Extensive autoimmune keratolysis with subsequent corneal perforation managed with tectonic endothelial keratoplasty

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ABSTRACT

Purpose: To report a case of corneal perforation secondary to an extensive rheumatologic corneal melt, that was successfully managed via systemic immunosuppression and internal tectonic endothelial keratoplasty (TEK).

Observations: A 55-year-old male with undiagnosed rheumatoid arthritis presented with a progressively enlarging area of peripheral ulcerative keratitis with extensive keratolysis which subsequently perforated despite treatment with oral steroids. The structural integrity of the globe was restored via a combination of cyanoacrylate glue and tectonic endothelial keratoplasty (TEK). This technique provided long term structural support and improved visual acuity.

Conclusions and Importance: TEK grafts represent a viable treatment option in a subset of patients with corneal perforation secondary to an extensive corneal melt. The familiarity and relative ease of the surgical technique along with a lack of corneal sutures represents an alternate technique when compared to full thickness or lamellar keratoplasty. Further, through the use of anterior segment spectral domain optical coherence tomography (SD-OCT) we demonstrate that the donor graft integrated within the host cornea. To our knowledge, this represents the first case in the literature of corneal perforation secondary to an inflammatory corneal melt that was successfully managed with internal tectonic endothelial keratoplasty.

1. Introduction

A number of autoimmune conditions have been reported to result in corneal melts including but not limited to: rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), Stevens-Johnson Syndrome and ocular cicatricial pemphigoid.\(^1,\)\(^2\) Of these systemic autoimmune diseases, rheumatoid arthritis is by far the most common disease associated with keratolysis. The spectrum of ocular disease associated with RA is vast and can vary from mild ocular surface disease to corneal perforation secondary to immunologic corneal melts.\(^3\) The exact pathophysiologic mechanisms which underly corneal melts in RA are not known, but it is hypothesized that a combination of a hostile ocular surface environment in conjunction with dysregulation of inflammatory cytokines can result in spontaneous keratolysis.\(^4\) Because of the potential for vision loss, patients with inflammatory corneal ulcerations and perforations represent some of the most complex and challenging cases faced by ophthalmologists. We present a case of a patient who suffered a corneal perforation as a result of undiagnosed RA. The structural integrity of the globe was restored via internal tectonic support provided through an endothelial keratoplasty.

2. Methods/report of the case

A 55-year-old African American male with past medical history of untreated hypertension and alcohol abuse, presented to the Beaumont Eye Institute with complaints of worsening visual acuity, photophobia, injection and pain in the right eye that had become progressively worse over the last month. Family history was notable for SLE in 2 of his sisters. His presenting visual acuity was 20/200 OD and 20/20 OS, intraocular pressure (IOP) and pupil exam were within normal limits in both eyes. Slit lamp biomicroscopy revealed an injected and chemotic conjunctiva. Corneal examination showed a large (8.0 mm by 3.25 mm) crescent of peripheral ulceration with a ledge of epithelial staining along with areas of stromal neovascularization. This area of corneal melt was notable for a 2 mm area of profound thinning (~95%) inferior and nasally (Fig. 1). The remainder of the anterior segment examination was notable for 2+
cyanoacrylate glue was used to plug the external ostium of the corneal perforation. A 6mm conjunctival peritomy was performed temporally in epithelial edge. The patient was taken to the operating room for urgent keratoplasty (TEK) graft was cut to a diameter of 8.0 mm and was introduced into the anterior chamber using a 30-gauge needle on a sheets glide. Air was injected and the TEK graft was positioned to plug the internal ostium of the corneal perforation without stripping any surrounding Descemet’s membrane. The sclera was closed with 10-0 nylon sutures and the conjunctiva was closed with buried 8-0 polyglactin sutures. The patient was placed in the supine position and admitted to the hospital for an urgent rheumatology consultation. He was subsequently started on an infusion of IV rituximab (1000 mg), oral (60 mg prednisone/day) and topical steroids. He received another IV rituximab infusion two weeks later and was tapered off his oral prednisone over 3 months. Over this period the glue remained intact and the graft remained under the perforation site (Fig. 2A); however, the edge of the graft started to detach temporally at the junction of normal and ulcerated cornea.

The patient was again lost to follow-up and presented one year later. The patient’s visual acuity at that time was 20/100. Slit lamp biomicroscopy revealed no cyanoacrylic tissue adhesive or signs of ocular inflammation. The cornea had re-epithelialized and the TEK graft was well positioned and continued to provide structural support in the area of corneal perforation. However, the temporal edge of the graft was not attached and remained in the visual axis (Fig. 2 B).

