Bilateral buphthalmos with congenital cataract in a floppy infant

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Key words: Buphthalmos, congenital anomaly, congenital cataract, glaucoma

Congenital glaucoma and congenital cataracts are well-known entities; however, their coexistence is rarely reported. We describe a case of bilateral combined buphthalmos with congenital cataracts in a floppy infant and the challenges involved in its management.

A 7-month-old male child was brought by his mother with complaints of whitish discoloration of both eyes, noticed since birth. The child had a global developmental delay and was not making any eye contact. The perinatal period was uneventful. The child was born out of a non-consanguineous marriage at full term by normal vaginal delivery and vaccinated until age. On examination, the vital parameters were stable. The central nervous system examination revealed a floppy infant with no neck holding. Other systems were within normal limits. There were no facial abnormalities. On ocular examination, the child has a blink response to light; however, the fixation to light was poor, unsteady, and not maintained. Bilateral enlarged, edematous corneas with central cataracts were noted [Fig. 1]. The intraocular pressure measured by the iCare rebound tonometer was 28 mm Hg in the right eye and 30 mm Hg in the left eye. Fundal glow was poor in both eyes due to the media opacity. Ultrasound B-scan showed anechoic vitreous with retina on. Then, the child was diagnosed with bilateral buphthalmos with congenital cataracts. Serology was negative for rubella, CMV, and toxoplasma. The urine test for reducing substances was negative. Serum calcium, phosphorous, electrolytes, renal and liver function tests were within normal limits. Echocardiography and ultrasound abdomen were within normal limits. The child was started on oral acetazolamide 10 mg thrice a day, and 0.25% Timolol eyedrops BD in both eyes and was planned for a two-staged surgical intervention. Evaluation under general anesthesia (EUGA) and bilateral trabeculectomy and trabeculotomy was followed by bilateral lens aspiration with posterior continuous curvilinear capsulorhexis and limited anterior vitrectomy in the second sitting. On EUGA, under sevoflurane, the intraocular pressure measured by Perkins's tonometer was 28 mm Hg and 30 mm Hg in the right and left eye, respectively. The corneal diameter measured was 11.5 mm vertically and 12.0 mm horizontally in the right eye, and 13.0 mm both horizontally and vertically in the left eye. Axial length measured by ultrasound A-scan was 20.82 mm and 20.79 mm in the right and left eyes, respectively. Bilateral trabeculotomy and trabeculectomy were done, and the child was managed with dorzolamide-timolol eyedrops TDS in both eyes along with other routine postoperative medications. Four weeks later, the cornea was noted to be clearer, and the anterior chamber was well-formed with a mildly elevated bleb [Fig. 2]. The intraocular pressure measured by the iCare tonometer was 9 mm Hg in both eyes. Subsequently, on EUGA under sevoflurane, IOP measured by Perkin's tonometer was 9 mm Hg and 7 mm Hg in the right and left eyes. Bilateral lens aspiration with posterior continuous curvilinear capsulorhexis and limited anterior vitrectomy was performed, and the child was left aphakic. On follow-up after 6 weeks, the child was following light, the visual axis was clear, and the intraocular pressures were maintained at 9 mm Hg in both the eyes [Fig. 3]. The child was prescribed aphakic spectacles and is on regular follow-up.

Discussion

Primary congenital glaucoma is a rare disease due to genetically determined abnormalities in the trabecular meshwork and anterior chamber angle, resulting in elevated intraocular pressure (IOP) without other ocular or systemic developmental anomalies. The coexistence of congenital glaucoma with congenital cataracts is a rare occurrence and has been reported in conditions such as congenital rubella syndrome, persistent hyperplastic primary vitreous (PHPV), and Peter's anomaly.[1,2] Infants with congenital rubella syndrome with microphthalmos may present with a raised intraocular pressure with an apparently normal-sized cornea. Both cataracts and glaucoma can also develop in disorders such as oculo-cerebral-renal syndrome of Lowe (OCRL or Lowe syndrome), Patau syndrome, and homocystinuria.[3,4] Also, complicated cataract develops following glaucoma surgery, and secondary glaucoma is a well-known complication after pediatric cataract surgery. Although congenital glaucoma has a more widespread angle malformation, these disorders may be associated with a partial abnormal angle development and a better prognosis.[2] The buphthalmic eye poses several challenges during cataract surgery. Clear corneal incisions must be used to prevent damage to the conjunctiva, sclera, and bleb area. A high viscosity ophthalmic viscosurgical device must be used to counter the vitreous upthrust. Low scleral rigidity, elastic anterior capsule, and instability of zonules make capsulorhexis difficult.[5,6] Trypan blue dye can migrate through the zonules and obscure the fundal glow. Possible variations in axial length measurements due to variations in intraocular pressure may affect the intraocular (IOL) power calculations.[7] The postoperative myopic shift and risk of retinal detachment may be exaggerated in children with glaucoma. The enlarged

