JAMA Ophthalmology Clinical Challenge

Acute Corneal Edema in a Middle-Aged Patient

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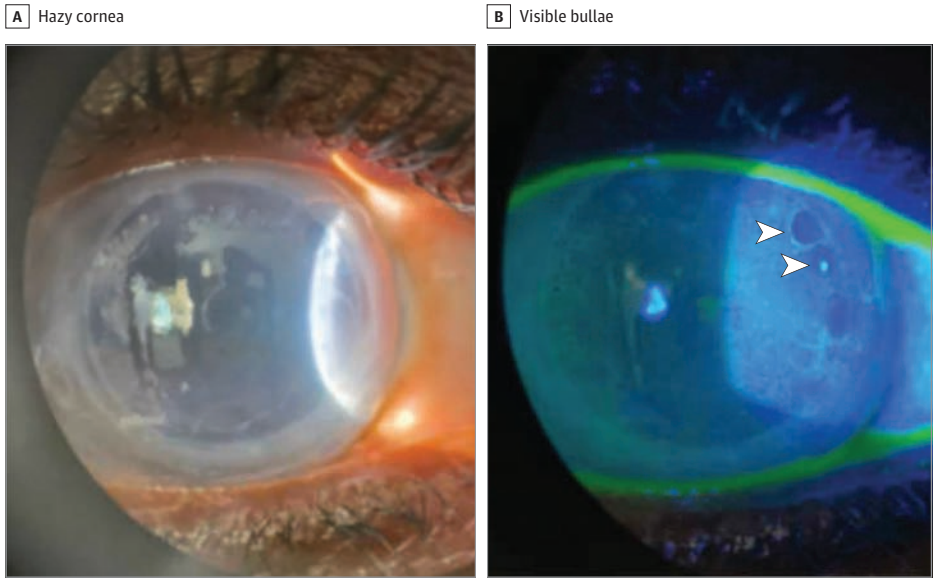

Figure 1. Slitlamp photographs of the right eye demonstrating a hazy post–penetrating keratoplasty cornea (A) and visible bullae (B, arrowheads). Note: the white opacities are likely due to stromal haze and do not represent frank Descemet membrane folds.

A middle-aged patient with history of bilateral penetrating keratoplasty (PKP) performed 20 years earlier for keratoconus presented with pain and blurriness of the right eye for 2 days. The patient reported adherence with the home regimen of prednisolone acetate 1%, 1 drop daily in both eyes for postoperative prophylactic immunosuppression. The patient reported no recent infectious illness or trauma. At a prior clinic visit, both corneal grafts were clear and compact, although both were ectactic. When using scleral contact lens (SCL), their best-corrected visual acuity was previously 20/40 OD.

On current presentation, the uncorrected visual acuity was counting fingers right eye, with no improvement on refraction and inability to tolerate a hard lens. Intraocular pressure was 18 mm Hg in the right eye. The eye had diffuse edema of the ectactic graft with microcystic edema, bullae, and folds in the Descemet membrane (DM) (**Figure 1**). There were no visible keratic precipitates or endothelial rejection lines. The anterior chamber was formed without visible inflammatory cells. Posterior segment examination could not be performed, though B-scan ultrasonography was unrevealing. Because of concern about acute graft rejection, the patient started prednisolone acetate 1% drops every hour (with taper over several weeks) and methylprednisolone, 24 mg (with taper over 6 days). Three weeks later, the patient had no improvement in the edema or vision and reported occasional pain.

WHAT WOULD YOU DO NEXT?

- **A.** Prescribe oral valacycolvir
- **B.** Prescribe the higher-potency topical corticosteroid topical difluprednate
- **C.** Perform anterior segment optical coherence tomography
- **D.** Prescribe a second course of oral corticosteroids

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Diagnosis

Descemet membrane detachment in recurrent ectasia after penetrating keratoplasty

What to Do Next

C. Perform anterior segment optical coherence tomography

Discussion

Initially, this patient with acute onset of corneal edema of a prior PKP graft was suspected to have acute graft rejection. Rejection is routinely treated with corticosteroid therapy (locally and sometimes also systemically). This patient did not respond to aggressive and prompt corticosteroid therapy, and additional corticosteroids (answers B and D) would be unlikely to improve their presentation. Given the preexisting ectasia of the PKP graft, the possibility of DM detachment causing corneal edema (corneal hydrops) was considered. The patient was reexamined, and a subtle DM break was detected on slit-lamp examination. The DM detachment was confirmed with anterior segment optical coherence tomography

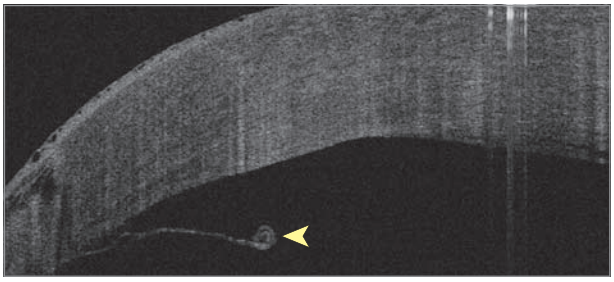

Figure 2. Anterior segment optical coherence tomography of the right eye demonstrating scrolled Descemet membrane (arrowhead) and overlying stromal thickening.

(answer C), establishing the diagnosis (**Figure 2**). Herpetic infection should also be included in a differential diagnosis for acute corneal edema, and this would be treated with oral antiviral therapy (answer A). However, in this patient, the graft ectasia would elevate clinical suspicion for DM detachment over a herpetic etiology.

DM detachment occurs rarely in recurrent ectasia after PKP,[1] so the incidence is unknown. The underlying mechanism for DM detachment after PKP is widely thought to be due to progressive ectactic changes that cause steepening past the elastic tolerance of the DM, ultimately leading to its rupture and subsequent corneal edema.[2] The causal mechanism for DM detachment in postkeratoplasty grafts is unknown, though it is most likely similar to that in treatment-naive keratoconus. As residual abnormal host keratocytes migrate into the corneal graft, they may cause subsequent steepening and tension on the DM within the deformed graft, leading to its rupture. Of note, pain with DM breaks is attributable to corneal edema and may be managed with medical therapy. Interestingly, late-onset DM detachment has been described in patients without ectasia following disruptive procedures such as transscleral cyclophotocoagulation[3] and laser iridotomy.[4] In our patient, the mechanical stresses of SCL use may have precipitated trauma via SCL-induced negative pressure[5] and via the insertion and removal of the lens.[6]

Distinguishing a DM detachment from a herpetic infection or graft failure secondary to endothelial rejection[7] in postkeratoplasty eyes can be difficult in the acute setting. All 3 conditions can be diagnosed clinically, and each can result in stromal edema with accompanying vision loss. Of note, breaks in DM in post-PKP eyes may be difficult to visualize as they tend to occur closer to the graft-host junction,[8] unlike in treatment-naive eyes with keratoconus. Nevertheless, it is important to consider these differential diagnoses and initiate treatment accordingly to avoid further visual complications.

DM detachment in ectactic PKP may sometimes spontaneously resolve or can be treated with pneumatic descemetopexy or repeat grafting (either endothelial keratoplasty or repeat PKP).[1] Pneumatic descemetopexy is less invasive than grafting but has variable outcomes in DM detachment in graft ectasia.[1]

Patient Outcome

The patient underwent pneumatic descemetopexy with incomplete re-apposition of the detached DM. They subsequently underwent a repeat PKP 2 months later.

ARTICLE INFORMATION

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