INTRODUCTION

Iridocorneal endothelial syndrome is described to be a unilateral ocular condition in young females. There are three known overlapping clinical variants of this condition namely, essential iris atrophy, Cogan-Reese and Chandler syndrome. We report a case of bilateral iridocorneal endothelial (ICE) syndrome with microspherophakia. A 25 years old female presented with microspherophakic lens dislocated into anterior chamber in right eye, and displaced inferiorly in left eye. She was also diagnosed with ICE syndrome and underwent lensectomies in both eyes. This unique combination has never been reported before.

CASE REPORT

A 25 years old female presented with the complaints of watering of both eyes and photophobia for last 2 months. The complaint of decreased vision in both eyes dated back to her childhood. Her past medical and surgical histories were unremarkable except for chronic urinary incontinence. Her family history did not reveal any ocular problem.

On examination, she had the best corrected Snellen visual acuity of 3/60 in the right eye and 6/60 in the left eye. Her highest recorded intraocular pressures were 14 mmHg OD and 12 mmHg OS. The right eye revealed multiple ciliary staphylomas, mild corneal edema and an anteriorly dislocated microspherophakic crystalline lens (Figure 1). Iris showed segmental atrophy. The left eye examination showed multiple ciliary staphylomas, clear central cornea and a band of peripheral corneal opacification (Figure 2). There was marked iris stromal atrophy with corectopia, polycoria and broad anterior synechiae. Though her pupil dilated poorly, it revealed microspherophakic crystalline lens displaced inferiorly, on slit-lamp examination which moved to mid-vitreous in supine position (Figure 3). Fundus examination in both eyes was unremarkable, with cup disc ratio of 0.3. There was no posterior embryotoxon, facial abnormalities and her family members had normal eye examination. Systemic examination showed no features suggestive of Marfan's syndrome, Weil-Marchesani and homocystinuria. Further work-up included A-scan, which showed normal axial length of 22.0 mm OD and 22.37 OS. The AC depth was unrecordable in right eye (due to anterior lens dislocation) and 1.95 mm OS. Specular microscopy showed endothelial cells dysmorphology, with loss of normal hexagonal pattern and areas of focal cell loss in both eyes. However, vision is surprisingly good probably due to lack of corneal edema. Endothelial cell count was 723 OD and 1051 OS. The central corneal thickness was 0.706 and 0.480 micrometer in the right and left eye respectively. Her family members had normal corneal endothelial counts and morphology.

On the basis of this clinical presentation and diagnostic work-up, she was diagnosed with iridocorneal endothelial syndrome with microspherophakia. Lensectomy were performed in both eyes through corneal approach as the crystalline lens had already dislocated into anterior chamber in right eye and there was a risk of same situation in left eye due to extensive zonular dehiscence.

DISCUSSION

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. 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. The most commonly described mechanism of this disease is an abnormality of the corneal endothelial cells, secondary spreading of the cells over the trabecular meshwork region causes anterior synechiae and elevated intraocular pressure (IOP). When it extends 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. Glaucoma and marked decrease in visual acuity despite corneal endothelial changes and iris abnormalities might not occur for a long-time as already reported. However, to the best of our knowledge (Pubmed search), ICE has never been described to be associated with microspherophakia. It is important to note that in essential iris atrophy, mainly iris changes are seen with minimal corneal involvement while in Chandler’s syndrome, mainly corneal involvement is seen with minimal iris changes. However, in this case both the cornea and the iris were markedly involved.

The etiology of ICE syndrome remains unclear, but viral origin (infection with Herpes simplex or Epstein-Barr virus) has been proposed. There might be a genetic basis for co-occurrence of ICE syndrome and microspherophakia, but it is difficult to attribute the microspherophakia to ICE syndrome or vice versa based on a single case.

This unique combination ICE syndrome with microspherophakia 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.

REFERENCES

Figure 1: Right eye showing ciliary staphyloma, anteriorly displaced microspherophakic crystalline lens.
Figure 2: Left eye showing peripheral corneal opacification, iris atrophy and polycoria.
Figure 3: Crystalline microspherophakic lens dislocated into vitreous cavity.