

Intraoperative optical coherence tomography-guided release of lenticulo-corneal adhesion and lens aspiration in anterior dislocation of lens with corneal edema

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Purpose: To describe the outcome of microscope integrated optical coherence tomography (MiOCT) guided removal of lenticulo-corneal adhesion and intralenticular lens aspiration (ILLA) in cases with anterior dislocation of the crystalline lens and corneal edema. **Methods:** MiOCT-guided ILLA was performed in three eyes of two cases of homocystinuria with spontaneous anterior dislocation of lens and corneal edema. Lenticulo-corneal adhesion was noted intraoperatively, which was not apparent pre-operatively. The lenticulo-corneal adhesion could be successfully peeled using intravitreal forceps and viscodissection with visco-dispersive viscoelastic under the guidance of MiOCT. **Results:** In all cases, the lenticulo-corneal adhesion could be successfully removed without any complication such as Descemet tear or worsening in corneal edema. Improvement in visual acuity was noted in all cases with resolution in corneal edema by 1 week. **Conclusion:** MiOCT-guided ILLA can be extremely useful in cases of lenticulo-corneal adhesion especially in cases with corneal edema.

Key words: Anterior dislocated lens, corneal edema, intralenticular lens aspiration, lenticulo-corneal adhesion, MiOCT

The management of spontaneous anterior dislocation of lens is difficult. The various described techniques for surgical removal of an anteriorly dislocated lens include intralenticular lens aspiration (ILLA), lensectomy and in the bag bimanual irrigation and aspiration.^[1-4] These procedures can be performed in pediatric cases without much difficulty when the corneal clarity is excellent. However, in the presence of corneal edema and lenticulo-corneal adhesion, the surgery becomes challenging. A blind attempt to remove the anterior capsule from the posterior surface of the cornea can result in descemet tear, descemet membrane detachment, and persistent corneal edema.

Microscope-integrated optical coherence tomography (MiOCT) has been reported to be extremely useful in various anterior segment surgeries. In cases of anterior dislocation of the lens with lenticulo-corneal adhesion and the presence of corneal edema, MiOCT can be a useful guide to the surgeon for careful peeling of the adherent anterior lens capsule.

Homocystinuria, an inborn error of metabolism, is an autosomal recessive disorder with elevated homocysteine levels.^[5] Ectopia lentis is the most common ocular manifestation noted in approximately 90% of the patients.^[6] Subluxation is most commonly infero-nasal; however, complete dislocation

of crystalline lens into the anterior chamber has rarely been reported.^[7]

We report the outcome of MiOCT-guided ILLA in two cases of homocystinuria with spontaneous anterior dislocation of the lens with lenticulo-corneal adhesion and corneal edema.

Methods

MiOCT (OPMI Lumera 700 and RESCAN 700, Carl Zeiss, Meditec, Germany) guided ILLA was performed in three eyes of two cases of homocystinuria that presented with spontaneous anterior dislocation of lens, lenticulo-corneal adhesion and corneal edema to the eye emergency. A 14-year-old female presented to the casualty services with bilateral anterior dislocation of the lens with lenticulo-corneal touch and corneal edema [Fig. 1a]. The best corrected visual acuity (BCVA) at presentation was 2/60 and 6/36 in the right and left eye respectively. The IOP was 12 mmHg and 16 mmHg in the right and left eye, respectively. The central corneal thickness (CCT) was 590 μm (OD) and 584 μm (OS) on ASOCT. On screening of other family members, her younger brother had posterior dislocation of the lens in the right eye and anterior dislocation of

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the lens with lenticulo-corneal touch and corneal edema in the left eye (second case) [Fig. 2a]. Ciliary staphyloma was visible superiorly. The BCVA in the right eye was counting finger at 1, and left eye was counting finger at 2 meters with an intra-ocular pressure (IOP) of 13 mmHg and 14 mmHg, respectively. The CCT was 598 μm (OD) and 569 μm (OS). None of the other family members had any ocular abnormality.

Comprehensive systemic examination revealed marfanoid habitus with global developmental delay in both cases. Chest X-ray, electrocardiography (ECG) and echocardiography was normal. Serum homocysteine level was increased ($>65 \mu\text{mol/L}$) with a decrease in serum folate and vitamin B12 levels. A diagnosis of homocystinuria was made.

