

Grand Rounds

A 29-year-old man with bilateral megalocornea

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History

A 29-year-old man presented at the Department of Health Eye Center, East Avenue Medical Center, Quezon City, for assessment of intermittent right-sided headache of 3 months' duration and gradual progressive blurring of vision in the right for the previous year. He had never been examined by an ophthalmologist. He had no hypertension or diabetes but did have a 10-year history of hyperthyroidism, with thyroid hormone levels presently controlled. There was no history of ocular trauma. Family history was unremarkable for other health conditions.

Examination

Best-corrected visual acuity in the right eye was 20/160 ($-2.75 -1.50 \times 120$); in the left eye, 20/20 ($-0.25 -1.75 \times 120$). Slit-lamp biomicroscopy showed bilateral symmetrical enlarged clear corneas, with a horizontal diameter of 14 mm in both eyes and a vertical diameter of 13 mm in the right eye and 12.5 mm in the left eye (Figure 1).

Anterior chamber examination was unremarkable, with no fibrillary material, iris atrophy, Krukenberg spindles, inflammatory/pigment cells, or midperipheral iris transillumination defects. Anisocoria was not present and the right eye pupil was atropinized after surgery. There was prominent iridodonesis and phacodonesis in both eyes (Video 1). Both lenses were clear and centered. Initial untreated intraocular pressure (IOP) was 56 mm Hg in the right eye and 18 mm Hg in the left eye. Four-mirror indentation gonioscopy showed 360° of open angles to ciliary body bands in both eyes, with more pigmented posterior trabecular meshwork (PTM) in the right eye (+4) compared with the left eye (+2). See Figure 2. There was no exophthalmos. Optic disc examination

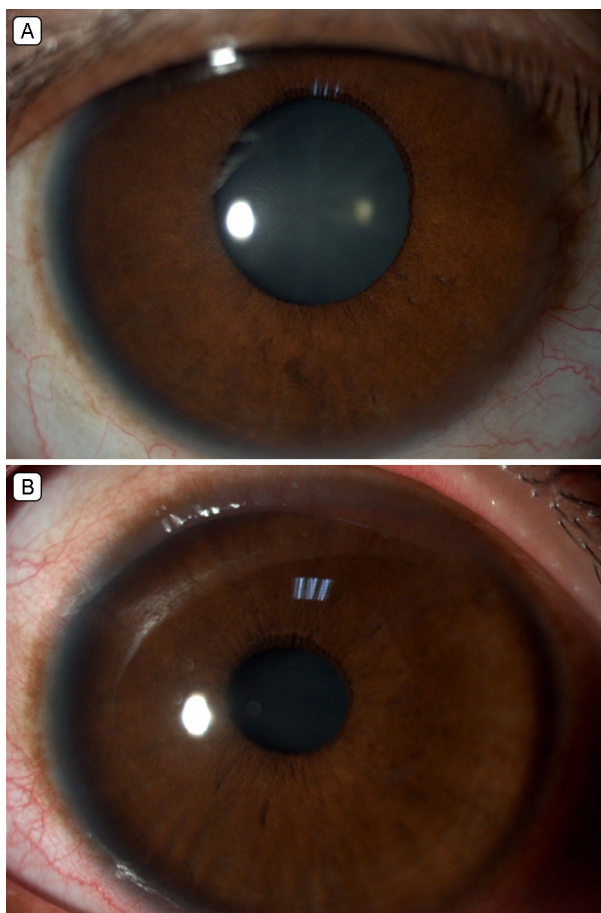


Figure 1. Anterior segment photographs. A, Right eye, after placement of glaucoma drainage device, showing an enlarged cornea, with a horizontal diameter of 14 mm (the pupil was dilated postoperatively). B, Left eye at presentation showing an enlarged cornea with a 14 mm horizontal diameter.

revealed advanced optic nerve cupping in the right eye and a normal disc in the left eye (Figure 3).

