Microspherophakia with secondary glaucoma

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Introduction
Microspherophakia is characterized by an increased anteroposterior diameter and a reduced equatorial diameter of the crystalline lens leading to a spherical configuration. It is seen either as an isolated anomaly or along with systemic disorders like Weill–Marchesani syndrome, Marfans disorder, Alports syndrome, Klinefelter syndrome, and Mandibulofacial dystostosis.1 We report a case of isolated microspherophakia with secondary angle closure glaucoma.

Case
A 23-year-old woman presented with complaints of gradually progressive diminution of vision in the left eye since 1 year. It was associated with occasional headache. She was on topical intraocular pressure (IOP) lowering agents for the past 5 months.

The best corrected visual acuity in the right eye was 6/18 [−6.00 diopter sphere/−1.00 cylinder at 40°] and 6/24 [−6.00 diopter sphere/−1.25 cylinder at 70°] in the left eye. IOP recorded with Goldmann applanation tonometer was 20 and 40 mmHg for the right and left eye, respectively.

Slit lamp examination showed a shallow anterior chamber [Van Herick grade 2] in both eyes. Gonioscopy revealed 360° of angle closure in both eyes (Fig 1.1). Relative afferent pupillary defect was noted in the left eye. Ultrasound biomicroscopy (Fig 1.2) revealed a decreased anterior chamber depth [1.3 mm OD, 1.5 mm OS], an increased lens thickness [4.3 mm OU], and a reduced equatorial lens diameter [6.3 mm OD, 6.2 mm OS] suggestive of microspherophakia.2 Peripheral laser iridotomy

Figure 1.1: (a) Gonioscopy showing synechial angle closure. (b) Slit-lamp photo showing shallow central anterior chamber.

Figure 1.2: UBM of left eye showing reduced equatorial lens diameter and shallow anterior chamber.
was performed in both eyes. The optic nerve head was healthy in the right whereas there was advanced cupping in the left eye (Fig 1.3). There was advanced field loss in the left eye (Fig 1.4). Since IOP was not under medical control she was advised surgical intervention in the left eye. Post-dilation, lens edge was visible all around (Fig 1.5)

**Discussion**

Microspherophakia is clinically characterized by a triad of angle-closure glaucoma, small spherical crystalline lens, and lenticular myopia. Visual compromise in patients with microspherophakia is attributed to refractive error or secondary glaucoma. Mechanism of glaucoma is due to pupillary

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**Figure 1.3:** Optic nerve head (a) Right eye with a healthy disc. (b) Left eye showing bipolar rim thinning.

**Figure 1.4:** Humphrey’s visual field.

**Figure 1.5:** Lens edge visible all around after dilation.
block and/or peripheral anterior synechiae. Acute angle closure can result from pupillary block due to anterior dislocation of the crystalline lens when associated with weak zonules. Such recurring pupillary block attacks result in synechial angle closure resulting in chronic angle closure. Initial treatment consists of relieving the pupillary block component by laser iridotomy followed by medical management of elevated IOP. Indications for surgery include poor control of IOP and/or lens dislocation. Scleral fixation of the intraocular lens may be required.

References

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