Both the patients were planned for ILLA under viscoat cover with anterior vitrectomy with primary aphakia under general anesthesia.

Surgical Technique

A single surgeon (PKM) under general anaesthesia performed all surgeries. (Supplemental video 1) Two 0.9 mm clear corneal side port incisions were made at 10, and 2 o' clock position with a 23G microvitoretinal blade (MVR; Alcon Laboratories, Inc.) and the same MVR was moved forward through the lens capsule to create capsular openings of approximately 0.9 mm size near the lens equator. The capsular opening margins but not the entire anterior capsule, was stained with trypan blue dye to avoid difficulty in identification of the capsulotomies, especially in presence of corneal edema. Although difficult to identify the capsulo-endothelial touch on microscope, real time OCT made it possible to identify the capsulo-corneal touch. The micro-adhesions were released by a combination of viscodispersive injection between the corneal endothelium and the anterior lens capsule in order to create a space between corneal endothelium and anterior capsule of the anteriorly dislocated lens [Fig. 3a and b]. After that 0.5% Pilocarpine was injected intracamerally to constrict the pupil. Guarded hydro-dissection and multiple hydrodelineation were performed using balanced salt solution (BSS) to create multiple cleavage planes in the crystalline lens. Lens matter was aspirated by using bimanual irrigation and aspiration with the cannula being placed within the capsular bag through two capsular openings created previously. The cannula of the irrigation probe was helpful in two ways. Firstly, it helped in stabilizing the bag and preventing the posterior displacement of the lens. Secondly, by hydrating the lens matter it ensured easy and fast aspiration of the lens matter in the capsular bag. The procedure was performed using the Centurion® vision system using low settings of IOP (30-40 mm of Hg), a low aspiration rate (24–30 mL/minute), and a moderately high vacuum (300–350 mm of Hg).

Following complete aspiration of the lens matter the remaining capsular bag was removed using a 23 G vitrectomy cutter. The anterior capsule was removed first followed by the posterior capsule. MiOCT was extremely useful in identifying the remaining capsular tags and ensured complete removal of the capsule.

Remaining vitreous strands were removed using a 23G vitrectomy cutter after injecting 0.1 ml Triamcinolone acetate (40 mg/ml) (Aurocort, Aurolab, India). Pilocarpine was reinjected

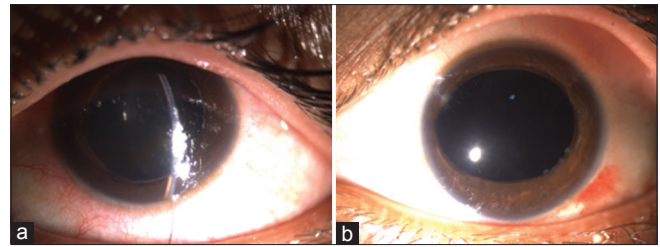


Figure 1: Slit lamp photograph of the first case (a) Pre-operative image showing anteriorly dislocated clear lens with circumcorneal congestion, corneo-lenticular touch, endothelial pigments, and dilated pupil; (b) Post-operative image showing aphakia with the clear cornea and superior-temporal peripheral iridectomy

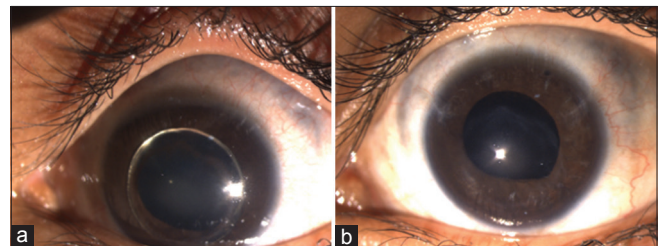


Figure 2: Slit-lamp photograph of the second case (a) Pre-operative image showing anteriorly dislocated clear lens with circumcorneal congestion, corneo-lenticular touch, endothelial pigments dilated pupil, and ciliary staphyloma; (b) Post-operative image showing aphakia with the clear cornea and superior-temporal peripheral iridectomy

