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A CASE STUDY OF ICE (IRIDOCORNEAL ENDOTHELIAL SYNDROME)

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Abstract : Irido corneal endothelial syndrome is typically unilateral, affecting middle aged women. It is due to the proliferation of abnormal corneal endothelial cell layer across the iris and angle, producing angle closure glaucoma. ICE syndrome consists of three very rare frequently overlapping disorders 1.progressive iris atrophy,2.Cogan-Reese syndrome,3.Chandler syndrome. We report here a case of ICE syndrome with progressive iris atrophy and glaucoma.

Keyword : ICE syndrome, progressive iris atrophy, abnormal corneal endothelium, glaucoma.

Introduction:

Iridocorneal endothelial syndrome typically involves one eye of middle aged women. ICE syndrome consists of 3 overlapping disorders, they are1. Progressive iris atrophy,2.Cogan Reese syndrome,3.Chandler syndrome2. The main pathology in all these variants is proliferation of abnormal corneal endothelial cell layer across the iris and angle. Glaucoma here is usually due to synechial angle closure secondary to contraction of this abnormal tissue3. The main clinicalfeatures of progressive iris atrophy are corectopia, pseudo polycoria, iris atrophy of varying severity4,broad based peripheral anterior synechiae(PAS), Increased intra ocular pressure(IOP) in 50%of individuals1,and abnormal specular microscopy5.

Case study:

Mr. Mahalingam, 34 years old(Fig-1&Fig-2), Mason by occupation, came with the complaints of defective vision, pain and discomfort in left eye of 1 week duration. There was no h/o injury or similar illness in the past. No other relevant history. On examination: The right eye appeared normal clinically (Fig- 3) with the visual acuity of 6/6 by Snellen's chart. In the left eye there was circum corneal congestion with minimal corneal edema. The anterior chamber was shallow with PAS. Iris showed severe atrophy with stretch holes, corectopia and pseudo polycoria(Fig-4). Fundus appeared normal with normal cup-disc ratio. Vision in the left eye was 6/36 NIP, and IOP was 40mmHg with Goldmann applanation tonometer

Fig 1-Clinical photograph of patient

Fig 2-Clinical photograph of patient

Fig 3-Slit lamp photograph of normal right eye

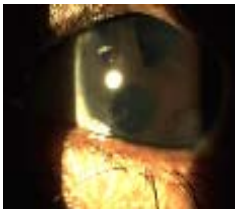

Fig 4- Left eye with stretch holes

GONIOSCOPY:

Revealed open angles in right eye and closed angles with PAS in left eye(Fig 5-8)

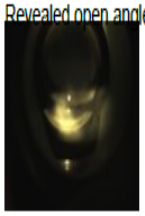

Fig-5 Gonio photo of closed superior angle

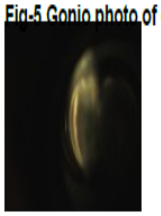

Fig-6-Gonio photo of closed temporal angle

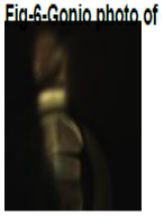

Fig 7-Gonio photo of closed inferior angle with PAS

Fig 8-Gonio photo of closed nasal angle with PAS

SPECULAR MICROSCOPY: RE- normal(Fig-9). LE- Complete loss of endothelial mosaic with loss of hexagonality .Pleomorphism and polymegathism were present(Fig-10).

Fig 9-Specular of right eye

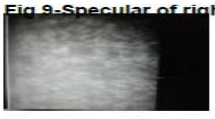

Fig 10-Specular of left eye

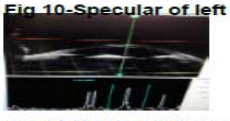

Fig 11-UBM of left eye slowing stretch holes and PAS

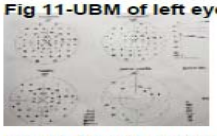

Fig-12 AP left eye with paracentral field defects

Ultra sound biomicroscopy of left eye:

Altered iris configuration with stretch marks from 9-12 clock position, broad based PAS from 5-7 0 clock position.AC diameter-2.89mm,lens-3.58mm and CCT-0. 52 microns(Fig 11).

Automated perimeter with octopus:

Normal fields in right eye. The left eye showed few scotomas in paracentral area with good reliability(Fig-12). This being his first field, patient was advised repeat fields.

Management:

This patient was treated with topical brimonidine and timolol eye drops twice daily and IOP was under control with medication. Patient is awaiting surgery on left eye.

Discussion:

ICE syndrome is common among females in middle age group. Three variants are 1.Progressive iris atrophy,2.Cogan-Reese syndrome,3.Chandler syndrome. The differentiation depends primarily on iris changes6.The main pathology in this syndrome is an abnormal corneal endothelial cell proliferation across iris and angle. Glaucoma is due to synechial angle closure secondary to contraction of this abnormal tissue, and 50% of patients with this syndrome will develop glaucoma. Glaucoma is severe in Cogan-Reese and Progreesive iris atrophy7.Patients are usually managed with medication, surgery(Trabeculectomy) combined with antimetabolite and shunt procedures are the next available option. The outcome of surgery is not good, failures are common due to endothelialization of filtering bleb.

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