Iridocorneal Endothelial Syndrome: A Case Report

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Introduction

Iridocorneal endothelial syndrome is a rare disease with a prevalence of less than one per two lakh population. Although associated with distinctive clinical features, 68% cases are misdiagnosed initially. Prevalence of glaucoma associated with ICE ranges from 46-82%. We are presenting this rare cause of glaucoma because of its diagnostic and therapeutic challenges.

Case Report

A 45 year old previously healthy woman presented with one month history of blurring of vision and pain in right eye. There was no history of coloured haloes, photophobia or trauma. At the time of presentation her best corrected visual acuity was 3m counting figures OD and 6/6 OS.

Slit lamp examination showed corneal oedema with aqueous flare. Intraocular pressure of right eye was 45mm Hg. She was started on oral and topical antiglaucoma medications. Her IOP got reduced to 25mm Hg and visual acuity improved to 6/9. When corneal oedema was reduced, slit lamp examination showed hammered silver appearance of corneal endothelium, few black iris nodules superonasally and corectopia towards 10'clock (Fig. 1, 2&3).

Fundus examination showed cd ratio of 0.7OD & 0.4 OS. Field examination showed superior arcuate scotoma OD (Fig.4). Gonioscopy showed peripheral anterior synechiae from 10-11'o clock with grade 2 angles in all other quadrants. Left eye was normal (Fig.5). Specular microscopy showed decreased cell count, polymegathism and dark cells with light central spot and light peripheral zone. Margins of the cells were not clear. These were suggestive of ICE (Fig.6&7). OCT showed superior field depression in TSNIT graph and loss of double hump pattern right eye (Fig.8&9). With the clinical presentation and investigation, we came to a diagnosis of iridocorneal endothelial syndrome [Cogan Reese type].

Topical anti glaucoma medications- timolol and brimonidine were given. Even with topical and systemic medications IOP remained high. So augmented trabeculectomy was done. IOP returned to normal and vision was 6/9.

Discussion

ICE is a spectrum of disease characterized by primary corneal endothelial abnormality. Typically a unilateral condition although sub clinical abnormalities may be seen in other eye. Usually it is seen in middle aged adult and have a female predilection. There is no systemic or genetic association. Three main variants are progressive iris atrophy, Cogan Reese syndrome and Chandler's. Cogan Reese is the commonest type in Orientals.

Corneal endothelial changes are typical having a hammered silver appearance. Specular microscopy will show characteristic ICE cells. [Dark cells except for a light central spot and light peripheral zone]. Clear hexagonal margins will be lost. Associated features are pleomorphism and decreased cell count. Corneal changes are predominantly seen in Chandler’s. In Chandler’s corneal oedema can persist even after controlling IOP.

Iris changes are more in progressive iris atrophy. Iris atrophy is more on the side opposite to pupillary distortion. Corectopia is always towards a prominent PAS. There will be ectropion uvea and iris holes [stretch holes or melting holes]. In Cogan Reese pedunculated iris nodules are seen. These nodules are normal iris tissue pinched by contracting endothelial membrane.

Glaucoma and corneal decompensation are serious sequelae of ICE. Glaucama can be due to extensive PAS or obstruction of anterior chamber angle by membrane. Prevalence of glaucoma is 46-82%. Younger patients are more affected.

Aetiology is unknown. Viral [Herpes] aetiology is the most commonly accepted theory. Campbell and associates proposed a membrane theory according to which abnormality of corneal endothelium is the primary defect.

Primary aim of the treatment is to reduce corneal oedema and glaucoma. Control of corneal oedema can be achieved by lowering IOP and by using hypertonic saline and soft contact lens. Persistence of corneal oedema can lead to corneal decompensation and keratoplasty.

Glaucoma in early stages can be controlled by aqueous suppressants. Miotics are not effective due to obstruction of trabecular meshwork. Long term medical management is usually ineffective. When no longer controlled medically, surgery is indicated, of which better options are augmented trabeculectomy and shunt surgeries. Surgical procedures have variable success rate. Late failure can occur due to obstruction of fistula by synechiae or by endothelialization. Failure can occur due to inflammatory response also. Failure rate is high in young individuals.
Summary

Though uncommon in routine practice, ICE syndrome has attracted much attention both for the enigma in its pathogenesis and challenges in its diagnosis and treatment. Failure rate of medical treatment is more than 70%. So it is better to intervene surgically as early as possible. But there is a high chance of failure even after surgery. We presented this case for its rarity and management difficulties.

References