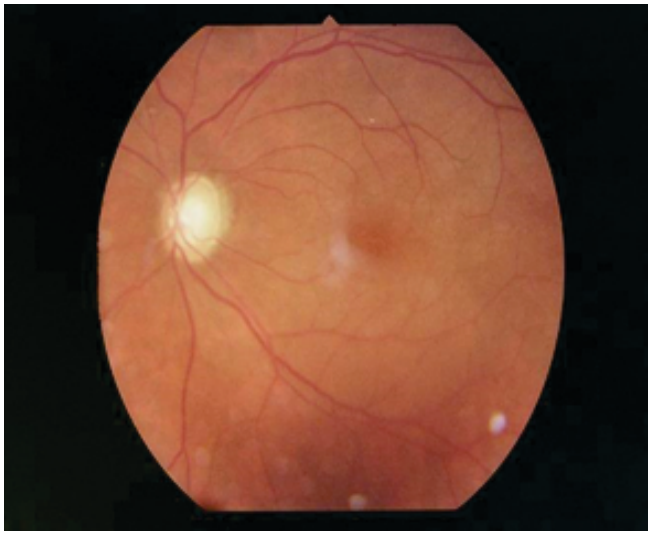

Figure 2 : Fundus examination of LE showing large cup-disc ratio and nasalisation of vessels

Serology was negative for Herpes simplex virus.

A final diagnosis of Esssential iridocorneal endothelial syndrome with secondary glaucoma of Right Eye was made based on the epidemiological data and clinical examination findings.

The patient was given medical therapy with 3 topical antiglaucoma medications (Latanoprost 0.005% , Brimonidine 0.15% & Timolol 0.5%). Surgical intervention i.e. trabeculectomy with peripheral iridectomy was advised since IOP remained uncontrolled in spite of maximal medical management to preserve vision for better quality of life and prevent from further damage to optic nerve due to glaucomatous optic atrophy.

Discussion

ICE syndrome is characterized by proliferation and structural abnormalities of the corneal endothelium, progressive obstruction of the iridocorneal angle and abnormalities of iris such as iris atrophy and iris hole formation[3]. The sequelae of these changes are decompensation of cornea and glaucoma, which represent the most common causes of vision loss in patients with ICE syndrome .[4]

Posterior polymorphous dystrophy, iris hypoplasia, and Axenfeld-Rieger syndrome are a few of the commonly considered differential diagnoses. These can be differentiated as follows:

Posterior polymorphous dystrophy

It is corneal dystrophy, with autosomal dominant inheritance. Three patterns which are observed are endothelial vesicle-like lesions, band lesions, and diffuse deep stromal opacities. It rarely results in corneal edema or elevated IOP.

Iris hypoplasia

It is easily distinguished from ICE syndrome by the typical finding of a hypoplastic iris. It may appear like aniridia, but rudimentary iris often revealed on gonioscopy.

Axenfeld - Rieger syndrome

This is again an autosomal dominant inherited condition with similar findings to that of ICE syndrome but presents with glaucoma in early infancy or childhood.

Trabeculectomy with antifibrotic agents is the surgery of choice for ICE syndrome. Shields et al. have reported a 69% success rate in a study on 33 eyes[5] while Yanoff and Duker reported a 64% of success rate 1-year postoperatively and a 36% at 3 years. [6]

Aqueous shunt surgery is considered by few as a popular first-line surgical intervention for glaucoma associated with ICE given the intractable nature of the disease. In pseudophakic eyes, the tube can be placed in the ciliary sulcus or pars plana. Kim DK et.al reported success rates with drainage device surgery to be about 70% at 1 year and 53% at 5 years.[7]

Conclusion

Medical therapy is usually given with 3 topical antiglaucoma medications and one systemic AGM. Surgery is recommended in cases of uncontrolled IOP in spite of maximal medical therapy. Trabeculectomy with antifibrotic agents is the surgery of choice for ICE syndrome. Corneal decompensation can likewise, be treated with surgery when medical management fails. Penetrating keratoplasty (PKP) or endothelial keratoplasty (such as DSEK or DSAEK) can be performed to replace the abnormal corneal endothelial cells and thereby improving corneal function.

A long-term follow-up is mandatory because of the progressive nature of the disease itself. Studies suggest follow-up at 2-3 months intervals when glaucoma is associated and depending on its severity.

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