

## Case report

# Management of bilateral microspherophakia with secondary angle closure glaucoma

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## Abstract

**Background:** Microspherophakia is characterized by lenticular myopia, late development of lens dislocation, shallow anterior chamber and angle-closure glaucoma. If it is associated with angle closure glaucoma, the management can be challenging. **Objective:** To report the management of a case of bilateral secondary angle closure glaucoma with isolated microspherophakia and right subluxated lens in a middle aged lady. **Case:** A 47-year-old female presented with bilateral secondary angle closure glaucoma with isolated microspherophakia, right subluxated lens and left operated trabeculectomy. The patient's best corrected log mar visual acuity improved from 1.0 to 0.2 and intraocular pressure (IOP) decreased from 24.9mm of Hg to lower teens following lensectomy with scleral fixation of posterior chamber intraocular lens in the right eye. The left eye IOP remained in mid teens without medication in two year follow up. **Conclusion:** This case demonstrates that microspherophakia should be ruled out even in middle aged patients presenting as angle closure glaucoma. The possibility of thickened cornea must be kept in mind requiring an IOP correction. Lensectomy with scleral fixation of posterior chamber IOL provides a satisfactory recovery of visual acuity and IOP control. Though in this patient, the lens was held in position by posterior synechiae in the left eye, future stability of zonules needs to be monitored.

**Keywords:** Microspherophakia, subluxated lens, angle closure glaucoma, lenticular myopia

## Introduction

Microspherophakia is a rare congenital malformation in which the lens has a small equatorial diameter and the zonular fibers are elongated and may be weak, causing excessive curvature of the anterior surface of the lens leading to lenticular myopia, late development of lens dislocation, shallow anterior chamber and angle-closure glaucoma.

The condition can either be isolated, idiopathic and familial anomaly or it may be associated with systemic defects like Marfan's syndrome, Weil-Marchesani syndrome, hyperlysinemia and congenital rubella (Bhattacharjee H et al, 2010).

Cycloplegics, laser-iridotomy, and trabeculectomy are used for relieving angle closure glaucoma in such patients (Imaizumi Y et al, 1999). However, lensectomy is usually required to lower the intraocular pressure (IOP) and preserve vision (Yasar T, 2003; Willoughby

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CE et al, 2002). No standard surgical technique has been adopted because of the variable laxity of the zonular fibers (Al-Haddad C et al, 2012). The different surgical procedures used in management of these cases are intracapsular extraction, phacoemulsification or lensectomy with a limbal approach or pars plana lensectomy and vitrectorhexis with lens aspiration. Intraocular lens (IOL) implantation is not always feasible in the absence of a stable capsular support, especially after complicated surgery (Al-Haddad C et al, 2012; Kaushik S et al, 2006; Wu-Chen WY et al, 2005; Kanamori A et al, 2004). Also, in the bag implantation of IOL is questionable due to unstable bag postoperatively, non-availability of a small capsular tension ring (CTR) and high rate of bag contraction (Al-Haddad C et al, 2012; Behndig A, 2002; Dufay-Dupar B et al, 2007).

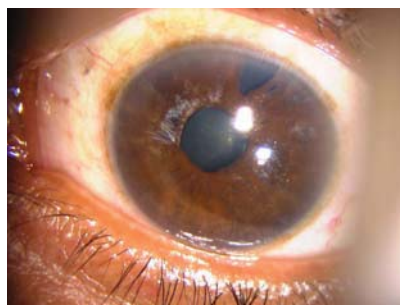
We present a case of a middle aged lady with isolated bilateral microspherophakia, thick corneas, angle closure glaucoma with right severe anteriorly subluxated lens. To the best of our knowledge thick cornea has not been reported with microspherophakia.

### Case presentation

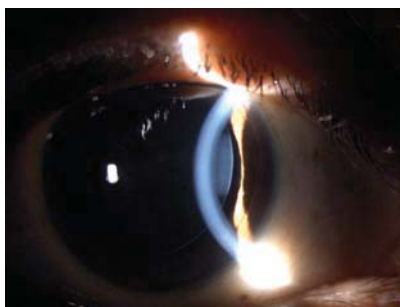
A 47-year-old lady, teacher by profession, presented with intermittent pain and diminution of vision right eye for the past six months. She had a history of severe pain, redness and decreased vision in the left eye a year back and was diagnosed as a case of left primary angle closure glaucoma and right angle closure suspect. She had been treated with oral acetazolamide, topical 0.5% timolol maleate and 4% pilocarpine in the left eye followed by left trabeculectomy and right laser iridotomy. She remained asymptomatic for the next four months without any medication. There was no history of similar episode earlier. She wore spectacles for distance for the past 20 years and did not have a family history of glaucoma.

