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Urrets-Zavalia syndrome following placement of scleral-sutured intraocular lens

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ABSTRACT

Purpose: To report a novel case of Urrets-Zavalia syndrome (UZS).
Observation: A 59-year-old man underwent removal of a dislocated intraocular lens and placement of a scleral-sutured intraocular lens. After surgery, the pupil in the operative eye was dilated, fixed, and unresponsive to constricting drops.
Conclusion: This case expands the known etiology of UZS. Possible preventative measures may include pre-operative screening for plateau iris and intra-operative use of iris hooks instead of pharmacological dilation.

1. Introduction

Urrets-Zavalia syndrome (UZS) is a rare postoperative complication characterized by a fixed and dilated surgical pupil that does not react to miotic drops.[1,2] The pupillary findings can be observed within the first few postoperative days, but onset occurring five months postoperatively has been reported.[1] A common finding in UZS is increased intraocular pressure secondary to angle occlusion and iris atrophy from a compromised blood supply. Here we report a novel instance of UZS developing after a removal of a dislocated intraocular lens and placement of a scleral-sutured intraocular lens.

2. Case report

A 59-year-old man presented to clinic with a recent onset of blurry vision in the right eye, in which he had previously undergone cataract surgery. This ocurred spontaneously, and he denied any preceding trauma to the eye or face. The uncorrected visual acuity was counting fingers in the right eye and 20/20 in the left eye; the intraocular pressure (IOP) was normal bilaterally. Examination of the right eye revealed a dislocated one-piece intraocular lens in the inferior vitreous; there were no retinal tears and the retina was attached. Examination of the left eye was reassuring, revealing a clear crystalline lens with no phacodonesis, and a normal fundus. A decision was made to explant the dislocated intraocular lens; because no lens capsule remnants were noted, sulcus intraocular lens placement was not possible, and a decision was made to implant a scleral-sutured intraocular lens.

On the day of surgery, the eye was dilated using a standard combination of 1% cyclopentolate, 1% tropicamide, and 2.5% phenylephrine. A complete 25-gauge pars plana vitrectomy was performed; of note, the vitrectomy fluid consisted of Balanced Salt Solution, supplemented with a 1000x dilution of epinephrine 1:1000 to maintain pupil dilation. The dislocated intraocular lens was moved into the anterior chamber using intraocular forceps, and then explanted via a 4-mm wide scleral tunnel using the previously described "Twist-and-Out" technique[7] using tying forceps and an iris spatula to protect the corneal endothelium.

The enVista MX60E intraocular lens was then implanted using a previously described technique.[8] In brief, small conjunctival peritomies were made inferotemporally and superonasally. Two sclerotomies were made within each peritomy, 4 mm apart from one another and 3 mm posterior to the limbus, as measured by calipers. The enVista lens was placed on the surgical field and CV-8 Gore-Tex suture was threaded through the opening at the junction between the lens optic and one of the haptics. The suture was then placed into the anterior chamber via the scleral tunnel, and externalized through the inferotemporal sclerotomies using intraocular forceps. This was then repeated with another CV-8 Gore-Tex suture, passed through the opening at the junction between the lens optic and the other haptic, with its ends externalized via the superonasal sclerotomies. The lens was then folded and inserted into the retroirideal space via the scleral tunnel. The Gore-Tex sutures were tightened and tied, the lens was noted to be in the correct orientation and centered, and the scleral tunnel and conjunctival peritomies were

