

A Case of Secondary Glaucoma due to Bilateral Anterior Chamber Shallowing Caused by Marfan's Syndrome

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Abstract

Purpose: To report the clinical manifestations and treatment of a case of secondary glaucoma evolving from bilateral anterior chamber flattening caused by Marfan's syndrome.

Methods: The ophthalmic and systemic features, B-scan and UBM characteristics were recorded. Therapy and efficacy were analyzed.

Results: Marfan's syndrome, in this case, caused bilateral iris-lens diaphragm anterior dislocation, anterior chamber flattening, pupillary block, angle closure, and finally resulted in persistent increased intraocular pressure (IOP). After undergoing pars plana vitrectomy and lensectomy combined with anterior chamber reformation, the visual acuity of the patient's right eye increased from 6/150 to 6/7.5 (best-corrected) and that of the left eye was improved from 6/100 to 6/10 (best-corrected). The IOP of the right eye fell to 18 mm Hg, and the left eye to 12 mm Hg.

Conclusion: Marfan's syndrome can cause bilateral anterior chambers flattening, and induce secondary angle closure glaucoma. Combined pars plana vitrectomy, lensectomy and anterior chamberplasty can re-form the anterior chamber, control IOP and maintain visual function. (*Eye Science* 2011;26:112-115)

Keywords: Marfan's syndrome; lens subluxation; secondary glaucoma

Marfan's syndrome is a congenital disorder of mesoderm maldevelopment, which may affect the eyes, cardiovascular system and skeleton¹. The most serious signs and symptoms in affected eyes include lens dislocation, ametropia, secondary glaucoma, and detachment of retina². However, bilateral

flattening of the anterior chamber is a rarely seen clinical sign. We report such a case treated in our hospital in July, 2010.

The patient was a 21 year-old chinese woman presenting to a local hospital with bilateral corneal edema, accompanied by decreased visual acuity two months ago. She complained of blurred vision in the morning, clearing somewhat in the afternoon. No other discomfort was reported. She had a history of poor vision since childhood, diagnosed as resulting from myopia, with a spherical equivalent power of -8.0 D bilaterally. Systemic examinations revealed a metacarpal ratio of 8.56 for the right hand, and 7.83 for the left. Ultrasonocardiographic (UCG) analysis indicated bicuspid and tricuspid regurgitation. No abnormality was observed in lungs. Her cousin had an atrial septal defect.

Examination of the eyes revealed visual acuity of 6/150 O.D, 6/100 O.S, with no improvement on refraction and an intraocular pressure (IOP) of 23 mm Hg O.D, 31 mm Hg O.S. The conjunctivae were white and quiet bilaterally. Staphylomata were present at the upper corneoscleral limbus of both eyes. The corneal diameter was 11 mm O.D, 12 mm O.S. Mild corneal edema was observed in the right eye. The cornea of the left eye was clear and compact. The anterior chamber was completely flat bilaterally (Figure 1). Further examination disclosed irregular posterior synechia and an irregular pupil with a diameter of approximately 4 × 4 mm, un-reactive to light. Ectropion uveae was noted. Glaukomfleken was present in both eyes. No view of the fundus was possible in either eye.

Ultrasound biomicroscopy (UBM) confirmed that the anterior chamber was flat bilaterally, with thinning of the ciliary body (Figure 2). In B-mode ul-

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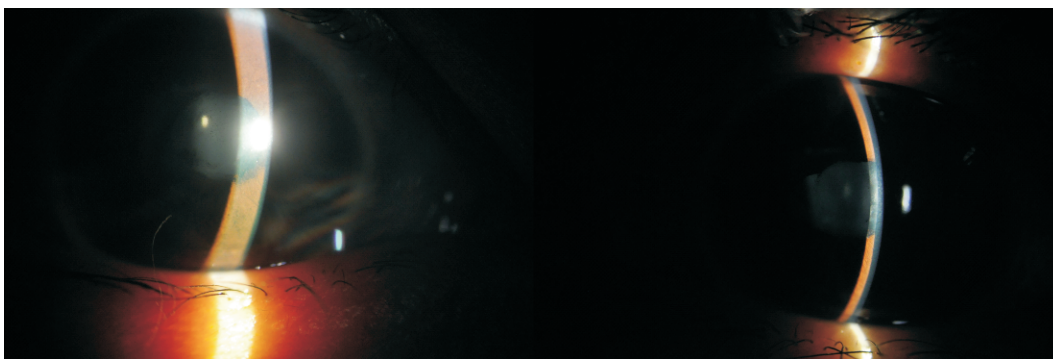


Figure.1 Photo of the anterior segment of both eyes at presentation; mild corneal edema is present on the right, while the left corneal is clear and compact. The anterior chambers in both eyes are completely flat.

