CASE REPORT

Hereditary anterior megalophthalmos with posterior vitreoretinopathy: a surgical challenge

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SUMMARY

Anterior megalophthalmos is an inherited condition characterised by enlargement of the anterior segment with associated glaucoma and vitreoretinopathy. These associations make surgical management very challenging. A 12-year-old boy, diagnosed and operated elsewhere for congenital glaucoma in both eyes, presented with raised intraocular pressure. Investigations revealed enlarged corneal diameter, increased anterior chamber depth with thick, clear cornea and no Haab striae while fundus evaluation revealed posteriorly dislocated cataractous lens with total rhegmatogenous retinal detachment in both eyes. The surgical challenges in management are highlighted in this case where appropriate investigations can aid in the selection of appropriate surgeries with good visual outcome.

BACKGROUND

Anterior megalophthalmos is often misdiagnosed as primary congenital glaucoma which is the most common differential of a child with increased corneal diameter and raised intraocular pressure (IOP). Recognition of associations, complications and differentiating features help us decide appropriate surgeries in such cases thereby ensuring long-term success and good visual outcomes.

CASE PRESENTATION

A 12-year-old boy presented with a 3-day history of loss of vision in the right eye. He had undergone bilateral trabeculectomy for primary congenital glaucoma diagnosed elsewhere 6 years ago. His best corrected visual acuity was accurate projection of rays in the right eye and 20/100 in the left eye with applanation IOP of 3 and 34 mm Hg, respectively. His cornea was enlarged and clear with corneal diameter measuring 15 mm and no Haab striae or limbal stretching in both eyes and an anterior chamber depth (ACD) of 4.7 and 4.9 mm in the right and left eyes, respectively. Both eyes had patchy iris atrophy with pin-point pupil in the right eye (figure 1A,C). Gonioscopy showed enlarged ciliary band in both eyes. Fundus examination showed bilateral posteriorly dislocated cataractous lens with total superior rhegmatogenous retinal detachment (RD) and lattice. Cup disc ratio was 0.9:1 with bipolar notch in the right and 0.8:1 with superior rim thinning in the left eye.

Review of family history revealed similar findings in the patient's elder sister with staphylomatous globe in her right eye.

INVESTIGATIONS

Axial length and corneal thickness measured were 28.27 and 26.57 mm, 661 and 628 μ , in the right and left eye, respectively. An orb scan did not reveal any signs of corneal thinning or aberrant curvature. Humphrey visual field was unreliable in the left eye.

DIFFERENTIAL DIAGNOSIS

The enlarged thick cornea and associated iris, angle and posterior segment anomalies, bilateral posterior dislocated cataractous lens with family history pointed to a diagnosis of hereditary anterior megalophthalmos which is distinct from primary congenital glaucoma.

TREATMENT

With a final diagnosis of anterior megalophthalmos, he was taken up for sequential (right followed by left) pars plana lensectomy, vitrectomy and silicon oil in both eyes. Postoperatively IOP was raised which mandated silicon oil removal and fluid gas exchange in both eyes. His IOP still remained high at 32 and 38 mm Hg in both eyes, which prompted maximal medical treatment with topical β -blockers, α -agonists, systemic glycerol and acetazolamide inhibitors. Persistently raised IOP despite maximum medical management, oil removal and fluid gas exchange 2 weeks after surgery with progressive increase in optic nerve head cupping, indicated need for definitive glaucoma surgery in both eyes.

OUTCOME AND FOLLOW-UP

Significant conjunctival scarring with shallow ACD due to overfilled gas in the right eye (figure 1B) warranted glaucoma drainage device (non-valved, aurolab aqueous drainage implant, tube was covered with corneal patch graft and fibrin glue) in the right eye. Intraoperatively, the gas was partially removed by paracentesis allowing chamber deepening. The tube was left unligated in view of intraocular residual gas which acted as a tamponade by itself obviating the use of ligature to prevent postoperative hypotony. In view of young age, relatively free conjunctiva in superotemporal quadrant necessitated limbus-based trabeculectomy with mitomycin C (0.02%, subconjunctival application for 3 min) in the left eye (figure 2) with no intraoperative or postoperative problems.

OUTCOME AND FOLLOW-UP

Postoperatively, he regained a vision of 20/80 with attached retina, no postoperative hypotony and IOP of 12 and 14 mm Hg in both eyes at 9-month follow-up.

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Figure 1 Slit lamp photograph of the right (A) and left eyes (C) postvitreoretinal surgery showing enlarged corneal diameter with aphakia, patch iris atrophy and small pupil. (B) Intraoperative photograph of the right eye showing superotemporal glaucoma drainage implant placement with residual gas in the area shown (black arrow).

Figure 2 Postoperative slit lamp photograph at 2 months showing patent tube with slit pupil (A) and raised diffuse bleb in the left eye (B) after limbus-based trabeculectomy with Mitomycin C.

