Bilateral Isolated Spherophakia with Lens Subluxation, High Myopia and Secondary Angle Closure: A Case Report

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Case Report

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Abstract

Background: Spherophakia is a rare condition and compared to its occurrence with familial and systemic disorders, isolated spherophakia is even more uncommon. This rare case of isolated spherophakia will raise the alertness of physicians when dealing with a high myopia patient with shallow anterior chamber and relatively normal fundus.

Case presentation: We report a case of a 17-year-old male who experienced painless decrease of vision in both eyes (OU) for 11 years, with progression of visual impairment and occasional ocular pain for one year. Examination revealed high myopia, increased intraocular pressures (IOP, 28 mmHg in the right eye (OD) and 33 mmHg in the left (OS)), shallow central anterior chambers, lenses of a spherical shape with superior subluxation, occludable angles without peripheral anterior synechiae, and healthy optic discs OU. A diagnosis of bilateral isolated spherophakia, lens subluxation, high myopia and secondary angle closure (AC) was made. Pars plana lensectomy with anterior vitrectomy and scleral suturing of an intraocular lens was performed. On postoperative follow-up at 19 days OD and 63 days OS, the visual acuity was 6/6 and the IOP was normal without any medication OU.

Conclusions: A presentation with high myopia, shallow anterior chamber with a normal retina should alert the clinician to the possibility of spherophakia, as compared to high myopia caused by elongated axial length. Surgery should be considered in cases of spherophakia with AC where the IOP cannot be controlled by noninvasive means.

Background

Spherophakia is a rare condition in which the crystalline lens assumes a spherical shape with an increased anteroposterior thickness and reduced equatorial diameter.1-2 Spherophakia can be familial or associated with systemic disorders such as Weill-Marchesani syndrome, homocystinuria and Marfan's syndrome.3-5 Isolated spherophakia is more uncommon.6

We report a case of bilateral isolated spherophakia combined with lens subluxation, high myopia and secondary angle closure (AC) in a male adolescent that was managed using lensectomy combined with anterior vitrectomy and a scleral-sutured intraocular lens (IOL) for both eyes.

There is no established standard management for spherophakia.7 Our study shed light on the diagnosis and treatment choosing when dealing with a spherophakia patient who present with high myopia and angle closure.

Case Presentation

A 17-year-old Chinese male presented with painless decrease of vision in both eyes (OU) of 11 years duration. The decrease in vision was first noticed at the age of six years and was not accompanied by pain or discomfort. He gave a history of progressive diminution of vision OU since then; spectacles were changed annually but no other examination was performed. A year ago the patient perceived a significant bilateral deterioration of vision and reported occasional ocular pain. At this time, he attended a local hospital where a diagnosis of bilateral lens subluxation was made. He declined treatment at that time; two months later he sought an opinion in our hospital.

The patient had a history of refractive error since the age of six; available records showed an average progression of -2.00D per year. There was no history of other ocular or systemic disease. He was a full-term normal delivery. There was no family history of spherophakia, Marfan's syndrome, glaucoma and other ocular or systemic disease.

The presenting Snellen visual acuities with correction were 20/63 in the right eye (OD) and 20/100 in the left (OS). Best-corrected Snellen visual acuity was 20/25 with -18.00D sphere and -1.00D cylinder at 800 OD and 20/25 with -18.00DS OS. The intraocular pressures (IOP) were 28 mmHg OD and 33 mmHg OS.

Slit-lamp examination revealed shallow central anterior chambers with a convex lens-iris diaphragm and a van Herrick grade of 0.1 OU. Both corneas were clear. The pupils reacted normally to light. Alphagan, Carteolol, and Brinzolamide were used prior to dilatation of pupils. The crystalline lens OU were clear but spherical in shape and subluxated superiorly (Fig. 1). The lens diameter was approximately 6.5 mm and the edge could be seen within the pupillary margin. The cup to disc ratio was 0.3 OU with healthy neuro-retinal rims. There were no other abnormalities noted in the anterior or posterior segment OU.

