CASE REPORT

Challenges in the implantation of a Boston type 1 keratoprosthesis and a glaucoma drainage device in a nanophthalmic eye

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SUMMARY

A 67-year-old man presented with hand motions vision, a decompensated vascularised cornea and a flat anterior chamber in the right eye 5 years following cataract and glaucoma surgery. He lost the left eye several years ago following a similar intervention. The intraocular pressure (IOP) was 19 mm Hg on four antiglaucoma medications and the axial length was 19 mm. In view of the high risk for graft failure, a Boston type 1 keratoprosthesis (Kpro) was planned for visual rehabilitation, and a glaucoma drainage device (GDD) for IOP control. Although a combined GDD with Kpro was indicated, we performed sequential surgeries to avoid vision threatening hypotony-related complications. He underwent pars plana vitrectomy, prophylactic lamellar sclerectomies and Kpro at the first stage, followed by a paediatric Ahmed glaucoma valve implantation 3 months later. Despite precautions, postoperative uveal effusion occurred but could be managed conservatively. His IOP was controlled; visual acuity improved to 20/50 and remained stable at 2 years.

BACKGROUND

Nanophthalmos or simple microphthalmos is a condition in which the total axial length of the eyeball is two SDs below the mean for age or less than 20 mm.^{1 2} In contrast to microphthalmos, a nanophthalmic eye is small in overall dimensions but not associated with developmental abnormalities.¹ The condition is characterised by short axial length, a shallow anterior chamber, narrow angles, thick sclera, macular hypoplasia and a normal-sized lens.¹

Owing to the large lens to anterior chamber ratio, nanophthalmic eyes are associated with a high incidence of angle closure glaucoma. The thick sclera in these eyes impedes vortex venous drainage and reduces the transscleral flow of protein from the eye contributing to spontaneous or postoperative choroidal congestion, uveal effusion and choroidal detachment.

It is known that cataract and glaucoma filtering surgeries in nanophthalmic eyes are associated with high rates of vision threatening intraoperative and postoperative complications such as severe uveal effusion, serous or haemorrhagic choroidal detachment, malignant glaucoma and corneal decompensation.^{1 2}

Although penetrating keratoplasty helps in restoring clear optical media in eyes with corneal decompensation, a shallow anterior chamber coupled with

high IOP in nanophthalmic eyes frequently leads to graft failure. Keratoprosthesis (Kpro) would be a better alternative for visual rehabilitation in these eyes. It is also known that a Boston type 1 Kpro is associated with a high incidence of glaucoma, and in an eye with pre-existing glaucoma, it would only worsen. Implantation of a glaucoma drainage device (GDD) is useful in controlling the intraocular pressure (IOP) in eyes with Kpro. Although a combined GDD with Kpro is a recommended option in eyes with pre-existing glaucoma, we performed sequential surgeries to avoid vision threatening hypotony-related complications in the nanophthalmic eye. The risk of sight threatening complications associated with intraocular interventions could be decreased with appropriate planning and meticulous sequential surgeries in these eyes.

CASE PRESENTATION

A 67-year-old man presented with decreased vision in his only seeing right eye due to secondary glaucoma and corneal decompensation. Visual acuity in the right eye was hand motions with accurate projection of rays and no light perception in the left eye. He had lost his left eye to complications following a surgical intervention for glaucoma. He had a flat anterior chamber following cataract and glaucoma surgery in the right eye, which resulted in corneal decompensation. Anterior segment examination of the right eye showed an oedematous vascularised cornea with epithelial bullae, a flat anterior chamber with 270° peripheral anterior synechiae and a posterior chamber intraocular lens. His left eye was phthisical with occlusiopupillae and a mature cataract. The right eye IOP was recorded as 19 mmHg on three topical and one oral antiglaucoma medications, and the left eye was phthisical with unrecordable IOP.

INVESTIGATIONS

The axial length was 19 mm in the right eye and 18 mm in the left eye. The ultrasound B scan showed a thickened choroid and sclera (2.4 mm) with attached retina in the right eye and a closed-funnel retinal detachment in the left eye. Anterior segment optical coherence tomography (ASOCT) was performed serially during the postoperative follow-up visits.

TREATMENT

As a part of staged procedure, two 4×4 mm anterior lamellar sclerectomies with open sclerectomies

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in the inferotemporal and inferonasal quadrants were performed. The iris tissue, intraocular lens and the capsule were excised and removed, pars plana vitrectomy was performed and a customised paediatric (7 mm back plate) aphakic Boston type I Kpro was implanted. As a measure to avoid serious intraoperative complications, special precautions were taken, including preoperative intravenous 20% mannitol (1 g/kg body weight) to decrease the vitreous volume, general anaesthesia to avoid excess retro-orbital pressure due to peribulbar anaesthesia and prophylactic lamellar sclerectomies and sclerectomies to prevent uveal effusion. The sclerectomies and sclerectomies were performed in a similar technique described by Wax *et al*³ obviating the need for vortex vein decompression.

Three months after Kpro, a silicone paediatric Ahmed glaucoma valve (AGV; FP8) was implanted under general anaesthesia, in the superotemporal quadrant for IOP control. A valved drainage implant was chosen to avoid severe postoperative hypotony and its associated complications, and a paediatric implant was chosen in view of a short eyeball. Intraoperative modifications in the surgery included a tight AGV tube entry (23 gauge) into the anterior chamber to prevent peritubular leakage, a longer tube in the anterior chamber for better visibility through the central optical cylinder of Kpro, and the use of tissue adhesive (Tissel, Baxter, Westlake Village, California, USA) to glue the scleral patch graft to prevent tube erosion and anterior chamber reformation with a high viscous viscoelastic to prevent hypotony.