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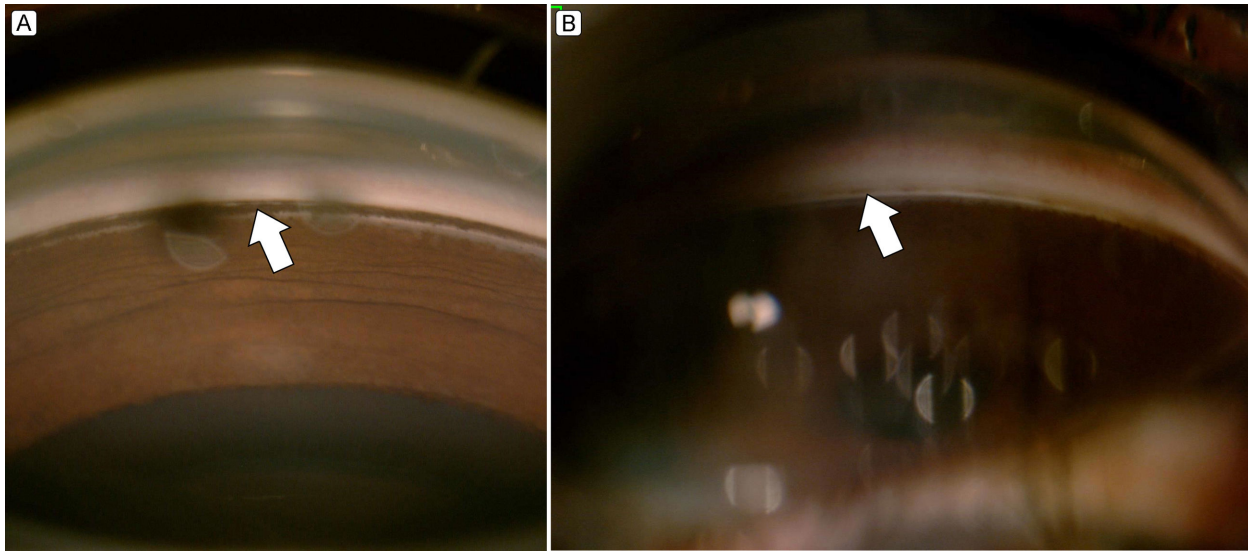


Figure 2. A, Gonioscopy of the right eye showing +4 posterior trabecular meshwork pigmentation (arrow). B, Gonioscopy of the left eye showing +2 posterior trabecular meshwork pigmentation (arrow).

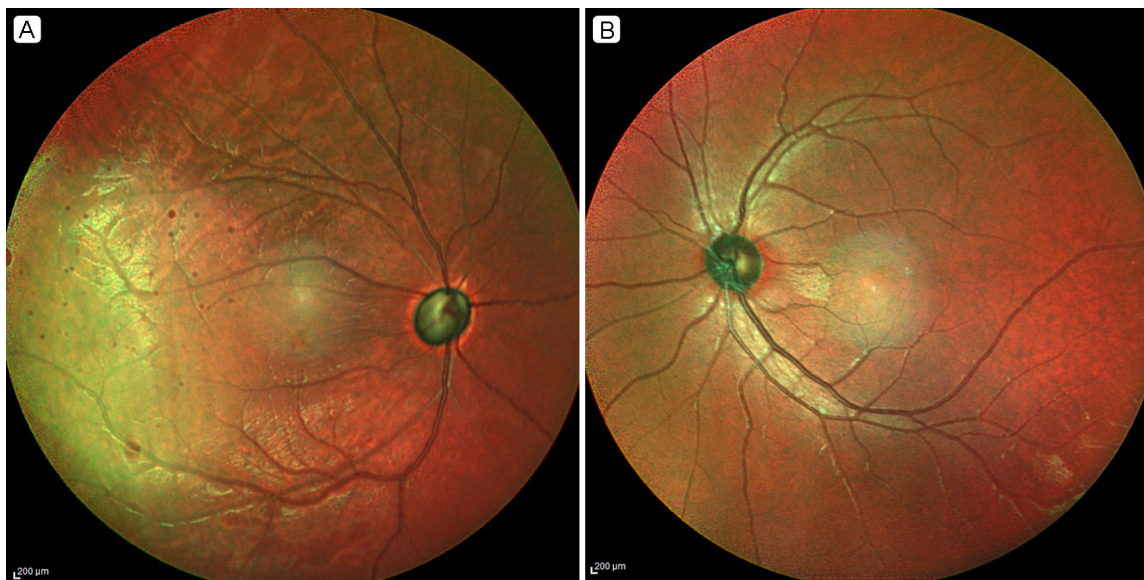


Figure 3. Multicolor optical coherence tomography imaging. A, Right eye, showing advanced glaucomatous optic neuropathy. B, Left eye, showing a normal optic disc.

