

Bilateral Fixed Dilated Pupil after Penetrating Keratoplasty

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Abstract

Purpose: To report a patient with Urrets-Zavalía (U-Z) syndrome after penetrating keratoplasty (PKP) in both eyes

Case report: A 22-year-old woman with keratoconus associated with a central full thickness scar in both eyes underwent PKP in her right eye that was postoperatively associated with a fixed dilated pupil and severe fibrin reaction, posterior synechiae and mild anterior sub capsular cataract. However he developed fixed dilated pupil leading to posterior synechiae after PKP in the second eye.

Conclusion: This case shows an intrinsic susceptibility for U-Z syndrome which alerts surgeons in the case of second eye surgery.

Keywords: Urrets-Zavalía Syndrome, Penetrating Keratoplasty, Dilated Pupil, Fixed Pupil, Inflammation

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Introduction

Urrets-Zavalía (U-Z) syndrome was first described as a syndrome consisting of a fixed, dilated pupil with iris atrophy following penetrating keratoplasty (PKP) in 1963.¹ Other manifestations that are not essential for the diagnosis are posterior synechiae, ectropion uvea, pigment dispersion, anterior subcapsular lens opacities and secondary glaucoma syndrome. At first this syndrome had been attributed to the keratoconus. However this syndrome was described after PKP for corneal dystrophy, deep anterior lamellar keratoplasty (DALK), descemet stripping endothelial keratoplasty (DSEK),

trabeculectomy, laser iridoplasty, iatrogenic mydriasis and after implantation of phakic intraocular lenses.²⁻¹⁰

Although Zavalía associated this syndrome with postoperative treatment with mydriatics, then Uribe supposed a process of postoperative spontaneous mydriasis, poorly responsive to miotics, in patients with keratoconus undergoing PKP.¹¹ Thus it could happen where mydriatics had not been prescribed. The precise contributing mechanism has not been known so far but fibrinous uveitis, iris vessel strangulation and pupillary block have been proposed.⁸

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Here in, we report a rare case of bilateral development of U-Z syndrome after PKP.

Case report

A 22-year-old woman with keratoconus associated with a central full thickness scar in her right eye referred to our cornea clinic because of severe visual loss. Slit-lamp biomicroscopy showed no other specific finding (such as any iris abnormality) rather than a central round full thickness scar of about 2 mm in a keratoconic cornea. She underwent PKP because her vision was not corrected even with contact lenses. During corneal transplantation, 7.75 mm donor cornea was sutured to a 7.50 mm bed by sixteen separate 10-0 nylon sutures under general anaesthesia. Although we used viscoelastic device to form anterior chamber and to protect donor endothelium, there was not any uveal prolapse during the operation. There was not any usage of intracameral acetylcholine chloride (Miochol). No mydriatic or miotic drops and mannitol were used during the operation or pre and postoperatively for the patient.

On the first postoperative day, the patient developed a dilated pupil. Subsequently, the patient developed a fixed dilated pupil and severe fibrin reaction and posterior synechiae and mild anterior subcapsular cataract. Although fibrin reaction brought under control by topical and systemic corticosteroids, posterior synechiae was remained without anterior synechiae (Figure 1). Intraocular pressure (IOP) was in the normal range during all follow-up visits. The patient underwent a PKP such as the first operation two years later because of a hydrops scar in the other eye. Again 7.75 donor cornea was sutured to a 7.50 mm recipient donor using sixteen separate 10-0 nylon sutures. During the operation, we used viscoelastic device associated with careful washout at the end of operation, because there was not any iris prolapse, we didn't use mannitol, also no mydriatic or miotic drops were used during the operation or pre and postoperatively, however on the first postoperative day, the patient developed fixed dilated pupil. The patient developed posterior synechiae without anterior synechiae (Figure 2). IOP was in the normal limits during all follow-up visits. Finally

best corrected visual acuity (BCVA) reached to $^{20}/_{40}$ in the right eye and $^{20}/_{30}$ in the left eye.

