A unique case of keratoconus with Cogan-Reese syndrome and secondary glaucoma

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Keratoconus (KC), though one of the most common corneal degeneration, still continues to be a mystique regarding its pathogenesis, diagnosis, associations, and management; with newer discoveries and evolutions being reported. We report, what we believe to be another new association of KC- Cogan Reese syndrome with secondary glaucoma. A 32-year-old male, diagnosed as bilateral KC, presented for examination. Unilateral Cogan-Reese syndrome and associated secondary glaucoma was identified. These associations had been missed by previous ophthalmologists. The patient was managed with a rigid contact lens for KC and topical antiglaucoma agents for glaucoma. He was advised regular reviews and is under observation till date. We describe the first case known to us of a new association with KC. This case not only highlights the ophthalmologist's need to look for multiple entities linked to KC; but may also pave way for future insights regarding pathogenesis and genetics of these associated diseases.

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Iridocorneal endothelial (ICE) syndrome is characterized by corneal proliferative endotheliopathy in which secondary corneal edema, peripheral anterior synechiae, and abnormalities of the iris stroma are seen. The disease complex, which includes essential iris atrophy, Chandler's syndrome, and Iris nevus (Cogan-Reese) syndrome, is almost always unilateral, nonfamilial, and typically occurs in females during young adulthood.[1] ICE syndrome is frequently complicated by secondary glaucoma and corneal decompensation.

Keratoconus (KC) is a progressive, bilateral (but usually asymmetric) ectatic corneal disease. Visual loss occurs from irregular astigmatism, myopia, and corneal scarring.[2]

Association of KC with ICE syndrome is not well reported. This case report describes a unique association of KC.

Case Report

A man in his early thirties presented with complaints of gradual diminution of vision in his right eye noted over a span of 3–4 years. The patient did not have any history of recurrent redness, itching or pain in eyes or any known history of trauma. He had been diagnosed as bilateral KC (with greater involvement of right eye) elsewhere and was referred for a second opinion.

On presentation, his uncorrected visual acuity was 20/400 OD and 20/60 OS. Both eyes were found to have compound myopic astigmatism. The right eye was being corrected to 20/60 with high myopic (−12 Diopter) and low cylinder correction

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(−2 Diopter). The left eye was corrected to 20/30. Retinoscopy showed a scissors reflex.

Anterior segment examination findings have been illustrated in Fig. 1. Slit lamp examination showed steep cornea both eyes with a faint apical corneal scar in the right eye. Fleisher's ring, prominent corneal nerves and Vogt's striae was present in both eyes, being more prominent in right eye. Right eye showed variable anterior chamber depth with iris nodules, atrophic areas on iris, ectropion uveae, and peripheral anterior synechiae. Pupil in right eye was approximately 5 mm with peaking at 10 o' clock position. Pupil was segmentally reacting to light owing to synechiae. Left eye had normal anterior chamber depth with circular pupil, normally reacting to light. Lens was clear in both eyes. Gonioscopy of the right eye showed peripheral anterior synechiae. Left eye gonioscopy showed open angles.

Intraocular pressure by Goldmann applanation tonometer was 28 mmHg in the right eye and 8 mmHg in left eye. Axial length was 22.79 mm in right eye and 22.23 mm in left eye. Optic disc examination [Fig. 2] showed a cup to disc ratio of approximately 0.4 in right eye with early glaucomatous changes suggested by inferior notching and inferior vessel bending (bayoneting sign). Left eye showed a healthy neuroretinal rim with a cup to disc ratio of approximately 0.3. Visual fields were unreliable but showed few scotomas in right eye and grossly normal left eye.