At presentation, the best corrected log mar

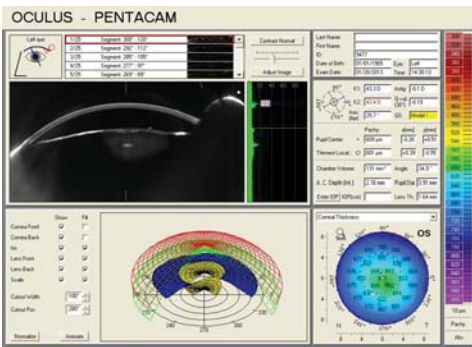
visual acuity in the right eye was 1.0 and that in the left eye was 0.3 with -4D spherical lens in both eyes. The central corneal thickness was 650 and 609 mm by ultrasonic pachymetry in right and left eye respectively. Applanation tonometry showed the intraocular pressure (IOP) of 34 mm of Hg and 22mm of Hg in the right and left eyes respectively. The corrected IOP of right eye was 24.9 mm of Hg and that of the left was 16.7 mm of Hg. The anterior chamber was shallow in both the eyes. There was a patent iridotomy at 1.30 and 11 o'clock positions in right eye. The left eye had a superior filtering bleb, patches of iris atrophy and patent surgical iridectomy at 1.30 o'clock (Figure 1). Dilated examination of the right eye revealed a small spherical lens with the lens edge seen within the pupillary margin (Figure 2). The lens was subluxated from 12 O'clock to 7 O'clock hour. The left pupil failed to dilate due to posterior synechiae and the lens had glaucomaflecken (Figure 3). The axial length of



**Figure 1:** Slit lamp photograph of left eye showing patches of iris atrophy and patent surgical iridectomy at 1.30 o'clock.



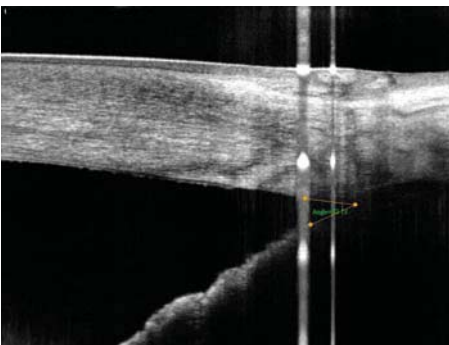
**Figure 2:** Slit lamp photograph of right eye showing subluxated cataract



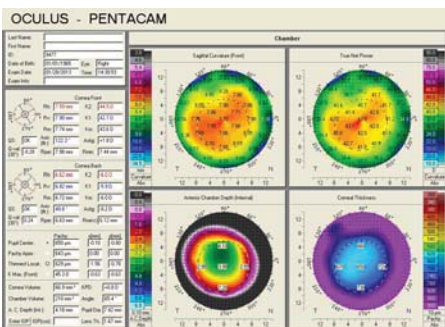
**Figure 3:** Pentacam of left eye showing gaucoma flecken



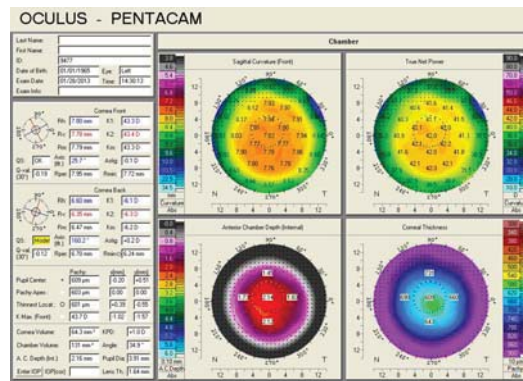
**Figure 4:** Postoperative photograph of right eye showing well centered IOL



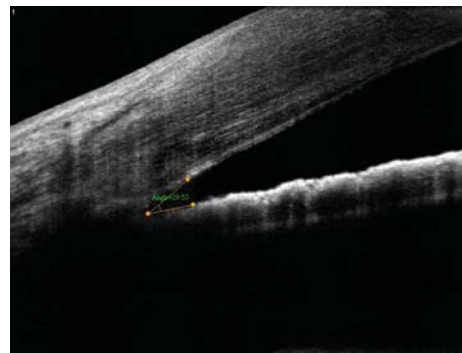
**Figure 5:** Post-operative anterior segment OCT of right eye showing anterior chamber angle width >30 degrees.



