• Letter to the Editor •

Combined phacoemulsification and anterior vitrectomy in a case of anterior megalophthalmos with open-angle glaucoma and high myopia

Cai-Xuan Huang, Xiao-Hui Zhao, Yi-Qiao Xing

Eye Center, Renmin Hospital of Wuhan University, Wuhan 430060, Hubei Province, China

Correspondence to: Xiao-Hui Zhao. Eye Center, Renmin Hospital of Wuhan University, No.238 Jiefang Rd., Wuchang District, Wuhan 430060, Hubei Province, China. zxh6981@126.com

Received: 2016-09-07 Accepted: 2017-01-17

DOI:10.18240/ijo.2017.07.26

Huang CX, Zhao XH, Xing YQ. Combined phacoemulsification and anterior vitrectomy in a case of anterior megalophthalmos with open-angle glaucoma and high myopia. *Int J Ophthalmol* 2017; 10(7):1178-1180

Dear Editor,

I am Dr. Cai-Xuan Huang, from the Eye Center, Renmin Hospital of Wuhan University, Hubei Province, China. I write to present a case report of bilateral anterior megalophthalmos with open-angle glaucoma, which was treated by combined phacoemulsification and anterior vitrectomy.

Megalocornea (horizontal corneal diameter more than 13 mm) can occur in the forms of simple megalocornea, anterior megalophthalmos and buphthalmos (infantile glaucoma)[1]. Anterior megalophthalmos presents with megalocornea, enlarged lens iris diaphragm and ciliary ring inherently. Its complications include iridodonesis, hypoplasia of iris dilator muscle, iris stromal atrophy, lens subluxation or luxation, and early cataract formation [usually not accompanied with raised intraocular pressure (IOP)][2]. Limited views, deep anterior chamber and the potential risk of intraocular lens (IOL) decentration bring surgical difficulties in anterior megalophthalmos[3].

The patient was a 38 year-old healthy woman without relevant family history. She had been myopic since childhood, and suffering from declining eyesight and distending pain in both eyes (OU) for the last six months. When she came to our Outpatient Department, her best corrected vision acuity (BCVA) was 20/200 OD and count fingers OS. Slit-lamp microscopy and anterior segment optical coherence tomography (AS-OCT) examinations revealed the presence of clear but large cornea (horizontal diameter was 13.34 mm OU; vertical diameter was 13.26 mm OD and 13.67 mm OS), deep anterior chamber, severe iridodonesis, iris atrophy, lens cortical opacity and lens subluxation, without phacodonesis. Her IOP was 41.0 mm Hg OD and 45.9 mm Hg OS (measured with Goldmann applanation tonometry). She was prescribed 0.2% brimonidine tartrate eye drops combined with 2% carteolo hydrochloride OU twice daily. When she came back two weeks later, her IOP reduced to 11.6 mm Hg OD and 13.9 mm Hg OS. She received a series of examinations. The pupils could not be dilated fully by tropicamide phenylephrine (Figure 1). B scan ultrasound confirmed lens subluxation, high myopia and vitreous opacity OU. Anterior chamber depth (ACD) and axial length (AL) was 5.37 mm and 32.4 mm OD; 5.77 mm and 31.75 mm OS (IOL-Master); the vitreous index (vitreous length/axial length×100) was 83.42% OD and 81.83% OS. Central corneal thicknesses (measured with AS-OCT) were 0.550 mm OD and 0.590 mm OS. Keratometric power was 37.88 at 94° and 40.42 at 4° OD, 38.84 at 68° and 39.85 at 158° OS. Endothelial cell density was 2017 cells/mm^2 OD and 1927 cells/mm^2 OS. Ultrasound biomicroscopy (UBM) showed iris depression and thinning, lens subluxation, extensive ciliary body atrophy and partial ciliary body disappearance (Figure 2). Due to the extreme deep ACD, neither B scan nor UBM could detect the diameters of capsular bag or lens thickness. Wide ciliary band and open anterior chamber angle were observed on gonioscopy, without obstruction in the trabecular meshwork OU. Fundus exams found pathological myopic changes like tessellated fundus and lacquer cracks; the ratio of cup to disc (C/D) was 0.6 OD and 0.7 OS. Cupping and pallor of the optic disc were also seen (the left eye was worse). Superior and inferior of per-papillary retinal nerve fiber layer (RNFL) were thinned on OCT images. On the basis of above findings, the diagnosis of bilateral anterior megalophthalmos with open-angle glaucoma, lens subluxation and high myopia was confirmed.