On gonioscopy (before the use of atropine) the trabecular meshwork was not visible over 3600 OU. The angles could be opened on indentation and no peripheral anterior synechiae (PAS) were detected. Corneal endothelial cell counts were 3085 cells per mm2 OD and 3170 cells per mm2 OS.

Ultrasonographic A-scan biometry recorded an axial length (AL) of 24.50 mm and a central anterior chamber depth (ACD) of 1.93 mm OD. The AL was 24.69 mm, and the central ACD was 1.98 mm OS. Keratometric readings were 40.86/41.36 D (mean = 41.11 D) OD and 40.56/41.36 D (mean = 40.96 D) OS, indicating relatively flat corneas. The radius of curvature was 8.21mm OD and 8.24mm OS.

Ultrasound biomicroscopy revealed a shallow anterior chamber, steep anterior lens curvature, anterior bowing of the iris, irido-lenticular contact, elongated zonules and an increased distance of the lens equator from the ciliary processes in both eyes, more inferiorly than superiorly (Fig. 2). The anterior chamber angle was closed with both the angle open distance at 500 μm and the trabecular iris angle of zero in all four quadrants OU - the image suggested a pupillary block mechanism.2 The corneal thickness was 0.58 mm and the central ACD 1.56 mm OU.

The patient was 184 cm in height and weighed 100 kg with normal skeletal proportions; there were no abnormalities of the skin, joints or fingers and toes. There were no features of Marfan's or Weill-

Marchesani syndromes. He did not have any anterior chest deformity, scoliosis, or osteoporosis and a cardiovascular examination was within normal limits.

A diagnosis of bilateral isolated spherophakia, lens subluxation, high myopia and secondary angle closure was made. Treatment with Alphagan one drop b.i.d, Carteolol one drop b.i.d and Brinzolamide one drop b.i.d was continued, but the IOP was measured at 27 mmHg OD and 30 mmHg OS. The addition of 1% Atropine sulfate one drop b.i.d decreased the IOP to 18-20 mmHg OU and lens extraction was planned. In view of the lack of zonular support, pars plana lensectomy with anterior vitrectomy and scleral suturing of IOL implantation were undertaken. Surgery was performed OS first followed by OD 44 days later.

The postoperative course was uneventful. Nineteen days following surgery on the second eye the presenting visual acuity with no correction was 6/6 OU with IOP's of 11 mmHg OD and 10 mmHg OS without any treatment. Slit-lamp examination showed clear corneas, deep anterior chambers and centered IOL's OU (Fig. 3). The patient is very satisfied with the outcomes so far and remains under close follow-up.

Discussion

Spherophakia is a rare congenital anomaly that involves weak zonules and a smaller, more spherical crystalline lens with an increased anteroposterior thickness of the lens and a reduced equatorial diameter.8 The excessive curvature of the anterior surface of the lens leads to a shallow anterior chamber and secondary angle closure glaucoma (ACG).5 Spherophakia is also associated with lenticular myopia, lens subluxation or dislocation.5,8

The features of spherophakia that have been reported include iridodonesis, phacodonesis, lens protrusion anterior to the pupillary plane, pupillary block, visibility of the lens equator on pupil dilation, zonular elongation or rupture and lens dislocation.1,6,9-10 Not all features occur in every patient with spherophakia.6,9-10 In our case, iridodonesis and phacodonesis were absent, probably due to the relative integrity of the zonules that caused a mild superior subluxation.

The exact mechanism of development of spherophakia remains speculative. The traction of the zonular fibers on the crystalline lens is necessary for its normal development and it has been suggested that spherophakia could be due to a congenital weakness of the zonules.11 As a result of such zonular weakness and lack of traction, the lens may retain its spherical form instead of gradually converting to a normal biconvex shape.11-12 An alternative theory suggests that spherophakia is due to arrested development of the secondary lens fibers or the insertion of abnormally thin secondary fibers, both resulting from a nutritional deficiency due to defects in the tunica vasculosa lentis.12-13 These defects are thought to cause cessation of growth of the lens with retention of the initial spherical shape from when the fetus is 5 to 6 months old.13

