Endothelial keratoplasty for bullous keratopathy in microcornea with anterior chamber intraocular lens: A clinical report with surgical challenge.

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Abstract

To explain how to treat eyes with an Anterior Chamber Intraocular Lens (AC IOL) and phakic or pseudophakic bullous keratopathy using thin Descemet-Stripping Endothelial Keratoplasty (thin-DSEK) or Descemet Membrane Endothelial Keratoplasty (DMEK), with or without removing the AC IOL. Secondary referral facility contrasting case studies. Descemet membrane endothelial keratoplasty, also known as thin-DSEK, was carried out in pseudophakic eyes fitted with iris-claw AC IOLs (group 1) or in phakic eyes fitted with angle-supported AC IOLs (group 2) in pseudophakic eyes. Except in eyes with insufficient corneal transparency or a significant risk for graft separation, DMEK was frequently carried out in both groups.

The endothelial cell density, postoperative Corrected Distance Visual Acuity (CDVA), preoperative surgical considerations, and complications were also recorded. All of the AC IOLs in group 1 were left in place. 90% of patients in group 2 involved the removal of AC IOLs. At six months, 36% of eyes in group 1 and 90% of eyes in group 2 had CDVAs of 20/40 (0.5 decimal) or better. 20% of eyes experienced graft detachment and 29% experienced de novo or worsening of glaucoma. DMEK made it possible to treat bullous keratopathy in eyes fitted with an AC IOL. If postoperative difficulties are predicted, removal of the intraocular lens may be necessary, but not for surgical purposes.

With DMEK in eyes with a phakic AC IOL and normal visual potential and thin-DSEK in eyes with low visual potential and/or concurrent disease, the surgical strategy may overall aim to minimise postoperative problems. Dutch ophthalmic USA and DORC international BV both employ Dr. Melles as a consultant.

Keywords: Eyes, Corneal transparency, Intraocular lens, Glaucoma.

Introduction

Descemet Stripping Endothelial Keratoplasty (DSEK) is being increasingly preferred in the management of bullous keratopathy. It is a relatively challenging procedure in presence of Anterior Chamber Intra-Ocular Lens (ACIOL) and microcornea with endothelial decompensation. DSEK with various techniques of IOL implantation have been well described in the literature with varying success rate. This case is unique in a sense that DSEK was performed in presence of microcornea as well as ACIOL, which poses a surgical challenge to a clinician. Herein, we report a case of bilateral pseudophakic bullous keratopathy in presence of microcornea with pre-existing ACIOL by DSEK with good visual outcome [1,2].

Case Presentation

A 36-year-old female presented with the complaints of pain, redness, watering, photophobia and diminution in vision both eyes for 6 months. She had a history of prior lens aspiration and Anterior Chamber Intra-Ocular Lens (ACIOL) implantation in next stage after 3 months of lens aspiration elsewhere. Her ocular examination revealed a distant best-corrected-visual-acuity of 6/36 in both eyes. Slit lamp

biomicroscopy revealed microcornea with horizontal diameter of 9 mm and vertical diameter of 8 mm from limbus to limbus. There was circumcorneal congestion, epithelial and stromal oedema with multiple bullae suggestive of pseudophakic bullous keratopathy both eyes. Superficial vascularisation was seen from 6 to 8 'o' clock in both the eyes. The anterior chamber depth was 2.73 in right eye and 2.72 mm in left eye; measured using Haag-Streit Pachymeter. There were no cells and flare visible. On distant direct ophthalmoscopy, a good fundal glow was seen in all quadrants. However, rest of the details could not be visualised. Digitally intraocular pressure appeared to be normal. Preoperative specular endothelial counts were 1241 and 1020 per mm² counts. Based on the evaluation and the clinical findings, patient was diagnosed as a case of pseudophakic bullous keratopathy with ACIOL with microcornea both eyes [3].

Patient underwent manual DSEK under local anesthesia in the right followed by left eye at our centre. Donor corneal endothelial counts measured were 2677per mm² right eye and 2823 per mm² left eye. First donor lenticule dissection was carried out on an artificial anterior chamber. After epithelial debridement 300 micron blade was used to mark the depth and lamellar dissection was carried out by curved and flat lamellar dissectors. The recipient bed was marked with a 7 mm trephine

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following which descemetorrhexis was performed using a reverse Sinskey's hook. A sheet glide was introduced into the anterior chamber through 4 mm superior limbal incision. A seven mm trephine was used to obtain the donor corneal button and the endothelial side was marked. The lenticule was placed over the sheet glide with viscoelastic (Viscoat) applied on endothelial side of donor lenticule and inserted into the anterior chamber using a bent 26G needle (cystitome). Subsequently the incision was closed using three 10-0 monofilament nylon sutures.

