CASE REPORT

Urrets–Zavalia Syndrome of Unresolving Mydriasis Following Endocyclophotocoagulation Combined with Phacoemulsification

Anny MS Cheng[1], Geetha G Vedula[2], Aarup A Kubal[3], Jillene Moxam[4], David TY Yang[5], Shailesh K Gupta[6]

Received on: 19 September 2023; *Accepted on:* 16 March 2024; *Published on:* 30 March 2024

Abstract

Aim and background: Combined endocyclophotocoagulation and phacoemulsification (ECP/Phaco) are uncommonly associated with complications. We present the first case of a rare complication following ECP/Phaco.

Case description: A 72-year-old patient with dense nuclear sclerotic cataracts and primary open-angle glaucoma (POAG) underwent bilateral surgery uneventfully. He experienced a brief episode of postoperative elevated intraocular pressure (IOP), but only one eye with a lower baseline IOP developed a dilated pupil. No pupillary response was observed after applying 4% pilocarpine. The fixed mydriasis persisted without reaction to light or near stimulus, and the best-corrected vision (BCVA) was 20/30 in the affected eye.

Conclusion: This case reports a possible rare complication when undergoing ECP/Phaco therapy. The pathogenesis of Urrets–Zavalia syndrome is unknown, but we hypothesized that eyes with more pronounced increases in IOP from baseline may be more susceptible to ischemic injury to the pupillary sphincter, resulting in a chronically dilated pupil.

Clinical significance: Even a modest transient rise in postoperative IOP in a glaucomatous eye with normal baseline IOP could result in a chronically dilated pupil.

Keywords: Case report, Endocyclophotocoagulation, Microinvasive glaucoma surgery, Mydriasis, Phacoemulsification, Urrets–Zavalia syndrome.

Journal of Current Glaucoma Practice (2024): 10.5005/jp-journals-10078-1431

Introduction

Microinvasive glaucoma surgery (MIGS) is a new class of glaucoma procedures that aim to reduce intraocular pressure (IOP) using less invasive methods that have a shorter visual recovery period than conventional surgery.[1] Endocyclophotocoagulation (ECP) is the only MIGS to suppress aqueous production by direct visualization and ablation of the nonpigmented ciliary epithelium through an endoscopic-guided technique. A recent systematic literature review of 81 publications reported that MIGS, especially when combined with cataract surgery, showed favorable safety and efficacy profiles.[1] In patients with concurrent glaucoma and cataracts, simultaneous ECP is a viable option due to its minimal adverse effect profile and ease of combination with cataract surgery.[2] Given that ECP/phacoemulsification (Phaco) is being advocated for use in a wider range of patients, it was paramount to publish our experience when undergoing this therapy.

Urrets–Zavalia syndrome,[3] in which the pupil remains fixed and dilated and may be associated with iris atrophy and glaucoma, is an uncommon complication after a surgical procedure. This rare complication is most frequently associated with intraoperative surgery, such as keratoplasty.[4] Few cases were reported after cataract or conventional glaucoma procedures; however, it is rarely associated with MIGS.[4] There has been a small case series report of Urrets–Zavalia syndrome following transscleral cyclophotocoagulation (TSCPC),[5] but not with ECP. To our knowledge, this is the first report of a patient who developed a persistently mydriatic pupil after combining ECP with Phaco. Institutional approval was waived as our single case report involves a retrospective medical record review of one patient, and the only interaction with the patient has been for the purpose of treating the patient, which does not meet the common rule definition of

[1]Department of Ophthalmology, Broward Health, Fort Lauderdale; Department of Ophthalmology, Specialty Retina Center, Coral Springs; Department of Ophthalmology, Florida International University, Herbert Wertheim College of Medicine, Miami, Florida, United States of America

[2,3]Department of Ophthalmology, Your Eye Specialists, Weston, Florida, United States of America

[4]Department of Ophthalmology, Specialty Retina Center, Coral Springs, Florida, United States of America

[5]College of Biological Science, University of California, Davis, Sacramento, California, United States of America

[6]Department of Ophthalmology, Broward Health, Fort Lauderdale; Department of Ophthalmology, Specialty Retina Center, Coral Springs, Florida, United States of America