3. Discussion

Corneal melt and perforation associated with RA represents a complex disease entity with guarded visual prognosis. In these cases, the primary objective of surgical intervention is to restore structural integrity to the globe. This can be done in a number of ways including through the use of tissue adhesive; however, this approach does not provide long term structural support for large corneal perforations. Consequently, tectonic support was traditionally provided through full thickness corneal patch grafts in cases of large corneal perforations. In the 1950s Paufique described lamellar keratoplasty (LK). LK is often considered the treatment of choice for noninfectious corneal melts because it provides structural stability with a relatively low risk of graft rejection. However the procedure is technically difficult, particularly in cases of large corneal melts and/or large perforations which require that crescentic shaped grafts be cut by hand.

While LK may be the procedure of choice, this case provides evidence that in a select subset of patients, internal structural support is a viable treatment option that provides some significant advantages over traditional full or partial thickness patch grafts. First, endothelial keratoplasty is the most commonly performed corneal transplant procedure in the United States. The widespread familiarity with performing this surgical technique is a significant advantage because it mitigates concerns regarding surgical complexity. Second, few if any sutures are required to secure the TEK graft. This is advantageous in patients, such as ours, who have a history of poor follow-up because the risk of suture related infections is minimized. In addition, sutures which may involve the visual pathway and or induce visually significant astigmatism can be avoided. Third, by providing internal tectonic support, the risk of Stromal neovascularization and lipid keratopathy at the graft-host junction is negligible.

Management of Descemet’s membrane ruptures in keratoconus, pellucid marginal degeneration and keratoglobus via an internal approach has been previously reported. These reports, although conceptually similar, are fundamentally different from our report because of the presence of a full thickness corneal macroperforation. In these previous reports, the authors were able to utilize the host’s corneal stroma to bolster the donor endothelial graft. In our case, support from the host cornea remained a question because of extensive corneal thinning and the presence of a corneal macroperforation. However, anterior segment spectral domain optical coherence tomography (SD-OCT) demonstrates not only support, but integration within the host cornea at the site of the perforation (Fig. 2 C).

Despite successful implementation in our patient, we recognize that there are limitations to the TEK graft technique. First, there is an increased risk of endothelial rejection and graft failure when compared to LK. While this is certainly an important consideration, graft failure is a secondary concern in the setting of a corneal perforation. Further it stands to reason, that using a smaller TEK graft may reduce the risk of endothelial rejection and in our case may have provided a clearer central visual axis. Second, TEK grafts cannot address irregular corneal...
astigmatism that occur as a result of a corneal melt. While LK possesses the potential to mitigate the irregular astigmatism, clinical experience suggests achieving satisfactory visual outcomes using LK is often unpredictable. Third, maintenance of the anterior chamber while inserting a TEK graft is more difficult than conventional endothelial keratoplasty. The use of cyanoacrylic tissue adhesive to plug the perforation to maintain a pressurized anterior segment greatly helps in the insertion of the tissue. Finally, the patient’s visual potential was limited in part by the fact that the TEK graft had detached from the host cornea at the junction of the normal and ulcerative cornea and become edematous. Currently, clear guidelines regarding appropriate TEK graft size and positioning are unavailable. Given the long-term stability, we offered our patient additional surgical intervention to clear the central visual axis (i.e. excision of the edematous corneal TEK graft at the time of cataract extraction). However, our patient declined further intervention given the potential for graft dehiscence. Regardless of these limitations, the TEK graft technique represents a viable alternative to traditional LK and may be a valuable therapeutic option in a select subset of patients.

We are unaware of previous reports which have utilized a TEK graft to restore structural integrity to the globe after corneal perforation. Further, slit lamp biomicroscopy and anterior segment SD-OCT images demonstrate the long-term stability and integration of the graft within the host cornea. Taken together we feel that this novel treatment strategy, in conjunction with systemic immunosuppression, is a viable option in patients with large corneal melts and perforations where LK may be technically difficult or where sutures may impede visual recovery.

Consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

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None.

Fig. 2. (A) Representative post-operative anterior segment photograph demonstrating conjunctival injection and progression of a large area of keratolysis. Cyanoacrylate glue can be seen plugging the external ostium of the previous corneal perforation (arrow). The tectonic endothelial keratoplasty (TEK) graft is well positioned posteriorly and is plugging the posterior ostium. Lastly a bandage contact lens is present. (B) Anterior segment photograph taken approximately one year after surgery, shows that the cyanoacrylate glue is absent, the cornea has re-epithelialized and that the TEK graft continues to provide structural support to the previous site of corneal perforation. (C) Anterior segment spectral domain optical coherence tomography (SD-OCT) taken at 1 year after surgery provides further evidence of corneal re-epithelialization. Additionally, it is difficult to distinguish between donor and host lamella providing compelling evidence of the TEK graft has integrated within the host cornea and continues to provide structural support. Finally, this image shows the temporal detachment of the TEK graft which is edematous and interfering with the central visual axis, likely contributing to the patient’s suboptimal visual acuity.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

No relevant conflicts of interests to report for any of the authors.

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