Management of Bilateral Pupillary Membrane and Congenital Cataract in a Patient with Microphthalmos

Mikroftalmili Bir Hastada Pupiller Membran ve Konjenital Kataraktın Yönetimi

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ABSTRACT

In this report, we evaluated the surgical management and visual outcome in both eyes of a 14-month-old patient with multipl ocular pathologies including microphthalmos, microcornea, microcoria, pupillary membrane, and congenital cataract. Membrane excision and cataract extraction were followed by secondary posterior chamber intraocular lens (PC-IOL) implantation in both eyes. No intra- or post-operative complications were seen. The patient followed by regular ophthalmologic evaluation during 5 years and appropriate visual rehabilitation techniques were applied. Her most recent visual acuity was 20/100 in both eyes. Our experience with this patient suggests that regular ophthalmic examination, refractive correction and visual rehabilitation besides selection of appropriate surgical method can be helpful in obtaining satisfactory visual results of managing patients with similar clinical findings.

Key Words: Pupillary membrane, congenital cataract, microphthalmos.

ÖZ

Bu yazıda her iki gözünde mikroftalmi, mikrokornea, mikrokori, pupiller membran ve konjenital katarakt gibi çok sayıda oküler patolojinin birarada bulunduğu 14 aylık hastada cerrahi tedavi yöntemi ve görsel sonuçları değerlendirdik. Membran eksizyonu ve katarakt cerrahisinin ardından arka kamaraya sekonder göz içi lensi (AK-GİL) implantasyonu uygulandı. Cerrahi sırasında ve sonrasında herhangi bir komplikasyon görülmedi. Hasta 5 yıl süresince düzenli oftalmolojik kontrollerle takip edildi ve uygun görsel rehabilitasyon yöntemleri uygulandı. Son görme keskinliği her iki gözde 20/100 idi. Bu hastadaki tecrübemiz uygun cerrahi metodun seçilmesinin yanısıra; düzenli oftalmolojik muayene, refraktif düzeltme ve görsel rehabilitasyonun bu tip hastalarda tatminkar görsel sonuçlar alınmasında yararlı olduğunu düşündürmektedir.

Anahtar Kelimeler: Pupiller membran, konjenital katarakt, mikroftalmi.

INTRODUCTION

Microphthalmos is a developmental ocular disorder defined as a small eyeball with an axial length of less than 15 mm in infants. Microphthalmos can be associated with abnormalities of the anterior and posterior segment, such as corneal opacities, microcornea, cataract, persistent hyperplastic primary vitreous and chorioretinal coloboma;[1] however, pupillary membrane and microcoria are not characteristic features of the disease.

Present patient had extraordinary characteristics including congenital bilateral pupillary membrane, microcoria and congenital cataract accompanying with microphthalmos/microcornea. To our knowledge, surgical management in a patient having these kind of features has not been previously described in the literature. Here we report the surgical approach and following therapies contributing to the visual rehabilitation of the patient.

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CASE REPORT

An otherwise healthy 14 month old female patient was brought to our clinic by her parents, who were concerned about the white pupillary opacities that had been noticed in both eyes at her birth. She was born by normal vaginal delivery following a healthy pregnancy. There was no family history of eye disorders similar to this.

On ocular examination, the patient did not look at or follow the objects, but she did appear to respond briefly to the light. The pupils were fixed and miotic and did not respond to the light. Pupillary dilation was attempted with mydriatics, but the pupils did not dilate. A horizontal nystagmus was observed in both eyes. There were no restrictions in the extraocular movements and the eyes appeared ortophoric. Under general anesthesia, an anterior segment examination with a surgical microscope revealed a brownish-white pupillary membrane in both eyes covered the entire pupil and adherent to the pupillary margins (Figure 1a,b). Intraocular pressures obtained via Schiotz tonometer were 15 mmHg in both eyes. Axial lengths measured via A-mode ultrasonography were 15.3 mm in the right eye and 15.6 mm in the left. Corneal diameters were 7.5 mm vertically and horizontally in both eyes. No abnormalities were observed on B-mode ultrasonography. On flash electroretinography, the amplitudes were decreased. On the flash visually evoked potential testing, the amplitudes were nearly normal with short P2 latencies.

Surgical Technique

The patient underwent surgery to remove pupillary membrane and to achieve pupillary aperture. Surgery was first performed in the right eye followed by the left in the same session.