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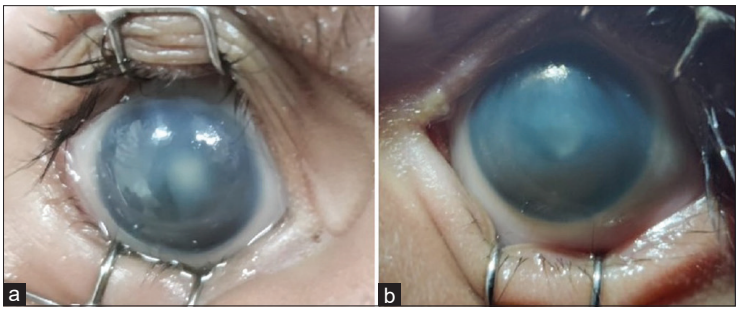

Figure 1: (a and b) Preoperative image of both eyes showing bilateral buphthalmos with central cataracts

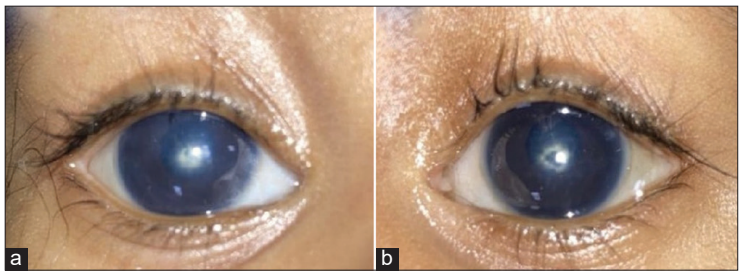

Figure 2: (a and b) Anterior-segment image of right and left eyes showing central cataract resolution of corneal edema following trabeculotomy with trabeculectomy

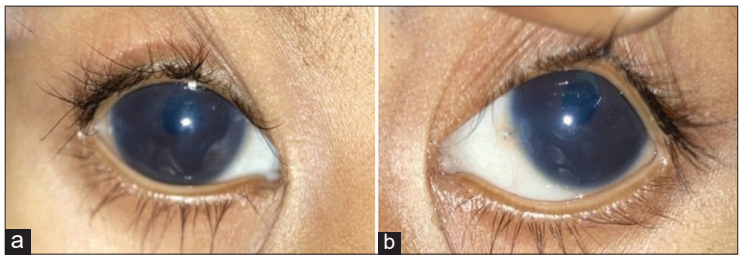

Figure 3: (a and b) Anterior segment image of right and left eyes showing clear cornea with aphakia following cataract surgery

capsular bag, expansion of the eye with raised intraocular pressure, and excessive capsular fibrosis pose an additional risk of decentration of an intraocular lens. For IOL stability and long-term centration, it is advisable to use a capsular tension ring, perform an optic capture, rhexis fixation or not perform a posterior capsulotomy in the same sitting.[8] Meticulous surgery is needed to prevent decontrol of IOP.

Buphthalmos with congenital cataracts may coexist in certain developmental anomalies. Ophthalmologists must be aware of the surgical challenges encountered in these cases. A two-stage bilateral simultaneous surgical approach is an effective treatment option.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Reddy UN, Chacham S, Rao JN, Priya AV, Mahmood A, Nagasravani J, *et al*. A rare case of buphthalmos and bilateral nuclear cataracts in a neonate with congenital rubella syndrome. J Evolut Med Dent Sci 2014;3:6965-69.
2. Khan AO. Buphthalmos in the setting of persistent hyperplastic primary vitreous cataract. Am J Ophthalmol 2003;136:945-7.
3. Ma X, Ning K, Jabbehdari S, Prosseda PP, Hu Y, Shue A, *et al*. Oculocerebrorenal syndrome of Lowe: Survey of ophthalmic presentations and management. Eur J Ophthalmol 2020;30:966-73.
4. Bunting R, Leitch J. Buphthalmos in trisomy 13. Eye 2005;19:487-8.
5. Sukhija J, Kaur S, Pandav SS, Kaushik S, Raj S, Ram J. Pediatric cataract surgery in buphthalmos. Eur J Ophthalmol 2015;25:260-2.
6. Morales-Fernández L, Martínez-de-la-Casa JM, Benito-Pascual B, Saénz-Francés F, Santos-Bueso E, Arriola-Villalobos P, *et al*. Cataract extraction in patients with primary congenital glaucoma. Eur J Ophthalmol 2020;30:525-32.
7. Khokhar S, Yadav D, Gupta S, Sihota R, Chaurasia AK, Gupta A, *et al*. Refractive outcomes of cataract surgery in primary congenital glaucoma. Eye (Lond) 2019;33:542-8.
8. Gupta PC, Singh RR, Pandav SS, Ram J. Rhexis fixation of intraocular lens in a case of buphthalmos with cataract. Nepal J Ophthalmol 2016;8:71-3.