

Late onset Descemet's membrane detachment: 15 years after limbal lensectomy with vitrectomy for ROP

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A case of stage 4B retinopathy of prematurity (ROP) after successful retinal reattachment surgery with maintained vision presented with hazy cornea with spontaneous Descemet's membrane detachment (DMD) 15 years after the surgery, requiring Descemet Stripping Endothelial Keratoplasty (DSEK) to restore vision. There are reports of late spontaneous DMD after phacoemulsification or previous corneal surgeries. This report is unique as there is no published literature of spontaneous DMD after limbal surgery for ROP when searched in PubMed. The immature Descemet's membrane (DM), surgical intervention and changes in immature DM with age would have contributed to spontaneous DMD and warrant a long-term follow-up of premature kids.

Key words: Descemet's membrane tear, DSEK (Descemet Stripping Endothelial Keratoplasty), retinopathy of prematurity

Retinopathy of prematurity (ROP) is a disorder describing an immature vascularization of a developing retina in low birthweight preterm infant.^[1] There is guarded anatomical and functional success of surgeries for stage 4 and 5 ROP.^[2] Scarcity of reports discussing long-term follow-up of surgically treated ROP cases prompted us to report a case of 16 years' outcome of a patient with bilateral ROP stage 4B, who was treated successfully with lens sacrificing vitrectomy in the right eye and went on to develop hazy cornea with spontaneous Descemet's membrane detachment (DMD) 15 years after the surgery, requiring Descemet Stripping Endothelial Keratoplasty (DSEK) to restore vision.

Case Report

A premature male baby, born at 26 weeks of gestation, with birthweight of 828 grams, diagnosed with hyaline membrane disease, managed in incubator for 2.5 months and presented to the vitreoretinal service of a tertiary eye care

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centre for management of stage 4B ROP in both eyes after laser photocoagulation done elsewhere. The retina was dragged anteriorly touching the lens and a lens sacrificing vitrectomy was done for the right eye through the limbal route in a closed chamber. Bimanual dissection with 20 G instruments was done and traction relief was adequate. Left eye had falciform retinal fold and was observed. There was a successful attachment of retina of the right eye at 6 weeks' follow-up with mild temporal dragging of the disc. The patient was regularly followed up thereafter and was given aphakic correction in the right eye. Left eye vision remained poor; at the age of 8 years, his vision was 6/60 in the right eye with +17D spherical correction. The intraocular pressure remained normal.

He presented again after a gap of 7 years, at the age of 15 years with complaints of diminution of vision in the right eye from 15 days with a vision of 2/60 and hazy cornea with DMD [Fig. 1a]. There was no history of trauma or any infection. Air injection was attempted after noticing a tear of Descemet's membrane (DM) [Fig. 1a], but was unsuccessful. DSEK was done after discussing all possible complications and after weighing the risk-benefit ratio. The age matched graft with pre-operative endothelial cell density of 2214 cell/mm² was selected. The donor dissection was performed manually after mounting the donor tissue on Barron's artificial anterior chamber (Katena, USA) and 60:40 'Taco' forceps technique was used for donor insertion. The unfolding of the donor tissue was performed by injection of balanced salt solution and air from the side port and patient was advised strict face-up positioning for first post-operative hours.

Patient was followed up at 6 weeks and subsequently at 6 months, and graft was found to be clinically stable [Fig. 1b] and confirmed by anterior segment optical coherence tomography (TOMEY SS-1000 CASIA OCT) [Fig. 2]. Fundus examination revealed attached retina with dragging of macula and some pigmentary changes at macula [Fig. 3]. Patient maintained a vision of 6/60 with +20 D spherical correction at 6 months post-surgery with normal intraocular pressure.

Discussion

This is a unique case spanning over 15 years post-ROP surgery landing with Descemet's tear leading to corneal oedema. ROP patients develop shallow anterior chamber and can land up in hazy cornea later on; moreover, the surgery through anterior route also theoretically predisposes to corneal decompensation. DM development in foetal eye is completed by 8 months of gestation and our report describes a child born at 26 weeks' gestation suggesting immature DM; the DM changes in thickness in the pre-natal period from 0.6 microns to 4 microns at 6 months and then undergoes stratification of basement membrane followed by post-natal increase in thickness.^[3]

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During the pre-natal period, the banded layer appears with cross-linking bridges. The non-banded layer appears after birth. Several changes were found in the endothelium: flattening out of the initially cube-shaped cells and changes in the intercellular junctions that can be seen from 20 weeks of foetal life. At 20 and 25 weeks of gestation, two layers of cells were observed

