Management of cataract in a patient with anterior megalophthalmos

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DESCRIPTION

A healthy 40-year-old man presented with reduced (counting fingers) right eye visual acuity (VA) due to a dense unilateral cataract (posterior subcapsular and nuclear sclerosis) (**figure 1A**). Examination revealed bilateral enlarged but thin corneas and deep anterior chambers (ACs) (**table 1**). Bilateral iridodonesis and phacodonesis were noted with no significant subluxation of the right crystalline lens. Iridocorneal angles were open bilaterally with normal intraocular pressures (12 and 14 mm Hg, right and left, respectively) (**figure 1C**). Corneal diameters were 14.5 mm/14.5 mm and 15 mm/15 mm, right and left, respectively, in horizontal and vertical meridians (**figure 1D**). There was no view of the fundus in the right eye, but the left eye posterior segment was normal without stigmata of vitreoretinal degeneration. B-scan ultrasonography excluded major right eye posterior segment pathology. Megalocornea, extremely deep AC and zonular laxity, without sequelae of congenital glaucoma (ie, raised intraocular pressure and optic neuropathy), suggested a diagnosis of anterior megalophthalmos (AM). Cataract surgery for visual improvement was discussed and consented for by the patient.

Cataract extraction in AM presents a number of surgical challenges. Extreme AC depth makes access to the cataract difficult, but this was bypassed by supracapsular prolapse of the lens nucleus out of the capsular bag also minimising shear stress on the weakened zonules of Zinn. Intraocular lens (IOL) choice in such a large eye was another challenging factor, and a +13.00 dioptre Rayner Superflex IOL (12.5 mm diameter, Rayner Intraocular Lenses Limited, UK) was implanted in the capsular bag with reverse optic capture technique. VA on the first postoperative day was 6/6, which was maintained at

Figure 1 (A) Slit lamp image of the anterior segment of the right eye showing a dense cataract in an eye with an exceptionally large cornea (megalocornea) and very deep anterior chamber. (B) Slit lamp image of the left eye showing megalocornea and deep AC with mild cataract. (C) Gonioscopy image of the right eye iridocorneal angle confirming a normal, open angle. (D) Vertical corneal diameter of the right eye, measured at 14.5 mm.

6-month review. This is the first case report of AM in Pakistan.

First reported in 1914, AM is defined as megalocornea with deep AC without abnormal axial length.¹ Elongation of the ciliary ring causes iris-lens-diaphragm instability (ie, phacodonesis and iridodonesis), predisposing to lens subluxation (crystalline lens and IOLs).² IOL choice is critical in such a large eyes and deep ACs with weak, stretched zonules. The main lens issues in AM are instability of standard diameter IOLs (ie, 13 mm) within the abnormally large capsular bag, lack of optic centration and postoperative posterior dislocation of IOLs.^{3–5} Cases of IOL dislocation with endocapsular fixation have been described.³ Multiple IOL strategies have been employed, including iris-claw

Table 1 Biometric parameters of the eyes in this anterior megalophthalmos case

Parameters	Right eye	Left eye	Normal range
Axial length	24.16 mm	24.19 mm	22–26 mm
AC depth	6.39 mm	5.81 mm	1.5–4 mm
Lens thickness	4.00 mm	4.04 mm	4 mm
White to white distance	15.0 mm	14.5 mm	11–12 mm
Central corneal thickness	475 µm	493 µm	555 µm
K1	45.2 D	44.3 D	42.00–44.00 D
K2	47.2 D	47.9 D	42.00–44.00 D
Preoperative refraction	Precluded by cataract	−2.50 DS/−0.50 DC × 120	−6.00 D to +3.00 D

The axial lengths (ultrasound for the right eye), anterior chamber depths, lens thickness, white to white distance, central corneal thickness and keratometry (K1 and K2) as measured by IOLMaster 700 V.1.14 (Carl Zeiss Meditec Ag, Jena, Germany) and refraction (in dioptre sphere, DS, and dioptre cylindrical, DC) are shown relative to normal reference ranges. AC, anterior chamber.

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Learning points

- Consider a diagnosis of anterior megalophthalmos in the presence of megalocornea, ultradeep anterior chamber and zonular laxity (ie, iridophacodonesis, ectopia lentis) without signs of congenital glaucoma or increased axial length.
- Preoperative diagnosis allows appropriate surgical planning to ensure appropriate intraocular lens (IOL) choices and promote optimal refractive and visual outcomes while avoiding IOL dislocation into the vitreous cavity.
- The surgeon must consider intraoperative challenges in anterior megalophthalmos, including accessing the cataract in an ultradeep anterior chamber, baseline zonular instability with difficult capsulorhexis and IOL considerations, in addition to standard cataract risks.

IOLs,⁶ iris-sutured IOLs and custom-made larger-diameter IOLs.⁵⁻⁷ AM is associated with vitreous abnormalities (eg, syneresis, synchysis and optically empty vitreous with strands)⁸ and vitreoretinal pathology (eg, lattice degeneration, associated with retinal detachment); thus, annual dilated fundal examination is recommended.⁸ Other notable ocular associations include anterior embryotoxon, mosaic corneal dystrophy, glaucoma due to goniodysgenesis, irregular astigmatism, and amblyopia due to strabismus and myopia.⁹ Mutations in the Xq12-q26 region have been implicated previously.¹⁰ Systemic associations include Marfan syndrome, trisomy 21, Apert syndrome, mucopolidosis type 2 and Walker-Warburg syndrome.¹¹

Recognition and diagnosis of AM allows planning for the monitoring of the posterior segment and preparation for the unique challenges of cataract surgery in the complex anterior segment.

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