A rare presentation of bilateral dislocated lens in a patient with isolated microspherophakia

Thandra Sai Shreya, Vijayalakshmi A Senthilkumar, Chitranjan Mishra, R Krishnadas

Key words: Dislocated lens, microspherophakia, secondary glaucoma

A 23-yr-old female presented with sudden diminution of vision in the left eye (LE) for 4 days associated with headache and vomiting. No history of ocular trauma or previous surgery or similar complaints in the family. Her best-corrected visual acuity (BCVA) was 20/50 with + 8.00 Dsph in RE, HM in LE. Anterior segment evaluation revealed a quiet eye with a deep anterior chamber (AC), iridodonesis, aphakia in RE, and focal corneal edema with a microspherophakic cataractous lens in the AC touching the corneal endothelium in LE [Fig. 1a and b]. Fundus evaluation of the RE showed an inferiorly dislocated cataractous lens in the vitreous cavity [Fig. 2a and b]. Ultrasound biomicroscopy (UBM) of the LE showed a small spherical lens in the AC touching the corneal endothelium with an increased anterioposterior diameter (3.72 mm) and decreased equatorial diameter (4.65 mm) [Fig. 3a and b]. B scan revealed dislocated microspherophakic lens into the vitreous cavity in RE and anteriorly dislocated lens in LE [Fig. 4a and b]. IOP in LE was controlled with oral carbonic anhydrase inhibitors and topical IOP-lowering agents.

Glucoma Services, Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Retina Consultant, Vitreoretinal Services, Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Madurai, Tamil Nadu, India

Correspondence to: Dr. Vijayalakshmi A Senthilkumar, Glaucoma Services, Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Madurai - 625 001, Tamil Nadu, India. E-mail: vijimmc@gmail.com

Received: 21-Oct-2019 Revised: 28-Nov-2019
Accepted: 10-Dec-2019 Published: 25-May-2020

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Cite this article as: Shreya TS, Senthilkumar VA, Mishra C, Krishnadas R. A rare presentation of bilateral dislocated lens in a patient with isolated microspherophakia. Indian J Ophthalmol 2020;68:1161-3.
Microspherophakia is a rare developmental condition characterized by increased anteroposterior diameter and decreased the equatorial diameter of the lens due to the simple arrest of lens development between the 5th and 6th months of intrauterine life. It can occur as an isolated entity or familial entity or associated with systemic disorders such as Weill Marchesani syndrome, Homocystinuria, Marfan syndrome, Alport syndrome, Klinefelter syndrome, Lowe syndrome, Peter’s anomaly, Cri-du-chat syndrome. Glaucoma in isolated microspherophakia can occur from spherical lenses causing a pupillary block, irritation of ciliary body by the dislocated lens or complete dislocation of the lens into AC.[1-4]

Figure 1: (a) Slit-lamp photograph of the left eye (LE) showing dislocated microspherophakic cataractous lens in the anterior chamber, (b) magnified view of microspherophakic cataractous lens

Figure 2: (a) Optos image of the right eye (RE) showing inferiorly dislocated microspherophakic cataractous lens in vitreous, (b) magnified view of microspherophakia lens in vitreous

Figure 3: (a) Ultrasound biomicroscopy (UBM) image of the LE showing small spherical lens in the anterior chamber touching corneal endothelium with completely wide open angles (b) white arrows showing small spherical lens

Figure 4: (a) B scan RE showing dislocated lens (white arrows) in vitreous, (b) B scan of the LE showing anteriorly dislocated lens (white arrows)

Our patient had no clinical features characteristic of any syndrome. She had corneolenticular touch in LE due to anteriorly dislocated lens with very high IOP which may lead to corneal decompensation if not treated appropriately. Microspherophakic lens usually causes secondary angle-closure glaucoma but our patient had open angles that can be clearly visualized with UBM. Elevated IOP probably due to the inflammation caused by the dislocated lens in AC as well as inverse pupillary block impairing the aqueous outflow.

Our patient underwent Pars plana lensectomy and scleral fixated IOL in LE. Two partial-thickness scleral pockets were
made, 180 degrees apart, at a distance of 1.5 mm from limbus with a 23G microvitreoretinal (MVR) blade for containing the IOL haptics. Three standard 23G pars plana vitrectomy ports and two ciliary sulcus-based sclerotomies were created close to the scleral pockets using a 24G needle, through which IOL haptics would be externalized. After performing lensectomy and core vitrectomy with 23G vitrectomy cutter, a three-piece non-foldable IOL (Aurolab, Madurai, India) was placed into AC through the tunnel and the haptics was externalized with a 25G end-gripping forceps (Alcon Laboratories, Fort Worth, Texas, USA) and tucked into the scleral pockets. Finally, the vitrectomy ports, the scleral tunnel, and conjunctiva were sutured, leaving the globe saline-filled. The postoperative period was eventful. Best-corrected visual acuity (BCVA) and IOP during the last follow-up were 20/70 and 20 mm Hg in the LE. She was advised to continue antiglaucoma eye drops in the LE.

We report this case because of an unusual presentation of isolated microspherophakia with bilateral simultaneous lens dislocation posteriorly into the vitreous cavity in RE and anteriorly into the AC in LE with secondary glaucoma.

Acknowledgement
Dr. Karthikeyan K, Retina Fellow, Department of Vitreoretinal Services, Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Madurai.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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