Unilateral keratoconus associated with iridocorneal endothelial syndrome: Case report

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Abstract:

We report a rare association of unilateral keratoconus (KC) and iridocorneal endothelial syndrome in a 34-year-old female. Slit-lamp examination showed advanced KC with faint apical scarring in her right eye. The pupil was superiorly displaced with superior peripheral anterior synechia. Specular microscopy showed abnormal endothelium with low endothelial cell count in the right eye. Corneal topography confirmed the unilateral KC diagnosis. As the patient did not tolerate hard contact lens, penetrating keratoplasty with pupilloplasty was performed with excellent outcome.

Keywords:

Iridocorneal endothelial syndrome, keratoconus, unilateral

Introduction

Keratoconus (KC) is a noninflammatory progressive corneal ectasia that usually manifests during the second decade of life. It is characterized by progressive corneal thinning, myopia, and irregular astigmatism.[1]

Iridocorneal endothelial (ICE) syndrome is a group of diseases characterized by abnormalities of the iris and the corneal endothelium. ICE syndrome comprises a spectrum of three clinical variants: Chandler's Syndrome, essential iris atrophy (EIA), and Cogan-Reese Syndrome. It is nonhereditary, and it is usually unilateral affecting adult women more often than men with onset at young-to-middle age.[2]

Few cases of bilateral KC and ICE syndrome have been reported.[3-6] This report describes the clinical features and management of a case of unilateral KC and ICE syndrome.

Case Report

A 34-year-old female presented complaining of a progressive decrease in vision in the right eye for the past 5 years. She denied any history of trauma, eye rubbing, allergic conjunctivitis, or recurrent eye inflammation. There was no significant medical or family history.

Examination revealed unaided visual acuity of counting fingers at two feet in her right eye and 20/30 in the left eye. Intraocular pressure measured by Goldmann applanation tonometry was 16 mmHg and 14 mmHg in the right and left eyes, respectively.

Slit-lamp examination showed advanced ectasia with faint central scar in the cornea of the right eye. The iris showed no transillumination defects. The pupil was horizontally oval and displaced superiorly with good reaction to light [Figure 1a]. Corneal sensation was normal for both eyes. Gonioscopy of the right eye showed that there is superior peripheral anterior synechia [Figure 1b], and it showed normal open-angle in the left eye. The rest of the right eye ocular examination was normal. The left eye examination was unremarkable.

Specular microscopy showed a low endothelial cell count of 558 cell/mm^2 in the right eye with abnormal morphology, mild rounding of endothelial cell angles, and few eccentric dark areas. The count was 2911 cell/mm^2 in the left eye with normal-looking cells [Figure 2a and b]. Corneal topography confirmed the diagnosis of

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advanced KC in the right eye, and it was within normal limits in the left eye.

The patient showed no improvement by refraction. Rigid gas-permeable lens fitting in her right eye was tried, but the patient could not tolerate the examination. Scleral lenses were not tried. Therefore, full-thickness penetrating keratoplasty and pupilloplasty to centralize the pupil was performed to the right eye. After the excision of the patient's cornea, an inferolateral crescent of the iris at the pupil margin was excised followed by limited suturing of the superonasal pupil. Then, the graft was sutured in place by interrupted sutures. Histopathology of the excised cornea showed discrete breaks in Bowman's membrane, corneal thinning, and scarring. Descemet's membrane showed a severe endothelial loss.

The patient was followed for 2 years with reported improved vision significantly. She had nonsignificant glare most likely due to the large postoperative pupil. Latest examination revealed unaided visual acuity 20/60 and 20/30 in the right and left eye, respectively. Intraocular pressure was within normal limits, and the corneal graft was clear [Figure 3]. The rest of right eye examination and left eye examination were unremarkable. Topography remained normal in the left eye until the last follow-up visit.

Discussion

KC usually presents as an isolated sporadic disease, but it can be associated with certain ocular and systemic conditions. It can be associated with atopic disease,[7] Down syndrome,[8] and eye rubbing.[9]

Our case describes a rare association of KC and ICE syndrome which is unilateral with a completely normal fellow eye. The other reported cases were bilateral. De Maria *et al*. reported a case of bilateral KC with unilateral EIA and advanced glaucoma.[3] Blair *et al*. described a case with bilateral progressive EIA and KC with coincident features of posterior polymorphous dystrophy.[4] Gus *et al*. described a case of unilateral posterior KC and iris atrophy that did not qualify as an ICE syndrome.[5] Chakrabarty's case had bilateral KC with Cogan-Reese syndrome and secondary glaucoma.[6] Table 1 shows a comparison between the current case and the previous cases.