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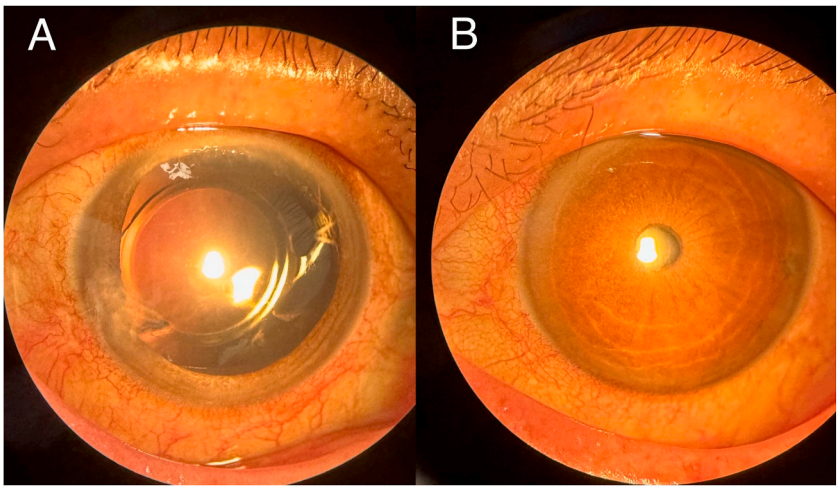

Fig. 1. Anterior segment photographs of the patient's eyes. Neither eye was pharmacologically dilated prior to the photographs being taken. A. Right eye. Note the extreme iris dilation. Corneal haze inferotemporally is from a corneal wound made during the patient's initial cataract surgery. The intraocular lens is centered and in proper orientation, with superonasl sutures visible in the photograph. B. Left eye.

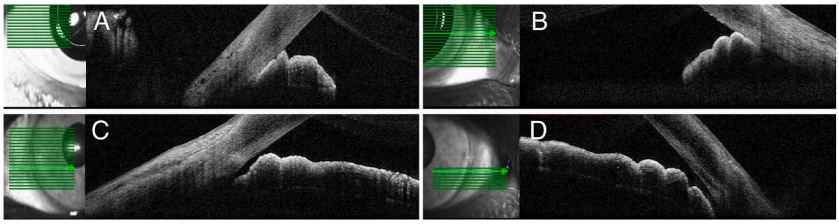

Fig. 2. Anterior segment OCT. A, B. Right eye, showing a permanently dilated pupil and closed angle nasally and temporally. C, D. Left eye, showing plateau iris configuration nasally and temporally.

closed with suture. Carbachol was instilled into the anterior chamber to cause pupillary constriction, but the pupil remained dilated.

On postoperative day 1, the best-corrected visual acuity (BCVA) in the right eye was counting fingers, IOP was 8, and the pupil remained dilated to 7.5 mm. The patient denied any episodes of post-operative severe eye pain, that might suggest a transient increase in intraocular pressure. The patient was placed on a regimen of moxifloxacin and 1% prednisolone acetate drops, four times daily. At the postoperative week 1 visit, BCVA improved to 20/60, IOP was 10, and the right pupil remained dilated. Moxifloxacin was stopped at the end of post-operative week 2 and prednisolone was tapered and stopped by post-operative month 1. The intraocular pressure remained within normal limits, the pupil remained dilated to 7.5 mm, and the patient continued to deny any episodes of severe pain in the operative eye. Throughout this period.

By postoperative month 2, the BCVA improved to 20/25. However, the right pupil remained dilated to 7.5 mm, and IOP was now noted to be elevated to 40. The patient was not in pain, suggesting the increase in intraocular pressure occurred gradually. The patient was started on brinzolamide (10 mg/ml) and Combigan (0.2% brimonidine and 0.5% timolol), as well as 1% pilocarpine.

By postoperative month 3, the IOP had decreased to 30, but the pupil remained dilated to 7.5 mm. The right angle was closed on gonioscopy, with no anterior synechiae noted. The anterior segment exam was otherwise normal, with the intraocular lens centered and in good position, and no signs of infection or inflammation (Fig. 1). Anterior-segment OCT was performed, confirming a closed angle in the right eye (Fig. 2A and B). Of note, the iris in the left eye was found to have a plateau configuration (Fig. 2C and D).