Corresponding Author: Shailesh K Gupta, Department of Ophthalmology, Broward Health, Fort Lauderdale; Department of Ophthalmology, Specialty Retina Center, Coral Springs, Florida, United States of America, Phone: +15613223588, e-mail: sgupta@specialtyretina.com

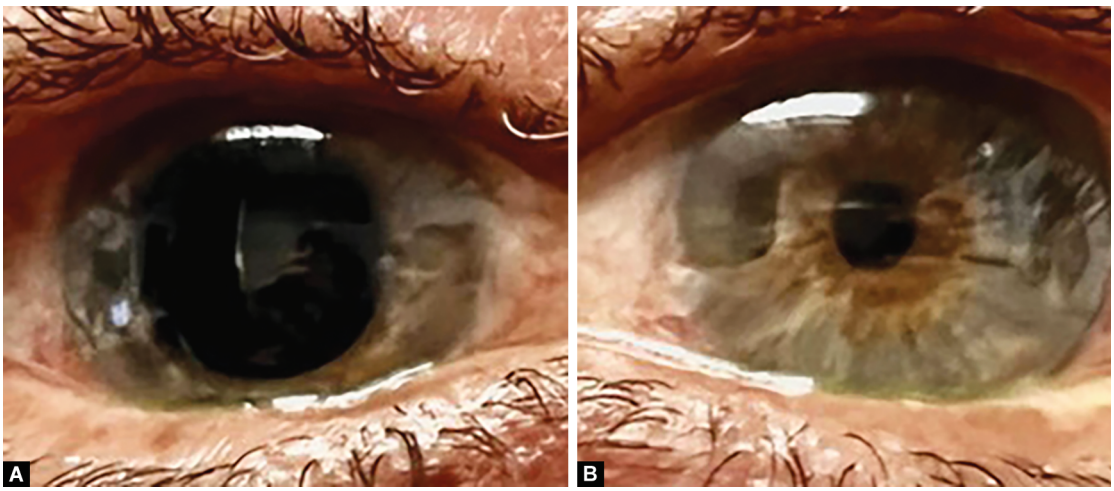
How to cite this article: Cheng AMS, Vedula GG, Kubal AA, *et al.* Urrets–Zavalia Syndrome of Unresolving Mydriasis Following Endocyclophotocoagulation Combined with Phacoemulsification. J Curr Glaucoma Pract 2024;18(1):28–30.

Source of support: Nil

Conflict of interest: None

Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details and related images.

© The Author(s). 2024 Open Access. This article is distributed under the terms of the Creative Commons Attribution 4.0 International License (https://creativecommons.org/licenses/by-nc/4.0/), which permits unrestricted use, distribution, and non-commercial reproduction in any medium, provided you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons license, and indicate if changes were made. The Creative Commons Public Domain Dedication waiver (http://creativecommons.org/publicdomain/zero/1.0/) applies to the data made available in this article, unless otherwise stated.



Figs 1A and B: The right eye (A) shows a fixed and uniformly dilated pupil, while the left eye (B) has no evidence of pathological signs

research (45 CFR 164.501). Although institutional approval was not required to publish the case details, we obtained informed consent from the patient for the publication of his case.

Case Description

A 72-year-old man with a history of primary open-angle glaucoma (POAG) was treated with latanoprost (Somerset Pharma, New Jersey, United States of America) bilaterally for 4 years. At the time of evaluation, he complained of blurred vision with a best-corrected vision (BCVA) of 20/50 bilaterally, while his IOP was 17 mm Hg in the right eye and 21 mm Hg in the left eye. Slit-lamp examination revealed bilateral dense senile nuclear cataracts. Fundus examination revealed symmetric glaucomatous optic nerve head cupping of 0.65 of both eyes. Gonioscopy revealed open angles in all four quadrants and no peripheral anterior synechiae (PAS) in both eyes. Visual field examination demonstrated nonspecific changes and nasal-step field defects with a mean deviation (MD) of –2.22 and –5.95 dB in the right and left eyes, respectively. Therapeutic options were discussed with the patient, who gave written informed consent after being informed about the nature, risks, and possible adverse consequences of the combined procedures.

