Too Tiny A Menace

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ABSTRACT

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Microspherophakia is a rare congenital anomaly wherein the anteroposterior diameter of the lens is increased and equatorial diameter of the lens is decreased. The most common syndrome associated with microspherophakia is Weill Marchesani syndrome. The common ocular issues in microspherophakia are high lenticular myopia, ectopia lentis and secondary glaucoma. Lens removal with appropriate intraocular lens implantation addresses the ocular issues in microspherophakia

Keywords: Microspherophakia, Weill Marchesani syndrome, Ectopia lentis

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CASE REPORT

A 32-year-old male, who is a daily wage labourer from West Bengal, India presented to the outpatient department with complaint of painless defective vision in both eyes (OU) since childhood. His Snellen uncorrected distance visual acuity was counting fingers at 1 meter in both eyes. His Snellen best corrected distance visual acuity was 6/60 in OU with a -20 Diopters spherical lens. On examination, the anterior segment of both eyes was within normal limits. Following pupillary dilatation, the lens in both eyes were observed to be small and spherical with the entire equator of the lens visualized within the pupillary margin **(Figure 1,2,3 and 4).** No phacodonesis was noted in the right eye while mild phacodonesis was noted in the left eye. Fundus in both eyes were normal with a cup-disc ratio of 0.2 and the intraocular pressure (IOP) with Goldmann Applanation Tonometry was 16mm of Hg in OU. Anterior segment Optical Coherence Tomography (AS-OCT) of both eyes showed the anterior surface of the lens in both eyes to be ahead of the plane of the iris **(Figure 5 and 6).**

Based on the above findings, a diagnosis of microspherophakia in both eyes was made. The patient was informed about his condition and the treatment required and he decided to pursue further treatment from his native place.

DISCUSSION

Microspherophakia is a congenital anomaly wherein the lens adopts an abnormal spherical shape instead of the normal biconvex shape. The anteroposterior diameter of the lens is increased while the equatorial diameter of the lens is decreased in microspherophakia.

The proposed etiopathogenesis is a developmental abnormality of the lens. During the 5th – 6th month of gestation, the lens is normally spherical in shape. Secondary lens fibers then begin to develop within the lens at this time thereby bringing about a change in the shape of the lens from spherical to biconvex. The tunica vasculosa lentis nourishes the lens during the fetal life and plays a vital role in the normal development of lens fibres during intrauterine life. In microspherophakia, defects develop within the tunica vasculosa lentis at around the 5th – 6th month of gestation, thereby causing a nutritional deficiency of the lens and a consequent defective development of the secondary lens fibers. The lens thus fails to attain its normal biconvex shape and continues to retain its spherical shape with no corticonuclear demarcation.[1] In addition, it is also speculated that in microspherophakia, the lens is never subjected to the forces of an optimally acting ciliary body and zonules during the embryonic period. Consequently, there is lack of tension exerted by the ciliary zonules onto the developing lens and thereby also con-

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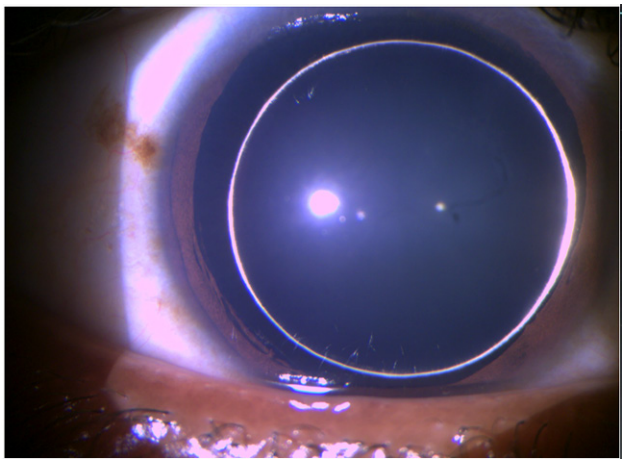