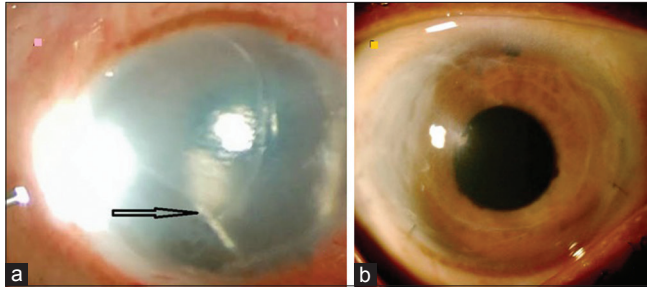


Figure 1: (a): Pre-operative picture of anterior segment of right eye showing hazy cornea with Descemet's membrane tear (black arrow) seen with light pipe illumination. (b): Post-operative picture of anterior segment 1 month following DSEK showing clear cornea

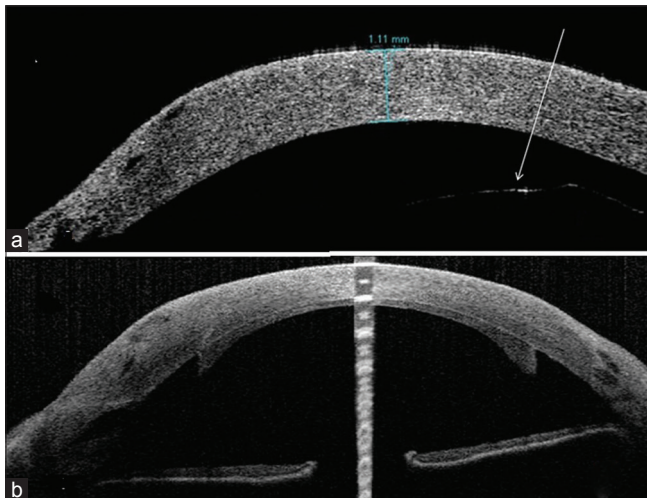


Figure 2: (a) Pre-operative anterior segment-OCT showing Descemet's membrane detachment with increased corneal thickness. (b): Post-operative AS-OCT showing attached graft with reduced corneal thickness

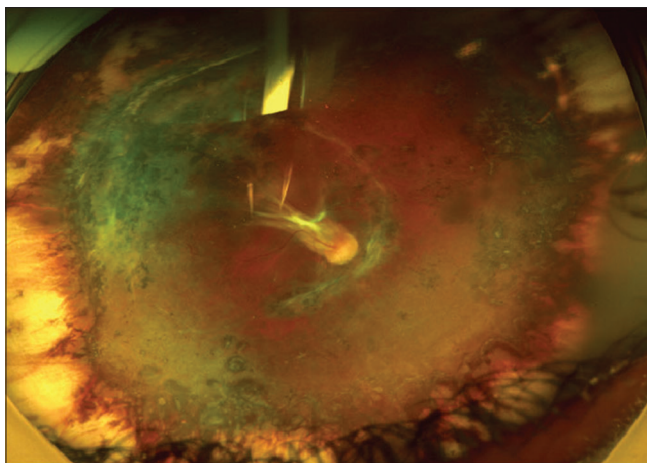


Figure 3: Wide angle fundus photo of right eye at last follow-up showing attached retina with dragging of macula and pigmentary alterations at macula

in the corneal midperiphery. The adult structure of the cornea was reached at about 6 months after birth. Premature birth would have resulted in immature DM, and surgery early in life through limbal route would have further interfered the normal development which could be the possible hypothesis for DM tear in our case, suspecting some trivial unnoticed trauma. Corneal endothelial cell loss during surgical manipulation would have further enhanced the risk along with the age-related attrition. This also explains the unsuccessful DM reattachment following attempted air injection.

Since retina was attached and visual prognosis was good, surgical intervention was planned after weighing the risk-benefit ratio. Scleral fixated intraocular lens was deferred, suspecting a high possibility of traction at the retina in view of previous fibrosis in pars plana area. Late retinal detachments are already a known complication after ROP surgery.^[4] The risk of posterior dislocation of corneal graft possible glaucoma and graft rejection was discussed.^[5]

In summary, we presented a case of Stage 4B ROP exhibiting tractional retinal detachment that was carefully addressed surgically, who was subsequently noted to have excellent anatomical outcome over 16 years of follow-up. The drop in visual acuity due to corneal oedema following spontaneous DM tear and its rectification with DSEK is of particular interest with no reports in literature.

Besides late retinal detachment due to retinal break formation, one need to be cautious in following these patients for corneal complications as well on a long-term basis. DSEK may be an option in such patients developing DM tear.

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.

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Conflicts of interest

There are no conflicts of interest.

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