Acute Hydrops following Penetrating Keratoplasty in a Keratoconic Patient

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Abstract

Purpose: To report a case with history of penetrating keratoplasty (PK) for keratoconus that developed acute hydrops in the recipient and donor cornea.

Methods: A 46-year-old man, with history of bilateral keratoconus, who had undergone corneal transplantation in his left eye, presented with complaints of sudden visual reduction, photophobia, redness and pain of the left eye.

Results: Review of his clinical course, slit-lamp biomicroscopy, laboratory evaluations including confocal microscopy and ultrasound biomicroscopy revealed acute hydrops in the graft. Second corneal transplantation was done for his left eye and pathologic examination confirmed the diagnosis.

Conclusion: Acute hydrops can occur after PK in patients with keratoconus. Although this condition is not common, it should be considered as a differential diagnosis of graft rejection.

Keywords: Acute Hydrops, Keratoconus, Corneal Transplantation

Introduction

Keratoconus is a degenerative, non-inflammatory disease of the cornea, with onset generally at puberty. It is progressive in 20% of cases and can be treated by lamellar or penetrating keratoplasty (PK). Its incidence in general population is reported to be about 1/2000. Changes in corneal collagen structure, organization, and intercellular matrix, and apoptosis and necrosis of keratocytes which prevalently or exclusively involve the central anterior stroma and the Bowman layer, are documented in the literature. These findings that show structurally weakened corneal tissue, are typically seen in keratoconus.

One of the complications of advanced keratoconus is acute hydrops. Affected patients suffer from acute visual loss with pain and photophobia. On slit-lamp examination, conjunctival hyperemia and diffuse corneal edema with intrastromal cystic spaces are visible. It is due to rupture of the endothelial layer and Descemet’s membrane allowing aqueous to enter the stroma and producing marked stromal edema. Corneal edema may last weeks to months and gradual improvement is usually associated with reduction in pain and redness. Corneal scar formation is a late complication of acute hydrops.

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Acute hydrops after corneal transplantation is rare, because cornea is usually harvested from a normal eye and extension of hydrops from recipient tissue to donor cornea is restricted by host-donor interface. Herein, we present a case of acute hydrops after PK in a keratoconic patient.

**Case report**

A 46-year-old man, diagnosed with bilateral keratoconus, presented to us with acute visual loss, pain, redness and photophobia of his left eye. He had undergone PK for the left eye 20 years ago. During these years, he had no problem with his operated eye and his visual acuity (VA) had been acceptable with glasses. One month before recent presentation, his problem had suddenly started and evolved within 10 to 12 hours. Asking his first surgeon, there had been no history of graft rejection and he had not used any topical or systemic medications.

At presentation, best corrected visual acuity (BCVA) of the right eye was 20/30 and of the left eye was counting finger at 1 m. On slit-lamp examination, Vogt's striae and Fleischer's ring with corneal thinning and ectasia were detected in the inferior paracentral area of the right eye indicating keratoconus. The left eye showed diffuse conjunctival congestion with severe corneal epithelial and stromal edema with cystic spaces that involved inferior part of the recipient cornea and a large part of the donor cornea. Also, there was superficial corneal vascularization inferiorly. There were no keratic precipitates (KPs). Because of severe and diffuse corneal stromal edema, it was not possible to evaluate anterior chamber in details (Figure 1). Intraocular pressure, measured by Tonopen XL instrument (Mentor, Inc, Norwell, Massachusetts, USA), was 8 mmHg in the right eye and 10 mmHg in the left eye.

Acute hydrops involving both the recipient and donor cornea was diagnosed. The patient received betamethasone 0.1% eye drop three times a day, homatropine 2% eye drop twice a day and hypertonic sodium chloride 5% four times a day. Three months later, the corneal edema significantly decreased, but because of permanent corneal scar and low VA, repeated corneal transplantation was done for the left eye.

**Results**

**Confocal microscopy**

Confocal microscopy (Confoscan 2, Nidek Technologies Srl, Vigouza, Italy) of the left eye revealed significant intra- and intercellular corneal epithelial edema with diffuse and severe corneal stromal edema and multiple intrastromal cystic spaces (Figure 2A). Also, focal subepithelial hyperreflective areas indicating fibrosis and stromal scar were reported (Figure 2B). A few polymorphonuclear leukocytes were visible in the anterior stroma. No endothelial cell layer was detected.

**Ultrasound biomicroscopy**

A high frequency ultrasound biomicroscope (UBM) equipped with a 50 MHz transducer (Paradigm UBM plus, Model p45; Paradigm Medical Industries, Salt Lake City, Utah) clearly demonstrated separated and widened spaces within the stromal lamellae, a tear in Descemet's membrane, and Descemet's
membrane detachment (Figure 3). Subepithelial fibrotic scar and bullae were also visible.

