Atypical presentation of macular corneal dystrophy managed by Descemet stripping endothelial keratoplasty

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Key words: Corneal guttae, descemet membrane, endothelial keratoplasty, endothelium, macular corneal dystrophy

Case Report

A 67-year-old home maker presented with gradual decrease in vision in both eyes for 3 years, more in the left eye compared to the right eye. Corrected distance visual acuity was 20/50 in the right eye and hand motions in the left eye. Slit lamp examination revealed whitish deposits at deep posterior stroma and Descemet membrane-endothelial complex scattered circumferentially in the peripheral cornea [Fig. 1a and b]. The Descemet membrane appeared thick with guttate changes. The lens was cataractous in both eyes. Specular microscopy was not readable in the right eye and showed reduced endothelial cell density and guttae in the left eye [Fig. 1c and d]. Anterior segment optical coherence tomography (OCT, Optovue) revealed a thick Descemet membrane with nodular changes [Fig. 1e and f]. A clinical diagnosis of macular dystrophy was made. Descemet stripping endothelial keratoplasty (DSEK) was performed along with cataract surgery in the left eye [Fig. 2a and b]. The postoperative visual acuity improved to 20/30 in the left eye. The histology of the stripped Descemet membrane showed alcian blue positive deposits along the endothelial cells [Fig. 2c].

Discussion

Macular corneal dystrophy is characterized by accumulation of glycosaminoglycans in stromal lamellae within keratocytes and endothelium.[1] Typically, the initial stage is seen as deposits in the superficial cornea as fleck-like opacities, which later coalesce to reach the limbus, deeper stroma, and the endothelium. On the basis of immunoreactivity, macular dystrophy is categorized into three different types-I, IA, and II.[2] Similar to other reports, we have observed that majority of patients with macular corneal dystrophy have good outcomes after deep anterior lamellar keratoplasty, while a few can develop primary graft failure after deep anterior lamellar keratoplasty.[3-6] This is explained by the relative degree of endothelial involvement in this condition. The endothelial changes are hard to discern on clinical slit lamp examination as the anterior stroma is also involved.

The case described here is unique as the deposits characteristic of macular corneal dystrophy were scattered more toward the peripheral cornea and predominantly localized to the posterior membrane level. The OCT confirmed

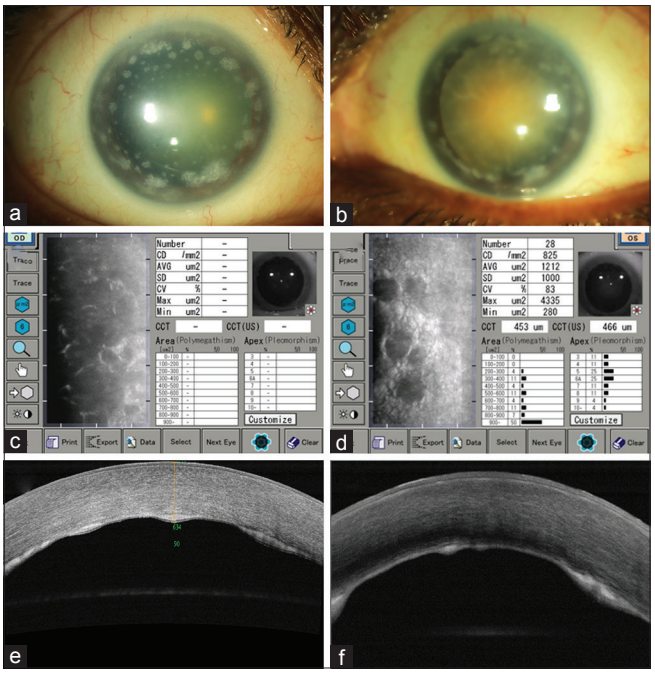

Figure 1: (a) Slit lamp photograph of the right eye showing whitish deposits at the peripheral cornea in a circumferential pattern; (b) slit lamp photograph of the left eye showing similar deposits in the left eye; (c) specular microscopy of the right eye showing a non-readable image in the right eye; (d) specular microscopy of the left eye showing reduced endothelial cell density, increased mean cell area and guttae; (e) anterior segment optical coherence tomography of the right eye showing thickened Descemet membrane and deposits at the posterior membrane level; (f) anterior segment optical coherence tomography of the left eye showing similar features as seen in the right eye