Ancillary Testing

Perimetry in the right eye showed a severely constricted field (Figure 4A); the left eye showed no visual field defect (Figure 4B). Optical coherence tomography of the left eye showed no thinning of the retinal nerve fiber layer. Corneas were thin (right eye, 466 μm ; left eye, 465 μm), with normal endothelial cell counts in both

eyes. Ultrasound biomicroscopy (UBM) revealed deep anterior chambers in both eyes, posterior midperipheral iris bowing, with prominent and long hyper-reflective zonules (Figure 5). Anterior chamber depths were above normal (right eye, 4.72 mm; left eye, 5.14 mm) as measured using UBM. Axial lengths of both eyes were above normal (right eye, 25.3 mm; left eye, 25.7 mm).

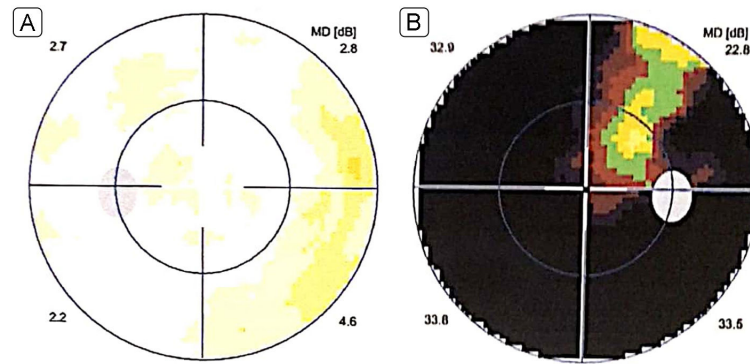
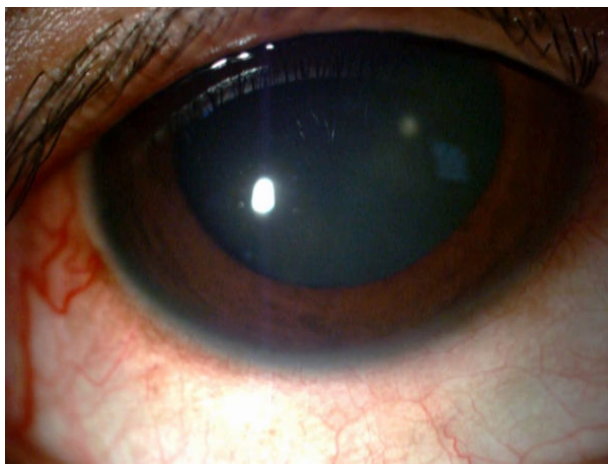


Figure 4. Perimetry performed after definitive management (surgery and laser iridotomy). A, Left eye, showing a normal visual field. B, Right eye, showing a severely constricted field sparing a superotemporal island.



Video 1. Slit-lamp examination of the right eye revealed very prominent iridonesis and phacodonesis, seen after each blink. This was also observed in the left eye.

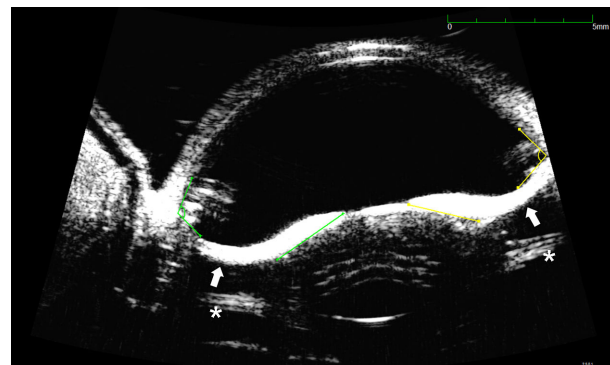


Figure 5. Ultrasound biomicroscopy of the left eye showing peripheral posterior bowing (arrows) of the iris prior to laser peripheral iridotomy, with hyperechoic zonules (asterisks).