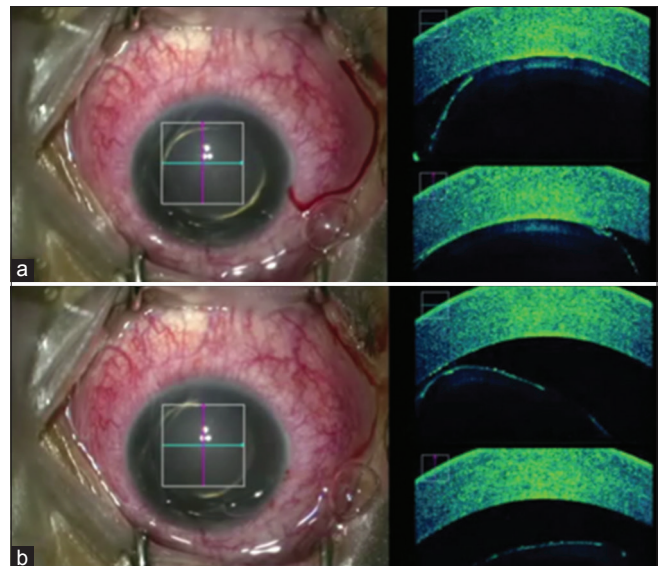


Figure 3: Microscope integrated OCT (MiOCT) image showing (a) Lenticulo-corneal adhesion before commencement of surgery; (b) Release of lenticulo-corneal adhesion intra-operatively under MiOCT guidance

and a peripheral iridotomy was performed. The wound was closed by applying 10-0 monofilament interrupted suture.

In the post-operative period, topical antibiotics (moxifloxacin 0.5% four times a day), topical steroids (prednisolone acetate 1% six times a day tapered over a period of 6 weeks), topical

cycloplegics, (homatropine bromide 2% four times a day for one week), and hypertonic saline 5% four times a day for two weeks was prescribed. The diet of both patients were supplemented with pyridoxine and folic acid along with a low methionine diet in consultation with the pediatrician.

Results

There was no intra-operative complication noted in any case. Post-operative visual rehabilitation was done in both cases with aphakic glasses. The first case achieved a BCVA of 6/12 OU with an IOP of 14 and 18 mm Hg in the right and left eye, respectively. Resolution of corneal edema was noted at 1 week follow up [Fig. 1b]. The second case achieved a BCVA of 6/9 OS with an IOP of 16 mm Hg. A central descemet scar was noted with iris atrophy suggesting long-standing anterior dislocation of the lens [Fig. 2b].

Discussion

Homocystinuria is an inborn error of methionine metabolism, occurring due to deficiency of cystathionine-b synthetase, an enzyme that converts homocysteine to cystathionine. The systemic features noted in this condition include marfanoid habitus, intellectual disability, seizures, osteoporosis and increased risk of thrombosis and atheroma formation. Ocular manifestations include ectopia lentis, myopia, glaucoma, iris atrophy, optic atrophy, and increased risk of retinal detachment. Ectopia lentis occurs in 90% of patients with homocystinuria. In contrast to Marfan's syndrome which is characterized by superotemporal subluxation of the lens, bilateral symmetrical inferior, and nasal subluxation of the lens is characteristic of homocystinuria.

Atypical subluxation of the lens has been previously reported in cases of homocystinuria.^[8] Elkington *et al.* previously reported anterior dislocation of the lens in five cases of homocystinuria which was managed conservatively by repositing the lens and keeping the patient on life-long pilocarpine.^[9] Lens extraction was performed in only one case that had acute glaucoma. Most of the cases had recurrent anterior dislocation of the lens for which repeated repositioning of the lens was required. Shafique *et al.*, in a case series of 20 patients, reported anterior dislocation of the lens in five patients.^[10] However, the management of these cases has not been described by the authors. Also, the presence or absence of corneal edema has not been highlighted in any of the above-reported cases. Both of our cases had an anterior dislocation of the lens with lenticulo-corneal adhesion and corneal edema. In the presence of corneal edema, a blind attempt to remove the adherent anterior capsule of the lens can result in inadvertent damage to the Descemet and endothelium. MiOCT was extremely useful at this step and helped in the removal of the adherent lens capsule with minimal manipulation.

Conclusion

We conclude that MiOCT is a handy tool in the management of cases of anterior dislocation of the lens with lenticulo-corneal adhesion and corneal edema.

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.

Compliance with ethical standards

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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