






Bilateral Angle Closure Glaucoma Due to Isolated Micro-spherophakia in a Child Patient

Çocuk Bir Olguda İzole Mikro-sferofakiye Bağlı Her İki Gözde Açık Kapanması Glokomu

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ABSTRACT This case report presents diagnosis and management of a 9 year old child patient with bilateral angle closure glaucoma caused by isolated microspherophakia. Biomicroscopic examination was normal in both eyes except for shallow anterior chambers. Crystalline lens was located slightly forward into the anterior chamber causing a pupillary block in both eyes. The intraocular pressure was 45 mmHg in her right eye and 37 mmHg in her left eye. Both irido-corneal angles were closed in gonioscopic examination. For the treatment of this condition; we had performed clear crystalline lens extraction and implanted intraocular lens by trans-scleral fixation method for both eyes. After the surgery her best-corrected visual acuity was 20/20 in both eyes. Anterior chamber depth was normal in both eyes. Irido-corneal angle was open without peripheric anterior synechia formation. Lensectomy and intraocular lens implantation may be a possible treatment for microspherophakia related glaucoma.

Keywords: Microspherophakia; glaucoma; lens extraction

ÖZET Bu yazıda, izole mikrosferofakiye bağlı her iki gözde açık kapanması glokomu olan çocuk bir hastanın tanı ve tedavisi sunulmuştur. Yüksek miyopisi olan 9 yaşında bir kız çocuğunun yapılan biyomikroskopik muayenesinde; her iki gözde ön kamaranın dar olduğu ve kristalin lensin hafifçe ön kamaraya doğru uzanarak pupiller bloğa neden olduğu görüldü. Göz içi basıncı sağ gözde 45 mmHg, sol gözde 37 mmHg olarak ölçüldü. Yapılan gonyoskopide, her iki gözde irido-korneal açılırların kapalı olduğu görüldü. Retina doğal olarak değerlendirildi. Bu durumun tedavisi için, her iki göze lens ekstraksiyonu ve trans-skleral fiksasyon tekniği ile göz içi lens implantasyonu yapıldı. Cerrahi sonrasında; en iyi düzeltilmiş görme keskinliği her iki gözde 20/20 olarak kaydedildi. Her iki gözde ön kamaranın derinliği normal idi. İrido-korneal açı periferik ön yapışıklık olmaksızın açık olarak değerlendirildi. Lensektomi ve intraoküler lens implantasyonu mikrosferofakiye bağlı glokomda uygun bir tedavi yaklaşımı olabilir.

Anahtar Kelimeler: Mikrosferofaki; glokom; lens ekstraksiyonu

Bilateral simultaneous angle closure glaucoma is a rare condition. Angle closure most commonly occurs in older hyperopic patients. Less commonly, it occurs in highly myopic patients.¹ Angle closure occurs much less commonly in younger patients. If angle closure glaucoma occurs in a young patient, it is usually associated with one or more developmental ocular anomalies. Spherophakia is a rare condition in which small, spherical shaped crystalline lens causes to pupillary block and secondary angle closure glaucoma.² Spherophakia may occur as an isolated anomaly or it may be associated with a systemic disorder.³ This case report presents diagnosis and management of a child patient with bilateral angle closure glaucoma caused by isolated microspherophakia.

CASE REPORT

A 9-year old girl with high myopia was examined in our outpatient clinic for routine refraction control. Her best-corrected visual acuity (BCVA) was 20/20 in both eyes with -9.00-1.00 x170 in the right eye and with -8.50-0.50 x10 in the left eye. Biomicroscopic examination was normal in both eyes except for shallow anterior chambers (Figure 1 and Figure 2). Crystalline lens was located slightly forward into the anterior chamber causing a pupillary block in both eyes. The intraocular pressure (IOP) was 45 mm Hg in her right eye and 37 mm Hg in her left eye. She had no pain. Gonioscopic examination revealed that both irido-corneal angles were closed but periferic anterior synechiae (PAS) was not observed by indentation. Central corneal thickness (CCT) was 554 microns in the right eye and 551 microns in the left eye. Corneal radius curvature was 7.30 mm in the right eye and 7.36 mm in the left eye. In the examination with full pupil dilation, both crystalline lenses were clear and the

margins of the lenses were visible. Retina was normal and there was no myopic fundus changes in both eyes. Cup to disc ratio was 0.3 in the right eye and 0.2 in the left eye. Anterior chamber depth was 2.02 mm in the right eye and 2.05 mm in the right eye. A-scan ultrasound biometry recorded axial length of 21.46 mm in the right eye and 21.24 mm in the left eye.

High myopia without myopic fundus changes, normal axial length and IOP elevation with shallow anterior chambers in both eyes, led us to diagnose microspherophakia. Also, in the examination with full pupil dilation, the spherical shape of the lens on slit-lamp biomicroscopy and the visibility of the entire lens margins in both eyes supported the diagnose (Figure 3 and Figure 4). Before the treatment, informed consent was obtained from the parents of the patient. For the treatment of this condition; after 5 mm capsulorhexis, we had performed clear crystalline lens extraction and implanted intraocular lens by trans-scleral fixation method by saving the capsular bag for both eyes.



FIGURE 1-2: Right and left eyes- Shallow anterior chambers.

