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CASE REPORT

Urrets-Zavalia syndrome after implantable collamer lens implantation

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Urrets-Zavalia syndrome (UZS), characterized by iris atrophy and a fixed, dilated pupil, is a rare postoperative complication that can occur after ocular surgery. We outline the case of a 19-year-old male patient in good health who was treated with an implantable collamer lens for extreme myopia and astigmatism in both eyes. During surgery, the patient's left eye had a fixed, mid-dilated pupil, and on the first postoperative day, his intraocular pressure (IOP) was elevated. With medical intervention, the high IOP was reduced within three days. Pilocarpine (2%) eye drops proved ineffective to constrict the pupil in the left eye. Up to one year of follow-up period, the pupil remained at a mid-dilated state and was unresponsive to both light and accommodative stimuli, and the best corrected vision was 6/9 in the right eye and 6/18 in the left eye. The patient has been diagnosed with UZS.

Keywords: iris atrophy, dilated pupil, implantable collamer lens, pilocarpine 2%, Urrets-Zavalia syndrome, extreme myopia

Introduction

In 1963, a fixed dilated pupil following ocular surgery, also known as Urrets-Zavalia's syndrome (UZS). This disease was first discovered in individuals who had undergone penetrating keratoplasty for keratoconus. Increased intraocular pressure (IOP) during or following surgery, the use of atropine or other mydriatic drugs, the presence of residual viscoelastic material in the eye, and anterior chamber inflammatory reactions during the postsurgical phase are all potential risk factors for UZS (1, 2).

In addition to this, UZS can also result from keratoplasty procedures like deep anterior lamellar keratoplasty (DALK) and Descemet stripping automated endothelial keratoplasty (DSAEK), cataract surgery, phakic intraocular lens (pIOL) implantation, trabeculectomy, iridoplasty, and goniotomy (2).

To our knowledge, the first association between UZS and implantable collamer lens (ICL) implantation was reported by Kummelil MK et al. (Poster P85, American Society of Cataract and Refractive Surgery, May 25–29, 2011, San Diego) (3). The two available models of posterior chamber phakic intraocular lenses (IOLs) are the phakic refractive lens (PRL) and the ICL (4). The most commonly implanted posterior chamber pIOL is ICL. These lenses are now being widely used as a treatment option for ametropia.

Case presentation

The Bangladesh Eye Hospital & Institute performed refractive surgery on a 19-year-old male patient with severe myopia. It was the first time he had undergone eye surgery. The patient had refractions of −14.00 −0.75 × 10 in the

right eye (OD) and −14.00 −1.00 × 180 in the left eye (OS), resulting in a Best Corrected Visual Acuity (BCVA) of 6/12 in the right eye (OD) and 6/18 in the left eye. A preventive barrage laser was conducted on the left eye after a fundus examination revealed a lattice hole between 12 and 1 o'clock. However, no retinal abnormalities were seen in the right eye. Tonometry revealed that each eye had an IOP of 13 mmHg non-contact tonometry (NCT).

Both eyes' Pentacams reported normal results. ICL (posterior chamber phakic implanted lens) surgery was recommended for the patient (ICL; STAAR Surgical, Nidau, Switzerland).

The right eye was operated on initially. The surgical procedure was carried out under a topical anesthetic. The anterior chamber was filled with 1% sodium hyaluronate, two superior and inferior paracentesis incisions were made, and a 3.2 mm clean corneal incision was made to allow for the implantation of the ICL. Once the ICL had been properly oriented and cared for, it was placed in the anterior chamber, with the footplates above the iris.

The ICL footplates were sequentially positioned behind the iris without applying pressure to the crystalline lens. The viscoelastic material was carefully removed. Following surgery, the patient received topical eye drops containing 1% prednisolone acetate and 0.5% moxifloxacin four times per day. A week after the right eye's smooth recovery, the left eye had an identical treatment. On the first postoperative day, the patient had a fixed, mid-dilated pupil with an elevated IOP of 35 mmHg in his left eye. To treat the patient's high IOP, anti-glaucoma medicines were administered topically (0.5% timolol and 0.2% brimonidine) and orally (acetazolamide).

Both eyes' ICLs are positioned correctly on anterior segment optical coherence tomography (AS-OCT), as seen in **Figure 1** (right eye and left eye).

