International Journal of Medical Ophthalmology

E-ISSN: 2663-8274
P-ISSN: 2663-8266
www.ophthalmoljournal.com
IJMO 2019; 1(2): 25-28
Received: 14-05-2019
Accepted: 18-09-2019

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Essential iris atrophy with secondary angle closure glaucoma: A rare spectrum of Irido-corneal endothelial syndrome

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DOI: https://doi.org/10.33545/26638266.2019.v1.i2a.16

Abstract
Iridocorneal Endothelial syndrome (ICE) is a rare unilateral progressive disease of unknown Etiology, predominantly occurring in middle aged females. It consists of three overlapping disorders namely progressive iris atrophy, Cogan Reese syndrome and Chandler's syndrome. Cogan-Reese syndrome is characterised by diffuse iris naevus or pedunculated iris nodules. Chandler's syndrome is associated with corneal changes portraying "hammered silver" appearance. Iris changes are prominent in progressive iris atrophy. The management is targeted to control corneal edema and secondary glaucoma. The following report is a case of essential iris atrophy, a rare condition under the spectrum of ICE syndrome. The proliferative Endotheliopathy results in characteristic iris changes, Irido-trabecular Synechiae, Corectopia and secondary angle-closure glaucoma. Gonioscopy revealed the characteristic "high peripheral anterior synechiae". The fundus examination showed unilateral glaucomatous cupping and intraocular pressure was managed with anti-glaucoma medications. Regular monitoring is required for early diagnosis of vision threatening complications in ICE syndrome.

Keywords: Irido-corneal endothelial syndrome, Essential iris atrophy, Chandler's syndrome, Cogan-Reese syndrome, high peripheral anterior synechiae

Introduction
Iridocorneal endothelial syndrome (ICE) is a rare disorder involving cornea and iris predominantly. ICE syndrome comprises of a spectrum of clinical entities namely essential Iris Atrophy, Chandler's syndrome and Cogan- Reese syndrome [1]. An abnormal corneal endothelium is postulated as the major pathogenesis and is associated with corneal edema, iris atrophy and secondary angle closure glaucoma [1]. It is a unilateral condition with a strong predilection for young women and is devoid of family history [2]. Though the etiology of ICE syndrome is largely obscure, it is believed to be of viral origin due to recurrent episodes of inflammation [3]. We present a case of ICE syndrome with predominant features of essential iris atrophy with secondary glaucoma.

Case report
A 34 year old female presented with complaints of photophobia and colored halos in the right eye for six months. On ocular examination, the Best Corrected Visual Acuity (BCVA) was 6/6 in both eyes. Slit-lamp examination of the right eye revealed a clear cornea with iris atrophy and multiple stretch holes at three o'clock, seven o'clock and 11 o'clock positions [Fig 1, 2 and 3].
Gonioscopy showed high peripheral anterior synechiae which was seen to extend beyond the Schwalbe's line. Intraocular pressure with Goldmann applanation tonometry was found to be 24 mm Hg in the right eye and 14 mm Hg in the left.
The fundus examination showed a cup to disc ratio of 0.6 in the right eye and 0.4 in the left eye [Fig 4 and 5]. The anterior segment examination and the fundus examination of the left eye was found to be within normal limits. Anti-glaucoma therapy was initiated with topical beta blocker in the right eye and the patient was advised to be on regular follow up.

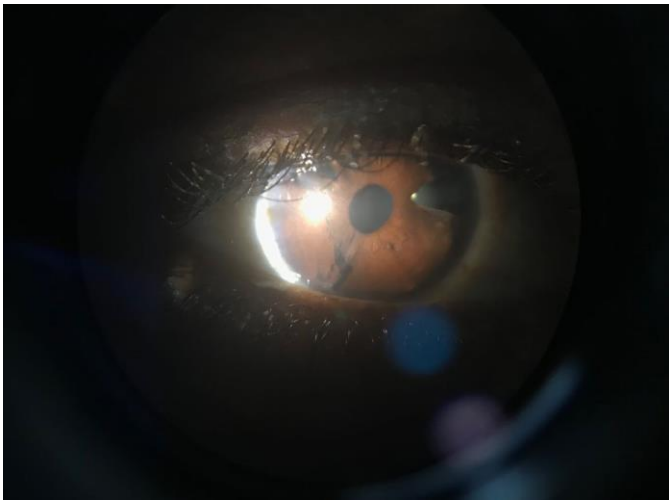

Fig 1: Slit lamp examination showing stretch holes at three o'clock, seven o'clock and eleven o'clock positions in the right eye.