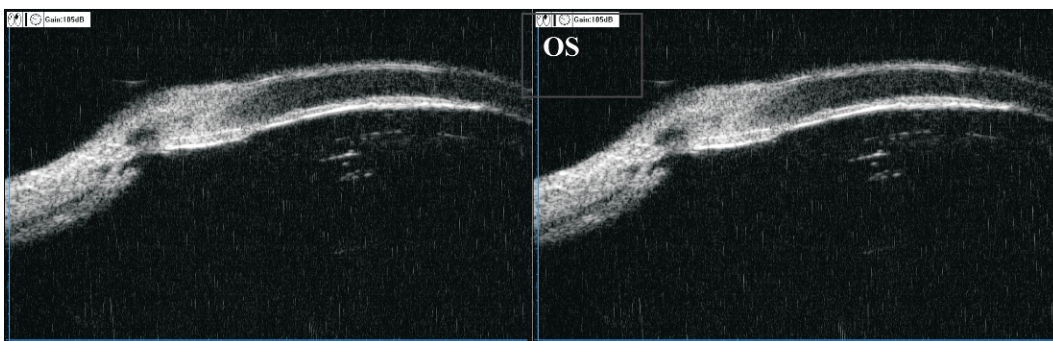


Figure.2 UBM imaging in both eyes; the anterior chambers in both eyes are flat, the superior corneoscleral limbus is thinned, a sclera staphyloma is present, and the ciliary body is thinned.

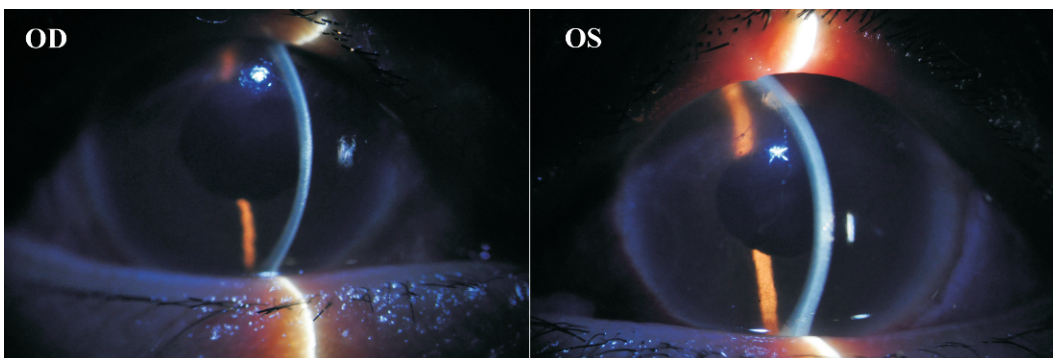


Figure.3 Post-operative photo of the anterior segment in both eyes; the central anterior chamber is deep bilaterally, with a peripheral anterior chamber depth > 1 CT, pupil diameter of 4x4 mm, and absent crystalline lens.

trasound imaging, mild vitreous opacity was observed in both eyes, and excavation of the optic nerve was identified. The thickness of the right lens was 3.53 mm, and 3.82 mm for the left lens. The axial length was 23.02 mm O.D, and 23.00 O.S. Based upon ocular and systemic abnormalities identified during examination, the patient was diagnosed with secondary glaucoma due to lens subluxation associated with Marfan's syndrome.

The patient was first treated with topical cyclo-

plegic agents, topical and systemic corticosteroids, and topical pressure-reducing medications with mannitol after admission. However, the anterior chambers remained flat despite these treatments.

The patient underwent left anterior chamberoplasty on 20th July, 2010. During the surgery, peripheral anterior synechiae was noted intermittently from 10:00 to 12:00, which were separated with viscoelastics, after which the left anterior chamber remained formed. One day postoperatively, the left central an-

terior chamber depth was 3.5 corneal thicknesses (CT), and the peripheral anterior chamber remained deep. The IOP was 11 mm Hg, and the best-corrected visual acuity was 6/18. Two days following the operation, the anterior chamber became shallow again, with a depth of 0.5–1 CT centrally. The patient underwent pars plan vitrectomy and lensectomy combined with anterior chamber plasty in the left and right eyes on 26th July, 2010 and 29th July, 2010, respectively. Both anterior and posterior synechia were separated intraoperatively. Post-operatively, the uncorrected visual acuity of the right eye was 6/60, which improved to 6/7.5 with an aphakic correction of, +10.0D+1.50D \times 150 $^\circ$, In the left eye was, uncorrected acuity of 6/18 improved to 6/10 with correction of +9.0D+1.75D \times 5 $^\circ$. IOP of the right eye was 18 mm Hg with twice-daily topical application of timolol, brinzolamide, and brimonidine tartrate drops. The IOP in the left eye was 12 mm Hg without pressure-lowering therapy. The central anterior chambers remained deep bilaterally, the peripheral anterior chamber was > 1 CT in both eyes, and the pupils were 4 \times 4 mm in diameter. The crystalline lenses were absent (Figure 3), the optic nerves were excavated with a cup-to-disc ratio of 0.9.