The triad of high myopia, shallow anterior chambers and AC/ACG should alert the clinician to a possible diagnosis of spherophakia.2 Khokhar et al reported that most of the patients with spherophakia had a history of insidious visual loss and, in some cases, pain resulting from secondary glaucoma.5 The visual compromise in these cases is attributed to refractive error and / or secondary glaucoma.5

Normal axial length and corneal curvature as well as with lack of characteristic fundus changes associated with high myopia (staphyloma, lacquer cracks, Fuch's spot or chorio-retinal atrophy) implicate the lens as the source of myopia in these cases.14 Our patient too was a high myope with normal axial length, fundus and relatively flat K-values; the characteristics of the lens and zonules as well as the anterior position confirm lenticular myopia.

As happened with our patient, those with spherophakia are easily misdiagnosed as simple high myopia; this is especially so in the early stage of the disease when there is no lens displacement or increased IOP. In this situation a normal retina with no changes usually associated with high or degenerative myopia along with a normal axial length and corneal curvature should raise the suspicion of spherophakia.

The mechanism of a shallow AC in spherophakia is attributed to pupillary block caused by the anterior lens curvature, increased anteroposterior thickness, and forward displacement of the lens.1,6 The anterior lens curvature and forward positioning of the lens leads to iridolenticular contact and pupillary block, pushing the iris forward to cause AC.2 In cases where the zonules are not intact, the lens dislocation and forward movement cause pupillary block and phacomorphic glaucoma.1,6 Unrelieved pupillary block may also lead to PAS formation and further damage to the trabecular meshwork with increase in IOP and glaucomatous optic nerve damage.1

Early diagnosis and management of AC with laser peripheral iridotomy (LPI) and medical treatment may prevent the onset of angle-closure glaucoma.2 When chronic AC with PAS is present, surgery may be required.1

Miotics cause ciliary muscle contraction and allow the lens to become more spherical and move anteriorly thereby aggravating the situation.15 Accordingly miotics are not recommended in the management of spherophakia.2 On the other hand, cycloplegic agents such as atropine relax the ciliary muscle, tighten the zonules causing posterior lens movement that can open the angle and decrease the IOP15-16

There is no established standard management for spherophakia.7 It has been suggested that LPI to relieve pupillary block should be performed in patients with spherophakia.2 LPI is noninvasive and relatively safe but does have occasional complications.17 In spherophakia there is the possibility of anterior vitreous prolapse following LPI.16 In some cases the angles may remain occludable or closed despite the iridotomy.10

Kaushik et al in a case report suggested lensectomy as an effective first-line strategy for managing a bilateral spherophakia patient with acute ACG.6 Willoughby et al reported a case of bilateral

spherophakia where phacoemulsification was required after iridotomy failed to control the glaucoma.1 Taylor et al have reported a case of spherophakia in the context of Weill-Marchesani syndrome where, after iridotomy and medical treatment had failed to control the IOP, removal of the lens was required to achieve control.16 Kanamori et al reported ACG with microspherophakia which required lensectomy, IOL implantation and goniosynechialysis to control IOP.10

Khokhar et al report that most of their cases were successfully managed with lensectomy with peripheral iridectomy.5 Rao et al found that lensectomy was effective in controlling IOP in about half of their patients with spherophakia; 40% needed anti-glaucoma medications in addition and only 7.7% needed glaucoma surgery.18 They also reported that younger age (≤6 years old) at presentation, higher IOP, the need for more anti-glaucoma medications and greater cup to disc ratio were risk factors for failure of IOP control postoperatively.18 In some cases trabeculectomy may still be needed for IOP control despite an initial lensectomy.9

Surgical intervention may be the only option when laser and medical treatment does not control the IOP. Early surgical intervention might prevent or limit visual loss from secondary glaucoma and lenticular myopia.4

The choice of removal of the lens alone or combined with glaucoma surgery should be tailored to individual case. In patients who have been diagnosed early and have no PAS or glaucomatous optic neuropathy but are not controlled with medical treatment, as was the case in our patient, lens extraction alone is likely to work. For those with long standing AC as determined by uncontrolled IOP, PAS and optic nerve/visual field changes, goniosynechialysis or trabeculectomy, depending on the extent of PAS, combined with lens extraction may be needed. While waiting for surgery, anti-glaucoma medications as well as cycloplegic agents can be used.

