Urretz-Zavalia syndrome after lamellar keratoplasty converting to penetrating keratoplasty

Penetran Keratoplastiye Dönülen Lamellar Keratoplasti Sonrası Gelişen Urretz-Zavalia Sendromu

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ÖZ

Bu makalede, Descemet membran rüptürü ile komplike olan ve penetran keratoplastiye dönülen, derin ön lameller keratoplasti sonrası göz içi basınç yüksekliği ile beraber fikse dilate pupillanın (Urretz Zavalia sendromu) geliştiği 2 vaka sunulmuştur.

Keratokonus nedeniyle opere edilen 32 yaşında erkek hasta ve granüler distrofi nedeniyle opere edilen 19 yaşında kadın hastada, Descemet membran rüptürü ile komplike olduğundan penetran keratoplastiye dönülmek durumunda kalınan derin ön lameller keratoplasti sonrası göz içi basınç yüksekliği ile beraber fikse dilate pupilla (Urretz Zavalia sendromu) gelişimi izlendi. Her iki olgu göz içi basıncını düşürmek ve greft saydamlığını korumak üzere tedavi edildiler, iyi sonuç görme keskinliklerinin yanında fikse dilate pupiller düzelmedi.

Sonuç olarak, kornea transplantasyonu geçiren tüm olgular Urretz Zavalia sendromunun erken tanısı için postoperatif dönemde yakın olarak takip edilmelidir. Uygun tedavi ile bu durumdaki sekel riski azaltılabilir.

Key words: Derin ön lameller keratoplasti, fikse dilate pupil, glokom, penetran keratoplasti, Urretz-Zavalia sendromu

ABSTRACT

Herein we report 2 cases of fixed dilated pupilla with intraocular pressure rise (Urretz Zavalia syndrome) that occurred after deep anterior lamellar keratoplasty surgery complicated with Descemet membrane perforation and converted to penetrating keratoplasty.

A 32 year-old male operated for keratoconus and a 19 year-old female operated for granular dystrophy experienced fixed dilated pupillae and intraocular pressure rise following deep anterior lamellar keratoplasty surgery complicated with Descemet membrane perforation and converted to penetrating keratoplasty. Both were treated to lower the intraocular pressure and to save the grafts with satisfactory visual acuity, however fixed dilated pupillae did not resolve.

In conclusion, all patients who underwent corneal transplant should be closely monitored postoperatively in order to timely diagnose Urretz Zavalia syndrome. Appropriate treatment may decrease the risk of sequels in this situation.

Key words: Deep anterior lamellar keratoplasty, fixed dilated pupillae, glaucoma, penetrating keratoplasty, Urretz-Zavalia syndrome

INTRODUCTION

Development of a fixed and dilated pupil with iris atrophy following penetrating keratoplasty (PK) for keratoconus was first described by Urrets-Zavalia in 1963 and named after him as "Urrets-Zavalia Syndrome (UZS)".[1] Ectropion uvea, pigment dispersion, posterior synechiae, anterior subcapsular lens opacities and late rises in intraocular pressure (IOP) are additional features of the syndrome. Although rare, this syndrome has also been reported in nonkeratoconus patients following non-penetrating keratoplasty surgeries such as deep anterior lamellar keratoplasty (DALK), trabeculec-

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tomy, phakic intraocular lens, argon laser peripheral iridoplasty, air-gas injection after DALK.[2-6]

DALK has been widely performed due to many advantages including lack of endothelial rejection risk, shorter need for steroids, and earlier suture removal. Main drawbacks of DALK are the steep learning curve and technical difficulty. The attempt to reach bare Descemet membrane can be complicated by its perforation leading to other complications such as double anterior chamber, or cause the operator to convert to PK surgery.

Herein, we report 2 cases of fixed dilated pupilla with IOP rise that occurred after DALK surgery complicated with Descemet membrane perforation and converted to PK.

