Management of Bilateral Ectopia Lentis et Pupillae Syndrome

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Abstract. A 52-year-old patient presented with signs clinically consistent with ectopia lentis et pupillae syndrome. The patient was treated successfully with vitrectomy, dislocated lens removal using perfluorocarbon liquid and phacofragmentation in the vitreous cavity, pupil reconstruction, and scleral-fixed intraocular lens implantation in both eyes. Despite the fact that the surgery was successful in technical terms, the final visual outcome was not as good as expected. This was caused by the optic nerve atrophy resulting from long-lasting glaucoma. Nevertheless, the described surgical techniques may be considered an effective method of treatment in cases of ectopia lentis et pupillae syndrome. [Ophthalmic Surg Lasers Imaging 2006;37:68-71.]

INTRODUCTION

Ectopia lentis et pupillae is a rare congenital inherited disorder, whose main features include lens and pupil displacement without any coexisting systemic disorders.1 It is speculated that it appears as a result of a neuroectodermal defect or persistence of fetal vasculature.2 Combined neuroectodermal and mesodermal origin is also postulated, but the exact cause remains unknown.3

Manifestations of this disorder may also include severe axial myopia, poor vision, cataract (usually before the age of 40 years), abnormal iris transillumination, poor pupillary dilatation, persistent pupillary membrane, and iridohyaloid membranes.4-6 Other ocular complications may include glaucoma, intraocular hypertensive crises, uveitis, and retinal detachment.1 The disorder is inherited in an autosomal recessive mode and is usually bilateral.4,7

We describe a case of bilateral ectopia lentis et pupillae syndrome that was treated surgically in both eyes.

CASE REPORT

A 52-year-old white woman was referred to the Department of Ophthalmology, Medical University of Lodz, Poland, to undergo planned surgery. At the time of referral, the patient complained of poor vision and dissatisfaction with the appearance of her eyes (displaced pupils). She presented with congenital bilateral pupil displacement and bilateral lens luxation into the vitreous cavity.

The patient had been treated for glaucoma for 7 years. Bilateral spontaneous lens luxation had been diagnosed 20 years earlier. The patient confirmed that one member of her family (sister) also had congenital pupil displacement, which was not present in either her parents or children.

Her previous ophthalmological history included several episodes of uveitis, as well as several procedures performed in both eyes due to difficulties in controlling glaucoma with medication. The procedures included YAG laser iridotomy, cyclocryotherapy (three times), and gonioplasty in the right eye, and YAG laser iridotomy (four times), cyclocryotherapy (twice), and gonioplasty in the left eye. Her general medical history was unremarkable; there were no systemic diseases present. Prior to the surgery, the patient was treated...
with the following topical glaucoma medications (both eyes): 0.5% timolol maleate two times daily, dorzolamide hydrochloride three times daily, 2% pilocarpine four times daily, and latanoprost once per day. The intraocular pressure (IOP) was well controlled with topical antiglaucomatous therapy in the preoperative period.

Preoperative visual acuity was hand motions in the right eye and counting fingers in the left eye. The IOP measured by Goldmann applanation tonometry was 12.2 mm Hg in the right eye and 22.0 mm Hg in the left eye. Because of very narrow and displaced pupils, it was impossible to perform either retinoscopy or autorefractometry before the surgery. Therefore, the preoperative refractive error was estimated subjectively; the patient estimated that +10 D eyeglasses were the best correction for her.

The biomicroscopic examination revealed bilaterally transparent corneas with few small persistent precipitates on the endothelium. Pupils were very narrow (approximately 1 mm in diameter) and were displaced inferonasally. Their dilatation, even with repeated use of mydriatic drops, was unsuccessful and ophthalmoscopic examination was not possible. Several lesions were seen on the iris due to previous laser therapy (Fig. 1). Bilateral crystalline lens luxation to the vitreous cavity was confirmed by B-scan ultrasonography examination (Fig. 2).