DISCUSSION

Anterior megalophthalmos is an inherited condition characterised by bilateral enlargement of the anterior segment structures including ciliary ring with associated lens subluxation/dislocation and vitreoretinopathy.^{1 2} Since stretching was confined to the anterior segment, there were no evident signs of posterior segment stretching (seen as chorioretinal atrophy or myopic changes in the fundus) in our case. Bilateral dislocated lens signifies enlargement of the ciliary ring commonly seen in this condition.^{3 4} Ocular biometry and corneal thickness helped us arrive at a correct diagnosis in our case which was wrongly diagnosed as primary congenital glaucoma elsewhere.^{3 5}

Goniodysgenesis in these cases can lead to secondary glaucoma which can be challenging to treat in view of other associations.^{1 3–7} Bilateral RD with posteriorly dislocated lens mandated definitive retinal surgery with injection of silicon oil with resultant raised IOP despite previously operated trabeculectomy. In our case, persistently raised IOP even after oil removal warranting glaucoma surgery signified additional angle dysgenesis responsible for glaucoma which was exacerbated with use of tamponading agents like silicon oil. These agents may be best avoided in such cases to avoid added insult to the trabecular meshwork.

Glaucoma surgery in patients undergoing retinal surgery can be very challenging due to conjunctival scarring, presence of silicon oil and young age of the patient.^{8 9} High chances of failure in such cases may warrant drainage implant as performed in the right eye while concerns for bilateral drainage implant at such a young age prompted us to try trabeculectomy in the other eye with relatively free conjunctiva.

Tube ligation is a recommended procedure to prevent postoperative hypotony in non-valved implants.^{8–10} In our case, shallow ACD in a gas filled eye suggested that the gas itself would help prevent hypotony thereby obviating tube ligation as recommended for non-valved implants. Yet, some gas had to be released for deepening of the chamber. Though these modifications have to be purely based on clinical findings on table and not as a standard, modifications tailored to the findings on table help us ensure successful outcomes even in challenging cases.

Learning points

- ▶ Anterior megalophthalmos can have associated features of ciliary ring enlargement, iris atrophy and subluxated or posteriorly dislocated lens with or without vitreoretinopathy with absence of limbal stretching or Haab striae that are typical of primary congenital glaucoma.
- ▶ Appropriate associations in posterior segment may warrant multiple vitreoretinal surgeries with tamponading agents. Silicon oil may cause secondary raised intraocular pressure in such cases with pre-existing goniodysgenesis and may be avoided in favour of other tamponading agents unless crucial for successful attachment of retina.
- ▶ A correct diagnosis and appropriate selection of glaucoma surgery in anterior megalophthalmos can help achieve successful outcomes despite visually threatening ocular associations and complications resulting in multiple surgeries.

Competing interests None.

Patient consent Obtained.

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REFERENCES

- 1 Kuchenbecker J, Baumann WB. Ciliary body dysplasia in megalophthalmos anterior diagnosed using ultrasound biomicroscopy. *Eye* 2002;16:638–9.
- 2 Ahmadi H, Banaee T, Javadi MA, et al. Vitreoretinal disorders in anterior megalophthalmos. *Jpn J Ophthalmol* 2006;50:515–23.
- 3 Vail DT. Adult hereditary anterior megalophthalmos sine glaucoma: a definite disease entity with special reference to the extraction of cataract. *Arch Ophthalmol* 1931;6:39–62.
- 4 Dufour R, Scouras J, Gailloud CL. Megalophtalmie, subluxation cristallienne et cataracte: décollement rétinien. *Ophthalmologica* 1971;165:409–16.
- 5 Meire FM, Delleman JW. Biometry in X linked megalocornea: pathognomonic findings. *Br J Ophthalmol* 1994;78:781–5.
- 6 Meire FM. Megalocornea. Clinical and genetic aspects. *Doc Ophthalmol* 1994;87:46–52.

- 7 Javadi MA, Jafarinasab MR, Mirdehghan SA. Cataract surgery and intraocular lens implantation in anterior megalophthalmos. *J Cataract Refract Surg* 2000;26:1687–90.
- 8 Gedde SJ, Schiffman JC, Feuer WJ, *et al.* Treatment outcomes in the tube versus trabeculectomy (TVT) Study After Five Years of Follow-up. *Am J Ophthalmol* 2012;153:789–803.
- 9 Tribble JR, Brown DB. Occlusive ligature and standardized fenestration of a Baerveldt tube with and without antimetabolites for early postoperative intraocular pressure control. *Ophthalmology* 1998;105:2243–50.
- 10 Gedde SJ, Herndon LW, Brandt JD, *et al.* Postoperative complications in the Tube Versus Trabeculectomy (TVT) study during five years of follow-up. *Am J Ophthalmol* 2012;153:804–14.

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