Given the present conditions, the patient was advised to take cataract surgeries successively. After fully learned the diagnosis, treatment options and complications, she chose to be aphakic and wearing glasses to correct refractive errors.

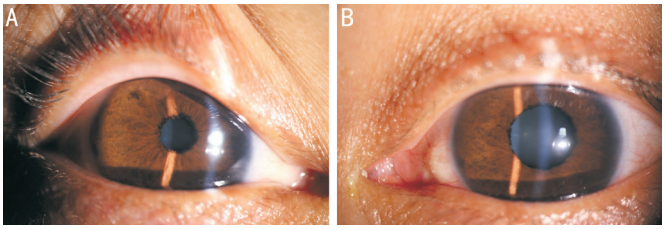

Figure 1 Slit-lamp appearances of anterior megalophthalmos after pupil dilation The extreme deepen of ACD in both eyes was clearly shown. The pupil of the right eye (A) could not be dilated. There was a slight mydriasis in the left eye (B).

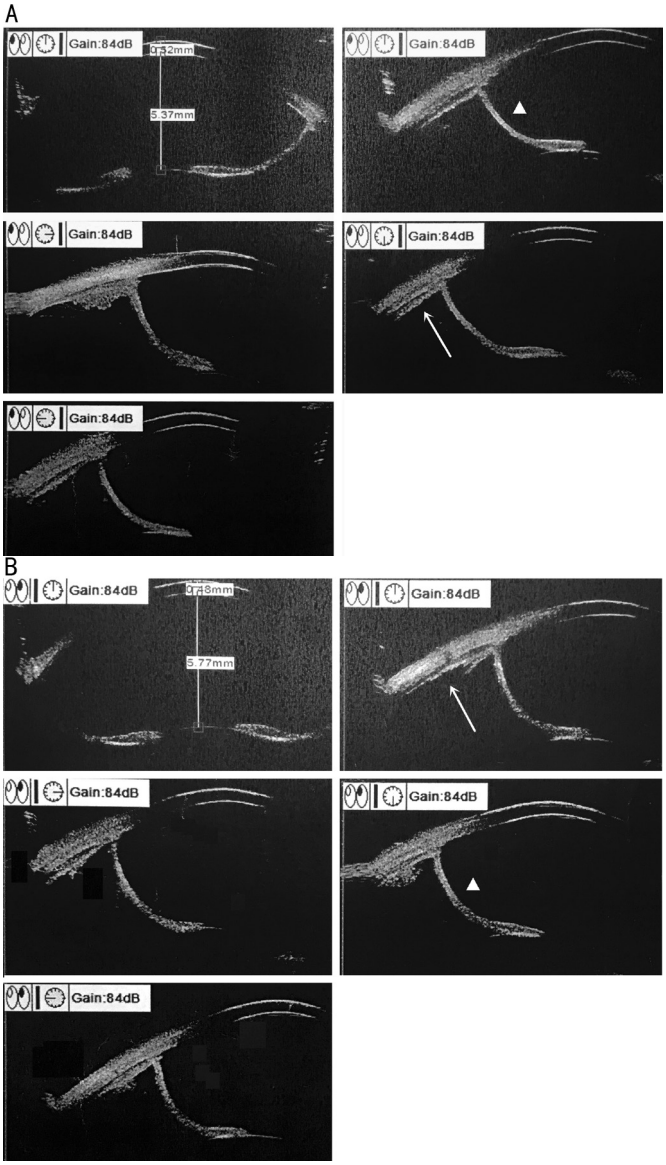

Figure 2 UBM results showed abnormities in iris, ciliary body and anterior chamber Anterior chamber angle was wide open (more than 90 degree) both in the right eye (A) and left eye (B). The iris exhibited thinning and posterior bowing (triangle) extensively. Partial ciliary processes were absent in the areas where they ought to exist (arrow).