**Histopathologic examination**

Histopathologic changes were focal disruption and fragmentation of Bowman's layer, which was replaced in affected areas with keratocytes and collagenous material. The epithelium itself was irregular in thickness and had an abnormal basement membrane in areas where Bowman's layer was destroyed (Figure 4A). A fibrotic area and handful polymorphonuclear cells were clearly visible in the central and anterior portion of the stroma (Figure 4B). In posterior stroma, another fibrotic area was seen. Descemet's membrane was detached and retracted and the torn edges rolled anteriorly (Figure 4C).

**Discussion**

Keratoconus is the most important degenerative corneal disorder that is characterized by progressive corneal thinning and ectasia. Sporadic form is the most common, but positive family history has been reported in a small percentage of patients. It can be isolated or associated with other diseases such as Down syndrome, connective tissue disorders, mitral valve prolapse, atopia, vernal keratoconjunctivitis, eye rubbing and use of contact lenses.

Due to significant scar formation or contact lens intolerance, about 10% to 20% of patients need corneal transplantation. In spite of 5 year survival rate of 93% to 97% for corneal graft in these patients, there is a risk of graft rejection especially endothelial one.

Acute hydrops is a complication of keratoconus that is caused by a tear in Descemet's membrane and is manifested by severe and acute edema of the corneal epithelium and stroma. Tear in Descemet's membrane results in aqueous leakage into the stroma. This complication, which occurs spontaneously or after trauma, is seen in 2% to 3% of the patients. The most common trauma is vigorous eye rubbing which is prevailing in patients with Down syndrome or severe ocular allergy.

Corneal edema secondary to acute hydrops is usually self-limited and improves within 6 to 10 weeks leaving stromal scar. Cycloplegics, topical steroids, or non-steroidal anti-inflammatory agents have been suggested to treat acute hydrops. Hypertonic sodium chloride eye drop or ointment is also effective. Recalcitrant ones may last several months and finally leave significant scar and up to sixty percent of them will eventually need corneal transplantation. Sometimes, corneal scar is out of the visual axis and by flattening the corneal surface makes contact lens wearing more convenient.

Herein, we introduced a keratoconic patient that had undergone corneal transplantation about 20 years ago and suffered from acute VA reduction secondary to acute hydrops. Confocal microscopy revealed cystic space that indicated intra- and intercellular epithelial and stromal edema. Since the patient was referred to us after a while, some chronic changes including vascularization of the...
cornea, areas of stromal fibrosis and scar also developed. Ultrasound biomicroscopy and histopathologic examination explicitly confirmed the detachment of Descemet’s membrane from the deep stroma.

Wickremasinghe et al.\textsuperscript{15} reported two keratoconic patients that had acute hydrops, 25 years after corneal transplantation. They suffered from acute visual loss and photophobia. On slit-lamp examination, stromal edema of the transplanted and adjacent recipient cornea was observed. Tear in Descemet’s membrane was not observed in these patients and they supposed that Descemet’s membrane tear and detachment at the donor-host interface was responsible for edema. Also, hydrostatic pressure might lead to fluid penetration into the recipient stromal cornea adjacent to the donor corneal stroma.

Thota et al.\textsuperscript{16} studied on patients with acute hydrops secondary to keratoconus. In histopathologic examination, interlamellar accumulation of fluid was more than intralamellar edema. Cornea responded to this fluid accumulation through formation of a cellular layer around these cystic spaces. This reaction is actually the response of avascular cornea to limit fluid dissemination and finally elimination of it.

Using UBM, Nakagawa et al.\textsuperscript{17} concluded although the presence of tear in Descemet’s membrane is a main factor for acute hydrops in keratoconic patients, it is not the single cause and other factors including intrastromal cleft formation are important. Intrastromal clefts are confirmed in all patients with acute hydrops. They concluded that UBM is helpful for detection of Descemet’s membrane tear and intrastromal clefts, especially when such tears can not be detected by slit-lamp biomicroscopy. These clefts may happen simultaneously or immediately after tear in Descemet’s membrane. In terms of resolving edema, the presence of cleft between Descemet’s membrane and corneal stroma postpones membrane reattachment and results in longstanding edema. Furthermore, intrastromal clefts expose large stromal surface to the anterior chamber and facilitate corneal edema.

One interesting finding in our patient was corneal vascularization that apparently developed after acute hydrops. Feder et al.\textsuperscript{18} pointed out the presence of the corneal vascularization as a rare complication of acute hydrops and considered hydrops adjacent to the limbal area with intrastromal cleft as a risk factor for development of corneal vascularization.

Several studies\textsuperscript{19-21} reported recurrence of keratoconus in transplanted cornea, even 22 years after surgery. In one case of recurrent keratoconus, acute hydrops occurred 20 years after surgery.\textsuperscript{22} In our patient there was no history of recurrence of keratoconus and he had good vision before hydrops.

**Conclusion**

Although there are few reports of acute hydrops in transplanted cornea of keratoconic patient, to the best of our knowledge, this is the first report of documented Descemet’s membrane detachment using UBM.

Differentiation between acute hydrops and graft rejection is very important because approach to these situations is completely different. In acute hydrops, although stromal scar may remain after resorption of the edema, there is no place for high dose of steroids and the condition is self-limited.

**References**