After 9 months of follow up, the best-corrected visual acuity remained stable at 6/7.5 in both eyes. The IOP was normal in both eyes with topical use of timolol, brinzolamide and brimonidine tartrate eye-drops in both eyes. The central anterior chambers remained deep bilaterally, and the peripheral anterior chamber was > 1 CT, in both eyes.

Discussion

Most patients with Marfan's syndrome present with upward lens dislocation, with occasional dislocation into the vitreous body. Anterior lens dislocation has rarely been reported, especially with secondary, bilateral flattening of the anterior chamber. The patient described in this report had no prior history of trauma or surgery. She presented to hospital complaining of decreased visual acuity, blurred vision in the morning and relatively clear eyesight in the afternoon, suggesting gradual changes in the anterior chamber depth associated with variability in the refractive power of the eyes. UBM imaging

failed to detect the echo of the suspensory ligament of lens, possibly indicating lens suspensory ligament dysplasia induced by Marfan's syndrome. Such suspensory ligament maldevelopment might possibly cause iris-lens diaphragm anterior dislocation, shallow anterior chamber, pupillary block, angle closure, and eventually persistent elevation of IOP³. During the early stages, it was possible that the suspensory ligament was tightened during the daytime when the sympathetic nerve was active, and thus the anterior chambers were deepened somewhat. By contrast, the suspensory ligaments might have been in a lax state at night, when parasympathetic activity was more prominent, leading to shallowing of the anterior chambers^{4,5}. This might in principle explain the patient's complaints of blurred vision in the morning, alleviated in the afternoon.

Developmental glaucoma can be excluded because no significant corneal enlargement was documented. In this study, lens subluxation evoked an elevation in IOP, and further induced secondary glaucoma. No evident abnormality in axial length or lens thickness was noted. The patient had modest myopia of -0.8D prior to surgery, but required +11D of aphakic correction postoperatively, which together with the normal axial length suggested that the myopia was mainly induced by anterior dislocation of the crystalline lens, as might be expected based on optical principles. Marfan's syndrome is a congenital disorder of mesoderm maldevelopment, which leads to connective-tissue defects, also affecting the sclera. Persistent increase in IOP was induced by a combination of anterior dislocation of the lens, flattening of the anterior chamber and posterior synechia, leading to scleral staphyloma. Postoperative examination showed that the patient's optic cup was enlarged bilaterally, suggesting that the IOP had been elevated for a relatively long time, leading eventually to severe optic nerve damage.

In this case, the patient was treated conservatively with cycloplegic agents anti-inflammatory medication and IOP-lowering therapy in an attempt to deepen the anterior chambers. However, these treatments were not successful due to severe anterior and posterior synechiae. Then, anterior chamber plasty was performed to promote anterior chamber reformation.

This operation was not sufficient to promote long-term deepening of the anterior chamber, due to the impact of lens subluxation and lens-iris diaphragm anterior dislocation. Therefore, we finally elected to perform pars plana vitrectomy in combination with lensectomy, which was ultimately successful in restoring physiologic anterior chamber depth in both eyes. Primary intraocular lens implantation was not attempted because of concerns over the health of the optic nerve and corneal endothelium. Following surgery, the patient was instructed to wear aphakic glasses for visual correction. This surgical approach achieved good visual recovery and stable IOP in both eyes.

References

- 1 Gonzales EA. Marfan syndrome. *J Am Acad Nurse Pract.* 2009;21(12):663–670.
- 2 Li J. Clinical symptom of Marfan syndrome and analysis of ophthalmic surgery. *Journal of Clinical Ophthalmology.* 2002;10(5):408–409.
- 3 Dagi LR, Walton DS. Anterior axial lens subluxation, progressive myopia, and angle-closure glaucoma: recognition and treatment of atypical presentation of ectopia lentis. *JAAPOS.* 2006; 10(4):345–350.
- 4 Duke-Elder S. *System of Ophthalmology.* London: Kimton. 1968;617–619.
- 5 Wang T, Liu L, Li ZH, et al. An ultrasound biomicroscopic study on changes of the structure of ocular anterior segment after topical application of cycloplegia. *Chinese Journal of Ophthalmology.* 1998; 34(2):137–140.