A 1.5 mm clear corneal temporal incision was first made with a 15 degree knife. After injecting a viscoelastic solution of sodium chondroitin sulfate (3.7%) and sodium hyaluronate (2.9%) combination into the anterior chamber, a second similar clear corneal incision was made at the superior side. An attempt was made to cut the tightly adherent and loose membrane with a vitreous scissor, but the membrane could not have been cut or separated from the pupillary margin. In the right eye, pupillary opening was achieved with sphincterotomy by using a vitreous scissor. After sphincterotomy, collar-button iris-retractor hooks were used to perform push-and-pull iris stretching, horizontally and vertically (Figure 1c).

In the left eye, the membrane and some of the adjacent iris were nibbled away to create a larger pupillary opening with an ocutome probe (Figure 1d). After having obtained a pupillary opening, white and dense cataract was noticed by the surgeon in both eyes. Anterior capsulorhexis was done by capsulorhexis forceps and then lens material was removed by an irrigation-aspiration device.

Posterior capsulorhexis was followed by anterior vitrectomy and then viscoelastic gel was removed from the anterior chamber. Incisions were closed with 10-0 nylon sutures. Subconjunctival injection of steroid and antibiotic were applied at the end of the surgery. Figure 1e and 1f show the immediate postopeartive appearance of the eyes.

Topical medication of antibiotics and hourly steroids were started to drop in the next morning after the bandages had been taken. Postoperatively, the anterior segments were clear and the pupillary diameters were 3 mm in the right eye and 2 mm in the left. Fundus examination revealed no abnormalities in both eyes.

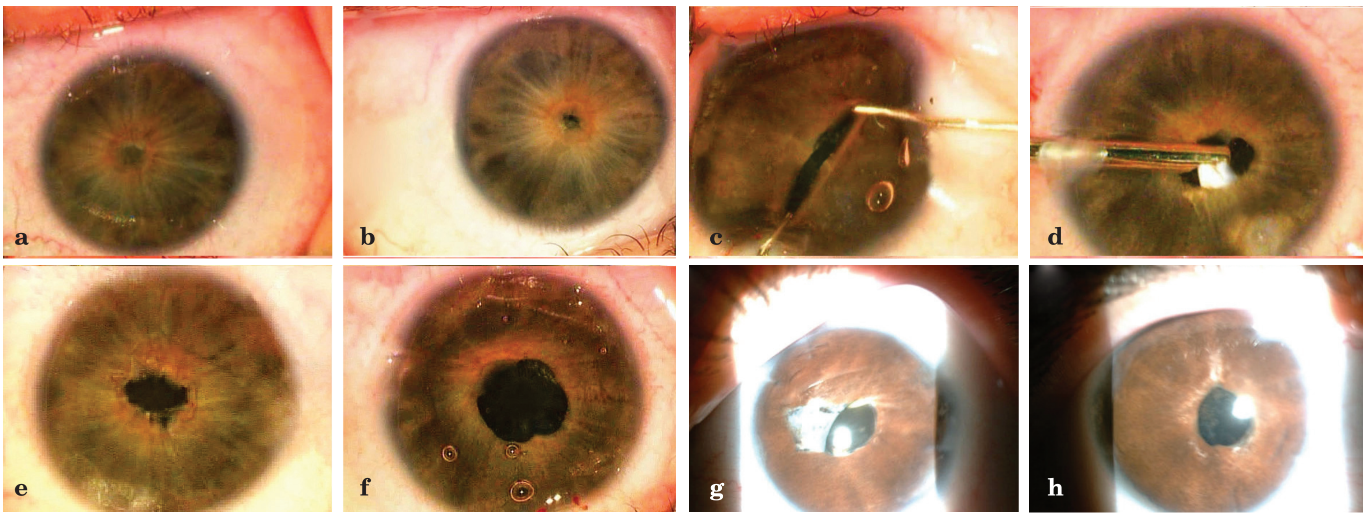

Figure a-h: *Pupillary membranes in the right and left eyes just before surgery (a,b). Vertical pupillary stretching with two collar-button iris-retractor hooks in the right eye (c). Removal of pupillary membrane with an ocutome probe in the left eye (d). Appearance of the right and left eye immediately after surgery (e,f). Biomicroscopic view of the right and left eye at the last visit (g,h).*

Two months after the surgery, the sutures were removed. Under general anesthesia, refraction examination revealed +18.00 -4.00 α180 in the right eye and +18.50 -4.50 α180 in the left eye via hand held autorefractometer. Intraocular pressure was 19 mm Hg in the right eye and 17 mmHg in the left. Bifocal glasses prescribed were +16.00 D for both eyes.

