Sterile Corneal Melt After Descemet Stripping Endothelial Keratoplasty in Patients With Previously Undiagnosed Sjögren Syndrome

Descemet stripping endothelial keratoplasty (DSEK) is an alternative to penetrating keratoplasty in the surgical management of endothelial disorders. The major advantage of DSEK over penetrating keratoplasty is that the integrity of the anterior cornea is largely maintained in DSEK. Hence, patients undergoing DSEK have a lower risk of complications such as suprachoroidal hemorrhage and postoperative traumatic wound dehiscence; these patients regain vision sooner and are more likely to achieve better uncorrected vision.1 Nevertheless, DSEK can be associated with complications, the most frequently reported being graft detachment. Graft rejection and pupillary block may also occur.1 We describe 2 patients who developed sterile corneal melting after DSEK. Medical evaluation revealed previously undiagnosed Sjögren syndrome (SS) as the underlying cause.

Report of Cases. Case 1. An 87-year-old man with corneal edema due to Fuchs endothelial dystrophy underwent DSEK in his left eye. The graft detached postoperatively, necessitating repositioning 2 weeks after DSEK. At that time, the surgeon removed the corneal epithelium using a crescent blade to enhance visualization of the anterior chamber. In the postoperative period, the graft remained attached. However, the patient experienced a delay in corneal epithelial healing. The surgeon noted the presence of significant dry eye syndrome and placed a punctal plug in the lower tear duct. Subsequently, a bandage contact lens was placed to enhance epithelialization. Despite treatment with topical antibiotic agents, lubrication, and oral doxycycline, the patient’s condition worsened. Six weeks after DSEK, the patient was referred to The Wilmer Eye Institute Ocular Surface Diseases and Dry Eye Clinic for further treatment. His visual acuity OS was counting fingers at 6 ft (180 cm). The cornea had significant edema and an area of 30% stromal thinning inferotemporally without an apparent infiltrate (Figure 1). The graft was attached, albeit slightly decentered. Corneal cultures were performed, and the patient was administered topical fortified antibiotics. Although the culture was positive for rare *Propionibacterium acnes*, the clinical findings did not improve with antibiotic drug therapy. Further thinning of the stroma occurred for several days. A detailed review of systems was positive for dry mouth, fatigue, and debilitating joint pains. Serologic testing revealed a significantly elevated C-reactive protein level but no autoantibodies. A right knee aspirate showed a small amount of white blood cells with no crystals. Based on the clinical findings, the patient was diagnosed as having seronegative rheumatoid arthritis and secondary SS. He was administered oral methotrexate and prednisone, as well as topical cyclosporine, 1%, and medroxyprogesterone acetate, 1%. One month later, his visual acuity was 2/200 OS with a failed graft, but there was complete resolution of the ulceration.

Case 2. An 83-year-old man underwent DSEK in his left eye for corneal edema due to complete Descemet membrane detachment after cataract surgery. The DSEK surgery was uncomplicated. However, the surgeon had to scrape the corneal epithelium to improve visualization. The presence of significant dry eye syndrome was also noted by the surgeon, and the lower tear duct was cauterized at the time of surgery. The patient was initially observed with “persistent corneal epithelial defect.” One month after
DSEK, his visual acuity was 20/400 OS. Examination revealed enlargement of the epithelial defect, with 50% stromal thinning nasally and inferonasally, without an apparent infiltrate (Figure 2). Corneal culture was positive for Corynebacterium pseudodiptheriticum in the enrichment broth only. A course of fortified antibiotic agents did not improve the clinical findings; there was further thinning of the stroma and impending perforation. The patient was then referred to The Wilmer Eye Institute Ocular Surface Diseases and Dry Eye Clinic. A review of systems was positive for dry mouth and significant joint problems. Serologic testing showed positive antinuclear, anti-Ro, and antiphospholipid antibodies; a low C3 level; and an elevated erythrocyte sedimentation rate. The patient was subsequently diagnosed as having primary SS. He was admitted to the hospital for pulse intravenous corticosteroid treatment, with transition to oral corticosteroids and hydroxychloroquine sulfate, as well as topical cyclosporine, 1%, and medroxyprogesterone acetate, 1%. One week later, amniotic membrane grafting was performed, and it was repeated 2 weeks later along with a tarsorrhaphy. After 6 weeks of cyclosporine treatment, his visual acuity was counting fingers at 30 cm with a failed graft, but there was no epithelial defect.

Comment. Sterile corneal melt in patients with primary or secondary SS is well recognized² and has been reported after cataract surgery and conductive keratoplasty.³⁴ To our knowledge, these are the first reported cases of corneal melt after DSEK. Both patients had previously undiagnosed SS. This syndrome is known to be widely underdiagnosed, especially in the male population and when ocular findings are the initial symptoms.

The corneal lesions associated with SS are characteristically painless epithelial defects and stromal ulcerations without an apparent infiltrate; they are commonly located in the central or paracentral regions of the cornea. The exact pathogenesis is not fully established and may involve the underlying inflammatory process, aqueous tear deficiency, denervation of the cornea from surgical trauma, or the use of topically administered medications that cause epithelial toxic effects or delayed healing.³ Mechanical epithelial scraping alone can also cause immediate damage to underlying anterior keratocytes, leading to their degeneration,³ and may have contributed to the development of corneal melting in both patients.

Although DSEK is known for maintaining corneal surface integrity, sterile corneal melt can occur, especially in patients with chronic dry eye syndrome. Therefore, it is prudent to perform a detailed review of systems and laboratory investigations as needed to uncover possible underlying collagen vascular disorders before any corneal surgery. Scraping of the epithelium should be avoided in patients with significant aqueous tear deficiency. Because systemic and local interventions are necessary in the treatment of these patients, a rheumatology consultation should be obtained promptly for appropriate management.

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Atropine Penalization for "Rescuing" Patching Failures

The prospective, multicenter Amblyopia Treatment Study I, performed under the direction of the Pediatric Eye Disease Investigator Group, demonstrated that atropine penalization of children with moderate amblyopia yielded improvement similar to that obtained with patching.¹ Among 419 children with visual acuities ranging from 20/40 to 20/100, the mean improvement after 6 months of treatment was 3.16 lines for children treated with occlusion, statistically indistinguishable from the 2.84 lines of improvement for children treated with atropine sulfate. Similar results

Figure 2. Patient 2. Penlight appearance of the left eye showing a large central epithelial defect and inferior and inferonasal thinning of the stroma.