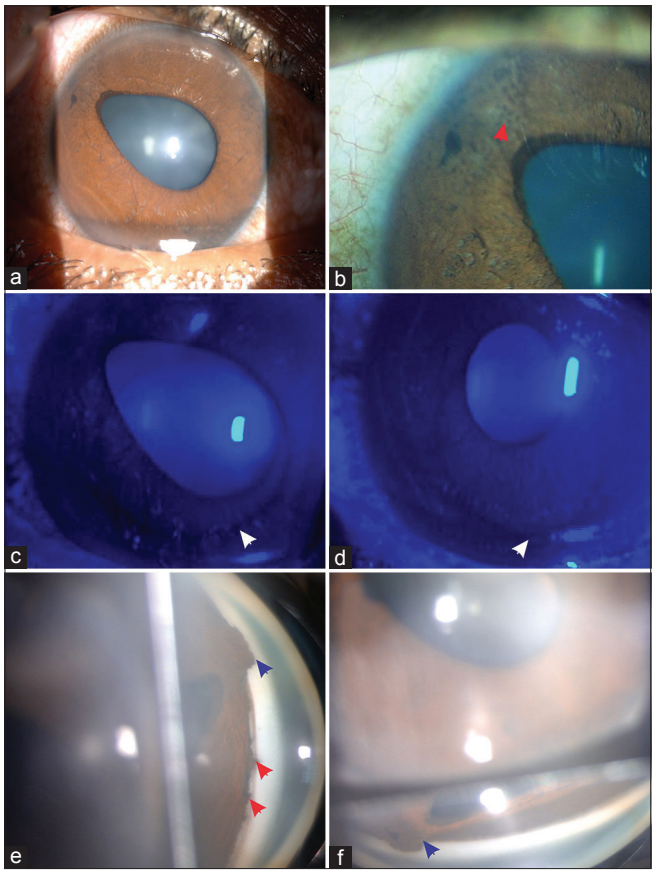

Figure 1: Anterior segment findings. (a) Diffuse illumination of right eye; (b) magnified view showing areas of iris atrophy and iris nodules (red arrow); (c and d) cobalt blue filter illumination of right eye (c) and left eye (d) showing Fleisher's ring (white arrow); (e) right eye gonioscopy of temporal angle showing broad peripheral anterior synechiae at 10–11 o'clock position (blue arrow) with smaller early adjacent synechiae (red arrows); (f) gonioscopy of temporal-superior zone showing temporal synechiae (blue arrow) and pigmented superior angle

Specular examination (Topcon SP-2000P, Topcon, Tokyo, Japan) of right eye revealed poor endothelial morphology with pleomorphism, polymegathism, and probable ICE cells. Left eye revealed normal corneal endothelium [Fig. 2]. Corneal topography (Orbscan IIz, Bausch and Lomb, Rochester, New York, USA) established the diagnosis of KC both eyes with the right eye being more affected [Fig. 3]. Inferior-Superior difference in both eyes in the central 1–5 mm zone was more than 1.2 Diopter. Pachymetry map shows reduced corneal thickness in both eyes with the central corneal thickness being 0.356 mm in right eye and 0.443 mm in left eye.

The patient was advised rigid contact lens for both eyes. In view of high myopia with a low cylinder refractive correction in right eye, surgical modalities in the form of keratoplasty was deferred. Keeping the high intraocular pressure, thin cornea and young age in mind; he was started on beta blocker (timolol maleate), and prostaglandin analog (latanoprost) eye drops for the right eye. On the patient's successive visits, intraocular pressure of right eye was lowered (16 mmHg on last visit) and the patient was comfortable with his visual rehabilitation.

Examination of patient's parents and sibling was undertaken. The ocular examination did not show any similar or significant findings. The patient has been advised a close monitoring to assess for KC and glaucoma progression. He is on follow-up ever since.

Discussion

We document a case of KC with Cogan-Reese syndrome and secondary glaucoma. In the past, Blair *et al*. have reported the apparent bilateral association of essential iris atrophy

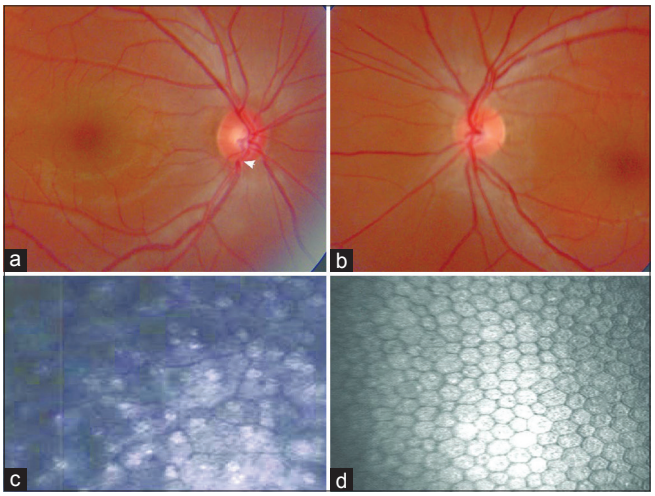

Figure 2: Color fundus photographs and specular microscopy image. (a) Right eye showing early notching in inferior neuroretinal rim and bayoneting of vessel (white arrow); (b) left eye showing healthy neuroretinal rim. (c) right eye magnified specular microscopy shows pleomorphism, polymegathism, light-dark reversal and a low cell density of 986 cells/mm^2; (d) normal polygonal endothelial cells of left eye with cell density of 2414 cells/mm^2