Treatment

Because of advanced glaucoma with uncontrolled IOP despite maximal medical therapy, the patient eventually underwent aqueous drainage device implantation (Auro-lab, Madurai, India; see Figure 6). Laser peripheral iridotomy (LPI) in the left eye reversed the posterior bowing of the peripheral iris.

Differential Diagnosis

Anterior megalophthalmos with megalocornea, pigment dispersion (pigmentary) glaucoma, and infantile glaucoma were in the differential diagnosis for the right eye.

Diagnosis and Discussion

Megalocornea, a rare nonprogressive enlargement of a normal cornea to 13 mm can occur, according to Waring et al,¹ in three patterns: (1) simple megalocornea unassociated with other ocular abnormalities, (2) anterior megalophthalmos with megalocornea, and (3) iris and angle abnormalities as well as buphthalmos in infantile glaucoma. Our case was associated with other ocular findings, and the enlargement of both corneas was probably congenital, but because they were clear, the corneal enlargement was most likely nonprogressive. These findings rule out infantile or congenital glaucoma. The recent symptoms of headache and blurring of vision in the right eye that prompted consultation were probably due to the high IOP and progressing glaucoma. Anterior megalophthalmos is characterized by bilateral congenital enlargement of the corneas and anterior segments; increased anterior chamber depth in both eyes was

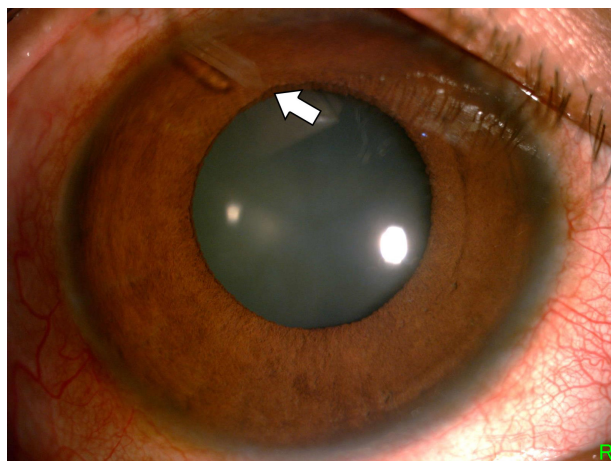


Figure 6. Anterior segment photograph of the right eye 1 week after glaucoma drainage device insertion showing the tube visible superotemporally (arrow).

observed in our patient.² Anterior megalophthalmic eyes present with a very deep anterior chamber and a vitreous index (vitreous length/axial length \times 100) below 69%.³ Our patient had a vitreous index of 60.75% in the right eye and 60.39% in the left eye.

Bilateral iridodonesis and phacodonesis in our patient was probably due to the zonular stretching caused by enlarged ciliary rings. The unilateral advanced glaucoma in the right eye (with unilateral hyper-pigmented PTM), without any of the other typical characteristics of pigment dispersion syndrome, was noteworthy, as was the lack of evidence of glaucoma in the contralateral eye, despite both eyes presenting with megalocornea, phacodonesis, and iridodonesis. We had difficulty making a definitive diagnosis of pigmentary glaucoma in the right eye, and we were uncertain whether the left eye would

develop glaucoma. During initial presentation with high IOP, there were no signs of active pigment dispersion. We hypothesize that the glaucoma in the right eye was probably due to another secondary open-angle glaucoma etiology, such as an abundance of trabecular meshwork mesenchymal tissue or goniodysgenesis⁴; however, we could not entirely rule out a pigment dispersion component because of the hyperpigmented PTM in the right eye. Weak zonules could cause angle closure glaucoma by forward movement of the lens-iris diaphragm, but the angles were wide open in our case. Our patient has been doing well with the implant in the right eye, with IOP of 18 mm Hg without glaucoma medication. After more than 1 year, the vision in the right eye has deteriorated because of cataract development.

This case highlights the fact that patients with megalocornea should be monitored for glaucoma, especially those with other ocular findings. LPI was performed in the left eye, even though we were unsure whether this was a PDS case, because this was the patient's good (glaucoma-free) eye, and the patient was very young. On UBM of the left eye, the reverse pupillary block seemed to have reversed after LPI.

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