Continuous irrigation from anterior chamber maintainer was used to maintain the depth intraoperatively. Air was injected into the anterior chamber for apposition of donor lenticule to the host stroma. Corneal massaging was performed for centration of donor graft into the host cornea. Patient was kept in strict supine position for at least 10 minutes following the procedure.

There were intraoperative challenges due to congenitally small cornea, less anterior chamber depth and presence of ACIOL; making the surgery a herculean task. These issues were addressed by adopting certain modifications such as using sheet glide owing to its thin and flexible nature avoiding contact to ACIOL or angle structures and transplanting smaller size of donor graft helped to overcome the difficulties in manoeuvring donor tissue in the recipient eye [4].

Postoperatively, patient was started on tapering dosage of topical steroids (prednisolone 1%), topical antibiotics (moxifloxacin 0.5%) and topical lubricants (Hydroxy propyl methyl cellulose 0.3%) and patient was followed up at regular intervals [5].

The patient was followed up daily for 1 week, weekly for a month and subsequently, monthly visits were continued for 1 year. On 1^{st} visit, the donor lenticule was in situ and well apposed with mild corneal oedema. The IOL was well centred and corneal oedema resolved gradually. At 1 year of follow up, she attained a BCVA of 6/12 in the right eye and 6/24 in the left eye.

Anterior Segment OCT (ASOCT) revealed donor lenticule thickness approximately 153 um RE and 167 um in LE well adhered to host cornea. The cornea was clear with no bullae and ACIOL was well centred as shown in Figure 1. Specular microscopy of right eye measured 2543 per mm² and left eye 2635 per mm². Anterior chamber depth of right eye was 2.55 mm and left eye was 2.45 mm. The IOP was 16 mm and 18 mm Hg with GAT. Fundus examination revealed a healthy optic disc and macula in both the eyes [6].



Figure 1: A, B) Post operative slit lamp images of the clear cornea with lenticule well apposed and ACIOL *insitu*; C) Image of ant segment OCT showing well adhered lenticule.

Results and Discussion

The management of PBK involves replacement of the diseased cornea and the current treatment options available are Penetrating Keratoplasty (PK) and Descemet Stripping Endothelial Keratoplasty (DSEK/DSAEK). DSEK has largely replaced conventional penetrating keratoplasty for managing the endothelial pathologies like aphakic and pseudophakic bullous keratopathy. Presence of ACIOL in smaller cornea, smaller size of lenticule making centration difficult, lesser anterior chamber depth makes intraoperative surgical manoeuvring of the lenticule and its placement tricky. There were associated features of damage to angle structures along with corneal decompensation in our case leading to secondary glaucoma, postoperative chronic uveitis and hyphaema which were taken into consideration while planning DSEK [7].

There have been several studies in the past reporting the successful outcome of combined PK with ACIOL, Iris claw lenses and transclerally fixated IOLs. However, limited studies have reported the outcome of DSEK combined with different IOL fixation techniques. In a case series of 9 patients who underwent DSEK combined with a retropupillary fixated iris claw lens, stable endothelial cell counts and an improvement in visual acuity of 0.6 log MAR units was reported at 6 months follow-up. A case series of 3 patients who underwent femtosecond assisted DSAEK with Fibrin glue assisted sutureless PCIOL has also reported an improved visual acuity post surgery. In a similar fashion, a technique using glued SFIOL has shown lower endothelial cell loss compared to sutured SFIOLs as the securely fixed haptics in the sclera instead of the usage of sutures prevent a spring like effect leading to pseudophacodonesis and provide a more stable configuration to the IOL. Endothelial keratoplasty has been found to be a feasible option for the treatment of bullous keratopathy in presence of ACIOL in a comparative case series by Liarakos et al. Rarely, there have been any report describing the management of PBK in presence of ACIOL by DSEK in a case of bilateral microcornea with operated congenital cataract [8].

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Conclusion

This case was unique and challenging as the surgeon had to face multitude of surgical difficulties at every step. Also, the post operative recovery was gradual due to the smaller size graft and early secondary glaucoma in this case. The decision to continue ACIOL was taken since there was an already preexisting iris schaffing and microcornea with lack of posterior support. At the end of 1 year postoperatively, both the cornea were clear with a stable and well centred ACIOL leading to a good visual outcome. To conclude, DSAEK/DSEK seems a viable option in patients with microcornea who develop pseudophakic bullous keratopathy following cataract surgery and ACIOL implantation where there is absence of capsular support as well as deficiency of iris tissue. However, a large scale randomized control trial especially in terms of graft survival and endothelial cell, is required to validate the use of DSEK as primary modality of treatment in such cases.

Conflicts of Interest

All authors have none to declare.

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