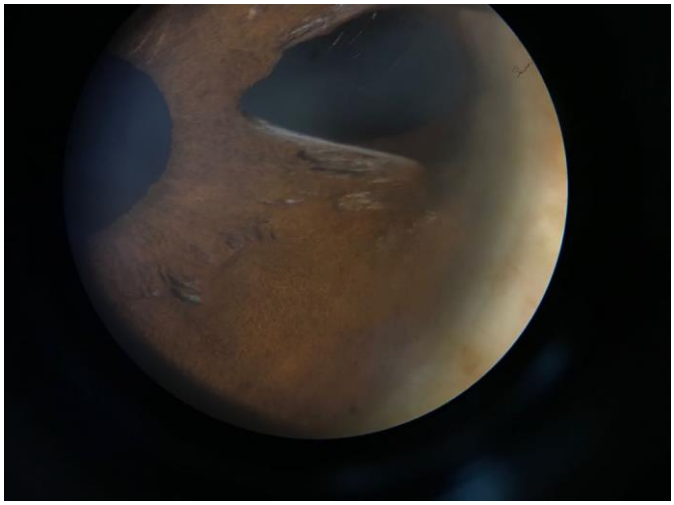

Fig 2: Slit lamp examination showing stretch hole and iris atrophy (magnified view)

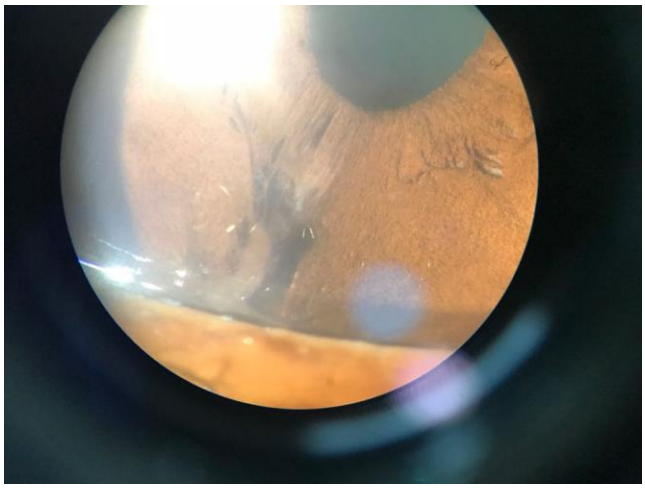

Fig 3: Slit lamp examination-mild Corectopia with iris atrophic patches

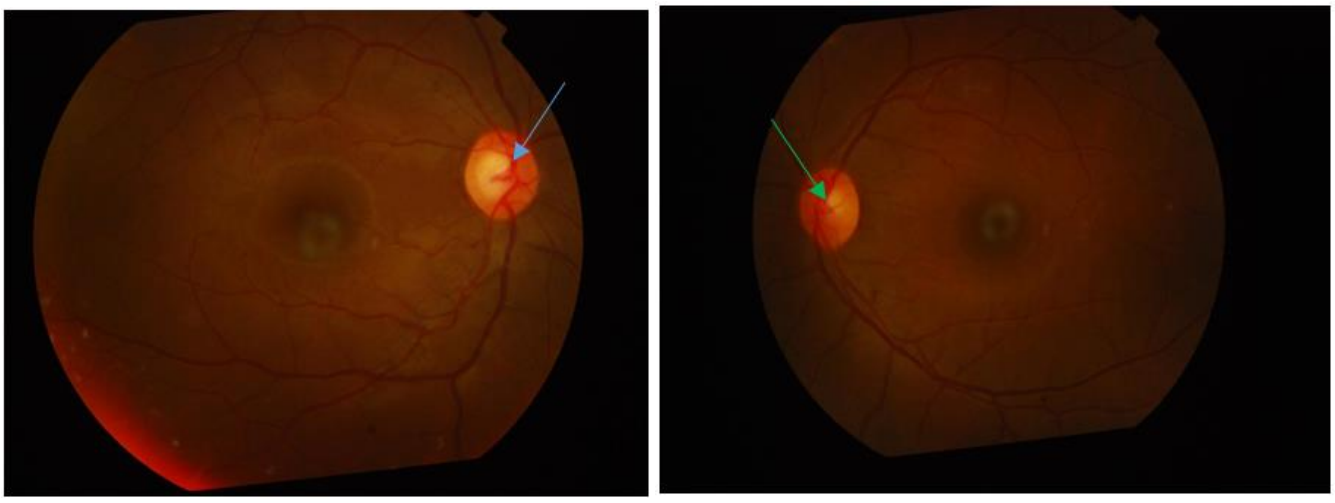

Fig 4, 5: Fundus picture showing glaucomatous cupping in the right eye (blue arrow) and normal optic disc in left eye (Green Arrow)

Discussion

ICE is a spectrum of disease characterized by primary corneal endothelial abnormality. It is typically unilateral although subclinical abnormalities may be demonstrable in the other eye. It is predominantly seen in middle aged adults and has a female predilection [2, 4]. There is no systemic or genetic association. It consists of three unusual and frequently overlapping disorders namely progressive iris atrophy, Cogan Reese syndrome and Chandler's syndrome [4].

