Complicated microspherophakia in a paediatric patient

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DESCRIPTION
An adolescent boy presented with pain, redness, watering, photophobia and loss of vision in both eyes for 1 month. His visual acuity was light perception both eyes (OU), and the intraocular pressures were 40 and 30 mm Hg in right and left eye respectively. The right eye had a microspherophakic cataractous lens in the anterior chamber with corneal decompensation and endothelial scarring (figure 1A). Ultrasonography revealed optic nerve head cupping in the right eye. The left eye had a clear microspherophakic lens dislocated anteriorly with lenticulo-endothelial contact (figure 1B), ciliary staphyloma and total glaucomatous cupping. A diagnosis of bilateral microspherophakia with secondary glaucoma and corneal decompensation was made. The patient underwent OU intralenticular lens aspiration under general anaesthesia under mannitol cover. Postoperatively, there was no improvement in vision possibly due to advanced glaucoma; however, the patient had symptomatic relief from pain and photophobia.

Microspherophakia is a congenital disorder, characterised by a decreased equatorial diameter and an increased anteroposterior diameter of the crystalline lens and variable degree of subluxation of lens due to zonular weakness. It may present as an autosomal dominant, autosomal recessive or isolated entity. Generally, it presents with high lenticular myopia, lens dislocation or subluxation (45%) and secondary glaucoma (51%).

Mechanism of glaucoma in microspherophakia includes pupillary block secondary to high anterior lens curvature, chronic pupillary block leading to peripheral anterior synechiae and possible angle dysgenesis. A study by Lim et al emphasised that more ‘micro-spheric’ the lens, the more mobile it is and hence, have higher probability of anterior subluxation and associated glaucoma. Thus, lenticular equatorial diameter inversely varied with the severity of lens-induced complications. In our case too, the right eye had a smaller lens (equatorial diameter <6.5 mm) compared with the left eye (figure 2A,B), and a worse clinical presentation with a calcified degenerated lens.

Learning points
► Microspherophakia, a developmental disorder of the crystalline lens, is associated with spectrum of complications that include lenticular myopia, lens subluxation or subluxation and secondary glaucoma. Secondary glaucoma is an irreversible cause of vision loss.
► Smaller the equatorial lens diameter, greater are the chances of complications, its severity and poorer visual outcomes.
► Timely removal of the lens with visual rehabilitation and glaucoma management are necessary to attain optimal visual outcomes in microspherophakic eyes.
Nagata et al have objectively described changes in the shape of lens in microspherophakia with affected lens fibres and induced hyaline degeneration.\(^3\)

To conclude, early diagnosis and timely management are essential to optimise the final visual outcomes in such eyes.

REFERENCES