EUROPEAN OPHTHALMIC PATHOLOGY SOCIETY (EOPS) MEETING 2022

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Case No. 53137, material distributed: 1 glass slide, PAS stain

Title: Corneal edema occurring 45 years after penetrating keratoplasty in a patient with keratoconus

Clinical history:

Case 1 (glass slide distributed)

A 66-year-old male patient presented with diffuse corneal edema in his left eye. He complained about a slowly decreasing visual acuity and foreign body sensation during the last 6 months. Penetrating keratoplasty had been performed in this eye 45 years ago for the diagnosis of keratoconus. Medical history revealed arterial hypertension, diabetes mellitus type 2, and obstructive sleep apnea.

Visual acuity at presentation was 0.8 in the right eye, and hand motions in the affected left eye. At slit lamp examination, a diffuse edema of the graft was seen. Anterior segment optical coherence tomography (AS-OCT, CASIA-2, Tomey Corporation) showed complete detachment of Descemet's membrane of the graft. A tear of Descemet's membrane could not be detected in any scan of the 360° visualization by AS-OCT. Focal attachments with splitting of DM in several thin layers were visible. Central corneal thickness was 1308 µm, but the stroma was thinned to 478 µm at the graft-host interface.

Penetrating repeat keratoplasty was performed (graft diameter 9.5 mm) and the excised corneal button sent for histological examination. Postoperative visual acuity increased to 0.1 and was limited by corneal astigmatism, cataract, and advanced glaucoma.

Case 2 (Electron microscopy images shown in presentation)

A 56-year-old female patient with keratoconus presented with subacute vision loss in her right eye, which had started 11 days ago. PK had been performed in this eye 29 years ago. Suspecting late graft rejection, treatment with frequently applied corticosteroid eye drops had been started by another clinic.

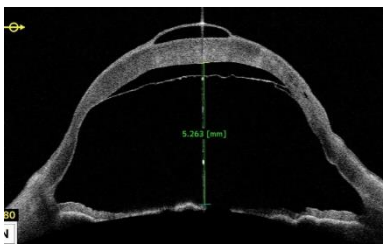
Using AS-OCT, detachment of Descemet's membrane could be visualized. Air injections to re-attach DM were tried twice, but without success. After the air injections, a tear in DM was detected by AS-OCT, which was not seen at time of presentation.

Because of increasing corneal edema, DMEK (8.0 mm graft) with air tamponade was performed. It succeeded without need of further air injections, resulting in clearance of the cornea and increase of visual acuity to 0.3. The removed DM was examined by electron microscopy.

Ophthalmic Pathology:

1. Macroscopy/Gross examination (case 1)

Corneal button, diameter 7 mm, opaque



2. Microscopy (case 1)

The epithelium of the excised cornea shows bullous detachment. The interface angle between donor and host is acute and measures about 35° degrees. Bowman's layer of the host is missing, whereas Bowman's layer of the graft is maintained continuously with a focal break in some slides. The stromal fibers in the host stroma appear loose. Descemet's membrane of the donor cornea is detached and has scrolled edges. Endothelial cells are nearly completely missing in the recipient's rim and are reduced and degenerated in the graft.

3. Transmission electron microscopy (case 2)

Transmission electron micrographs shows a normal anterior banded layer (ABL) and an abnormal posterior nonbanded layer (PNBL) containing abnormal vacuolar inclusions and banded collagenous material (BM) in its anterior and posterior portions, respectively. Lamellar splitting between ABL and PNBL can be observed along the zone of vacuolar inclusions.

Diagnosis

Corneal ectasia with late detachment of Descemet's membrane 45 years after penetrating keratoplasty for keratoconus

Comment

Detachment of Descemet's membrane (DM) is a rare event which can occur spontaneously decades after penetrating keratoplasty. Only about 30 cases have been published so far, but an underdiagnosis might be possible since the detachment cannot always be seen at the slit lamp. Nearly all cases in the literature (except for two) have been described in eyes with keratoconus.[1-14] Usually, DM detachment happens decades after PK: in our case series of 7 eyes, the mean interval between PK and DM detachment was 36 years (range 29-45 years).[14]

Clinically, this condition presents usually as (sub-)acute painless vision loss, but slowly progressive decrease of vision can occur as well (2 out of 7 cases in our series). [14] The most important differential diagnoses are graft failure and graft rejection. However, the acute onset of symptoms is not typically for graft failure due to endothelial cell loss. Graft rejections occur usually in the first months and years after keratoplasty and are very uncommon after decades. Anterior segment optical coherence tomography (OCT) is a useful tool to detect DM detachment and to confirm the diagnosis. We found several typical OCT signs in eyes with late DM detachment after PK: detached donor's DM with focal attachments and splitting into several layers, stromal alterations (loose fibers, caverns), and a thinned and steep (i.e. ectatic) host corneal rim. [14]

The etiology of DM detachment is unknown: mechanical stress as in eye rubbing or floppy-eyelid syndrome might be a risk factor for late DM detachment.[12,13] The exact pathomechanism of late DM detachment remains unclear: Some authors call it "acute hydrops" since ruptures in Descemet's membrane have been found.[8,9] However, histological detection of DM ruptures does not prove this theory since the rupture might take place intraoperatively or during histological processing. We found primary DM tears in AS-OCT scans in 2 out of 7 cases only, whereas alterations of the stroma in the interface region and host's stromal rim were far more common (n=6).[14] Similar stromal clefts and thinning have been described by other authors, too.[4, 9, 10]

Dursun et al examined the removed corneal buttons histologically and found ruptures of DM.[12] However, since they did not provide OCT scans (which were not available at time of publication), it is possible that this was an artifact.[12] DM tears can be induced by air injections as in our case 2 and as reported in the literature.[4]

We further investigated the detached Descemet's membranes, which were removed during DMEK, by transmission electron microscopy. We found a normal anterior banded layer (ABL), but vacuolar inclusions and collagenous material in the anterior and posterior parts of the posterior non-banded layer (PNBL). The lamellar splitting of DM, which had been observed preoperatively by OCT, is located in the zone of vacuolar inclusions between the ABL and PNBL.

Several surgical techniques have been described to treat eyes with late DM detachment after PK: descemetopexy with air or sulfur-hexafluoride gas, repeat PK, DSAEK, DMEK, or partial descemetorhexis with air injection.[1-10] DMEK is technically challenging and the corneal ectasia persists afterwards, but has the advantage of fast visual rehabilitation.

References

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