One of the theories regarding ICE syndrome's etiology is the association with herpes simplex virus infection, but most likely

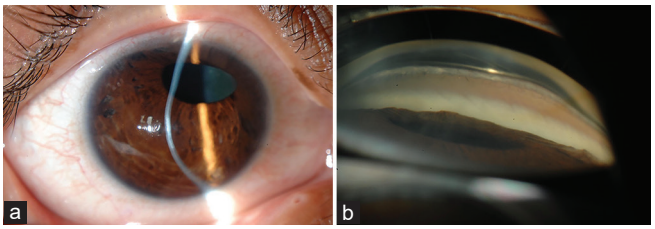

Figure 1: (a) Slit-lamp photograph showing steep cornea with apical scar and superiorly displaced pupil, (b) gonioscopic photograph showing peripheral anterior synechia superiorly

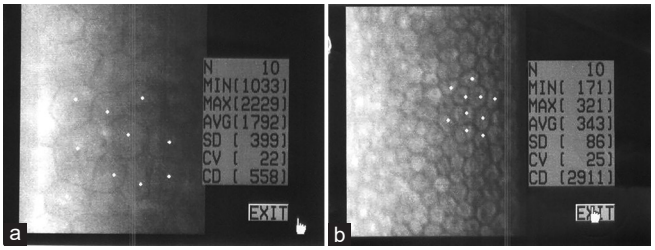

Figure 2: Specular microscopy photographs showing (a) right eye with decreased endothelial count, mild rounding of endothelial cells angles, (b) normal endothelium of left eye

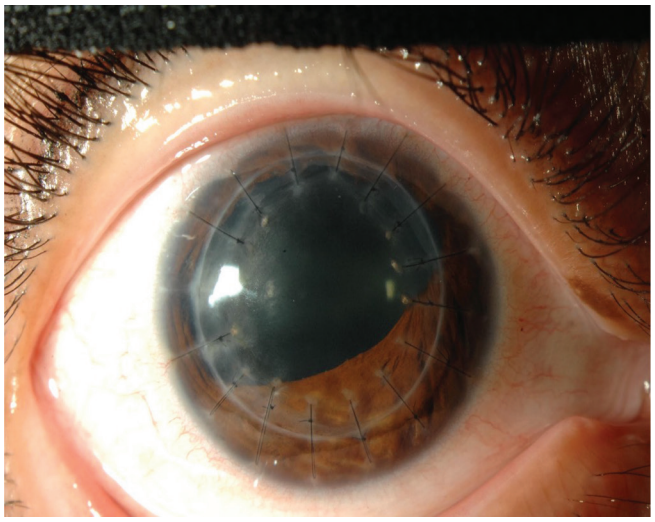

Figure 3: Photograph showing clear graft with central dilated pupil 2 years after the surgery

Table 1: Comparison of clinical features of reported cases

	Age	Sex	KC	ICE	Specular microscopy	Glaucoma	Management
Current case	34	Female	Unilateral	Unilateral	Changes of ICE	None	Penetrating keratoplasty, pupilloplasty
De Maria *et al*.[3]	38	Male	Bilateral	Unilateral EIA	Changes of ICE	Present	Glaucoma drainage device implantation
Blair *et al*.[4]	47	Female	Bilateral	Bilateral EIA, posterior polymorphous atrophy	Changes of both ICE and PPD	Present	Initially beta-blocker shifted to neptazane after development of wheezing, RGP lenses
Gus *et al*.[5]	47	Female	Unilateral	Unilateral mild peripapillary iris atrophy	Not done	None	Not mentioned
Chakrabarty[6]	32	Male	Bilateral	Unilateral Cogan-Reese syndrome	Abnormal endothelial morphology	Present	RGP lenses, topical antiglaucoma

ICE: Iridocorneal endothelial, PPD: Posterior polymorphous dystrophy, RGP: Rigid gas permeable, KC: Keratoconus, EIA: Essential iris atrophy

this was not the cause in our case as there was no history of recurrent redness, and the corneal sensation was normal.[10]

Full-thickness penetrating keratoplasty was performed instead of deep anterior lamellar keratoplasty because the endothelium was already low in count in this case. Despite the low endothelial count of 558 cell/mm^2, the cornea was still clear with no edema. Furthermore, as the pupil was not central, it was important to perform a pupilloplasty to address the centralization of the pupil.

In conclusion, KC can rarely be associated with ICE syndrome unilaterally, the management of which can be a clinical and a surgical challenge, yet the patient can have an excellent outcome.

Declaration of patient consent

The authors certify that we have obtained appropriate patient consent forms. In the forms, the patient has given her consent and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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