The patient underwent femtosecond laser-assisted cataract surgery in the left eye first. A standard Phaco surgical technique with topical anesthesia and a main temporal clear corneal incision was performed. After placement of the intraocular lens in the bag, Provisc (Alcon, Ft Worth, TX) was injected to inflate the ciliary sulcus 360° to enhance visualization of ciliary processes. ECP is then performed with 0.2 W energy for 270° while avoiding 3 and 9 o'clock to blanch and shrink the ciliary processes. The surgery on the left eye was uneventful. Standard topical therapy with moxifloxacin, 1% prednisolone acetate, and prolensa 0.07% was applied 4 times daily in the early postoperative period. On postoperative day 1, BCVA was 20/40, and IOP was 36 mm Hg, which reduced to 12 mm Hg following wound burp. Around 1 week later, the VA was 20/25, and the IOP was 15 mm Hg. One month after the procedures, he was presented with a satisfactory IOP of 12 mm Hg and a BCVA of 20/25.

The same methodology was used to perform ECP/Phaco in the right eye 2 months later by the same experienced surgeon (G.V.). There were no immediate intraoperative complications. His BCVA was 20/400 on postoperative day 1, and IOP was elevated to 36 mm Hg, which reduced to 12 mm Hg immediately following the wound burp. The anterior chamber had a minimal postoperative reaction. One week later, the VA was 20/25, and IOP was 18 mm Hg, slit lamp exam revealed minimal intraocular inflammation and no hyphema. However, the right pupil remained dilated at 8 mm despite topical pilocarpine at 4% at the office visit. Topical pilocarpine 1% four times a day was started. The patient was followed every 2–3 weeks, then monthly, and thereafter. Approximately 6 months after the procedure, the patient was not on glaucoma medications, IOP was 10 mm Hg in the right eye and 12 mm Hg in the left eye, and BCVA was 20/30 bilaterally. No new PAS or iris atrophy was seen in our follow-up. A persistently dilated pupil without reaction to light or near objects was present only in the right eye (Fig. 1).

Discussion

Endocyclophotocoagulation (ECP) is a laser-based MIGS treatment option that is gaining popularity, accounting for 11% of all MIGS procedures, and 90% of ECP is performed in conjunction with Phaco.[6] ECP/Phaco has a high reported safety profile; reported complications were uncommon in 1-year, 3-year, and 6-year follow-ups.[7] Of the reported complications, including cystoid macular edema and uveitis, the vast majority resolve with medication. Hyphema is rare, and hypotony has not been reported.[7] To the author's knowledge, this is the first reported case of Urrets–Zavalia syndrome after ECP/Phaco. Urrets–Zavalia syndrome is uncommon, with approximately only 100 published case reports to date.[4] Risk factors and mechanisms remain unclear. One of the presumed risk factors is intraocular inflammation, such as toxic anterior segment syndrome or cataract remnants, which can lead to the release of inflammatory substances.[8] However, our patient presented with a minimal inflammatory reaction after surgery that responded appropriately to topical steroids, and no remnants of lens material were noted on intraoperative or postoperative examination.

A small case series has reported this rare complication after TSCPC.[5] It was hypothesized that TSCPC might damage the short ciliary nerve fibers that innervate the ciliary muscle and the iris in one of their patients with dilated and peaked pupils. TSCPC delivers laser energy indirectly through the sclera, and individual variations in axial length, position of ciliary processes, and probe orientation may result in laser energy being misdirected to the peripheral iris. However, in our patient, ECP allowed direct visualization for focused and precise treatment without destroying deep ciliary

body tissue. This was done to avoid direct thermal laser damage to the pathway of the short ciliary nerves to the pupillary sphincter muscle. It is also hypothesized that a circumferential 360° laser could result in a fixed and dilated pupil if the ciliary nerves, which run radially from the posterior pole to the iris sphincter muscle, were to be injured in the treatment. A similar phenomenon of a fixed and dilated pupil has been described after panretinal photocoagulation and 360° argon laser peripheral iridoplasty.[4] In fact, the ECP technique used in our patient specifically spared the 3 and 9 o'clock meridians and treated only 270° of circumference with preservation of short ciliary nerves.

