

Unilateral Iridocorneal Endothelial Syndrome In Young Female

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Abstract: Iridocorneal endothelial syndrome is described to be a unilateral condition in young females in its common presentation. However, it has also been reported to be bilateral. There are three known overlapping clinical variants of this condition namely, essential iris atrophy, Cogan-Reese and Chandler syndrome. Hereby we describe a case of unilateral ICE syndrome in young female with retinal detachment in other eye.

Keywords: Cogan Reese, Chandler syndrome, Glaucoma

INTRODUCTION

Iridocorneal endothelial (ICE) syndrome encompasses a group of ocular pathologies characterized by corneal proliferative endotheliopathy in which secondary corneal edema, peripheral anterior synechiae, and abnormalities of the iris stroma are the common hallmarks [1, 2]. The disease complex, which includes essential iris atrophy, Chandler's syndrome, and iris nevus (Cogan-Reese) syndrome, is usually unilateral, non-familial, and typically occurs in females during young adulthood. However, it has been reported to be bilateral [2]. We need to differentiate it from Rieger's anomaly and posterior polymorphous corneal dystrophy; however, there is no corneal involvement in the former and autosomal dominant inheritance pattern in the later [1].

CASE REPORT

We describe a case of unilateral ICE syndrome in young female with retinal detachment in other eye which has never been reported in literature.

A young female 28 years old presented with complaints of defective vision in both eyes. The complaint of decreased vision in left eye dated back to her childhood, since birth and right eye since last one year. Her past medical and surgical histories were unremarkable. Her family history did not reveal any ocular problem.

On examination, she had visual acuity of hand movements in the right eye and no PL in the left eye.

Her recorded intraocular pressures were 7.1 mm Hg in both eyes. The right eye revealed normal anterior segment including clear cornea, normal pupil reacting to light, normal anterior chamber and clear lens (Fig. 1). The left eye revealed anterior synechiae, areas of peripheral iridocorneal touch, iris stromal atrophy, pupillary membrane and a dense elevated mass extending between 11 to 4 o'clock position in anterior hyaloid phase (Fig. 2). Other details could not be made out.

Fundus examination in right eye revealed hazy view with cup disc ratio of 0.8 with area of chorioretinal atrophic patch temporal to disc, around 1 DD. Inferiorly there was retinal detachment with presence of 2 retinal holes at 6 o'clock position, probable reason for poor vision (Fig. 3-5).

On the basis of this clinical presentation, probable diagnosis of iridocorneal endothelial syndrome been made.



Fig. 1: Right Eye showing normal anterior segment



Fig. 2: Left Eye showing anterior synechiae, areas of peripheral iridocorneal touch, iris stromal atrophy, pupillary membrane and a dense elevated mass extending between 11 to 4 o'clock position in anterior hyaloid phase.

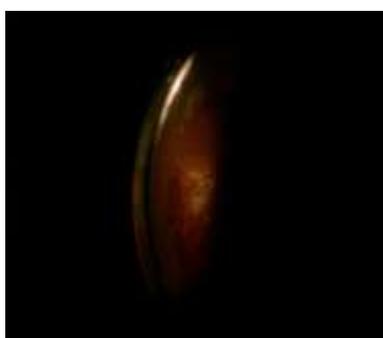


Fig. 3: Right Eye fundus showing CD ratio of 0.8

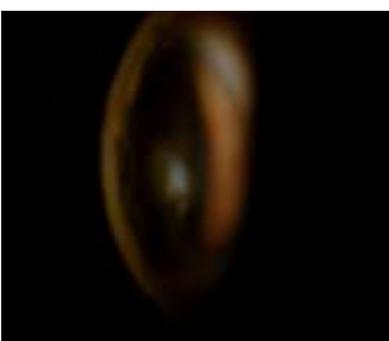


Fig. 4: Chorioretinal atrophic patch temporal to disc



Fig. 5: Inferior RD

DISCUSSION

The most commonly described mechanism of Iridocorneal Endothelial Syndrome is an abnormality of corneal endothelial cells, secondary spreading of the cells over the trabecular meshwork region causes anterior synechiae and elevated intraocular pressure, after extension across the surface of the iris, it results in polycoria and corectopia. In literature, visual loss in ICE syndrome has been attributed to corneal edema and glaucoma [4, 5]. Glaucoma and marked decrease in visual acuity despite corneal endothelial changes and iris abnormalities might not occur for a long-time as already reported [3, 6]. However, to the best of our knowledge unilateral ICE has never been described to be associated with retinal detachment. The etiology of ICE syndrome remains unclear, but viral origin (infection with Herpes simplex or Epstein-Barr virus) has been proposed [7].

This unique combination of unilateral ICE syndrome with retinal detachment has never been reported before, therefore, it needs to be ascertained that whether it was a coincidence or an association. Nevertheless, we emphasize the careful examination of anterior as well as posterior segment in this syndrome.

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