Urrets-Zavalia Syndrome After Posterior Chamber Phakic Intraocular Lens Implantation: An Unusual Complication

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Dear Editor,

Urrets-Zavalia syndrome (UZS), also known as Castroviejo syndrome, is characterized by a fixed dilated pupil and is a recognized complication of various anterior segment surgeries, including cataract surgery, deep anterior lamellar keratoplasty, Descemet stripping automated endothelial keratoplasty, trabeculectomy, iridoplasty, goniotomy, C3F8 injection into the anterior chamber (AC), and phakic intraocular lens (P-IOL) implantation.[1,2] The reported incidence in the published literature ranges from 0% to 17.7%, depending on the type of surgery performed and numerous intraoperative and postoperative factors.[1] The pathophysiology of UZS involves iris ischemia causing sphincter muscle atrophy or damage to the radial parasympathetic fibers that innervate the pupil constrictor muscles. Neuronal injury can result from direct trauma or alteration in the acetylcholine mechanism leading to parasympathetic dysfunction. Atrophy of the iris sphincter muscle may be due to surgical injury, use of mydriatic agents, AC inflammation, and raised intraocular pressure (IOP) which can be secondary to retained viscoelastic material or intracameral gas injection.[1,3,4] This report presents a case of unilateral UZS in a young patient after posterior chamber P-IOL surgery. The unusual presentation and its significant educational value make this case particularly noteworthy. It highlights the need for awareness of this potential complication, in light of the growing popularity of refractive surgeries, to optimize management strategies. Prior to publication, written informed consent was obtained from the patient for the use of his clinical history and images for academic purposes in established medical journals.

A 25-year-old male patient presented with high myopic astigmatism seeking refractive surgical correction. Subjective correction was –12.00 diopters (D)/-2.25 D × 180° in his right eye (OD) and -7.00 D/- 2.00 D × 180° in the left eye (OS), with a best-corrected visual acuity of 6/6 bilaterally. Keratometry readings were 40.25 D @ 171° and 42.25 D @ 81° for OD and 40.75 D @ 176° and 42.5 D @ 86° for OS. Corneal thickness, white-to-white distance, and AC depth measured 529 µm, 12.12 mm, and 3.33 mm in OD and 526 µm, 12.16 mm, and 3.33 mm in OS. Anterior and posterior segment evaluations were unremarkable for both eyes.

Based on these parameters, implantation of the Eyecryl phakic toric aspheric IOL (Biotech Vision Care; Ahmedabad, India) was planned, with the OD operated on first. The surgery was uneventful, and the patient achieved a visual acuity of 6/6 on the first postoperative day. A week later, the OS was operated without any complications. However, within an hour of surgery, the patient reported increasing pain in the OS. IOP was measured at 40 mmHg (applanation tonometry), and slit-lamp evaluation showed corneal edema, a grade 3+ AC reaction, and a mid-dilated, fixed pupil unresponsive to light. Posterior segment evaluation was normal, with no sign of inflammation. An initial diagnosis of toxic anterior segment syndrome (TASS) was made. The patient was prescribed systemic prednisolone (1 mg/kg

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body weight; Omnacortil tablet, Macleods Pharmaceuticals Pvt Ltd, Mumbai, India), acetazolamide (250 mg every 8 hours; Diamox tablet, Sun Pharmaceutical Industries Ltd, Mumbai, India), and homatropine eye drops (twice daily; Homide 2% ophthalmic drops, Indoco Remedies Ltd, Mumbai, India), along with the standard postoperative regimen, which included a combination of topical moxifloxacin (Moxicip ophthalmic drops, Cipla Ltd, Mumbai, India) and prednisolone acetate (Pred Forte ophthalmic suspension, Allergan India Pvt Ltd, Mumbai, India) with lubricating drops. On the first postoperative day, OS visual acuity was limited to counting fingers close to the face, IOP was 24 mmHg, and grade 3+ AC inflammation persisted. The pupil was mid-dilated, irregular, nasally deviated, and nonreactive to light. On the second postoperative day, anterior segment optical coherence tomography was performed to assess the vault and rule out any inadvertent iris capture. The P-IOL was positioned correctly, with a vault of 650 µm, and the AC angle was wide open. Pentacam tomography (Oculus Optikgeraete GmbH; Wetzlar, Germany) confirmed these findings (Figure 1). Given the tomographic finding of an open angle and absence of pupillary block, AC inflammation or retained viscoelastic material were considered the likely causes of the elevated IOP.

With continued topical and systemic treatment, AC inflammation resolved by the seventh postoperative day, and visual acuity improved to 6/9. IOP was 12 mmHg with twice-daily topical timolol (Timolet ophthalmic drops, Sun Pharmaceutical Industries Ltd, Mumbai, India) as the sole antiglaucoma medication. The pupil was mid-dilated, slightly deviated nasally, and sluggishly reactive to light. The patient reported photophobia and night-time glare. On the 14th day, topical pilocarpine (Pilocar 2% ophthalmic drops, FDC Ltd, Aurangabad, India) eye drops were administered every 15 minutes for 1 hour as a trial application, but the pupil remained unresponsive. Surgical mechanical manipulation of the iris followed by intracameral pilocarpine (Carpinol injection, Sun Pharmaceutical Industries Ltd, Mumbai, India) was attempted to restore a regular pupil shape, but the effect was temporary.