The IOP normalized on the third postoperative day. On the other hand, the left pupil was found to be mid-dilated, with no direct or consensual reaction to light or restriction to accommodation. Furthermore, eyedrops containing 2% pilocarpine caused no effect. The pupil to the right stayed normal. After 2 weeks of surgery, the IOP was 15 mmHg, and the left eye's uncorrected visual acuity had improved to 6/18. The ICL was properly positioned, as shown by the significant space between it and the crystalline lens. AS-OCT of both eyes indicated a normal vault and a properly positioned ICL (**Figure 1**). The patient had a terrible evening glare following surgery.

Throughout the 1-year follow-up, his left pupil remained mid-dilated and did not react to pilocarpine. However, without the need for medication, IOP remained within normal limits (**Figure 2**).

When the patient's 1-year follow-up period was completed, their right eye vision was 6/9, and all other findings in the right eye were within normal limits. The left eye had 6/18 vision and an IOP of 15 mmHg without treatment; however, the pupil remained mid-dilated. The cause of the mid-dilated pupil in the case is still unknown.

Discussion

Urrets-Zavalia Syndrome (UZS) was initially characterized fifty years ago in individuals who, subsequent to atropine administration, had a fixed and dilated pupil after penetrating keratoplasty for keratoconus. The condition known as UZS is not well understood. Various theories have been proposed to clarify the mechanisms preceding UZS, and it is now widely recognized that iris ischemia is the primary etiology of the condition (1, 5–7). A possible cause of UZS is potentially biochemical changes in iris innervation (7, 8).

According to theories, UZS may be related to

1. Elevated IOP after surgery:
    - Viscoelastic agent retention after phakic IOL implantation (9).
    - Following anterior segment surgical operations such as DALK, DSAEK, and intracameral C3F8 injection for the treatment of acute corneal hydropsies, a pupil block is caused by air or gas bubbles in the anterior chamber (2).
2. Injury to the iris directly during penetrating keratoplasty (2).
3. Intraocular substances forcing the lens-iris diaphragm on the host cornea's edge can cause vascular strangulation (1, 10).
4. Extremely poisonous anterior segment syndrome (11).
5. Hormone toxicity to the iris sphincter or vasculature caused by surgery (12).

These are a few case studies with dilated pupils after ocular surgery, with their results (13).

1. Two distinct case reports with Iris-claw implanted as an IOL were published by Yuzbasioglu E et al. (14) in 2006 and Park SH et al. (10) in 2008. Both patients had no known causes of increased IOP and preexisting corneal disease. Neither patient responded to light or pilocarpine in the initial days after surgery. The pupil was irreversible even after 2–6 months of recovery from surgery, respectively. Li K et al. (15) in 2025 reported on toric ICLs implanted in both eyes. The pupil of the left eye tends to dilate. The cause of pupil dilation was pupillary sphincter paralysis caused by elevated IOP and the excessive size of the toric ICL. The toric ICL was replaced after 2 months and the pupil gradually returned to normal.
2. ICL with a PRL as an IOL was used in two case reports of dilated pupils, reported by Kummelil MK et al. (3) and Perez-Cambrodi RJ et al. (4). The two study participants had no prior history of corneal disease. They did not respond to pilocarpine or light in any way. In both cases, the follow-up results were irreversible at 3 months and 3 years, respectively.