For the combined phacoemulsification and 23-gauge (23G) anterior vitrectomy in both eyes, the surgeon (Zhao XH, who owns 20y of experiences in cataract surgery) performed the following procedures: 0.04% oxybuprocaine on the cornea, a 3.2-mm temporal clear corneal incision, followed by injection of a dispersive ophthalmic viscosurgical device (OVD), continual curve capsulorrhexis (the pupil of either eye could not be fully dilated, therefore iris hooks were used to expand the visual scope), hydrodissection, nuclear fracturing and cortical cleanup; then conducted posterior capsulorhexis and anterior vitrectomy with a 23G vitreous cutter through clear corneal incision and aspirated OVD at last. With the help of iris hooks, a quarter of zonular dialysis was seen evidently in both eyes (lens suspension ligament fractured from 7 o'clock to 10 o'clock OD and from 4 o'clock to 7 o'clock OS). No adhesion between iris and anterior capsule existed. She had administered 0.3% tobramycin and 0.1% dexamethasone ophthalmic suspension, 0.2% brimonidine tartrate eye drops for the first two weeks. Follow-up was performed at 1, 3, 4, 6, and 12wk postoperatively. Her IOP maintained at a normal level throughout follow-up. Her BCVA was 20/25 with +2.25 D sphere and +2.0 D cylinder at 5° OD, 20/100 with +2.75 D sphere and +0.50 D cylinder at 145° OS at the third months.

Anterior megalophthalmos is characterized by the presence of megalocornea and ciliary ring enlargement with a very deep anterior chamber. Its secondary effects include iridodonesis, iris stromal atrophy, zonular anomalies and cataract formation[4]. X-linked genetic transmission is found as the most frequent cause of anterior megalophthalmos (possibly located on Xq21.3-q22)[5-6]. High myopia brings pathological changes to the eyeball, including Schlemm's canal, trabecular network and sclera, increasing the morbidity rate of primary open-angle glaucoma[7].

This patient has deep anterior chamber, lens subluxation and ciliary body dysplasia, corresponding with Kuchenbecker's descriptions of anterior megalophthalmos[8]. What's more, she suffered from high IOP and optic disc atrophy as well, which are not the typical symptoms. So it is essential to differentiate her disease with other types of glaucoma. Clinical manifestations of congenital glaucoma include poor vision, corneal edema, Descemet's membrane opacity and mild mydriasis; in the late stage, the C/D increases and eyeball expands. Our patient's eye pain and declining eyesight started 6mo ago, elevated IOP had been left unnoticed. However, her corneas always stayed clear, anterior chamber angle was wide open; a normal angle view also seen by gonioscopy; IOP could reduce to a normal level by administering eye drops. These indicate the absence of congenital glaucoma and lens dislocation-induced pupil-blocking glaucoma, fitting well with the diagnosis of primary open-angle glaucoma and high myopia in anterior megalophthalmos.

Despite the lens dislocated to a certain extent, a standard IOL could have been inserted into the capsular bag during the surgery, even implanting a capsular tension ring. Since her

ideal IOL diopter was around +4.0 D OU by preoperative exams, which was approaching +0.0 D, the oculists did not conduct IOL implantation. It turned out her postoperative spherical equivalent refraction was between +2.25 D and +2.75 D. This confirmed measurement errors existing in the IOL-Master exams and SRK-T formula calculation, probably result in the overly long ACDs and ALs.

Weakened zonules, lens subluxation and small pupils all burden the cataract extraction surgery in anterior megalophthalmos patients[9]. Phacoemulsification should be the first choice[10], by which IOP could remain stable intraoperatively. If there is a wide range of lens dislocation, surgeons should move to extracapsular cataract extraction procedure. Custom IOLs and standard IOLs had been reported implanted in capsular bags successfully, with certain risk of IOL decentration[11-12]. Also note that another method of IOL implantation, posterior IOL sulcus fixation, cannot guarantee the centration of IOL because of the deepened and enlarged posterior space[13].

In summary, anterior megalophthalmos is a rare disease. Potential occurrences such as lens subluxation, iris stromal atrophy and glaucoma should be kept in mind. Special consultations must be carried out for patients before cataract surgery.

ACKNOWLEDGEMENTS

Foundation: Supported by the National Natural Science Foundation of China (No.81400420).

Conflicts of Interest: Huang CX, None; **Zhao XH,** None; **Xing YQ,** None.

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