SUMMARY OF THE SURGERY

Surgery was started as DALK using the big-bubble technique described by Anwar and Teichmann[7] in both cases under general anesthesia. A Hessburg Barron (JedMed Instrument Co., St. Louis, Missouri, USA) suction trephine (7.00 mm to 7.5 mm) was used for partial thickness trephination of the host cornea up to 60% to 80% depth. A 30-gauge needle bent 60ș (bevel facing downward) at 5 mm from the tip was attached to a 3-cm^3 air-filled syringe. The needle tip was introduced at the edge of the partial thickness trephination, deep into corneal stroma, and gradually advanced to the midperipheral cornea while keeping the tip under direct visualization. A sudden easing of the resistance was accompanied by the appearance of while a whitish circular semiopaque disk (big-bubble). Air was further injected gradually to enlarge this disk, reaching up to the edge of trephination mark. The superficial stromal layer, approximately 40% to 50%, was removed by lamellar dissection using a blade (Mani MGL24, Japan) and a very thin layer of corneal stromal tissue over the large air bubble was left. At this stage, peripheral paracentesis was performed using a knife (Mani MVR21A, Japan)to lower the IOP. A small opening was then created in the stromal tissue overlying the air bubble using a knife (Mani MVR21A, Japan). Anterior stromal layers were then dissected over a blunt iris spatula and introduced into the space created by the big bubble. At this stage both of the eyes had large Descemet membrane perforations that did not allow to continue the procedure so that the perforated Descement membrane was excised and a routine PK surgery using single 10/0 monofilament sutures was performed. Postoperatively topical 0.1% dexamethasone (Maxidex, Alcon, USA) eye drops and 0.3% tobramycin (Tobrex, Alcon, USA) eye drops were instilled at 6-hour intervals.

CASE 1

The 32 year-old male operated for keratoconus was stable on the postoperative first day (Table 1). The graft was clear, the anterior chamber depth and the pupilla were normal. On the second postoperative day, he complained of headache and severe pain in the left eye. The graft was edematous, anterior chamber was shallow, pupil was dilated and unresponsive to light (Figure 1). IOP was 40 mmHg with Goldmann applanation tonometry. He was put on intravenous mannitol 20% and peroral acetazolamide (Diazomid, Sanofi, Turkey) therapy as well as topical dorzolamide-timolol fixed combination (Cosopt, Merck Sharp&Dohme, USA). Laser iridectomy was performed under topical anesthesia. The iridocorneal touch at the superior quadrant resolved and IOP decreased to 26 mmHg. However, the pupil remained dilated and the iridocorneal touch at the inferior quadrant insisted. On the sixth postoperative day the patient was discharged with an IOP of 25 mmHg. On follow-up IOP increased to 32 mmHg despite antiglaucomatous therapy (intravenous mannitol, per oral acetazolamide, topical dorsolamide-timolol fixed combination) so trabeculectomy was performed. IOP decreased to 14 mmHg with no medication after trabeculectomy and remained stable for 23 months.

CASE 2

The 19 year-old female operated for granular dystrophy experienced severe headache at the first postoperative day. The IOP was 45 mmHg on Goldmann applanation tonometry. On biomicroscopic evaluation the anterior segment was shallow, the pupilla was middilated and unresponsive to light with accompanying nasal and inferior peripheric anterior synechias. She was put on intravenous mannitol 20% and acetazolamide (Diazomid, Sanofi, Turkey) therapy as well as topical dorzolamide-timolol fixed combination (Cosopt, Merck Sharp&Dohme, USA). Laser iridotomy under topical anesthesia was attempted however was ineffective. Besides medical treatment the IOP did not decrease under 35 mmHg and the shallow anterior chamber threatened the clarity of the graft. For this reason to resolve the refractory peripheric anterior synechias and to reconstitute the anterior chamber a revision operation under general anesthesia with

Table 1. Pre-, per-, post-operative patient data.

Patient	Age/ Gender	Indication	Big bubble	Intracamaral air/ viscoelastic	Preoperative mannitol	Postoperative cyclopentolate	Highest IOP (mmHg)	Fixed dilated pupilla
AS	32/M	Keratoconus	+	-	+	-	40	Day 2
MK	19/F	Granular dystrophy	+	-	+	-	45	Day 1