Lens extraction is frequently the intervention of choice to manage lens subluxation and/or angle closure / angle closure glaucoma in patients with spherophakia.1,3-4,6 The procedure is not without complications that are related to the small capsular bag with a relatively high equatorial diameter and instability of zonular fibers.19 Despite the risks and possible complications, which were discussed with the patient during informed consent, in this case we felt surgical intervention was required in this instance.

As the laxity of zonular fibers varies in patients with spherophakia the choice of technique for lens extraction, extracapsular cataract extraction (ECCE), phacoemulsification or pars-plana lensectomy depends on the individual case. ECCE with IOL in spherophakia carries a high risk of complications especially vitreous loss.16 Intracapsular lens extraction combined with a scleral-sutured sulcus-fixated posterior chamber intraocular lens (PC IOL) has been recommended if the zonules are judged to be very weak.3 The long term risk of dislocation of an IOL implanted in the bag even with use of a capsular tension ring (CTR) makes this approach controversial in spherophakia.10 Both phacoemulsification combined with implantation of a PC IOL plus CTR and lensectomy with scleral-fixated PC IOL implantation have been reported to be effective in lowering the IOP and enhancing the visual acuity.7

We felt that the best approach in our patient would be a lensectomy, anterior vitrectomy with a scleral-sutured a PC IOL placed in the sulcus. At least in the short term this intervention has resulted in good vision as well as IOP control and could be used in other patients with spherophakia who have a similar clinical picture.

A limitation of this case report is the short follow up. Another is that while the thickness of lens was measured but unfortunately not recorded; this did not however affect the diagnosis or the decision making.

Conclusion

We present a rare case of isolated bilateral spherophakia with angle closure that was successfully managed with surgical intervention. A presentation with high myopia, shallow anterior chamber, AC/ACG with a normal retina should alert the clinician to the possibility of spherophakia.

Surgery should be considered in a spherophakia patient with angle closure where the IOP cannot be controlled by noninvasive means. A pars-plana lensectomy with anterior vitrectomy and scleral fixation of the IOL was successful in our case and appears to be a reasonable approach.

Declarations

Ethics approval and consent to participate All procedures conformed to the Declaration of Helsinki. This study was approved by the ethics committee of the Beijing Tongren Hospital.

Consent to publish Written informed consent was obtained from the patient and his mother for publication of this case report and accompanying images.

Availability of data and materials All the data generated or analyzed during our study are contained within the manuscript.

Competing interests

The authors declare that they have no competing interests.

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Authors' Contributions

All authors read and approved the final manuscript. Study concept and design (XYS); data collection (YZ, CW); analysis and interpretation of data (CW, XYS); writing the manuscript (YZ); critical revision of the manuscript (RT).

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Figures

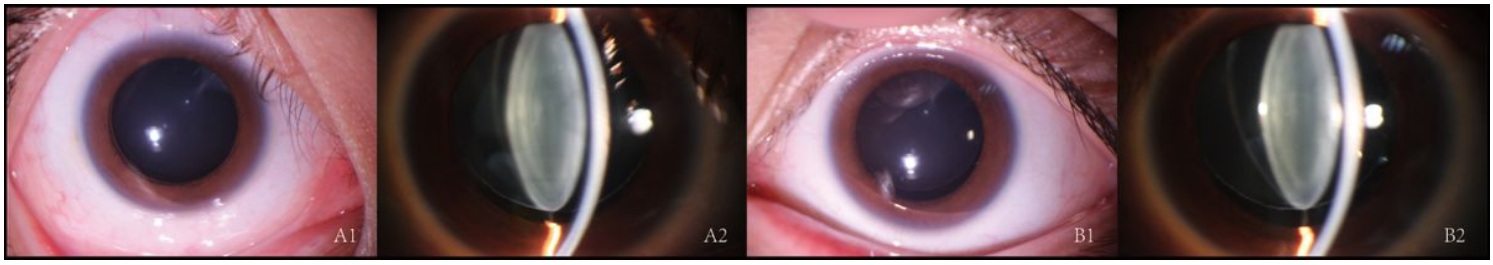

Figure 1

Slit-lamp photographs. (A) The right eye shows spherophakia of the lens; the margins of the lens are visible when the pupil is dilated. (B) The left eye shows similar findings

