CASE REPORT

Lens coloboma treated with lens surgery

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SUMMARY

A 5-year-old boy was referred to our clinic due to an abnormal visual acuity test at school. His corrected visual acuity was counting fingers in the left eye. A nasal side deficiency of the lens substituted by a membrane was found. Lens coloboma was diagnosed. After making a 3 mm limbal incision, the colobomatous lens was removed by anterior continuous curvilinear capsulorhexis and lens aspiration. Posterior capsulorhexis and anterior vitrectomy on the side of the lens was performed to prevent posterior capsular or anterior hyaloid opacity. As the defect in the lens was very large, intracapsular placement of an intraocular lens was not feasible. A three-piece acrylic soft intraocular lens was placed in the ciliary sulcus. Since amblyopia was diagnosed by poor corrected visual acuity as 20/800 1 month after the operation, occlusion therapy with correcting eyeglasses was started at 6 h a day on the contralateral eye. The patient's corrected visual acuity improved to 20/125 7 months after the operation.

BACKGROUND

Lens coloboma is defined as an anomaly of lens shape, and is mainly a result of defects in zonules and ciliary bodies during ocular development.¹ Lens coloboma is a congenital problem, and may accompany other ocular defects or systemic disorders. Surgical treatment is required for visual improvement. We present a case of lens coloboma treated successfully with a lens operation.

CASE PRESENTATION

A 5-year-old boy was referred to our clinic due to an abnormal visual acuity test at school. He was a healthy, well-developed boy with no systemic disease. His corrected visual acuity was 20/20 and counting fingers in the right and left eye. Intraocular pressure, anterior segment and fundus examination were normal in both eyes. No strabismus was found. A nasal side deficiency of the lens substituted by a membrane was found in the left eye (figure 1). A normal crystalline lens was noted in the contralateral eye. Lens coloboma was diagnosed.

TREATMENT

After making a 3 mm limbal incision, the colobomatous lens was removed by anterior continuous curvilinear capsulorhexis and lens aspiration. Posterior capsulorhexis and anterior vitrectomy on the side of the lens was performed to prevent posterior capsular or anterior hyaloid opacity. As the defect in the lens was very large, intracapsular placement of an intraocular lens was not feasible. A three-piece acrylic soft intraocular lens

Figure 1 Lens coloboma presenting as nasal side deficiency of lens.

(MA60MA, Alcon Labs, Texas, USA) was placed in the ciliary sulcus.

OUTCOME AND FOLLOW-UP

Mild temporal decentration of the intraocular lens was noted after the operation, without changes of intraocular lens position during follow-up (figure 2). Since amblyopia was diagnosed by corrected visual acuity as 20/800 fully corrected by myopia −3 dioptres and astigmatism −2.5 dioptres 1 month after the operation, occlusion therapy with correcting eyeglasses was started at 6 h a day on the contralateral eye. The patient's corrected visual acuity improved to 20/125 7 months after the operation.

DISCUSSION

Lens coloboma results from the failure of fetal fissure closure during ocular development, usually unilaterally. Once the zonule or ciliary body fails to develop, the tension is released and the lens contracts segmentally a notch. So it is actually a coloboma of the zonule rather than a defect in the lens itself.¹

Figure 2 Mild temporal decentration of a sulcus-placed three-piece acrylic soft intraocular lens.

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Lens coloboma occasionally appears in combination with other ocular defects, including iris, choroid or optic disc coloboma.²⁻³ Besides, there is also a higher incidence of cataract and retinal detachment in these patients. The retinal detachment commonly involves the nasal quadrant, and is the result of a ciliary body defect, which might induce abnormal adherence between lens and peripheral retina. Moreover, fluid might gain access to the subretinal area through the break in the ciliary body.¹ Apart from ocular defects, lens coloboma might be one of the manifestations of systemic diseases, including Marfan syndrome and Marshall syndrome.⁴⁻⁶ Several case reports have shown the association between Marfan syndrome and lens coloboma, and it is postulated that increased transforming growth factor β might be the link behind this.⁵ In our case, the patient had lens coloboma without an accompanying congenital cataract, and no other systemic diseases were noted.

In order to enhance surgical planning, a detailed imaging of the lens, zonulae and ciliary body is needed. High-resolution ultrasound biomicroscopy has earlier been used for in vivo study of the human zonular apparatus and lens. The use of Scheimpflug images can be obtained with a Pentacam to depict a more accurate view of the lens coloboma morphology and to anticipate the potential complications during surgery.⁷ Since our young patient did not cooperate, we could not perform a Pentacam examination.

Treatment of lens coloboma is mainly surgical but limited literature is available. Nordlund and colleagues reported seven cases of lens coloboma treated by phacoemulsification and intraocular lens placement. Six of the seven patients showed improvement of visual acuity and only one patient developed retinal detachment after surgery.⁸ The surgical management of a lens coloboma is frequently challenging, since there is a high risk of capsular fornix aspiration, zonular dialysis extension, vitreous herniation into the anterior chamber, intraocular lens decentration and closure of the capsular opening. Hence, the implantation of a capsular tension ring is preferred before the emulsification of the nucleus to prevent capsular bag deformity, and to enable nucleus rotation and avoid capsular collapse; thereby, reducing surgical complications. In another study by Gurler and colleagues, 13 cases of lens coloboma under treatment of phacoemulsification, along with capsular tension ring and intraocular lens placement, were presented. Significant improvement in visual acuity was noted after surgery without major complication. Thus, the paper concluded that phacoemulsification with capsular tension ring and intraocular lens implantation is a safe and effective option.⁹

In our case, the patient's family could not afford a capsular tension ring. Owing to the enormous defect in the lens, an intra-capsular intraocular lens placement was not feasible. Thus, we

chose a sulcus three-piece intraocular lens as an alternative. Posterior capsulotomy and anterior vitrectomy were also performed to avoid posterior capsular opacity, which is a common complication in children undergoing cataract surgery.¹⁰ In our patient, visual acuity only mildly improved due to prolonged deprivation amblyopia.

In summary, lens coloboma is a rare congenital disease with anomaly in lens shape. Surgical treatment can achieve visual improvement.

Learning points

- ▶ Lens coloboma is a defective lens as a result of defects in zonules and ciliary bodies during ocular development.
- ▶ Lens coloboma can be treated by removal of the defective lens.
- ▶ An intraocular lens can be placed in the capsular bag along with a capsular tension ring, or positioned in the ciliary sulcus.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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