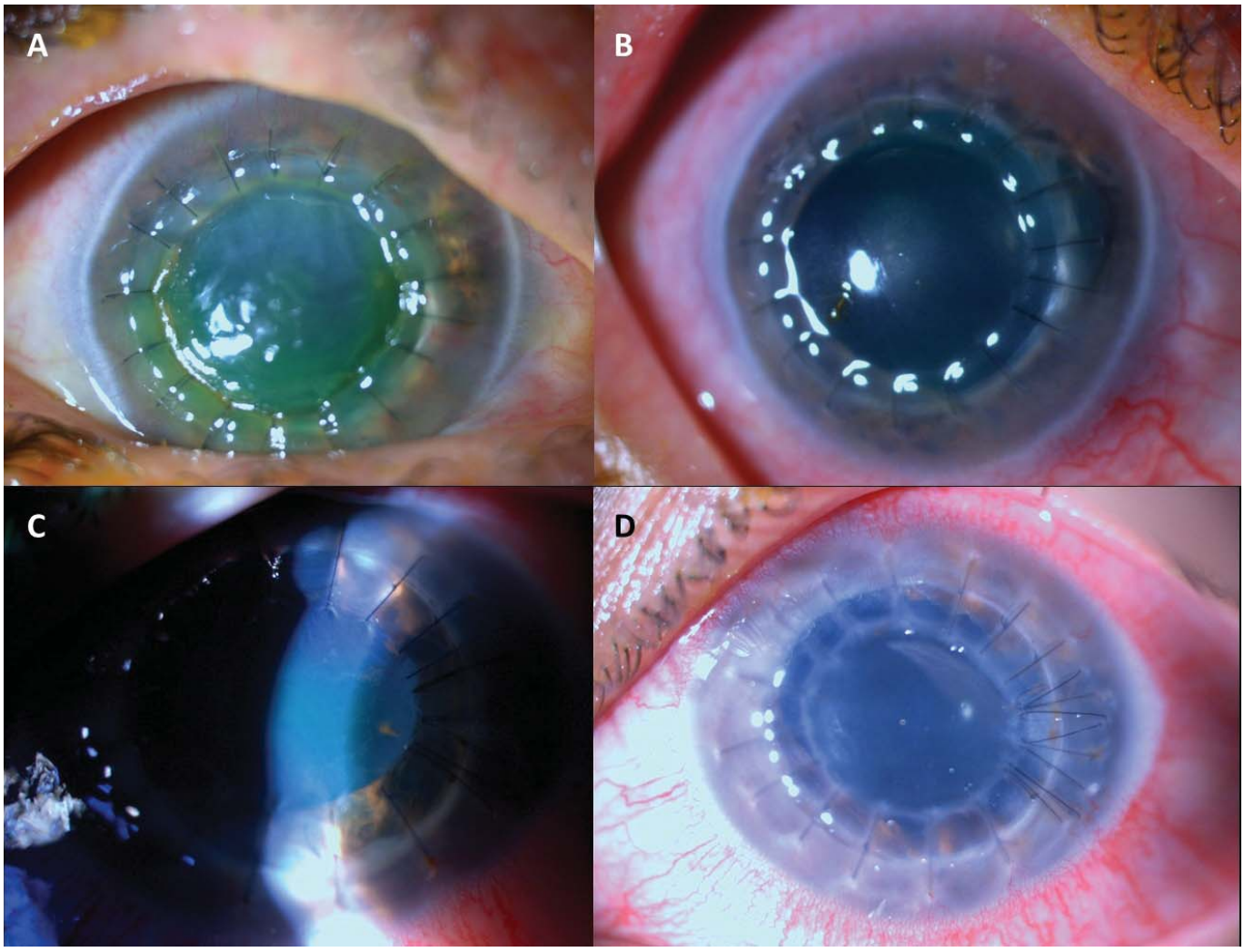

Figure 1. *A 32 year old male operated for keratoconus;* ***A.*** *Edematous graft with shallow anterior chamber is evident.* ***B.*** *Clear graft with accompanying fixed dilated pupilla on the third month of penetrating keratoplasty.*
A 19 year old female operated for maular edema; ***C.*** *Shallow anterior chamber with fixed dilated pupilla.* ***D.*** *Clear graft, normal anterior chamber depth with a fixed dilated pupilla after viscodissection.*

viscoelastic material was performed. After this operation IOP dropped to 18 mmHg with no antiglaucomatous medication and the peripheric anterior synechias resolved. However, the pupilla remained fixed dilated. The patient is stable for 18 months with no need of antiglaucomatous therapy.

