OLGU SUNUMU / *CASE REPORT*

Urrets-Zavalia Syndrome After Penetrating Keratoplasty

Penetran Keratoplasti Sonrası Urrets-Zavalia Sendromu

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ABSTRACT

Urrets-Zavalia syndrome (UZS) was first described in 1963 as a case of atrophic dilated pupil accompanied by secondary glaucoma, which developed after penetrating keratoplasty (PKP) using mydriatics in patients with keratoconus. Urrets-Zavalia syndrome is a surgical complication in which the pupil remains fixed and dilated after an intraocular surgical intervention. Although it is thought to be especially related to penetrating keratoplasty, it can also be seen after other intraocular surgical procedures (Trabeculectomy, deep anterior lamellar keratoplasty, cataract, goniotomy). The most important risk factor is iris ischemia due to increased intraocular pressure during or after surgery. The study aimed to present the management of Urrets-Zavalia Syndrome, which developed after penetrating keratoplasty in two patients with left eye corneal granular dystrophy and left eye keratoconus.

Key words: *penetrating keratoplasty; Urrest-Zavalia syndrome; keratoconus; corneal dystrophy*

ÖZET

Urrets-Zavalia sendromu (UZS) ilk olarak 1963 yılında keratokonuslu hastalarda midriyatikler kullanılarak yapılan penetran keratoplasti (PKP) sonrası gelişen, sekonder glokomun eşlik ettiği atrofik genişlemiş pupilla olgusu olarak tanımlanmıştır. Urrets-Zavalia sendromu, göz içi cerrahi girişim sonrası pupillanın sabit ve geniş kalmasıyla oluşan bir cerrahi komplikasyondur. Özellikle penetran keratoplasti ile ilişkili olduğu düşünülmekle birlikte diğer göz içi cerrahi girişimlerden sonra da (Trabekülektomi, derin anterior lameller keratoplasti, katarakt, goniotomi) görülebilmektedir. En önemli risk faktörü, ameliyat sırasında veya sonrasında göz içi basıncının artmasına bağlı olarak oluşan iris iskemisidir. Çalışmanın amacı, sol göz kornea granüler distrofisi ve sol göz keratokonusu olan iki hastada penetran keratoplasti sonrası gelişen Urrets-Zavalia Sendromu'nun yönetimini sunmaktır.

Anahtar kelimeler: *penetran keratoplasti, Urrets-Zavalia sendromu, keratokonus, kornea distrofisi*

Introduction

Urrets zavalia syndrome (UZS) was first defined in 1963 as a syndrome in which a dilated pupil is formed with iris atrophy following penetrating keratoplasty[1]. Although this condition was initially thought to occur only in keratoconus patients, it can also be seen after penetrating keratoplasty (PKP), deep anterior lamellar keratoplasty (DALK), descemet stripping endothelial keratoplasty (DSEK), goniotomy, laser iridoplasty, iatrogenic mydriasis and implantation of phakic intraocular lenses[2].

Other features of this syndrome include ectropion uvea, pigment dispersion, iris atrophy, anterior subcapsular lens opacities, and secondary glaucoma. The incidence of urrets-zavalia syndrome after keratoplasty has been reported to be between 2.2% and 17.7%[1].

The etiology of UZS has not been fully elucidated. Pupillary block caused by intraoperative air or gas tamponed is thought to cause increased intraocular pressure and secondary ischemia in the iris, so the fixed dilated pupil can be seen[3]. Intraoperative or postoperative strong mydriatic use, intraoperative iris trauma, parasympathetic nerve injury, argon laser peripheral iridoplasty, and iris-claw phakic intraocular lens implantation may cause the development of UZS[1,3–5].

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Trauma to the iris caused by trephines or scissors can result in a fixed dilated pupil[6]. While some surgeons think that the pressure behind the lens during PKP may cause permanent damage to the iris and pupil due to the lens being pushed forward[7], some surgeons think this is not the case[8].

Many studies have reported that fixed and dilated pupil development mostly develops on the penetrating keratoplasty background[1,2,4,7,9]. The reason why this situation occurs more frequently after PKP compared to other operations has not been fully explained. It is thought that the application technique of the surgery performed may cause this. It is thought that iris ischemia may occur in PKP, as iris vessels may be more prone to compression in the area of the incision margin of the host cornea[4].