The pathogenesis of Urrets–Zavalia syndrome is not fully understood, but the existing literature mostly attributes it causally to iris ischemia due to compression of the iris vasculature by a postoperative IOP spike.[4] Interestingly, our patient who underwent the same surgery in the fellow left eye did not develop pupillary abnormalities, although IOP in both eyes was elevated to the same level on postoperative day 1. It is possible that eyes with more dramatic IOP increases from baseline, IOP may be more likely to induce iris ischemia. In this case, the right eye had a lower baseline IOP (17 mm Hg) than the fellow left eye (21 mm Hg) at baseline. We hypothesize that when IOP is acutely elevated in eyes with a high baseline IOP, subclinical iris ischemia may occur, but it may not manifest clinically. While some studies showed that iris fluorescein angiography in patients with Urrets–Zavalia syndrome revealed reduced filling and similar morphology changes of iris vessels as severe iris ischemia,[9] other studies showed a normal radial pattern of the iris vasculature with no areas of iris ischemia.[5] Iris atrophy is a sequelae that is described in Urrets–Zavalia syndrome[3]; however, some patients with persistently dilated pupils following procedures do not exhibit signs of iris atrophy or PAS that could be related to iris ischemia, neither in our patients nor in the majority of published literature on the Urrets–Zavalia syndrome. Hence, the mechanism of Urrets Zavalia syndrome cannot be fully determined and is probably multifactorial.

Conclusion

Endocyclophotocoagulation and phacoemulsification (ECP/Phaco) is well established and is the procedure of choice as a microinvasive adjunct to Phaco. Our brief report of Urrets–Zavalia Syndrome in combined ECP/Phaco may indicate that even a modest transient rise in postoperative IOP in a glaucomatous eye with normal baseline IOP could cause ischemic damage to the pupillary sphincter and lead to a permanent dilated pupil.

Clinical Significance

Acknowledgment of our patient outcome and close monitoring postoperatively would allow for strategizing ways to avoid potential side effects.

Orcid

Anny MS Cheng https://orcid.org/0000-0001-7035-8567

References

1. Cantor L, Lindfield D, Ghinelli F, et al. Systematic literature review of clinical, economic, and humanistic outcomes following minimally invasive glaucoma surgery or selective laser trabeculoplasty for the treatment of open-angle glaucoma with or without cataract extraction. Clin Ophthalmol 2023;17:85–101. DOI: 10.2147/OPTH.S389406
2. Rathi S, Radcliffe NM. Combined endocyclophotocoagulation and phacoemulsification in the management of moderate glaucoma. Surv Ophthalmol 2017;62(5):712–715. DOI: 10.1016/j.survophthal.2017.01.011
3. Urrets Zavalia a Jr. Fixed, dilated pupil, iris atrophy and secondary glaucoma. Am J Ophthalmol 1963;56:257–265. DOI: 10.1016/0002-9394(63)91861-0
4. Magalhães OA, Kronbauer CL, Müller EG, et al. Update and review of Urrets-Zavalia syndrome. Arq Bras Oftalmol 2016;79(3):202–204. DOI: 10.5935/0004-2749.20160059
5. Vieira GM, Vieira FJ, Ritch R. Urrets-Zavalia syndrome after diode laser transscleral cyclophotocoagulation. J Glaucoma 2017;26(7):678–682. DOI: 10.1097/IJG.0000000000000666
6. Yang SA, Mitchell W, Hall N, et al. Trends and usage patterns of minimally invasive glaucoma surgery in the united states: IRIS® registry analysis 2013-2018. Ophthalmol Glaucoma 2021;4(6):558–568. DOI: 10.1016/j.ogla.2021.03.012
7. Feinberg L, Swampillai AJ, Byles D, et al. Six year outcomes of combined phacoemulsification surgery and endoscopic cyclophotocoagulation in refractory glaucoma. Graefes Arch Clin Exp Ophthalmol 2023;261(5):1339–1347. DOI: 10.1007/s00417-022-05906-0
8. Kurtz S, Fradkin M. Urrets-Zavalia syndrome following cataract surgery. Case Rep Ophthalmol 2021;12(2):659–663. DOI: 10.1159/000513959
9. Jain R, Assi A, Murdoch IE. Urrets-Zavalia syndrome following trabeculectomy. Br J Ophthalmol 2000;84(3):338–339. DOI: 10.1136/bjo.84.3.337b