OUTCOME AND FOLLOW-UP

One month after implantation of the Boston type 1 Kpro (figure 1), the visual acuity improved from hand motions to 20/50. Glaucomatous optic disc damage, a 0.8 cup with a very thin inferior rim (figure 2), was noted and the visual fields showed a superior arcuate defect. The high IOP post-Kpro was managed with three topical antiglaucoma medications (a combination of dorzolamide 2%+timolol maleate 0.5% and brimonidine tartarate 0.1%) and supplemental oral acetazolamide. The plan was to perform the second surgery after 3–4 weeks; however, owing to logistical reasons and at the patient's request, it was performed 3 months later. Post-AGV implantation, there was minimal serous choroidal detachment, which subsided with conservative management. The visual acuity stabilised at 20/50, and the IOP was digitally normal with a well-placed tube (figure 3) and good bleb. Two years post-Kpro and AGV, the patient is enjoying 20/50 vision with a digitally normal IOP on two topical antiglaucoma medications (a combination of

Figure 1 One-month postoperative slit lamp photograph showing a well-placed Boston type 1 keratoprosthesis.

Figure 2 Fundus photograph of the right eye showing a 0.8 cup with an inferior thin rim and pallor.

brimonidine 0.2% and timolol maleate 0.5%), a stable optic disc and a visual field.

DISCUSSION

We report a case with a sequential Boston type 1 Kpro and AGV in a nanophthalmic eye; to the best of our knowledge, this has not been reported so far. There is one case report on sequential cataract surgery followed by a paediatric AGV in a nanophthalmic eye, which helped to achieve a stable IOP and vision.⁴

Any intraocular surgery in a nanophthalmic eye comes with its inherent risk. This risk is further increased following glaucoma surgery as lowering the IOP changes the globe dynamics and integrity. Sudden decompression of the globe during surgery may trigger massive uveal effusion, which may be associated with exudative retinal detachment, suprachoroidal effusion/haemorrhage, malignant glaucoma and corneal decompensation. The margin of safety is very narrow and these postoperative complications may lead to permanent visual loss. Any surgical intervention in these eyes needs meticulous planning and implementation to avoid serious intraoperative and postoperative complications.

Figure 3 Postoperative slit lamp photograph showing a patent tube behind the optical cylinder (arrow) 1 month after Ahmed valve implantation.

Singh *et al* first reported the results of intraocular surgery in nanophthalmos. They found that 60% of the 15 patients who underwent glaucoma filtration surgery failed to achieve IOP control and 86.6% suffered visual loss.⁵ Yalvac *et al*² in their series of 28 eyes with mitomycin C trabeculectomy and inferior sclerotomy reported uveal effusion in 50% of cases. In our case, the technique described by Wax *et al* helped to prevent intraoperative choroidal detachment and open sclerotomies helped in resolution of uveal effusion in the early postoperative period.

Managing glaucoma post-Kpro is very challenging as IOP measurement with currently available tonometers is not possible in these eyes with artificial corneas. Various tonometers like tonopen and the diaton transpalpebral tonometer have been tried; however, none provide a reliable measurement. Currently, the tactile estimation of IOP is the most practical method that provides fairly reliable IOP estimation in experienced hands.

Conventional filtering surgery is not an option in eyes with Kpro. Transscleral cyclophotocoagulation is a useful adjunct to

control IOP; however, the unpredictability in IOP control, postoperative inflammation and uveal effusion are concerns and risks have to be weighed before considering it in a nanophthalmic eye. Owing to better safety and predictability, the obvious choice in our case was a valved GDD for IOP control. Post-Kpro and GDD, owing to the limited anterior chamber and angle view in these eyes, serial ASOCT can be performed to assess the angle, the retroprosthetic membrane and to evaluate the position of the drainage tube.

AGV being a valved implant, theoretically should not be associated with postoperative hypotony. However, sudden decompression, intraoperative hypotony and peritubular leakage could lead to hypotony even in eyes with a valved implant and the problem would be compounded by severe uveal effusion in nanophthalmic eyes. The postoperative complications could, to a great extent, be prevented by appropriate preoperative and intraoperative precautions and meticulous surgical technique. Despite all the precautions, postoperative choroidal effusion can occur. However this can be managed conservatively with lesser severity.

Learning points

- ▶ Implantation of paediatric Boston type 1 keratoprosthesis could be considered for visual rehabilitation in eyes with nanophthalmos and severe corneal disease.
- ▶ Great caution should be exercised in managing coexisting glaucoma in these patients. Meticulously planned sequential intraocular procedures may help to prevent serious vision threatening complications.
- ▶ A valved paediatric glaucoma drainage implant is a good option in nanophthalmic eyes.
- ▶ Appropriate preoperative precautions such as intravenous mannitol, general anaesthesia, and intraoperative modifications such as prophylactic lamellar sclerectomies with full-thickness sclerotomies could prevent serious complications such as severe choroidal effusion, suprachoroidal haemorrhage and exudative retinal detachment.

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