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Sir,

Bilateral juvenile glaucoma with iridotrabecular dysgenesis, congenital ectropion uveae, and thickened corneal nerves

Congenital ectropion uveae (CEU) is a rare, nonprogressive neural crest cell disorder resulting from proliferation of iris pigment epithelium on the anterior surface of iris from the pigment ruff. The association of glaucoma in this primary iris pigment epithelial hyperplasia, which may present at birth, infancy, or later stages in life has been well documented in the literature.[1–6] Systemic associations often reported are neurofibromatosis, facial hemihypertrophy, Prader–Willi syndrome, and Rieger's syndrome.[2]

It is characteristically unilateral with ipsilateral glaucoma, with only one prior case reported of bilateral congenital ectropion uveae with bilateral glaucoma.[4] There is one previous case report of unilateral CEU with associated thickened corneal nerves.[5]

We present here a case of bilateral juvenile glaucoma with iridotrabecular dysgenesis, congenital ectropion uveae, and thickened corneal nerves, which has not been reported earlier.

Case report

A 22-year-old male presented with painless gradual loss of vision in both eyes during the previous 7 years. His best-corrected visual acuities were 20/60 in right eye and 20/80 in left eye. The applanation intraocular pressure (IOP) on examination was 30 mmHg in right eye and 37 mmHg in left eye.

Congenital ectropion uveae was present in both eyes, extending 360° around the pupil to the mid-periphery of the iris, where it extended in a sharply demarcated scalloped border with the rest of the iris having smooth, cryptless appearance (Figure 1a, b). The pupil itself was

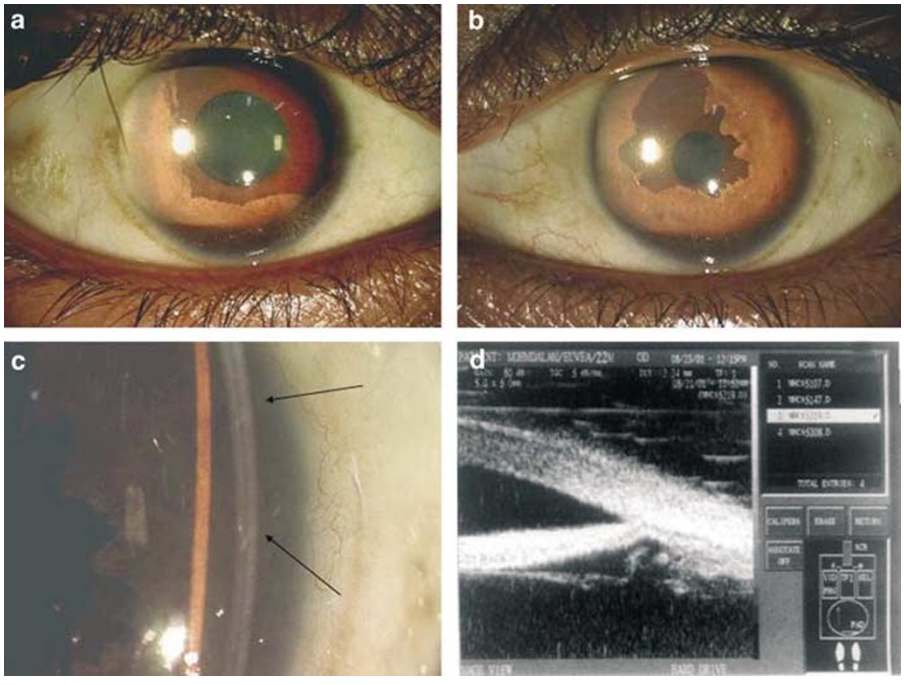

Figure 1 (a) Anterior segment photograph of right eye showing congenital ectropion uveae 360 degrees around the pupil and extending to the superonasal periphery. (b) Anterior segment photograph of left eye showing congenital ectropion uveae extending 360° around the pupil to the mid-periphery of the iris. (c) Anterior segment photograph of left eye (magnification × 25) showing greyish thickened corneal nerves in temporal quadrant (arrows). (d) Ultrasound biomicroscopy showing anterior insertion of iris root in left eye.

