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Case Report

Urrets-Zavalia Syndrome After Implantable Collamer Lens in a Systemic Lupus Erythromatosis Patient

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Abstract

Urrets-Zavalia syndrome (UZS) is a rare complication of ophthalmic surgery. In this paper, we report a case of UZS in a young lady who is a known case of systemic lupus erythromatosis (SLE) which was in remission for few months before surgery who underwent a bilateral consecutive implantable collamer lens (ICL) implantation for high myopia. The patient underwent the first surgery in the right eye uneventfully; three days later she underwent the operation in the left eye, and presented in the first post operation day with high intraocular pressure (IOP) and mid-dilated pupil. She was managed properly with intraocular pressure-lowering agents, but unfortunately, the pupil remained dilated after 2 years of follow up, even with all measures taken properly trying to constrict it back.

Keywords: Urrets-Zavalia Syndrome; ICL; Phakic Lens; Implantable Collamer Lens; Implantation; High Myopia; SLE

Introduction

Urrets-Zavalia syndrome (UZS) is characterized by atrophic and mydriatic pupil associated with secondary glaucoma after ophthalmic surgery, although patients may present with peripheral synechiae, posterior subcapsular lens opacity, iris ectropion, pigmentary dispersion, and glaukomflecken [1].

Although It was classically described after penetrating keratoplasty, Other procedures were shown to be associated with UZS; these include trabeculectomy, deep anterior lamellar keratoplasty, Descemet-stripping automated endothelial keratoplasty, cataract surgery, goniotomy for congenital glaucoma, phakic intraocular lens implant, argon laser peripheral iridoplasty, octafluoropropane injection in the anterior chamber to manage acute hydropes and after laser-assisted in situ keratomileusis refractive surgery [1,2].

To the best of our knowledge, in the context of ICL implantation, this syndrome has been only reported three times before [3,4] and this is the first case to be identified in SLE patients who underwent an ICL implantation.

Case Report

25 years old female patient, seeking refractive surgery. She is known case of SLE for the last 7 years and in remission without medications for the previous 12 months. Best corrected visual acuity is 6/9 vision with -18.0 +1.5 x 100 in the right eye and -14.0 + 2.00 x 57 diopter in the left eye. Intraocular pressure (IOP) was 15 mmHg in both eyes. A corneal topography (Pentacam, Oculus Optikgeräte GmbH, Wetzlar, Germany) showed a central corneal thickness of 522 micrometer in the right eye and 518 micrometer in the left eye, with non-suspicious keratometry map and good looking anterior and posterior floats maps. Pupils were regular, round and reactive to light and accommodation in both eyes with a diameter of 4.1 mm in the right eye and 4.3 in the left eye (as shown by Pentacam). Anterior chamber depth was 3.29 mm in the right eye and 3.34 in the left eye. White to white as measured by pentacam was 11.4 mm in the right eye and 11.8 mm. A decision was made to implant a bilateral ICL with central hole in two separates occasions, 3 days apart.

The surgical procedure was performed under topical anesthesia. A standard surgery through a temporal clear corneal incision was made. Given her SLE condition, she was given prophylactic dose of oral steroids (60 mg prednisolone 3 days before the surgery and to be given for 1 week after surgery with abrupt discontinuation of the drug after that). Three days after an uneventful postoperative course in the right eye, the left eye was operated. On the first postoperative day of the left eye, the patient was found to have high IOP (48 mmHg) and a fixed, mid-dilated pupil. There was moderate corneal epithelial edema, with mild anterior chamber reaction. The anterior chamber was deep both centrally and peripherally. The patient was treated with full anti- glaucoma medications. The second day, the IOP dropped to 18 mmHg. However, the left pupil remained mid-dilated. There was no reaction to pilocarpine eye drops. Two weeks postoperatively (Figure 1), the uncorrected visual acuity in

the left eye improved to 6/12, and the IOP was 14 mm Hg. But the patient was complaining of photophobia. The ICL was well positioned, with a visible space between the crystalline lens and the ICL. Anterior-segment optical coherence tomography (OCT) (Figure 2) of the left eye showed well-positioned ICL with normal vault. Her left pupil remained fixed and mid-dilated throughout 2 years of postoperative follow-up, with no response to pilocarpine with different concentrations ranging from 1-4%. The patient was managed in the morning period with colored contact lenses. She was offered a pupilloplasty surgery to constrict the pupil but she refused to undergo any further surgical procedure.

Figure 1: Slit lamp photo showing the dilated pupil and the absence of iris atrophy.

Figure 2: Anterior segment OCT.

Figure 3: Angle OCT.

Discussion

ICL refractive surgery entails advantages such as the ability to correct a wide range of refractive errors, shorter visual recovery times, being reversible, more stable refractive outcomes and superior quality of vision [5]. There are two models of the ICL, one without a hole and needs a peripheral iridtomy to be done before the planned surgery; and the other with a central hole and needs no peripheral iridtomy [6].

The underlying mechanism of UZS is still a topic of speculations; the most widely accepted theories are acute rise in IOP, ischemia of the iris, and presence of iris abnormalities, which may be more common in keratoconus. The instillation of strong mydriatic and the intraoperative contact between the iris and peripheral cornea are other presumed triggers [7].

Biochemical changes in iris innervation have also been suggested as a cause for UZS, including injury to parasympathetic nerve fibers causing denervation of the constrictor muscles [8].

Three types of pupillary dilatation were identified in patients with UZS; reactive pupil with anisocoria, at least 1.5 mm larger than the fellow eye; an unreactive pupil that returns slowly to its normal state; and irreversible pupil dilatation with iris atrophy [9].

Several cases have been reported after the implantation of different types of IOL; few after anterior chamber IOL and even fewer after posterior chamber IOL [3,4].

In our case; the pupil remained fixed and mid-dilated. No patches of iris atrophy were observed. There was no evidence of keratoconus. Adie's tonic pupil was ruled out, because there was no constriction to any concentration of pilocarpine instillation.

Few modalities of treatment are available to address the problem of fixed dilated pupil and the bothersome associated glare and photophobia; these include corneal tattooing, colored contact lenses, and artificial iris implants; in addition miotics such as guanethidine and pilocarpine have demonstrated efficacy in treating some cases of UZS and attenuating mydriasis [1].

Conclusion

For the first time, a case of UZS is reported in patient with SLE after ICL implantation. Whether the underlying disease has a causative relationship with the development of this rare complication or not needs further studies.

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