**Figure 6:** Post-operative Pentacam of right eye showing anterior chamber depth and thick cornea.



**Figure 7:** Pentacam of left eye showing shallow anterior chamber and thick cornea



**Figure 8:** Anterior segment OCT of left eye showing angle width of nasal, inferior, temporal and superior angles

the globe was 22.67 mm on both sides.

She was of average height (154 cm) and thin built. She had normal skeletal proportions with no evidence of arachnodactyly, short and stubby fingers or reduced joint mobility. There was no anterior chest deformity or scoliosis. The cardiovascular examination was within normal limits. Urine chromatography was negative for homocystinuria.

In light of the above findings, a diagnosis of bilateral microspherophakia with secondary glaucoma was made. The patient underwent right limbal lensectomy with ab-externo scleral fixation of +22.0 D posterior chamber intraocular lens under local anesthesia. Postoperatively, the patient's vision improved and her IOP remained between 12 and 18 mm of Hg without any drugs till her last visit. Two year follow up showed a logmar BCVA of 0.2



and 0.3 with -2D cylindrical x120 degrees and -4D spherical respectively in right and left eyes. The IOL was well centered, the angle had opened to more than 30 degrees in the right eye (Figure 4, 5 and 6). In the left eye, the peripheral anterior chamber depth by pentacam showed low values of 1.45 mm (Superior), 1.83 mm (Temporal), 1.73mm (Nasal) and 2.12 mm (Inferior). But the anterior segment OCT of left eye showed open angles namely 23.28° (Superior), 29.53° (Temporal), 29.18° (Nasal) and 31.82° (Inferior) in spite of the forward push by the lens (Figure 7, 8). The fundus examination revealed normal disc in both the eyes.

Lensectomy was, therefore, deferred in the left eye and the patient was advised to come for regular follow-ups every 3 months.

### Discussion

The decreased equatorial diameter of a microspherophakic lens measures an average of 6.75 to 7 mm (normal, 9.0 mm), and the anteroposterior diameter is larger than normal (>5.0 mm, normal: 3.4 to 4.5 mm) (Tindall, 2008). Glaucoma occurs due to pupillary block by the spherical lens or irritation of the ciliary body by the dislocated lens in the anterior chamber or complete luxation of the lens in anterior chamber (Kaushik et al, 2006).

Most patients present in adolescence or early childhood (Senthil et al, 2012). As our patient became symptomatic at a much later age, the diagnosis had possibly been missed and she was treated as primary angle closure glaucoma and prescribed pilocarpine eye drops in both eyes. Due to the phenomenon of inverse glaucoma, miotics are known to worsen the pupillary block by relaxing the weak zonules and promoting anterior migration of lens and increasing the contact between the iris and lens. (Urbanek, 1930).

Cycloplegics are the treatment of choice. The diagnosis was clinched on dilating the right

eye but the left eye failed to dilate because of the posterior ring synechiae which held the lens in place. The treatment protocol in this patient aimed towards management of myopia, glaucoma and subluxation. Lensectomy alone in the right eye could have controlled the glaucoma but would have resulted in anisometropia. The patient was reluctant for any further surgical intervention in the left eye as her IOP was controlled and vision was satisfactory with the anterior chamber angles having opened up following trabeculectomy. The IOL implantation in the right eye was possible by using a combination of capsular tension segments to fix the bag along with the capsular tension ring (CTR). Though described by some authors (Khokhar et al, 2012), the CTR is not the treatment of choice because of the small sized bag. Scleral fixation is the alternative procedure (Fan et al, 2003) which was successfully performed in this patient. Bilateral iris fixated phakic intraocular lens implantation has been used effectively for correction of high myopia in spherophakia. However, they are recommended only in patients with appropriate anterior chamber depth, absence of lens displacement and compliance with annual examinations. During follow up, the authors suggest monitoring the endothelial cell count, changes in IOL location, and development of cataract or corneal decompensation (Moshirfar, 2010).

The angles do open up partially following lensectomy which relieves pupillary block but it may not be sufficient to control the IOP in presence of extensive peripheral anterior synechiae (Kaushik S et al, 2006). In our patient, lens extraction with scleral fixation of PC IOL in the right eye effectively managed the subluxation of lens, myopia and glaucoma. The IOP of the left eye was satisfactorily controlled by trabeculectomy at a follow-up of two years. However, a careful watch is required for especially left eye with regards to control of IOP and development of cataract.

## Conclusion

Angle closure glaucoma due to microspherophakia needs to be differentiated from primary angle closure glaucoma as pilocarpine administration further worsens the condition. Corneal thickness measurement is required to correctly measure the IOP. A blanket treatment cannot be followed as each eye may require a tailor made approach.

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