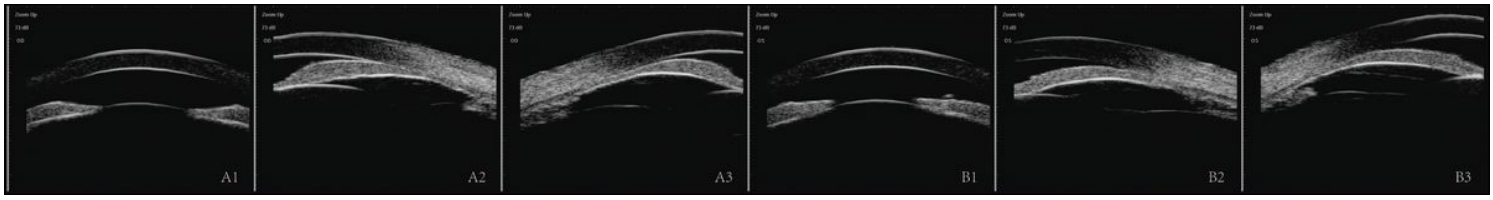

Figure 2

Ultrasound biomicroscopy. (A) The right eye shows shallow anterior chamber and AC due to pupillary block mechanism. (B) The left eye shows similar findings

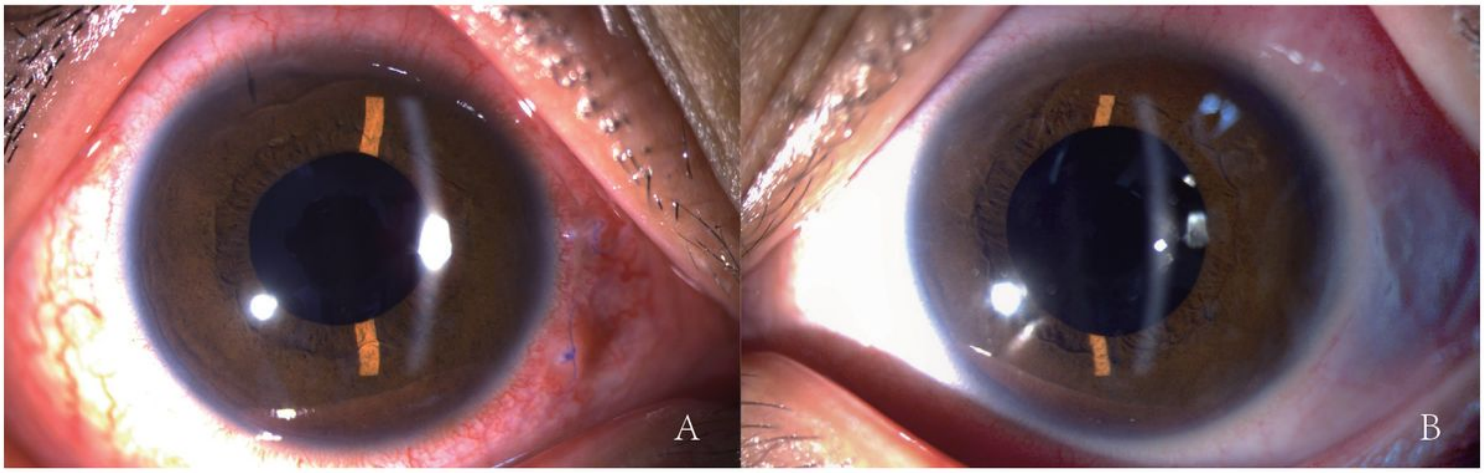

Figure 3

Postoperative photographs. (A) The right eye shows clear cornea and deep anterior chamber. (B) The left eye shows similar findings.

Supplementary Files

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