DISCUSSION

The exact aetiology of UZS is not clear. Most of the proposed mechanisms are related to IOP rise and iris ischemia: ischemia resulting from the compression of iris between lens and cornea, high IOP and low ocular rigidity leading to occlusion of the vessels at the root of iris, direct surgical trauma to the iris vessels in the midperiphery, damage to the radial nerve fibers of the iris, sympathetic nervous system and iris abnormalities.[8]It has also been suggested that strong mydriasis due to atropine causes direct contact between the iris and the peripheral cornea leading to angle closure.[1,9] Herein, both surgeries were started as DALK to form big-bubble and cause a temporary IOP rise. The iris is compressed against crystalline lens by expanding intrastromal bubbles in Anwar's big-bubble technique.[3]Although preoperative mannitol was given, the sudden IOP rise caused by big-bubble formation might overcome the decreasing effect of mannitol and cause iris ischemia. Moreover, after Descemet membrane perforation the urging air to the anterior chamber might trigger a plato iris like mechanism to cause both iris ischemia and IOP rise (Figure 2). What is more the air bubbles might also cause a block in humour aqueous outflow.

As reported earlier the presence of peripheric anterior synechia formation which address severe anterior chamber inflammation also causes IOP rise.[1,4] Plasmoid aqueous occurring as a response to iris ischemia may be another reason for obstruction of aqueous flow to cause IOP increase.[10]

Mydriatic use is reported as an effective method to prevent development of pupillary block glaucoma in these cases. However, it is not successful in every case and might even ease pupillary block in eyes with air bubble in anterior chamber.[4]Therefore, this attempt to avoid pupillary block

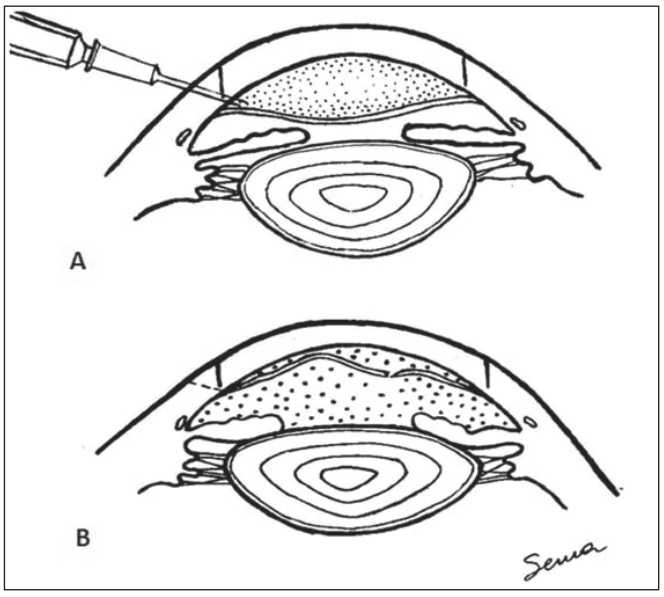

Figure 2. A. *Big bubble formation might cause an IOP rise.* ***B.*** *Descemet membrane perforation causes air to urge into the anterior chamber, which might trigger a pseudopupillary block to cause both iris ischemia and IOP rise.*

may in fact act as a trigger for development of UZS.

The following recommendations may decrease the incidence of UZS. First, hyperosmotic agents lower IOP and shrink vitreous body. Therefore, during air injection or bubble formation, there is more chance for the crystalline lens to sag backward; the risk of iris compression between air bubble/posterior layer of big bubble and crystalline lens is reduced. We routinely give 200 mL of 20% mannitol 30 minutes prior to surgery to patients, unless contraindicated. Also, preoperative application of mydriatic drops are reported to decrease the risk of temporary pupillary block during surgery.

All patients that have undergone corneal transplant should be closely monitored during the first postoperative week. Timely diagnosis and treatment may decrease the risk of some sequels, such asrefractory posterior synechiae and optic nerve head damage due to high IOP. Whenever the UZS is suspected full dose antiglaucomatous therapy (intravenous mannitol, peroral acetazolamide, and topical antiglaucomatous agents such as dorzolamide-timolol fixed combination) should be started. If possible peripheric iridectomy should be tried to release the pupillary block.However, if the IOP rise persists anterior chamber revision with viscoelastic agent dissection to resolve the refractory peripheric anterior synechias and to relieve the anterior chamber angle should be performed.

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