Figure 1. **Slit lamp image of right eye under diffuse illumination.**

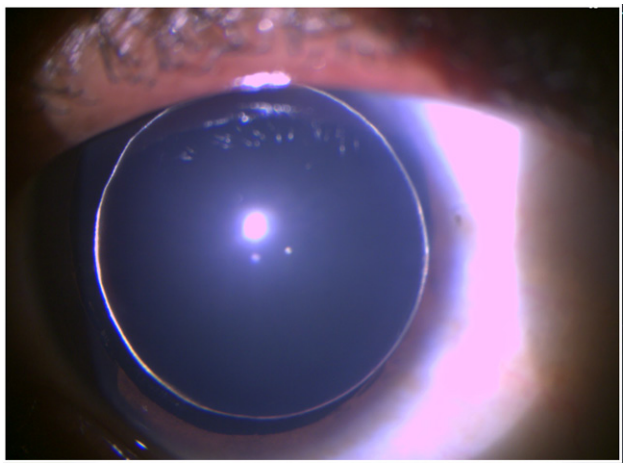

Figure 2. **Slit lamp image of left eye under diffuse illumination**

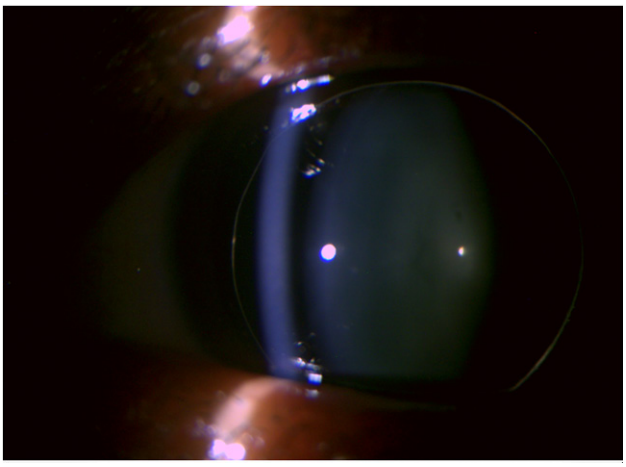

Figure 3. **Slit lamp photograph of right use showing optical section of the lens**

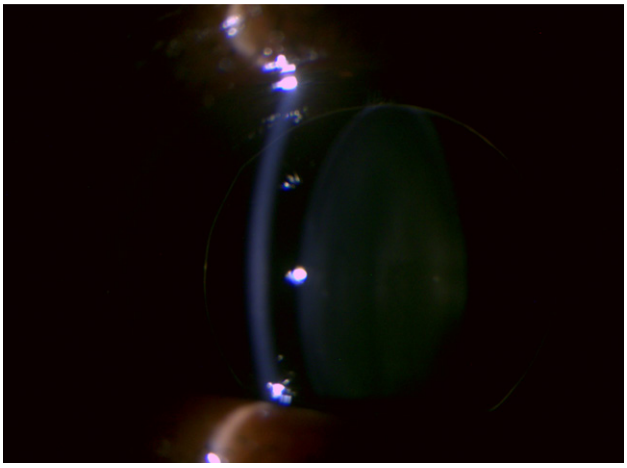

Figure 4. **Slit lamp photo of left eye showing optical section of the lens**

tributing to an abnormal spherical shaped lens.[2]

Microspherophakia can occur sporadically or as an inherited disorder. When inherited, the commonly implicated genes are the LTBP2 gene, ADAMTS gene and the FBN1 genes. Multiple systemic associations are noted in microspherophakia with the most common being Weil Marchesani syndrome[3] and homocystinuria.[4] The commonly encountered ocular associations are megalocornea, aniridia, iridocorneal endothelial syndrome and optic disc colobomas.

The ocular issues in microspherophakia can discussed under the following headings

1) Lenticular Myopia

2) Secondary Glaucoma

3) Ectopia Lentis

LENTICULAR MYOPIA

The abnormal spherical shape of the lens with an increase in the anterior and posterior curvatures causes a high degree of lenticular myopia. This high myopia needs optical correction with appropriate spectacles/ contact lenses. Refractive lensectomy is a good surgical option as it not only corrects the lenticular myopia but also the other issues in microspherophakia viz secondary glaucoma and ectopia lentis.[5]

ECTOPIA LENTIS

The ciliary zonules in microspherophakia are found to be long, lax and weak. The zonules, especially those on the posterior surface of the lens, are usually unattached to the ciliary processes. Because of the frailty of the zonules, these lenses are prone to subluxation or dislocation either anteriorly or posteriorly, which may be incited with a trivial trauma or even spontaneously.[5]

The management of ectopia lentis in microspherophakia is by lensectomy and implantation of an appropriate intraocular lens (IOL). The approach for the lensectomy can be through the limbal route or through the pars plana route. The indication, timing and mode of extraction of the subluxated/dislocated lens is still

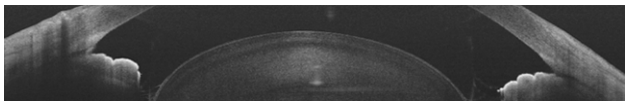

Figure 5. **AS OCT image of right eye**

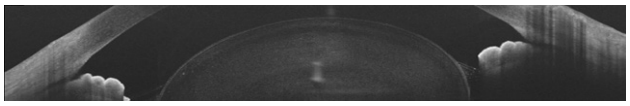

Figure 6. **AS OCT image of left eye**

debatable and there are no controlled trials or retrospective studies that have specifically identified an ideal age for intervention.[6] Before embarking on a particular route of approach for the lensectomy , it is essential to examine the patient in both sitting and supine position as lenses that seem to be approachable through the limbal route might subluxate posteriorly on adopting the supine position.

The management for the ectopia lentis in microspherophakia can be decided after taking into consideration the patient's visual symptoms, best corrected visual acuity, degree of lens subluxation and the presence/absence of secondary glaucoma.