round and sluggishly reactive, in both eyes. The corneal diameters were 11.50 mm horizontally with greyish thickened corneal nerves seen in the temporal quadrant of both eyes (Figure 1c). Anterior chamber was of normal depth in both eyes. Fundus evaluation revealed near total glaucomatous cupping in both eyes. On gonioscopy, both eyes revealed an anterior insertion of the iris root over the pigmented trabecular meshwork that was confirmed on ultrasound biomicroscopy (UBM) (Figure 1d). Various ocular dimensions on UBM were as follows: trabecular iris angle 27.2°, angle opening distance 0.166 mm at 250 μm and 0.277 mm at 500 μm, trabecular-ciliary process distance 0.766 mm, iris ciliary process distance 0.220 mm, anterior chamber depth 3.148 mm, and iris thickness 0.336 mm. No evidence of neurofibromatosis (like Café-au-lait spots, fibroma molluscum, plexiform neurofibromas, central nervous system tumours, iris nodules, etc), ptosis, and other systemic or ocular abnormality was present. Humphrey visual field analysis of both eyes showed advanced defects sparing central 5–7° of the visual field.

After informed consent, uneventful trabeculectomy with mitomycin-C application for 3 min was performed in both eyes. The patient developed diffuse, elevated, avascular bleb with IOP of 8–10 mmHg both eyes at 1 week, with IOPs ranging from 10 to 12 mmHg at 3 months follow-up.

Comment

CEU is a very uncommon malformation of the iris, which may be associated with iridotrabecular dysgenesis with glaucoma, blepharoptosis and other ocular and systemic anomalies.[1–7] Not all cases have systemic associations, however. None of the nine patients reported by Dowling *et al*[3] had any systemic associations.

Glaucoma is present in most of the cases reported. Ritch *et al*[2] found glaucoma in seven out of eight cases of CEU, Dowling *et al*[3] in eight of nine cases, and it was also noted frequently in other reported cases of CEU.[1,4,6,7] The onset of glaucoma is usually in childhood or early adolescence as was seen in cases in this report but cases with late onset also have been reported.[3,5,6] IOP shows an initial decrease when the medication is administered but rises again soon after and filtering surgery is required in most of the cases.[3]

There is one case report of unilateral CEU with prominent corneal nerves and glaucoma.[5]The differential diagnosis of prominent corneal nerves include corneal disorders like anterior keratoconus, Fuch's dystrophy with bullous keratopathy, posterior polymorphous dystrophy, herpes zoster and simplex keratitis and failed grafts, and may be associated with systemic disorders like congenital icthyosis, Refsum's disease, multiple endocrine neoplasias, leprosy, and neurofibromatosis.[5]

The present case had bilateral CEU with late onset primary glaucoma with bilateral prominent corneal nerves, which has not been reported earlier. The triad of primary developmental glaucoma, CEU, and prominent corneal nerves is embryologically related and points to a neural crest cell theory of anterior segment dysgenesis.[8]

Although CEU is a benign ocular abnormality, a high degree of caution should be exercised so as to detect associated glaucoma (which may be early or late) as early as possible to prevent the irreversible optic neuropathy caused by it.

Acknowledgements

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Sir,

Greenish discoloration of a CeeON 911A silicone intraocular lens

Since the advent by Charles Kelman in 1967, phacoemulsification has improved a lot. Foldable intraocular lens implants (IOL) are one of the most significant breakthrough after phacoemulsification. We here report a case of greenish discoloration of a silicone IOL (CeeOn 911A, Pharmacia Corporation). Exact cause of this discoloration is still unknown.

