Cataract surgery in a case with bilateral iridolenticular coloboma associated with microphthalmos and relative anterior microphthalmos

Nilgün Solmaz, MD, Feyza Önder, MD

We present the case of a 35-year-old woman with bilateral congenital coloboma of the iris and the lens. In the right eye, an approximately 130-degree zonular absence and excessive notching of the lens and associated microphthalmos were present and in the left eye, zonular deficiency from 7 to 10 o'clock and relative anterior microphthalmos. Cataract surgery was performed in the left eye first. Phacoemulsification was uneventful, but it was almost impossible to rotate the 3-piece acrylic intraocular lens (IOL) in the capsular bag. Because of the additional zonular dialysis in the inferior quadrant, a Cionni capsular tension ring (CTR) was inserted with transscleral fixation, achieving capsular bag stabilization and IOL centration. In the right eye, the equatorial contour was improved by placing a classic CTR following hydrodissection. Subsequently, lens aspiration and implantation of a 1-piece acrylic IOL were completed without complication despite the large zonular defect.

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Lenticular coloboma is a rare congenital anomaly characterized by notching in the crystalline lens at the equator due to segmental zonular absence.1,2 It is often associated with uveal coloboma and microphthalmos or relative anterior microphthalmos.1,2 Glaucoma1 and retinal detachments with giant retinal tears1,2 are seen more frequently in eyes with coloboma, and cataract development occurs at relatively young ages.1–5

Eyes with congenital coloboma are at greater risk for complications during cataract surgery. Zonular failure increases the risk for vitreous loss,4–6 and notching in the equator complicates intraocular lens (IOL) implantation and centration in the capsular bag.6 In addition, the eccentric pupillary aperture and associated pathologies make cataract surgery more complex. The literature on the surgical experience with the colobomatous eye is limited due to the condition’s infrequent occurrence.3–10 In recent years, several authors have reported that capsular tension ring (CTR) implantation has reduced the risk for complications and made the cataract surgery safer.6–9,11 We report our surgical experience and the challenge faced in the management of a case with bilateral iris and lens coloboma associated with microphthalmos in the right eye and relative anterior microphthalmos in the left eye.

CASE REPORT

A 35-year-old woman presented to our clinic with complaint of gradual deterioration of vision. The corrected distance visual acuity (CDVA) was 20/70 in both eyes. The slitlamp examination revealed bilateral cataract with iris and lens coloboma associated with microcornea, shallow anterior chamber, stromal iris atrophy, and ectropion uveae on the pupillary margins (Figure 1). On gonioscopy, the anterior chamber angles were open and intraocular pressure (IOP) was 19 mm Hg in the right eye and 20 mm Hg in the left eye.
eye. The horizontal corneal diameter and the mean keratometry were 10.0 mm and 51.5 diopters (D), respectively, in the right eye and 10.5 mm and 50.0 D, respectively, in the left eye. The axial length (AL) and anterior chamber depth (ACD) measurements with ultrasonic biometry showed the presence of microphthalmos in the right eye (AL, 19.64 mm; ACD, 1.83 mm) and relative anterior microphthalmos in the left eye (AL, 24.48 mm; ACD, 2.03 mm). The corneal endothelial cell count was 4167 cells/mm² and 3460 cells/mm² in the right eye and left eye, respectively. Retinochoroidal coloboma was not detected on examination of the fundus. There was also no systemic anomaly except a 1.0 mm ptosis of the left eyelid.

Cataract surgery was performed under general anesthesia. The IOP was reduced with intravenous mannitol 20% infusion preoperatively. The less malformed left eye was operated on first. Slow-motion phacoemulsification with low fluid and low vacuum parameters was completed without problems, but it was almost impossible to rotate the 3-piece acrylic IOL (Acrysof MA60AC 25.5 D, Alcon Laboratories, Inc.) in the capsular bag during implantation. The iridolenticular synechia in the colobomatous region broke out and additional zonular dialysis occurred in the lower quadrant while the surgeon was attempting to place the upper haptic. A Cionni CTR (type 2C, Morcher GmbH) was implanted with transscleral fixation at the 9 o’clock position with no vitreous loss. This facilitated IOL rotation but not centration. By locking 1 of the IOL haptics under the loop of the Cionni CTR, IOL centration was possible in the field of the eccentric pupil (Figure 2, B).