Definitive indications for refractive lensectomy are as outlined below [5]

1. Diplopia which is not amenable to optical correction.
2. Progressive subluxation of the lens
3. Severe subluxation of the lens (where lens edge uncovers more than 50% of the dilated pupil)[7]
4. Significant lens opacification.
5. Secondary Glaucoma
6. Anterior / Posterior dislocation of the lens
7. When the best corrected visual acuity (BCVA) obtained with optical correction (spectacles/ contact lenses) does not provide a good vision related quality of life for the patient

SECONDARY GLAUCOMA

Secondary glaucoma in microspherophakia comes under the purview of angle closure glaucoma. The mechanisms responsible for the secondary angle closure glaucoma are

1. The increased anterior curvature of the lens causing an increase in the relative pupillary block, iris bombe formation and angle closure.
2. The weak zonules in microspherophakia causing an anterior subluxation or anterior dislocation of the lens resulting in a forward push on the iris and the resultant angle closure glaucoma.
3. Posterior dislocation of the lens with vitreous blocking the pupil and bringing about a pupillary block and angle closure glaucoma.

Abnormal development of the angle structures resulting in an open angle glaucoma is also now recognized as a probable mechanism for secondary glaucoma in microspherophakia.[8]

The presentation of the patient can be either with

a. An acute elevation of the IOP resembling a primary angle closure attack
b. Chronic glaucomatous changes of the optic nerve

When the patient presents with an acute elevation of the IOP, the management initially resembles that of a primary angle closure attack wherein antiglaucoma medications are used to lower the IOP. However, later the management differs from that of a primary angle closure attack in that pilocarpine shouldn't be instilled prior to the laser peripheral iridotomy (LPI). This is because, apart from the increased anterior curvature of the lens causing a pupillary block there is also an added element of weak zonular apparatus causing a possible anterior subluxation of the lens in microspherophakia. In this context, if a miotic agent like pilocarpine is used (as is conventionally done in primary angle closure glaucoma with pupillary block), followed by the laser peripheral iridotomy, the pupillary block will be relieved by the LPI but the anterior subluxation of the lens would have worsened. This is because the accompanying ciliary body relaxation induced by the miotic agent will lower the tension exerted by the zonules on the lens thereby further accentuating the anterior displacement of the lens and worsening the angle closure brought about by the anterior push of the iris into the angle structures. Thymoxamine, an alpha-adrenergic antagonist, is preferred as the miotic prior to the LPI in microspherophakia as it only causes pupillary constriction and does not affect the ciliary body.[9]

A mydriatic-cycloplegic agent on the contrary is a double-edged sword. If there is adequate zonular support to the lens, the use of a mydriatic cycloplegic agent will cause relaxation of the ciliary body, thereby increasing the tension on the zonules and bring about a posterior displacement of the lens thereby simultaneously relieving the 2 mechanisms responsible for angle closure (viz pupillary block and the anterior push on the iris by the subluxated lens). However, if the zonular support of the lens is not adequate, then use of the mydriatic-cycloplegic agent will on the contrary cause

a dislocation of the lens into the anterior chamber courtesy of the dilating action on the pupil brought about by the mydriatic-cycloplegic agent.

The safest approach is to avoid both miotics and mydriatic-cycloplegic agents in these patients who present with acute elevation of IOP. It would be prudent to initiate the patient on antiglaucoma drugs especially hyperosmotic agents which will ensure the shrinkage of the vitreous and thereby the lens can move posteriorly simultaneously reducing or relieving the pupillary block and the anterior push on the iris. Placing the patient in a supine position can also aid in the posterior displacement of the lens away from the pupil. Then an LPI can be safely performed to relieve the element of pupillary block and break the attack of acute IOP rise without causing any untoward mishaps as mentioned above. The caveat is that the relative contribution of the pupillary block mechanism and anterior pushing mechanism on the iris towards the acute IOP rise will determine the success of the LPI in breaking the acute angle closure attack.

Inability to break the attack of angle closure glaucoma with LPI signifies either synechial closure of the angle or an increased element of anterior push on the iris as the mechanism responsible for angle closure attack. In these circumstances lensectomy with IOL implantation and if needed an appropriate glaucoma surgery will be needed for the management of the patient.

The pertinent question, especially when the patient presents with chronic glaucomatous optic neuropathy is whether to perform lensectomy with IOL implantation alone or glaucoma surgery alone or to combine both. A study conducted by Senthil et al[10] showed that out of 29 microspherophakic eyes with glaucoma that was managed with trabeculectomy alone, 45% later required a lensectomy. Rao et al[11] analyzed the efficacy of lensectomy alone in microspherophakic eyes with glaucoma and found that nearly half of the eyes had their IOP well controlled with lensectomy alone, 40% of the eyes had IOP control on adding antiglaucoma medications along with the lensectomy while 7.7% of the eyes needed further surgical intervention for IOP control. The authors of the study conducted by Rao et al[11] recommended that lensectomy with IOL implantation would suffice to manage the secondary glaucoma in microspherophakia with early glaucomatous optic neuropathy and in the event of advanced glaucomatous optic neuropathy in microspherophakic eyes, they recommend to combine lensectomy and glaucoma surgery.

END NOTE

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