The 'ICE' cell is an altered endothelial cell, which shows proliferative and structural abnormalities and the ability to migrate into the surrounding tissues. They are abnormally large cells with severe pleomorphism [5]. Desmosomes, tonofilaments and numerous microvilli have been identified by electron-microscopy and substantiates that ICE cell

mimics epithelial-like characteristics [3]. The abnormal endothelial cells may migrate posteriorly forming a membrane that covers the iris and trabecular meshwork [6]. The contraction of this membrane leads to characteristic iris changes, Irido-trabecular synechiae, corectopia and secondary angle-closure glaucoma [5]. Glaucoma may appear in the absence of synechiae because of membrane migration and functional closure of the angle [2].

The abnormal corneal endothelial cell layer is the common link between these three variants. It has the ability to proliferate across Schwalbe's line, obliterating the angle and then proceeding on to the iris, where it forms an abnormal basement membrane. This eventually contracts triggering iris atrophy, corectopia and formation of synechiae. This justifies the term 'proliferative Endotheliopathy' [7]. The angle obstruction results in an increase of intraocular pressure (IOP) and development of glaucoma in 46% to 82% of patients [8].

Alvarado first postulated that the underlying Endotheliopathy could have a viral origin. The endothelial alterations in ICE syndrome are similar to those observed in viral infections. Polymerase Chain Reaction has demonstrated the DNA of Herpes simplex virus in a substantial percentage of corneal specimens.[9] Campbell and associates proposed the membrane theory, according to which the abnormality of corneal endothelium is the primary defect [7].

The iris – naevus syndrome or Cogan-Reese syndrome is characterised by diffuse iris naevus or pedunculated iris nodules, surrounded by matted appearance of stromal iris [10]. These nodules are normal iris tissue pinched off by contracting endothelial membrane. Corectopia is found to be severe in this variant. Iris atrophy is mild and absent in 50%. Ectropion uveae may be present [11].

Chandler's syndrome is particularly associated with corneal changes which typically show "hammered silver" appearance [8]. Iris alterations are minimal. Specular microscopy reveals characteristic ICE cells which are dark cells, except for a light central spot and light peripheral zone. The clear hexagonal margins of endothelial cells are lost. The associated features include pleomorphism and decreased endothelial cell count [12]. In Chandler's syndrome, corneal oedema can persist even after control of IOP.

Iris changes are prominent in progressive iris atrophy. Iris atrophy is prominent on the side opposite to pupillary distortion [13]. Corectopia points towards a prominent peripheral anterior synechiae. Iris holes can be of two distinct subtypes termed as stretch holes and melting holes. The stretch holes result from iris thinning on the other side of corectopia and melting holes are due to tissue ischemia. Gonioscopy discloses "high" peripheral anterior synechiae (PAS) [13].

Glaucoma and corneal decompensation are serious sequelae of ICE syndrome. Glaucoma can be due to extensive PAS or obstruction of anterior chamber angle by membrane. The primary aim of treatment is to reduce corneal edema, manage glaucoma and prevent irreversible visual impairment [13]+ The control of corneal edema can be achieved by lowering IOP, use of hypertonic saline and soft contact lens [14]. The persistence of corneal edema can lead to corneal decompensation. In advanced cases of corneal edema, penetrating keratoplasty (PK) should be considered [15]. Deep Lamellar Endothelial Keratoplasty (DLEK) and Descemet Stripping Endothelial Keratoplasty (DSEK) are successful in patients with ICE syndrome [16].

Glaucoma can be controlled by aqueous suppressants initially but long term medical management is ineffective. When indicated, surgical options are augmented trabeculectomy and shunt surgeries.[8] Antifibrotic agents have been proposed to increase the success rate of filtering surgery [17]. The failure of filtering surgery may be due to the progressive growth of endothelial membrane over the trabecular meshwork and filtration site. Late failure can occur due to obstruction of fistula by synechiae or endothelialisation [18]. Failure of surgery can occur due to an exuberant inflammatory response in the young.

There are various anterior segment mimics which may be complicated by complications such as corneal edema and glaucoma. The differential diagnoses include Posterior Polymorphous Dystrophy (PPCD), Fuchs endothelial dystrophy, Axenfeld-Rieger syndrome, iris melanoma, inflammatory iris nodules and aniridia [14].

Conclusion

ICE syndrome should be considered in the differential diagnosis of a young female presenting with unilateral iris anomalies, corectopia, high peripheral anterior synechiae and glaucoma. Regular monitoring is required for early diagnosis and treatment of vision threatening complications like corneal edema and glaucoma.

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