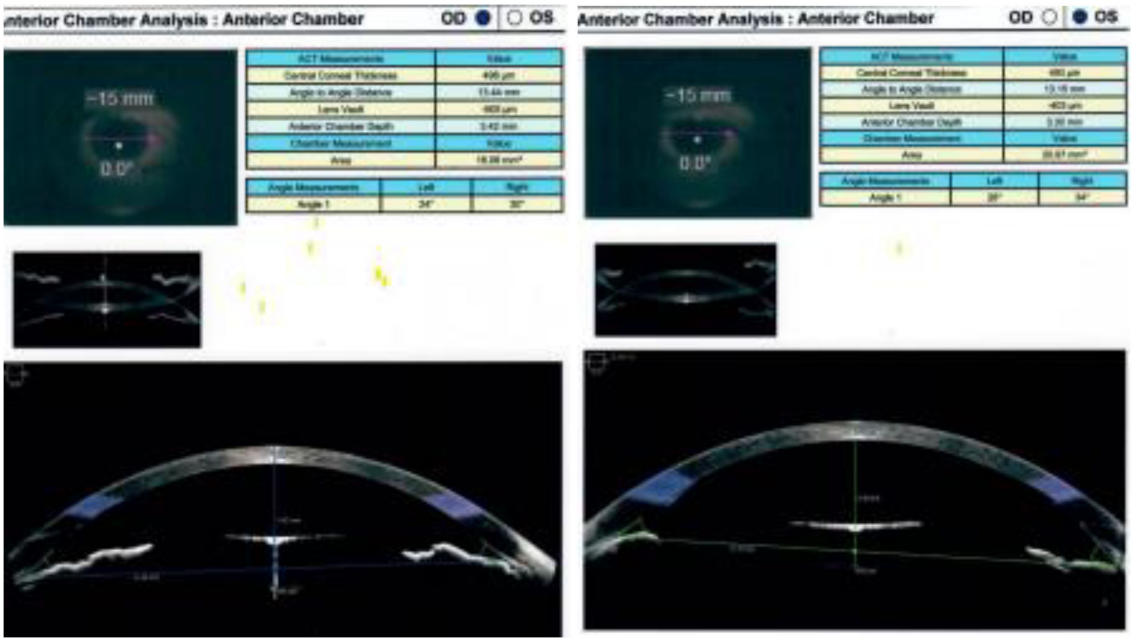

FIGURE 1 | Implantable collamer lenses (ICLs) are positioned correctly on anterior segment optical coherence tomography (AS-OCT), as seen in both eyes.

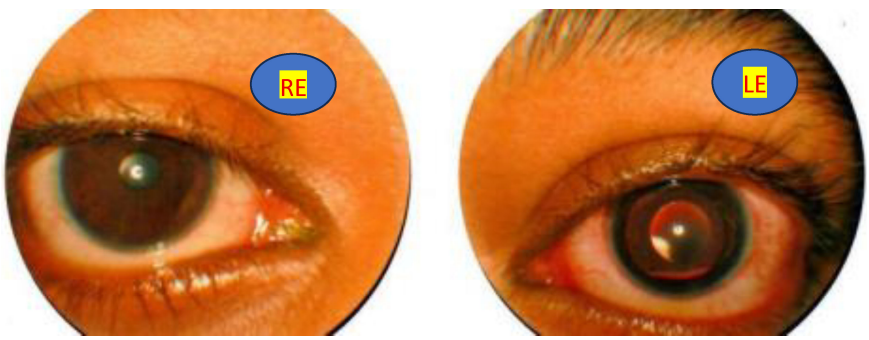

FIGURE 2 | While the right eye's pupil was normal in size at the end of the year-long follow-up period, the left eye's pupil remained mid-dilated.

3. The use of ICL as a phakic IOL is documented in case studies (16, 17). Before their surgeries, every patient in the research was free of corneal diseases. Upon initial recovery, they showed no reaction to either light or pilocarpine. According to the most recent follow-up results, the condition was found to be partially reversible two to three months following surgery (13, 14). Three years following surgery, it was demonstrated that the patient in the Niruthisard D and Kasetsuwan N (13) study had a permanently reversible condition.

Our patient came with extreme myopia, and the routine surgical procedure of ICL implantation was done.

Under topical anesthesia, our patient's implantation surgery proceeded without a hitch. Habash AA et al. (16) reported a case that was comparable to ours. On the day following the surgery, the patient exhibited a non-reactive, moderately dilated pupil in the left eye along with elevated IOP. The IOP in our patient was controlled medically within 3 days. The pupil, however, stayed mid-dilated and fixed. Another two case reports showed that the dilated pupils were not responsive to light and 2% of pilocarpine eye drops (17).

This case study is the first in Bangladesh to explain the UZS after collamer lens implantation. Anterior subcapsular cataracts, elevated IOP, pupillary block, and endothelial cell loss are the main postoperative consequences following ICL implantation. Our case study emphasizes the potential consequence of this posterior chamber pIOL implantation modality: a fixed, dilated pupil in conjunction with higher IOP. Following PIOL implantation, we need to be careful regarding the patient's potential risk of developing UZS.

Ethical approval

The ethical approval has been granted from the ethical review board of the Institute (No. BEHI/2023/N/026)

Funding

Nil.

Consent of the patient

Informed written consent has been taken from the patient to publish the case scenario with images for a journal.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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