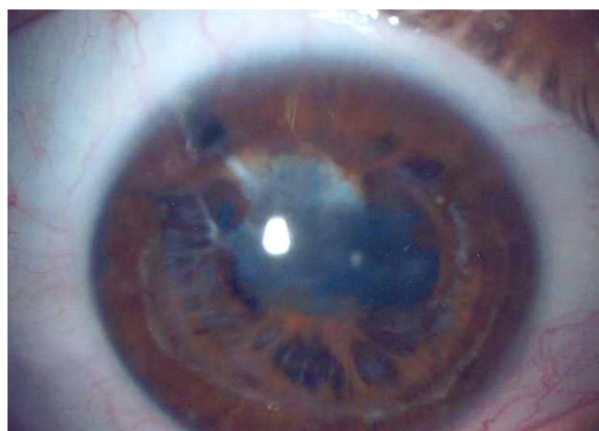


Figure 1. Right eye of the patient

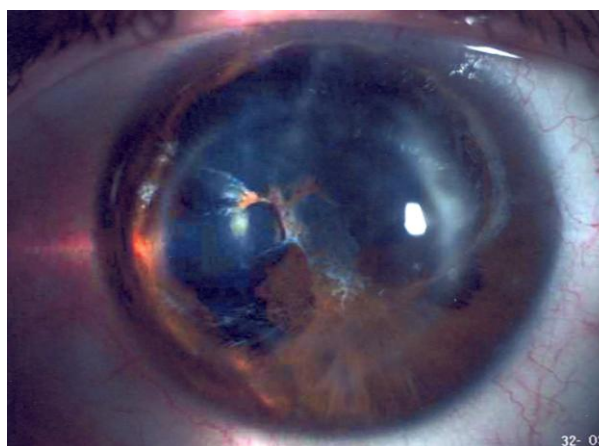


Figure 2. Left eye of the patient

Discussion

The precise contributing mechanism of U-Z syndrome has not been known so far. It was first attributed with mydriatic drops specially atropine drops in keratoconic eyes. Mydriatics could produce this condition by peripheral movement and apposition of the iris and/or strangulation of iris vascular supplies. Other etiologies such as paralysis of local parasympathetic system and injury to iris innervations have been proposed. Ischemic trauma to iris vessels because of rise of IOP or gas injection and mechanical trauma probably by corneal scissors have been suggested.¹²

The other contributing factors could be introducing chemical materials including preservatives in ophthalmic solutions specially mydriatics, viscoelastic materials and

intraocular air/gas injection into the anterior chamber during the operation.¹³

However it has been proposed that there may be an intrinsic iris abnormality in patients with keratoconus predisposing these patients to U-Z syndrome more than the others. Such abnormality may be more prominent in patients with down syndrome.¹⁴ To our knowledge, it was the first reported case of bilateral development of U-Z syndrome in a patient with keratoconus. This case alerts the surgeon about the intrinsic tendency toward this syndrome which necessitates preventive managements during the operation of the other eye. However the exact etiology of U-Z syndrome remains unknown. This may be achieved by the preventive strategies against this syndrome, such as decreasing the positive posterior pressure preoperatively and also intraoperatively lowering the risk of iris prolapse and subsequent iris ischemia.

Therefore, use of a Honan balloon, digital massage, or intravenous mannitol is recommended preoperatively as well as the control of IOP both during the intraoperative and postoperative period. The other recommendation is discontinuation of the use of both intraoperative and postoperative topical atropine¹⁵ and application of mild, short-acting agents instead if necessary and diminishing air/gas tamponade during and after procedures. A careful washout of such materials from the anterior chamber is logical.

Conclusion

The exact etiology of U-Z syndrome is unknown and different mechanisms have been proposed. This may complicate preventive strategies for this syndrome. This case shows an intrinsic susceptibility for U-Z syndrome which alerts surgeons in the case of second eye surgery.

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