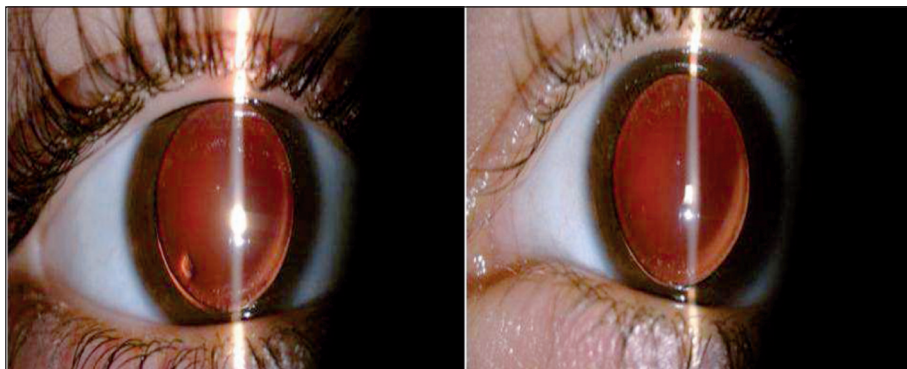


FIGURE 3-4: Right and Left eyes- Margins of the crystalline lenses with dilatation.

To prevent capsular phimosis, IOL optic was captured through the 5 mm anterior capsulorhexis opening.

Postoperative first week IOP was 18 mm Hg in the right eye and 17 mm Hg in the left eye. After the surgery her best-corrected visual acuity (BCVA) was 20/20 in both eyes with -1.00-1.50 x180 in the right eye and with -1.25-1.00 x15 in the left eye. Anterior chamber depth was normal in both eyes. Iridocorneal angle (ICA) was open without PAS formation. The patient has been followed up at regular intervals for 2.5 years with excellent IOP control.

DISCUSSION

Spherophakia is a rare condition in which small, spherical shaped crystalline lens causes to pupillary block and secondary angle closure glaucoma.² Glaucoma associated with isolated spherophakia in a child patient is a very rare condition and its management is much rarely reported and discussed in the literature. Phacodonesis, iridodonesis, zonular dehiscence, lens dislocation and pupillary block are some of the features of the spherophakia.⁴ Spherophakia is usually associated with systemic disorders, mostly with Weill-Marchesani syndrome.⁵ Angle closure glaucoma, shallow anterior chamber and myopia association should direct the clinician to the possible diagnosis of spherophakia.⁶ In our patient, spherophakia occurred as an isolated condition without any systemic disorder. She had the triad of angle-closure glaucoma, shallow anterior chamber and high myopia.

For management of this condition, we had performed clear crystalline lens extraction and implanted intraocular lens by trans-scleral fixation method for both eyes. Scleral fixation of the IOL is a commonly performed and preferred technique, especially in cases with Marfan disease. In the bag IOL placement is not possible in such cases due to weakness of the zonules. In our case, the reason we preferred scleral fixation of the IOL is that the capsular bag is not large enough to implant IOL due to microspherophakia. After the surgery, IOP control was succeeded. Crystalline lens extraction and in-

traocular lens implantation appears to be an effective management for these patients. However, the management of glaucoma in isolated spherophakia has not been established. Reports in the literature are controversial. Willoughby and Wishart reported a case of microspherophakia with glaucoma. They have performed lensectomy and controlled IOP without any medication after surgery.³ Yasar described a case of microspherophakia with high IOP in whom lensectomy was temporarily successful. Later on, antimetabolite augmented trabeculectomy was needed in both eyes to control high IOP.⁷ In an other study, Senthil et al. had reported that nearly half of the microspherophakia patients with glaucoma who underwent trabeculectomy surgery required lensectomy during the follow up period.⁸

The role of lensectomy for IOP control in a microspherophakia patient is to relieve pupillary block and increase the anterior chamber depth. Cases with extensive PAS may not benefit from the lensectomy surgery. In our patient, PAS was not observed by indentation gonioscopy. This point may play the key role while deciding which surgical management to perform in a microspherophakia patient with glaucoma. Also, this point may explain different surgical approaches and their success rates in the literature. One must take into account the possibility of IOP failure after lensectomy surgery, especially in the presence of extensive PAS.⁴

In our case, clear crystalline lens extraction and intraocular lens implantation for both eyes was beneficial for the patient. Since the cause of pupillary block and the angle closure glaucoma was the spherical shaped crystalline lens, lens extraction was a good choice for our case. Other options were trabeculectomy and peripheral iridectomy but it was reported in many studies that lensectomy is the most suggested choice for lens caused angle closure glaucomas.^{9,10} The patient has been followed up at regular intervals for 2.5 years with excellent IOP control without any medication. We suggest that lensectomy may be a treatment for microspherophakia related glaucoma.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, ex-

pertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Ayşe Sevgi Karadağ, Nusret Özdemir; **Design:** Ayşe Sevgi Karadağ, Burak Bilgin, Nusret Özdemir; **Control/Supervision:** Nusret Özdemir; **Data Collection and/or Processing:** Nusret Özdemir; **Analysis and/or Interpretation:** Ayşe Sevgi Karadağ, Burak Bilgin, Nusret Özdemir; **Literature Review:** Havva Gül Özdemir, Ali Şimşek; **Writing the Article:** Burak Bilgin, Havva Gül Özdemir, Ali Şimşek; **Critical Review:** Ayşe Sevgi Karadağ, Burak Bilgin, Nusret Özdemir.

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