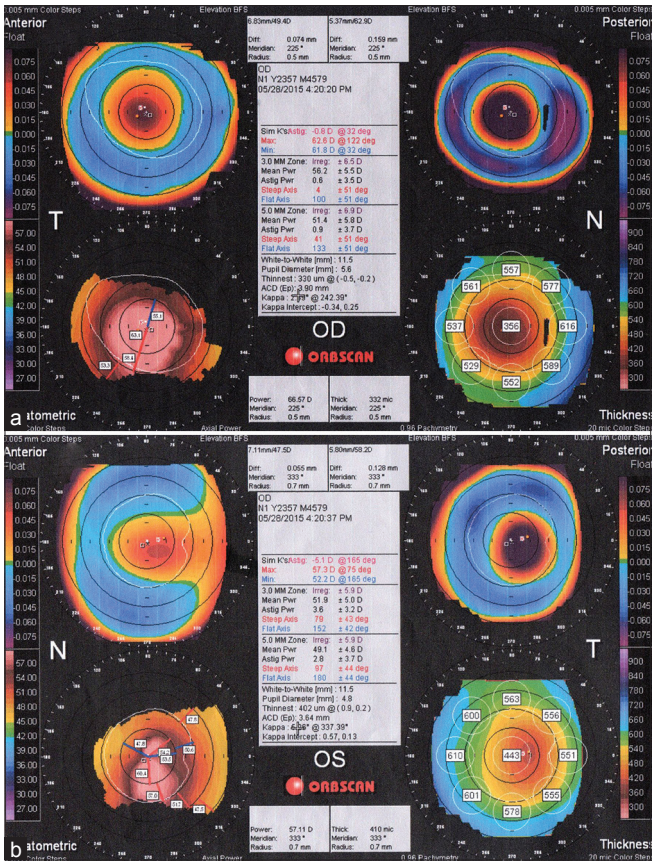

Figure 3: Corneal topography. (a) Right eye showing advanced keratoconus. (b) Left eye showing moderate keratoconus. Maximum simulated keratometric value was 62.6 Diopter in right eye and 57.3 Diopter in the left eye. Posterior elevation was 0.159 mm in right eye and 0.128 mm in left eye

and KC with coincident features of posterior polymorphous dystrophy (PPD).[3] They have also proposed a hypothesis similar to the genetics of retinoblastoma for the pathogenesis of the ICE syndrome with associated KC and/or PPD. The study of Cremona *et al*. presented a 10 year data of associations of KC and corneal dystrophies.[4] ICE syndrome was not documented in same. Although Chandler's syndrome has been postulated to be associated with KC, there are limited published reports. KC has been reported to be associated with PPD, iris heterochromia, and band keratopathy.[4-6] We consider our case as unique since there are no published reports of KC with Cogan-Reese syndrome.

ICE and PPD are believed to be closely related diseases with overlapping features.[7,8] Our case appeared more like ICE rather than PPD because of its unilaterality, nonfamilial nature, specific corneal morphology, presence of synechiae and glaucoma. Because of the presence of iris nodules and pigmented iris lesions, we diagnosed it to be Cogan-Reese syndrome. Although KC has multiple reported associations, our case appears to be the first reported case of a unique association. If ophthalmologists are aware of the entities linked with KC, the management can be rendered better by timely diagnosis.

Past studies have reported associations of KC and have postulated on the pathogenesis. Many studies have focused on the association of PPD claiming a common embryological origin together with simultaneous chromosomal alterations.[3,9,10] Eiferman *et al*. reported a case of iridoschisis and KC in the same eye and suggested a related pathogenesis since the posterior layers of the cornea, and the iris stroma have a common embryological origin.[11] Still, there is much to be learnt about the pathophysiology of KC and its related disease complexes. Chromosomal analysis would have given more insight to this case; which could not be done and account as a limitation of our study. Future work may focus on the possible pathophysiologic or genetic links between these entities, although a chance association cannot be excluded from this study.

Conclusion

This report documents, what we believe to be a new association of keratoconus- CoganReese syndrome. Possible pathophysiology triggering the association needs better understanding.

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Conflicts of interest

There are no conflicts of interest.

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