Case report

A 35-year-old woman underwent uneventful phacoemulsification with in the bag implantation of a +22.0 D silicon foldable IOL (CeeON 911A, Pharmacia Corporation, Serial No. 6295 07 211) under topical anaesthesia with 2% xylocain (Barrett- Hodgson, Pakistan) on 15 November 2001. No intraocular dye (trypan blue, indocyanine green) for staining of anterior capsule was used. Her postoperative recovery and follow-ups were unremarkable and at her last visit on 31/01/2002, her visual acuity (VA) in her operated eye was 1.0 with –1.00 DS. She then lost to follow-up.

On 10 March 2004, she presented with complaint of decreasing vision in her left (other) eye. Examination revealed cataract in her left eye, resulting in decreased VA. Interestingly her right eye, which was operated for cataract in 2001, caught more attention. The IOL in the right eye was positioned in the bag, well placed and well centred with anterior capsular opacification. There was no posterior capsular opacity, but one-third of both the anterior and the posterior of the IOL were green in colour. (Figure 1). The centre was clear and so were the haptics. VA of the patient in this particular eye was same as recorded earlier in 2002, that is, 1.0 with –1.0 DS. Contrast sensitivity was not carried out. Captivatingly, the patient was unaware of the change in colour of IOL and there were no visual complaints.

In order to determine the cause of discoloration of IOL, she was inquired comprehensively. She was asked questions pertaining to the use of medication, systemic illness, and any invasive procedure especially indocyanine green angiography or fundus fluorescein angiography during the preceding years. Nothing affirmative was reported, which could be suspected as a cause for greenish colour of IOL.

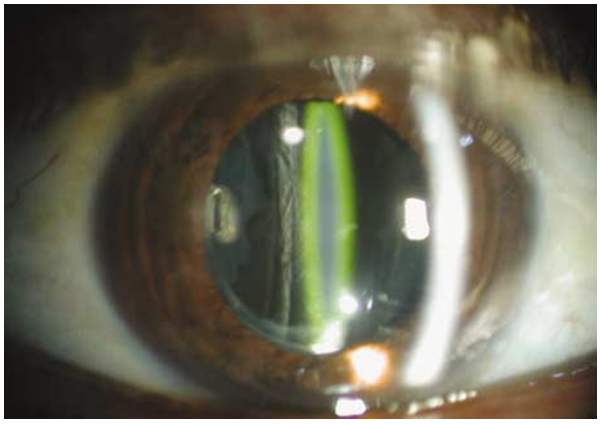

Figure 1 Slitlamp photograph of greenish discoloration of CeeOn 911A IOL.

Discussion

Even when the most common intraocular surgery is performed uneventfully, something unexpected can jump up into the scene. The case cited above is a unique example of 'an extraordinary development in an ordinary treatment'

Foldable IOLs are in use since mid-1980s. In this particular case, CeeOn 911A was implanted in the patient's eye that turned green in almost 3 years. CeeOn 911A is a UV-absorbing foldable lens having silicone optic and haptics made of polyvinylidene fluoride homopolymer (PVDF). Although large case series of silicone IOL implantation has demonstrated excellent visual outcomes,[1,2] various ophthalmologists reported cases of adverse events with silicone lenses. To the very best of our knowledge, **no case of greenish discoloration of CeeOn lens has ever been reported earlier**.

In 1991, Milauskas[3] reported brownish discoloration of varying intensity of silicone IOLs (manufactured by STAAR Surgical and IOLAB Corporation) in a series of 15 cases, 4–5 years after surgery. VA of all the patients under observation remained stable. At that time, Milauskas questioned the long-term stability of silicone IOLs in human eyes. Koch and Heit[4] in 1991 reported two cases of brownish discoloration of silicone IOLs. However, use of silicone IOLs in human eyes continues till today. Manufacturers claim use of newer and improved generation silicone material in these lenses. However, recently, Tanaka *et al*[5] reported brown haze in an Allergan SI-40NB silicone IOL on the very next day of surgery.

Exact causative factor has not yet been identified; few speculative mechanisms can, however, be addressed.