In the right eye, 2 paracenteses were made at the 10 o’clock and 12 o’clock positions and a dispersive ophthalmic visco-surgical device (OVD) was injected over the colobomatous area. A 2.4 mm clear corneal tunnel incision was made at the 10 o’clock position and continuous curvilinear capsulorhexis was performed using the Utrata forceps. A classic CTR (type 14, Morcher GmbH) was inserted following hydrodissection. The lens nucleus was very soft and was aspirated manually with low fluid flow for fear that excessive fluid passage could seep into the vitreous during phacoemulsification. The zonular defective area was often tamponaded with the dispersive OVD to prevent vitreous prolapse. A 1-piece acrylic IOL (Acrysof SA60AT 28.0 D, Alcon Laboratories, Inc.) was implanted using an injector and centered with the haptics positioned at the 1 o’clock and 7 o’clock positions (Figure 2, A).

Postoperatively, the CDVA improved to 20/50 (−1.50 −0.50 × 140) in the right eye and to 20/32 (−1.75 D) in the left eye. The limited increase in visual acuity in the right eye was linked to amblyopia. Although pupilloplasty was not performed, the patient did not complain of photophobia or glare. No problems developed during the 12-month follow-up period.

DISCUSSION

In eyes with lens coloboma, cataract surgery becomes challenging because of the higher risk for complications.4–9,10 In these eyes, complications such as vitreous herniation into the anterior chamber, zonular dialysis extension, capsular fornix aspiration, and IOL decentration are frequent outcomes.4,5 In addition, there is the risk for intraoperative malignant glaucoma resulting from positive pressure from microphthalmos and excess fluid transition into the vitreous.5 The development of phacoemulsification techniques and CTRs have improved the surgical results.3,5 Capsular tension ring usage protects against capsular bag collapse and prevents passage of excessive fluid behind the cystoid macular edema.
In conclusion, cataract surgery in eyes with coloboma requires extra care and planning at each stage. Capsular tension ring implantation decreases the risk of complications and facilitates cataract surgery in eyes with lenticular coloboma. However, iris coloboma repair is more problematic. Not insisting on closure of iris defects in microphthalmic eyes with lens coloboma and avoiding methods with risks for complication may be more appropriate.

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lens, expanding the capsular bag on the vitreous. In addition, even if phacoemulsification is completed uneventfully, forming the capsule contour is also necessary for IOL rotation and centration. Although there was no significant notching at the lens equator in the left eye in our case, the 3-piece IOL could not be rotated in the capsular bag. This shows that even small equatorial irregularities may prevent the movement of the haptics, especially hard haptics.

Capsular tension ring implantation can be performed before and after phacoemulsification depending on the width of the zonular defect. Implantation of a CTR before phacoemulsification is preferred because it facilitates nucleus rotation and protects against capsular fornix aspiration and collapse. We also inserted the CTR after hydrodissection and completed the surgery uneventfully in the right eye despite the large coloboma.

In eyes with coloboma, the closure of iris defects depends on the size of the defect and the patient’s degree of preoperative photophobia, particularly prior to the progression of the cataract. Several surgical techniques to repair iris coloboma have been described. In addition, postoperative anterior capsule fibrosis or cosmetic contact lenses can overcome photophobia and glare symptoms. We also considered that the anterior capsule fibrosis would provide an adequate functional and cosmetic result and that there would be no need for pupilloplasty in the left eye. In the right eye, several alternatives to close the iris defect were evaluated. We initially thought of implanting a partial aniridia ring such as the Morcher type 94G instead of the classic CTR. This was not done because we could not predict the degree of expansion of the capsular bag diameter. Iris sutures could not be performed as the colobomatous region was too wide. However, the patient did not suffer from photophobia or glare in the postoperative period.

In conclusion, cataract surgery in eyes with coloboma requires extra care and planning at each stage. Capsular tension ring implantation decreases the risk for complications and facilitates cataract surgery in eyes with lenticular coloboma. However, iris coloboma repair is more problematic. Not insisting on closure of iris defects in microphthalmic eyes with lens coloboma and avoiding methods with risks for complication may be more appropriate.