Since the etiology of UZS is not clearly known, it is not possible to prevent its development. It is evaluated that preoperative use of mannitol, ensuring good anterior chamber stabilization during surgery, avoiding iris trauma, complete clearance of viscoelastic substances, and control of postoperative eye pressure can prevent the development of UZS.

Case 1

A 15-year-old male patient was examined for vision loss in both eyes. In the ophthalmological examination, bilateral vision was evaluated as CF 5 meters. His intraocular pressure was in the normal range. In the slit lamp examination, he had bilateral granular dystrophy in the cornea. Keratoplasty was planned for the patient due to corneal dystrophy. Penetrating keratoplasty was performed on the patient's left eye. On the first day after surgery, we observed that he had a fixed dilated pupil in the left eye. No other abnormalities were detected on examination. The patient was discharged with topical droplets: steroid, antibiotic, and autologous serum. In the following visit, visual acuity in the left eye reached 1/10. Intraocular pressure was normal. The slit lamp examination of the anterior segment was normal, and the number of endothelial cells was 1450 mm^2. In the follow-up visit after 2 months of operation, the fixed dilated pupil on the left eye persisted, and posterior synechiae developed. In the consecutive control examinations, fixed pupil dilatation and posterior synechiae remained. The patient who developed an increase in lens opacity in the 6th month was followed up for 1 year. After 1 year from the keratoplasty, the patient underwent cataract surgery, combined lens extraction and intraocular lens implantation into the capsule. In addition, pupilloplasty surgery is performed for fixed dilated pupils. After the procedures, the visual acuity in the left eye was 2/10, and intraocular pressure was 14 mmHg. The graft was clear, and sutures in the iris were intact. After the cataract surgery, the endothelial cell count was 1280 mm^2. Preoperative and postoperative anterior segment photographs of the patient are shared in the Figure 1.

Case 2

A 25-year-old female patient was examined with the complaint of visual impairment in the right eye. In the ophthalmological examination, the visual acuity was CF 5 meters, and intraocular pressure was 11 mmHg in the right eye. In the slit lamp examination, there was keratoconus in both eyes. Penetrating keratoplasty was performed with the diagnosis of left keratoconus. During the procedure, the hypotonic iris was noticeable. On the first day after surgery, we observed that she had a fixed dilated pupil in the left eye. No other abnormalities were detected on examination. Topical pilocarpine 2% was given to the patient to relieve the mydriasis. The patient was discharged with topical droplets: steroid, antibiotic, and autologous serum. In the first month's visit after penetrating keratoplasty, the visual acuity was CF 5 meters, and IOP was normal. The graft was clear, 10.0 nylon sutures were

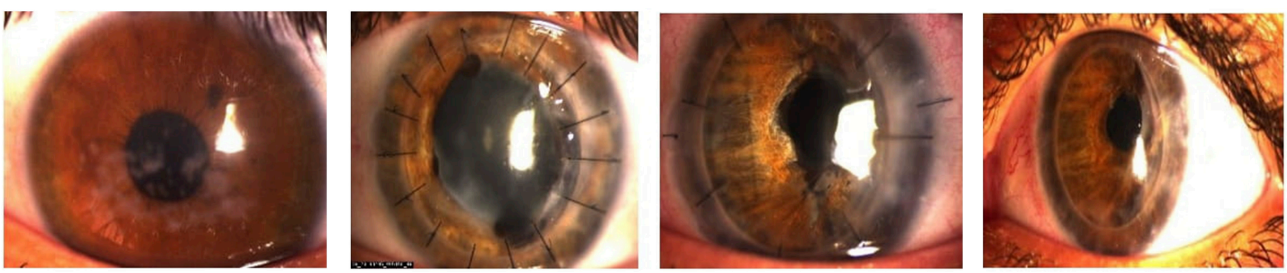

Figure 1. *Preoperative, postoperative 6th month, postoperative cataract and pupilloplasty surgery, the last situation.*