Ultrasound biometry (A-scan) showed the axial length of 26.23 mm in the right eye and 25.72 mm in the left eye. Corneal curvature values measured with Javal keratometer were $K_1 = 44.25$ D, $K_2 = 43.05$ D in the right eye and $K_1 = 42.50$ D, $K_2 = 42.50$ D in the left eye. These data indicated axial myopia.
The surgeries were performed under local anesthesia in both eyes in the interval of 2 months. All of the steps of the procedure were similar in both eyes. Standard three-port pars plana vitrectomy was performed to completely excise the vitreous. A hole of approximately 5 mm in diameter was made in the center of the iris with the vitrectom tip, to create a new pupil. Perfluorocarbon liquid (DK-line) was then injected to elevate the crystalline lens into the retropupillary space, where phacofragmentation was performed with the US Oerlii phaco tip (2 mm diameter, 90° angle, no irrigation sleeve) (Oerlii Instrumente AG, Berneck, Switzerland). A posterior chamber, all-polyethylene acrylic intraocular lens was implanted into the ciliary sulcus using a simple, previously described outside-in method for scleral fixation.8

The power of implants was determined by ultrasound biometry and calculated for a posterior chamber intraocular lens using SRK II formula. The calculated values were +13.5 D in the right eye and +15.5 D in the left eye. The patient had implanted intraocular lenses of the following power: +15.0 D in the right eye and +16.0 D in the left eye.

Postoperative visual acuity without any correction was 1/60 in the right eye and 2/60 in the left eye. Time of follow-up was 2 months. The IOP was 12 mm Hg in both eyes without glaucoma medication. The postoperative refractive error measured by autorefractometry amounted to approximately -2.0 D sph -1.5 D cyl in both eyes.

On the slit-lamp examination, the pupils were somewhat oval in shape, centrally located, with slightly irregular margins. Intraocular lenses were correctly positioned (Fig. 3). The fundus examination revealed pale optic nerve disc with typical, severe glaucoma cupping. Retinal vessels and the macula appeared normal. Areas of chorioretinal atrophy were seen in the periphery.

DISCUSSION

Ectopia lentis et pupillae syndrome is generally thought to be inherited in an autosomal recessive mode. Neither our patient's parents nor her children suffered from this disorder; however, her sister did. This is in accordance with the above mentioned pattern of inheritance.

The patient fulfilled the criteria of ectopia lentis et pupillae syndrome, including displacement of pupils and cataractous crystalline lenses, axial myopia with myopic fundus abnormalities, poor pupillary dilatation, and poor vision. Moreover, the abnormalities were congenital and bilateral, with no signs of any systemic illness. She also had other associated ocular complications (i.e., glaucoma and uveitis).

Both lenses were luxated without any apparent cause when the patient was 32 years old, so it is a logical assumption that they must have been ectopic previously.

Pars plana vitrectomy has become an efficient method of dislocated lens management. The use of ultrasonic phacofragmentation and perfluorocarbon liquids allows for safe crystalline lens removal in the vitreous cavity and it minimizes the incision to the diameter of the intraocular lens optic. These techniques together with posterior chamber, scleral-fixated intraocular lens implantation create a complex surgical method that is effective in such cases.
Vitrectomy was used by Rossiter et al. for the treatment of secondary phacoalytic glaucoma in a patient with ectopia lentis et pupillae. In our case, the IOP also normalized after surgery and glaucoma medication was no longer necessary.

The surgery was successful in both eyes regarding luxated crystalline lens removal, implantation of a sclerally fixated intraocular lens, and pupil reconstruction. However, the final visual outcome was not as good as might be expected. This was due to the long-lasting glaucoma resulting in optic atrophy, which was impossible to evaluate prior to surgery because of very small and ectopic pupils. Despite this, the patient was satisfied with the results, which improved her visual abilities considerably. In our opinion, vitrectomy with phacoemulsification may be considered an effective method of treatment in cases of ectopia lentis et pupillae syndrome.

REFERENCES