3. Discussion

Urrets-Zavalia syndrome (UZS) is a rare complication of intraocular surgery, in which the surgical pupil remains fixed, dilated, and unresponsive to miotic drops.[1,2] When the condition was described initially, it was considered a complication of penetrating keratoplasty.[3] Later, UZS was observed following deep anterior lamellar keratoplasty (DALK), Descemet stripping endothelial keratoplasty (DSAEK), trabeculectomy, and phakic intraocular lens implantation.[1,3–5] Several cases of UZS following cataract surgery have also been reported, all associated with toxic anterior segment syndrome (TASS).[6]

In this report, we describe the first case of UZS following explantation of a dislocated IOL and placement of a scleral-sutured IOL. At the conclusion of his otherwise uncomplicated surgery, our patient was noted to have a dilated pupil that was unresponsive to carbachol treatment; it remained fixed and dilated post-operatively.

This presentation is consistent with UZS. After conducting a literature review on 10/29/2023 utilizing PubMed using the key words "Urrets-Zavalia syndrome," "secondary intraocular lens," "intraocular lens exchange," and "scleral-sutured intraocular lens," we did not find any prior reports of Urrets-Zavalia syndrome following implantation of a scleral-sutured intraocular lens.

The pathophysiology of UZS remains unclear, and multiple mechanisms have been proposed. Common factors associated with UZS include the use of longer-acting mydriatic medications, post-operative IOP elevation, and placement of an air bubble following corneal transplantation (DALK, DSAEK); all of these have been suggested cause iris ischemia leading to compromised pupil constriction.[2,3,9–12] Our patient developed an inability to constrict the pupil immediately after his surgery, but his IOP remained normal throughout at least his first post-operative month. However, the surgery to replace his dislocated

intraocular lens with a scleral sutured intraocular lens did require a relatively long period of dilation. Furthermore, epinephrine has the additional effect of small vessel constriction,[13] and its use in the vitrectomy fluid may have resulted in intraoperative iris ischemia and thus its subsequent inability to constrict.

Anterior segment OCT imaging at post-operative month 3 revealed a closed angle in the operative eye. Intriguingly, the iris in the fellow eye was in a plateau configuration (Fig. 1B), raising the question of whether a similar iris anatomy in the operative eye contributed to the development of UZS. Plateau iris configuration is characterized by the narrowing of the irido-trabecular angle, due to an anterior iris insertion on the ciliary body or an anterior rotation of the ciliary body itself.[14–16] The close apposition of the peripheral plateau iris to the cornea may predispose peripheral iris vessels to compress with dilation; prolonged dilation – and compression of these vessels - may thus result in severe ischemic damage to the constrictor pupillae muscle and result in iris atrophy, as observed in UZS.[16]

Interestingly, a UZS-like phenomenon has been reported following argon laser peripheral iridoplasty in patients with plateau iris syndrome[17] though the authors note that mydriasis in their patients was likely due to laser-mediated damage to the parasympathetic fibers innervating the constrictor pupillae muscle. The mydriasis in those patients resolved and pupillary responses improved within ~1 year, likely as their nerve fibers recovered; such functional recovery has not been reported in patients with "classic" UZS[1,18] and has not been observed in our patient to date.

4. Conclusion and importance

In summary, we describe a patient who developed Urrets-Zavalia syndrome following replacement of a dislocated intraocular lens with a scleral sutured intraocular lens. Compromised pupil constriction, consistent with UZS, was observed immediately after surgery, leading us to suspect that prolonged iris dilation led to ischemia of the constrictor pupillae muscles. The finding of plateau iris configuration in the fellow eye also suggests that iris anatomy may predispose patients to iris ischemia and UZS with iris dilation. Surgeons may consider screening patients scheduled for pro longed intraocular procedures for plateau iris configuration and in such patients to use mechanical dilation, such as with iris hooks, instead of pharmacological dilation methods.

5. Patient consent

The patient consented to publication of the case.

Claims of priority

After conducting a literature review on (10/29/2023) utilizing PubMed using the key words "Urrets-Zavalia syndrome," "secondary intraocular lens," "intraocular lens exchange," and "scleral-sutured intraocular lens," we did not find any prior reports of Urrets-Zavalia syndrome following implantation of a scleral-sutured intraocular lens.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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