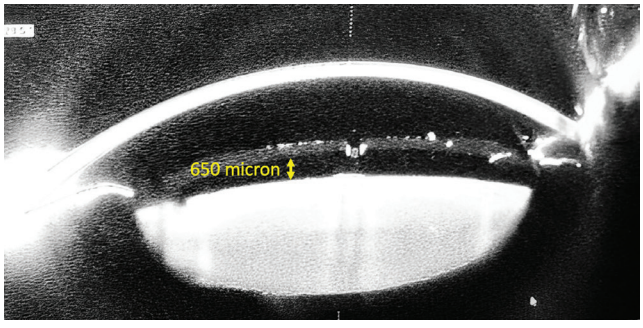

Figure 1. Pentacam tomography (Oculus Optikgeraete GmbH; Wetzlar, Germany) image showing the phakic posterior chamber intraocular lens

By the following day, the pupil had returned to its mid-dilated, nasally deviated position.

A provisional diagnosis of UZS was made. As IOP was within normal range (12-14 mmHg) and baseline glaucoma evaluation showed normal results, the timolol eye drop was stopped and the other postoperative medications were gradually tapered and ceased as per standard protocol. The patient was counselled about the prognosis, and with regular follow-up, symptoms improved significantly. Although the pupillary dilation improved slightly, the pupil maintained a more dilated, nasal configuration. Atrophic patches were seen on the iris, along with pigment dispersion on the P-IOL (Figure 2).

The occurrence of UZS following posterior chamber P-IOL implantation has been sparsely reported in the literature,[4,5,6,7,8] particularly in cases associated with TASS. Potential pathogenic mechanisms for the development of UZS include genetic predisposition to iris tissue injury due to mechanical, neurological, or inflammatory processes.[2] Iris fluorescein angiography in affected patients suggests areas of ischemia and nonperfusion.[9] Although UZS usually manifests unilaterally, rare cases affecting both eyes have been documented, suggesting a potential underlying anatomical predisposition in these eyes.[1,10,11,12] The exact mechanism causing its unilateral or bilateral presentation remains unclear.

In this case, an uneventful surgery was followed by TASS and raised IOP. Both inflammatory and IOP-induced damage have been linked to UZS.[4,8] The association between TASS and UZS in cataract patients has been reported by Nizamani et al.[13] and Ganesan et al.[14], with the latter suggesting that TASS may represent an aborted form of ischemic damage preceding UZS. The clinical events and the P-IOL used in this case closely resembled those reported by Balparda et al.[8] However, unlike their cases, where surgeries were performed at different centers with possible variations in sterilization and handling procedures, the surgeries

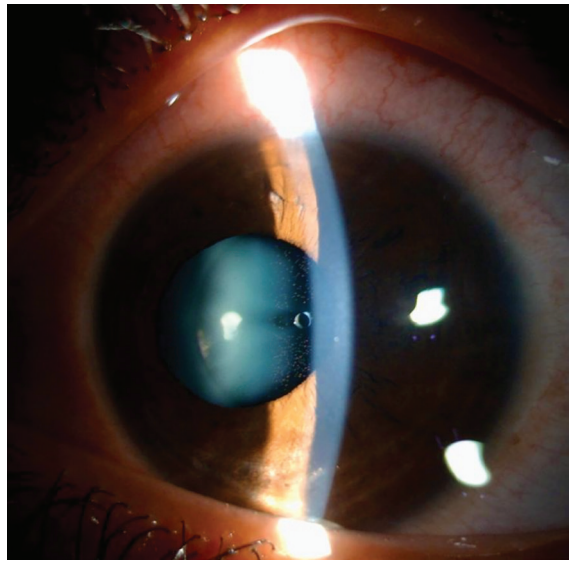

Figure 2. Mid-dilated, nasally shifted pupil, iris atrophic patches, and iris pigment dispersion on the phakic intraocular lens

in this case were performed by the same surgeon at a single center. Early systemic and topical corticosteroid administration controlled the inflammatory cascade by the seventh day in our case. In contrast, cataract formation, endothelial damage, and UZS in their case were likely due to delayed resolution of corneal edema and AC inflammation.

A similar case report of TASS following P-IOL implantation suggested that the etiology may involve viscoelastic residues or an idiosyncratic inflammatory response to intracameral pilocarpine.[7]

Topical pilocarpine has been reported to have a therapeutic role in the UZS pupil, causing its constriction and restoration of light reflex.[4] However, in this case, the pupil did not respond to topical pilocarpine. Given the significant improvement in the patient's subjective symptoms two months post-surgery, any further intervention was temporarily postponed. Pupillary recovery following UZS possibly depends on the spectrum of muscular damage. Patients with marked atrophy of both the anterior and posterior layers of the iris present with irreversible mydriasis. Between one-third and two-thirds of patients with milder damage recover partial pupillary activity within 1 to 18 weeks.[1]

UZS after P-IOL implantation is an uncommon but potentially vision-impairing complication. With the growing popularity of refractive surgeries, it is important to be aware of this clinical entity as a potential complication. Optimal visual outcomes in such cases are dependent on early diagnosis and prompt control of IOP and AC inflammation.

Ethics

Informed Consent: Written informed consent for publication obtained.

Declarations

Authorship Contributions

Surgical and Medical Practices: M.S., A.R., Concept: M.S., Design: M.S., A.R., N.H., Data Collection or Processing: A.R., Analysis or Interpretation: M.S., A.R., Literature Search: M.S., N.H., Writing: M.S., A.R., N.H.

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