Visual acuity examination revealed 8 cpcm using Lea Grating Paddles in both eyes when measurement was possible. Alternating occlusion therapy was performed during the course of observation. One year after the first surgery, +32 D three-piece foldable hydrophobic acrylic intraocular lenses that were calculated according to SRK-T formula were placed in sulci in both eyes at several days interval. At the first day after the surgery, a mild inflammation which was controlled by frequent topical steroid instillation was noted in both eyes. No intraocular pressure elevation at subsequent visits was observed.

The patient was examined by regular visits and appropriate bifocal glasses were prescribed during the follow-up period. At 6 year of age, her best spectacle corrected visual acuity was 20/100 in both eyes with +5.75 -1.00 α20 D and +6.75 -2.00 α20 D for the right and left eye, respectively. Axial lengths were 19 mm and corneal diameters were 8 mm in both eyes. Strabismus or fixation preference has not been observed since the surgery. Although the amplitude of the nystagmus decreased during follow-up, it has not resolved completely. Figure 1G and 1H show the appearance of anterior segment of the eyes.

DISCUSSION

Severe visual impairment in infants must be detected as early as possible to initiate treatment to prevent deep ambliyopia.[2] In the present patient, multipl factors contribute the visual impairment including congenital cataract, pupillary membrane, microcoria and microphthalmos/microcornea. A pupillary opening of less than 1.5 mm can produce significant decrease in visual acuity because of decreased retinal illumination.[3] The cause of the pupillary anomaly is probably related to presence of the pupillary membrane in this patient.

Forceps and scissors have been used to remove pupillary membranes in previous studies. [4] Lambert et al used an ocutome probe to create pupillary opening in three of five eyes which had microcoria caused by pupillary membrane.[5] In the present patient, cutting or peeling the fibrotic and loose membrane adherent to the pupillary margin with forceps were not possible. Therefore we used pupillary stretching in the right eye and ocutome probe in the left. Two procedures resulted in obtaining an adequate pupillary opening without any complication.

Microphthamos is a rare condition which can have additional pathologies in the globe and/or systemic anomalies.[6] Congenital cataract is the most common cause of severe visual impairment in microphthalmic eyes. Surgical management of cataract in microphthalmic eyes is characterized by several intraoperative difficulties including a shallow anterior chamber and a poorly dilating pupil when compared to that in normal sized cataractous eyes.[6]

Postoperative complications include glaucoma and secondary membrane formation, which might be heightened in cases of microphthalmic eyes.[7] Previously, lensectomy by pars plana approach was the standard procedure in such cases; however, recent improvements in limbal surgical techniques such as using a closed-chamber surgical technique with viscoelastic agents offers advantages to create enough space during the cataract surgery and protect cornea and anterior chamber structures.[7] The complications of cataract surgery in severe microphthalmic eyes with a corneal diameter of less than 9 mm, which included corneal opacity for the limbal approach and secondary membrane formation for the pars plana approach were reported by Yu et al.,[8] In the present patient, limbal approach was a better option to remove the pupillary membrane. After then, cataract surgery was performed through the same incision without any difficulty.

Primary posterior capsulotomy and anterior vitrectomy are highly recommended procedures to prevent posterior capsular opacification which is seen almost a hundred percent in infant eyes after cataract surgery. Implantation of a PC-IOL for the surgical management of congenital cataracts without microphthalmia in patients older than two is commonly accepted procedure.[9] However, safety of IOL implantation during infancy in microphthalmic eyes has not been proved. Some studies suggest that microphthalmos is not too severe, PC-IOL implantation can be applied.[9] However, the timing intraocular lens implantation and determining of the exact power of the lens in microphthalmic eyes are highly controversial.

Inserting PC-IOLs secondarily in both aphakic eyes of a 2.5-year-old patient who had microphthalmos with a 9 mm corneal diameter was reported by Sinskey et al., The patient showed a late secondary membrane formation after surgery and required re-operation. In the present patient, the insertion of PC-IOL of optic diameter 6 mm and total length 13 mm was achieved, without any major difficulty and postoperative complication. Inatomi et al.,[10] reported that the best predicted refraction was calculated using the SRK/T formula compared to SRK, SRK II, S-SRK, Holladay, and Hoffer Q formulas for microphthalmos with axial lengths less than 19.0 mm.

In the present study, IOL power was calculated using the SRK-T formula. The last refraction of present patient was slightly underestimated, which could be managed by wearing glasses.

This report was undertaken to evaluate the management of microcoria, pupillary membrane and congenital cataract in a patient with microphthalmos/ microcornea who underwent pupillary membrane removal, cataract surgery and secondary PC-IOL implantation. It is important to remember that selection of appropriate surgical methods and regular follow-up examinations as well as vigarous visual rehabilitation therapies are needed to obtain satisfactory vision in eyes with multiple abnormalities which deteriorate vision.

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