Cataract surgery in child with congenital microphthalmos and orbital cyst
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An 8-year-old boy presented with low vision in his left eye and no vision in his right eye since birth. The diagnosis was bilateral microphthalmos with orbital cysts and a dense cataract in the left eye. To improve the vision in the left eye, cataract extraction and anterior vitrectomy were performed. After the surgery, the visual acuity improved from light perception to counting fingers. During the follow-up of 3 months, no severe complications occurred. To our knowledge, this is the first case of cataract surgery in a case of microphthalmos with orbital cysts.

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Microphthalmos with orbital cyst is a rare, severe developmental abnormality of the eye that results from defective closure of the embryonic fissure at an early stage. The condition is usually unilateral. Treatment may consist of observation only or, in severe cases, may involve surgical removal of the globe and/or cyst. We report a case in which cataract extraction in the better eye of a child presenting with bilateral microphthalmos with orbital cysts improved the vision.

CASE REPORT

An 8-year-old boy complained of low vision in his left eye and no vision in his right eye since birth. It was said that the vision in the left eye had deteriorated 4 years earlier. Heart surgery had been performed the previous year because of congenital heart disease. Otherwise, the child was healthy and had no family history of any similar developmental disease.

The head and face had an abnormal appearance (Figure 1, A). The corrected distance visual acuity (CDVA) was no light perception in the right eye and light perception in the left eye. The right eye was displaced superiorly under the upper eyelid by a large cyst, which made the right eye nearly invisible and also caused lower eyelid ectropion (Figure 1, B). The left eye had enophthalmos and nystagmus. Slitlamp examination of the anterior segment showed a microcornea, a formed anterior chamber, temporal iris coloboma, posterior synechia, peripheral anterior synechia, and a dense and calcified cataract (Figure 1, C). The binocular fundus could not be visualized. Ultrasonography showed severe bilateral microphthalmos (12.7 mm in diameter of the right eye and 13.6 mm of the left eye) and large orbital cysts (27.6 mm in diameter in the right orbit and 11.7 mm in the left orbit). Choroidal coloboma and tractional retinal detachment were also detected in the left eye by ultrasonography. Ultrasound biomicroscopy of the left eye showed a thickened cornea (800 μm), a normal-depth anterior chamber, peripheral anterior synechia, a closed anterior chamber angle, and an organized anterior capsule. Computed tomography confirmed bilateral microphthalmos with orbital cysts, a small left orbit, and craniofacial skeletal dysplasia (Figure 2).

Based on clinical and imaging findings, the bilateral microphthalmos with orbital cysts was diagnosed. To improve the vision in the left eye, cataract extraction, posterior capsulotomy, and anterior vitrectomy were performed. The corneal diameters, measured during surgery, were 4.0 mm in the right eye and 8.0 mm in the left eye. Following surgery, the boy’s CDVA improved to counting fingers (Figure 1, D). The CDVA remained stable during the postoperative following-up of 3 months.

DISCUSSION

Microphthalmos with orbital cyst is a congenital abnormality caused by a defect in the closure of the embryonic fissure at the 7 mm to 20 mm stage of development during weeks 6 to 7 of gestation. The cysts project through a congenital defect in the wall of the microphthalmic eye and are lined by a neuroectoderm. The condition is usually unilateral; bilateral cases are relatively rare. Microphthalmos with orbital cyst,
especially bilateral microphthalmos and orbital cysts, can be associated with systemic abnormalities involving the cardiovascular system, central nervous system, pulmonary tract, digestive tract, and the kidney.\textsuperscript{1} In our case, the patient with bilateral microphthalmos and orbital cysts also had craniofacial skeletal dysplasia and congenital heart disease.

To our knowledge, the treatment of microphthalmos and orbital cyst has focused primarily on achieving good cosmesis because the visual potential is poor.\textsuperscript{1,3–6} We found no report of cataract surgery performed in these patients. In our case, both eyes had microphthalmos and an orbital cyst, so care was taken to prevent binocular blindness. Cataract extraction, posterior capsulotomy, and anterior vitrectomy were performed in the better eye. The visual acuity in that eye improved from light perception to counting fingers.

We decided not to implant an intraocular lens (IOL) for several reasons. First, according to the rough result of ultrasound using the Haigis formula,\textsuperscript{7} the IOL power required was approximately 42.0 diopters, which was unavailable. Second, the anterior and posterior capsules could not be separated. Therefore,
the IOL could not be implanted in the capsular bag. Third, because of the temporal iris coloboma, posterior synechia, and peripheral anterior synechia, the IOL could not be stably implanted in the sulcus, which might cause corneal endothelial decompensation and secondary glaucoma.

The potential complications of cataract surgery in microphthalmos and orbital cyst have been rarely reported, although some studies have shown complications of cataract surgery in nanophthalmos such as cystoid macular edema, uveal effusion, retinal detachment, choroidal hemorrhage, malignant glaucoma, and vitreous hemorrhage.8,9 Nanophthalmos is a rare ophthalmic condition characterized by small eyes, shallow anterior chambers, narrow angles, a high lens to eye volume ratio, thick sclera, short axial length, and moderate to high hyperopia.10 During surgery, a sudden decrease in intraocular pressure may result in uveal effusions, secondary retinal detachment, vitreous hemorrhage, or malignant glaucoma.11 It has been postulated that the thick sclera seen in nanophthalmos impedes venous drainage by the vortex system.10 We believe that different mechanisms may exist in nanophthalmos and microphthalmos with orbital cyst. Nanophthalmos is an eye that is anatomically intact, while microphthalmos with orbital cyst is a colobomous small eye. In microphthalmos and orbital cyst, the sclera is not thick. Therefore, these patients may not be prone to develop uveal effusion.

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