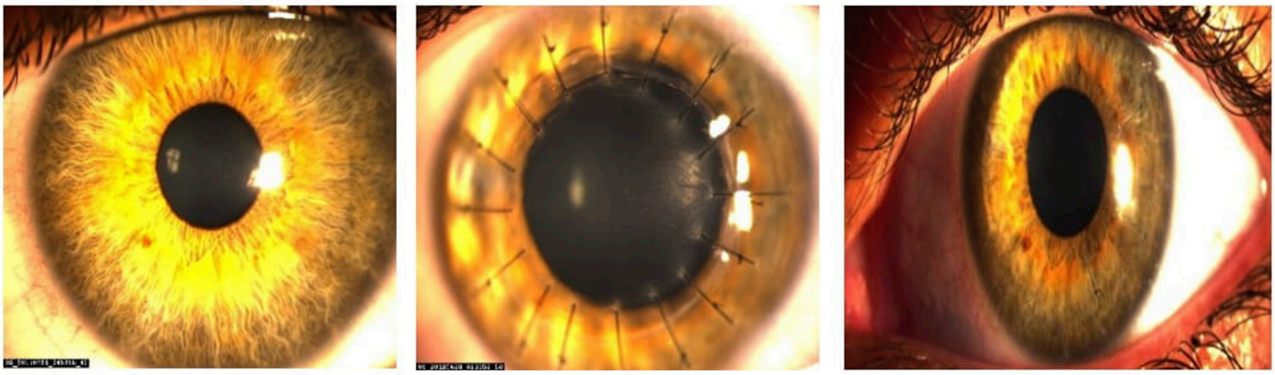

Figure 2. *Preoperative, postoperative 1st day, the last situation.*

intact, and the other structures were normal in the anterior segment evaluation. Apart from this, no additional examination findings were observed, such as posterior synechiae or increased intraocular pressure. Pupil diameter decreased in response to topical pilocarpine treatment. After the 1 year post-op visit, the visual acuity in the left eye was 2/10, and the IOP was 16 mmHg. In the slit lamp examination, the graft was clear, and the pupil size was normal. Other structures were evaluated as normal. The number of endothelial cells was 1750 mm^2. Preoperative and postoperative anterior segment photographs of the patient are shared in the Figure 2. Informed consent forms were obtained from both cases.

Discussion

Although UZS was initially thought to develop only after penetrating keratoplasty in keratoconus patients, it can also be seen after deep anterior lamellar keratoplasty, descement membrane endothelial keratoplasty, goniotomy, laser iridoplasty, iatrogenic mydriasis and phakic intraocular lens implantation[2]. Considering that most of the cases in which UZS developed were phakic, it can be better understood why the cases mostly developed after keratoplasty[10]. Although intraoperative or postoperative intraocular pressure elevation has been confirmed in almost all cases, no clear way to control this situation has been demonstrated[11]. Iridectomy and using hyperosmolar agents have been noted as preventive measures against UZS[2,12]. The frequency of surgical interventions has been reported as 51.8% in PKP, 18.1% in DALK, 8.2% in DSAEK, 8.2% in cataract, 1.9% in trabeculectomy and 0.9% after goniotomy[13,14]. In the cases in which UZS was reported, 42.5% was accompanied by keratoconus, 23.7% stromal dystrophy, 9.4% Fuch's dystrophy, 8.5% plateau-iris syndrome, 8.5% senile cataract, 1.9% primary open-angle glaucoma, 1.9% high myopia, 0.9% congenital myopia[14].

Various mechanisms have been proposed for the etiology of UZS. Urrets-Zavali suggested that the use of atropine during surgery causes the iris to contact the peripheral cornea, leading to peripheral anterior synechiae and secondary glaucoma1. However, a study by Geyer et al. in 1991 concluded that the occurrence of UZS after keratoplasty was unrelated to atropine[15].

Sharif and Casey reviewed data from 100 patients who underwent PKP for keratoconus[16]. They concluded that the incidence of UZS decreased from 4% to 1.5% after a preoperative intravenous injection of 20% mannitol. They noted that the hypertonic solution effectively reduced vitreous volume and compressed the iris.

Although Urrets-Zavalia syndrome is a well-defined clinical entity, it is not considered a preventable condition because its predisposing factors have not been demonstrated. When it develops, there is no specific treatment. However, miotic agents and pupillary reconstruction are used as treatment alternatives. Fixed dilated pupils, which appeared immediately after penetrating keratoplasty, were observed in the patients included in the study. In one of the patients, cataract surgery was performed due to lens opacity, and pupilloplasty was performed in the same session. Another patient's response to pilocarpine was considerable.

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