

## Primary graft failure after big bubble deep anterior lamellar keratoplasty in macular corneal dystrophy

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Macular corneal dystrophy is autosomal recessive dystrophy characterized by deposits of abnormal glycosaminoglycans in stromal lamellae and within endothelial cells. Deep anterior lamellar keratoplasty is successful in the management of this dystrophy. We herein describe three cases of primary graft failure after uneventful big bubble deep anterior lamellar keratoplasty for macular corneal dystrophy.

**Key words:** Deep anterior lamellar keratoplasty, endothelium, macular corneal dystrophy

Macular corneal dystrophy is characterized by the accumulation of glycosaminoglycans in stromal lamellae, within keratocytes and endothelium.<sup>[1,2]</sup> The endothelial changes are hard to discern on clinical slit-lamp examination. A recent report highlighted the utility of ultrasound biomicroscopy in detecting these changes in the posterior membrane level.<sup>[3]</sup> Three variants of dystrophy are described based on immunoreactivity. Despite the Descemet membrane (DM)- endothelial changes, good outcomes have been reported after deep anterior lamellar keratoplasty (DALK) in macular dystrophy.<sup>[4-7]</sup>

We herein report three cases of primary graft failure after uneventful big bubble DALK in macular corneal dystrophy.

### Case Reports

#### Patient 1

A 65-year-old male diagnosed with macular corneal dystrophy [Fig. 1a] had penetrating keratoplasty in the left eye in the year 1997. In 2010, he underwent a big bubble DALK in the right eye. Postoperatively, the vision was counting fingers 1 m. There was no DM detachment on slit-lamp biomicroscope and optical coherence tomography (OCT, Visante). The lamellar graft

showed edema even at 2 months postoperatively [Fig. 1b] and hence a decision to do penetrating keratoplasty was made. The histology of the DM revealed thickening and loss of endothelial cells [Fig. 1c]. Following penetrating keratoplasty, the cornea cleared, and visual acuity improved to 20/30 at 3-month follow-up period.

#### Patient 2

A 58-year-old female with macular corneal dystrophy presented with a visual acuity of hand motions in both eyes. The central corneal thickness was 560 and 552  $\mu$  in the right and the left eye. She underwent big bubble deep anterior lamellar keratoplasty in the right eye. Postoperatively, her vision was to 20/200. The graft was edematous. The DM was well attached as seen on slit-lamp biomicroscope and OCT evaluation. The graft failed to recover even after 6 months. She also developed cataract in her right eye. She was advised to undergo a full-thickness keratoplasty in the right eye along with cataract extraction. As vision in the right eye was still better over the left eye, the patient opted to undergo a penetrating keratoplasty with cataract extraction in the left eye first. The histology of the left eye showed a thickened DM and absent endothelial cells. The stroma showed deposits staining blue with Alcian blue stain. She was advised repeat keratoplasty in the right eye, to be performed electively.

#### Patient 3

A 45-year-old female with macular corneal dystrophy presented in 2011, with a vision of counting fingers 1 m in both eyes. The central pachymetry was 548 and 550  $\mu$  in the right and the left eye, respectively. She underwent an uneventful big bubble deep anterior lamellar keratoplasty in the right eye. Postoperatively, her vision was counting fingers 2 m. DM was well apposed, but there was persistent graft edema, which recovered incompletely over 3 months. At 1 year, the best-corrected visual acuity improved to 20/50, but there was subtle graft edema. The patient maintained the same vision for 5 years after which the visual acuity dropped in both eyes. There were significant cataract and the increased graft edema in the right eye. In view of poorer vision in the left eye, a penetrating keratoplasty was first performed in the left eye. Postoperatively, the vision in the left eye improved to 20/30. In her right eye, she underwent phacoemulsification with intraocular lens implantation with Descemet stripping endothelial keratoplasty. Postoperative vision improved to 20/30 in the right eye.

### Discussion

DALK is a preferred option in corneal dystrophies where endothelium is unaffected. In macular corneal dystrophy,

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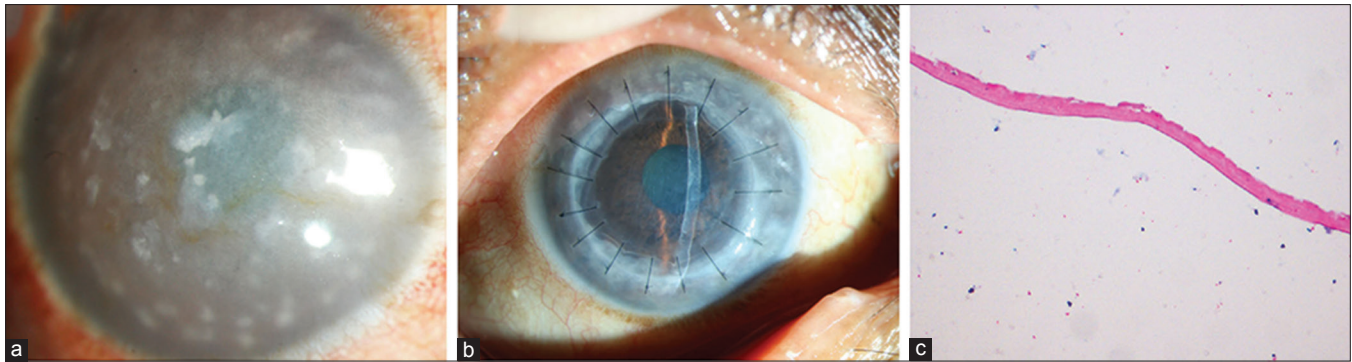
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**Figure 1:** (a) Pre-operative photograph of Patient 1 showing typical stromal deposits suggestive of macular corneal dystrophy, (b) Post-operative photograph of the same patient at 1 month following DALK showing persistent graft edema, (c) Photomicrograph of the Descemet membrane showing thickening and absence of endothelial cells(x10)

glycosaminoglycan deposits are also present in the corneal endothelial cells. However, the correlation between the deposits and endothelial function is not very clearly understood. Kawashima *et al.*<sup>[6]</sup> reported increased endothelial cell loss after DALK in macular corneal dystrophy in comparison to lattice corneal dystrophy. Furthermore, among the 10 cases of macular corneal dystrophy that underwent DALK with a mean age of  $42 \pm 10$  years, they reported two cases of endothelial decompensation at 1.5 and 6 years postoperatively.

In our center, the mean age at which DALK was performed for macular corneal dystrophy was 31 (15–65) years. While those <40 years had a good recovery after DALK (unpublished data), three patients who had a primary graft failure were above 40 years of age. In two of the patients described here, the pachymetry was over  $540 \mu$  (documentation not available in one patient). In those with good visual outcomes after DALK, the mean pachymetry was 462 (range 394–510)  $\mu$ . The deposits in macular dystrophy increase with age at all levels of the cornea, and so the age at surgery may have a bearing on the outcome after DALK. It is likely that endothelial function is well preserved until the deposits become numerous. As the DM was well apposed, it is believed that the endothelial dysfunction may have been subnormal and not necessarily absolute in these three cases. At present, there is no clarity as to what point in time, endothelial function gets compromised in patients with macular corneal dystrophy. Pre-operative pachymetry may be useful in identifying eyes with poorer endothelial function.

## Conclusion

Patients who undergo DALK at an older age for macular corneal dystrophy should be explained about the possibility of suboptimal outcomes. Furthermore, as cataract extraction may be needed post-DALK in these patients, the possibility of secondary endothelial failure should also be discussed. Studies should be carried out to assess the effect of age and pachymetry on outcomes after DALK in macular corneal dystrophy.

## 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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