A case of spontaneously resolved primary congenital glaucoma

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Primary congenital glaucoma usually presents as enlarged and hazy cornea at birth or early childhood. The diagnosis is based on a thorough clinical examination under anesthesia. Most cases require surgical intervention as the definitive treatment. In very rare instances, primary congenital glaucoma may arrest and resolve spontaneously. We describe a case of spontaneously arrested and resolved primary congenital glaucoma in a 37-year-old male presenting with large cornea, Haab’s striae, and normal intraocular pressure in one eye. Such a case has not been previously described from the Indian subcontinent.

Key words: Arrested congenital glaucoma, congenital glaucoma, glaucoma, megalocornea, primary congenital glaucoma

Primary congenital glaucoma is a rare cause of childhood blindness.[1] The disease is usually manifested at birth or early childhood (before 3 years of age). Only a few cases of spontaneously resolved primary congenital have been reported previously.[2‑6] Herein, we report a rare case of spontaneously arrested primary congenital diagnosed in an adult.

Case Report

A 37-year-old male visited our center for a routine eye examination. His chief complaint was decreased vision in the right eye for which he was using spectacles. He gave a history of a larger right eye since early childhood.

On examination, his best corrected visual acuity was 6/9 (+1.00Dx160) OD and 6/6 OS. The intraocular pressures on Goldman applanation tonometer were 11 mmHg OD and 13 mmHg OS. The right eye cornea was larger with a diameter of 13.5 mm when compared with 11.75 mm for the left eye [Fig. 1]. Anterior segment examination revealed the presence of Haab’s striae in the nasal half of the right eye cornea, which was otherwise clear [Fig. 2]. Gonioscopy revealed prominent iris processes in the right eye [Fig. 3]. The anterior segment examination of the left eye was normal. The cup disc ratio was 0.3 for the right eye optic disc and 0.2 for the left eye optic disc. The posterior segment examination was normal. Anterior segment optical coherence tomography was done which also demonstrated the Haab’s striae [Fig. 4]. The central corneal thickness was thinner in the right eye at 449 microns, than 500 microns in the left eye. Visual field testing [Fig. 5] and optical coherence tomography - retinal nerve fiber layer [Fig. 6] demonstrated defects in the right eye.

On further examination of history, the patient revealed that the color of the right eye was initially whitish in early childhood.
Figure 2: Anterior segment photograph right eye shows the presence of Haab’s striae in the nasal half of the cornea and along the nasal limbus.

Figure 3: Gonioscopy right eye demonstrates open angle and prominent iris processes (block arrow).

Figure 4: Optical coherence tomography of cornea in the region of Haab’s striae.

Figure 5: Visual fields shows minimal and early defects in the right eye and a normal visual field in the left eye.

Figure 6: Disc photographs and corresponding optical coherence tomography - retinal nerve fiber layer scans both eyes. Reduced retinal nerve fiber thickness is observed in the right eye supero-temporal and infero-temporal quadrant.

Discussion

Only a few cases of spontaneously resolved primary congenital glaucoma have been reported previously.[2‑6] Nagaö et al. reported a series of 14 eyes with spontaneously resolved primary congenital glaucoma.[3] At the time of diagnosis, large corneas were observed in all affected eyes and Haab’s striae in 10 of 14 eyes. Intraocular pressures were normal in all 14 eyes. Mastropasqua et al. reported novel confocal microscopic changes in an adult eye with megalocornea and regressed congenital glaucoma.[6] These findings included reduction of keratocyte density in the mid- and rear-stroma, an abnormal “clew-shaped” morphology of stromal nerves, and the presence of hyperreflective structures overhanging the endothelial layer at the level of the Descemet membrane. Severe polymegathism, pleomorphism, and a markedly decreased cell density were observed in the endothelium. The present case which got restored to normal on its own. A diagnosis of right eye spontaneously resolved primary congenital glaucoma was made, and further follow-up advised.
had enlarged cornea, Haab’s striae, and normal intraocular pressure. Continued postnatal development and maturation of the drainage angle might be a mechanism for spontaneous resolution. Reversal of optic disc cupping with normalization of intraocular pressure is a well-known phenomenon in primary congenital glaucoma. Long-term follow-up is important for these eyes, due to subtle angle abnormalities and the possibility of later development of glaucoma.

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Conflicts of interest
There are no conflicts of interest.

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