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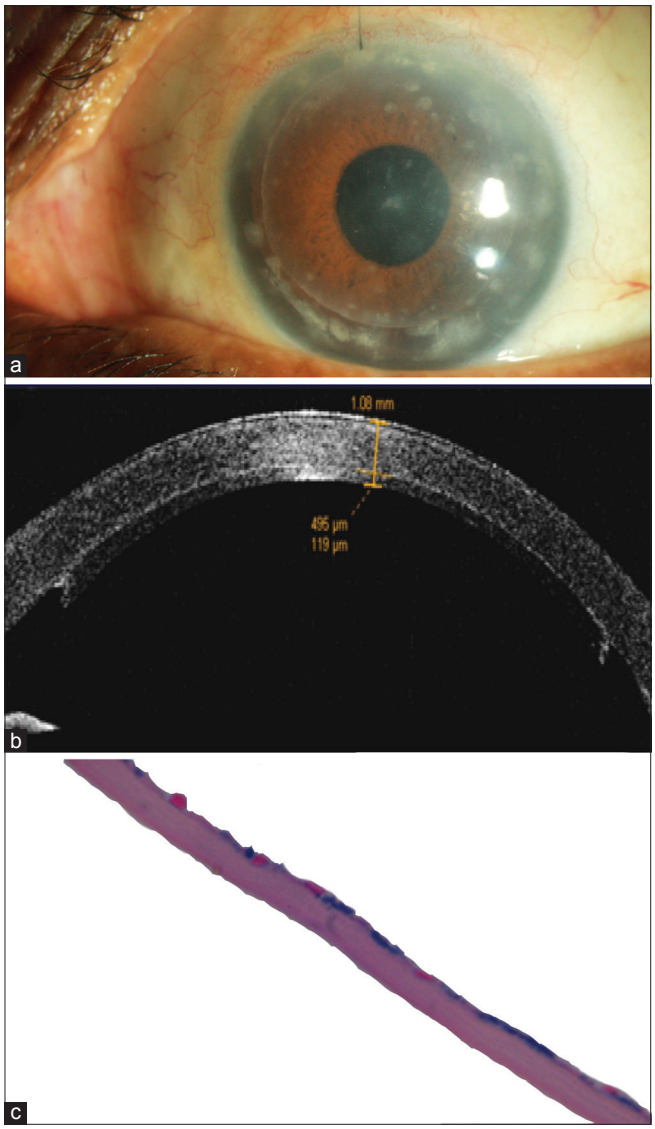

Figure 2: (a) Slit lamp photograph of the left eye on postoperative day 5 after simultaneous phacoemulsification with intraocular lens and Descemet stripping endothelial keratoplasty; (b) anterior segment optical coherence tomography showing the Descemet stripping endothelial keratoplasty graft in place; (c) histology of the stripped Descemet membrane showing alcian blue positive deposits of glycosaminoglycans consistent with the clinical diagnosis of macular corneal dystrophy

the relative paucity of involvement of the anterior stroma. In view of the thickened Descemet membrane and endothelial involvement, a decision of DSEK was undertaken along with cataract surgery in the left eye.

Some authors have proposed that the endothelial changes in macular corneal dystrophy are secondary to stromal lesions, while others believe that they are the primary features of the disease itself.[1] This patient at 67 years of age did not show significant anterior stromal involvement in the central cornea. In addition, the lesions were predominantly located at the posterior membrane level. Hence, it is conjectured that the endothelial changes in macular corneal dystrophy are a primary feature of the disease that occurs variably.

Conclusion

Although the genetics of macular corneal dystrophy is well characterized, the phenotype–genotype correlation needs to be studied. This will help in understanding the disease better and selecting the exact type of keratoplasty required in a specific situation.

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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