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CASE REPORT

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Malignant glaucoma presenting with uncontrolled intraocular pressure and myopic refractive surprise after cataract surgery

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Abstract

We present a seemingly typical case of bilateral angle closure with elevated intraocular pressures. After cataract surgery, there was axial shallowing, escalating intraocular pressure, anterior displacement of the IOL, and myopic shift in the left eye. Irido-zonulo-hyaloido-vitrectomy resolved the angle closure, normalized intraocular pressure, and corrected the myopic shift.

KEYWORDS

aqueous misdirection syndrome, malignant glaucoma

1 | INTRODUCTION

The term malignant glaucoma was first described by von Graefe[1] in 1869 as a rare and unique form of recalcitrant glaucoma, unresponsive to treatment, presenting with a shallow anterior chamber despite a patent peripheral iridectomy. Malignant glaucoma is a bit of a misnomer as it has nothing to do with malignancy, but rather to the catastrophic nature of vision loss. Since then, successful surgical treatment modalities have been developed, but the name has persisted. Aqueous misdirection, ciliary block glaucoma, and lens block angle closure[2,3] are more descriptive terms to highlight the mechanism of action, but the many names imply this is still a poorly understood disease.

We present a case of bilateral angle closure with improvement in intraocular pressure (IOP) after laser peripheral iridotomy (LPI) in the right eye but with development of malignant glaucoma after cataract surgery in the left eye.

2 | CASE REPORT

A 74-year-old Caucasian gentleman was seen for a routine examination complaining of poor vision ever since getting new glasses from 1 year prior.

Past medical history was significant for insulin-dependent diabetes mellitus, hypercholesterolemia, and hypertension. Past family history and ocular history were negative. Review of symptoms was significant for glare with night driving, but the patient denied any eye pain, photophobia, or visual disturbance. The patient denied being on topiramate, cholinergics, or sulfa-containing drugs.

Manifest refraction was +1.00 + 0.75 × 075 with 20/40 in the right eye and +1.00 sph with 20/25 in the left. BAT (brightness acuity testing) was 20/70 in the right and 20/50 in the left. IOP was 40 mmHg in the right eye by Goldmann applanation. IOP was 30 mmHg in the left eye by Goldmann applanation. Slit-lamp examination revealed narrow angles with peripheral iris-corneal touch in

both eyes, 2–3+ nuclear sclerosis in both eyes, and 1–2+ anterior capsule haze in the right eye only. Dilated examination was deferred due to narrow angles, but non-dilated examination with a 75D lens showed a cup-to-disc ratio (CDR) of 0.3 in both eyes with intact rims. Gonioscopy revealed no angle structures seen, with a steep approach 360 in both eyes (Shaffer Grade 0 both eyes), and opened only to anterior trabecular meshwork with compression in both eyes.

Pupillary examination was 4 mm and symmetrical with no afferent pupillary defect (APD) but with minimal reaction in both eyes. Confrontation visual fields, alignment, and extraocular motility were within normal limits.

The diagnosis of angle closure was made in both eyes, and the patient was referred for a same day YAG laser peripheral iridotomy (PI) of the right eye. Laser PI was successfully performed temporally but was difficult due to extreme peripheral iris-corneal touch. The patient was started on timolol maleate/brimonidine tartrate BID and travoprost qhs samples in both eyes.

On postoperative day one, the IOP improved to 15 mmHg and the left eye was 28 mmHg on timolol maleate/brimonidine tartrate and travoprost eyedrops in both eyes. Gonioscopy in the right eye showed a steep approach but was now open to posterior trabecular meshwork (Shaffer Grade 2). Compression gonioscopy was open to scleral spur. Since the patient's IOP was not dangerously elevated in the left eye and since he had cataract-related symptoms along with a phacomorphic component, the decision was made to bypass YAG PI and proceed directly with cataract extraction with intraocular lens implant (IOL) placement in the left eye. Optical biometry with the Lenstar showed an axial length of 21.81 mm OD, 21.69 mm OS, anterior chamber depth of 2.13 mm OD, 2.10 mm OS, lens thickness of 5.05 mm OD, 5.13 mm OS, and keratometry 46.55 × 47.37@067 OD, 46.51 × 46.95@083 OS. A plano target was chosen, and the surgery was performed uneventfully with an in-the-bag single piece acrylic IOL (Alcon SA60AT) in the left eye.

Postoperative day one after uneventful cataract surgery of the left eye, uncorrected visual acuity was 20/400 with pinhole 20/70, and the IOP was 21 mmHg. Slit-lamp examination showed a clear cornea, but the chamber was visibly shallow not just peripherally but also centrally with anterior displacement of the IOL optic against the iris. Manifest refraction was −3.50 spherical with visual acuity of 20/60. Gonioscopy in the left eye revealed no angle structures seen with a steep approach and compression did not significantly change the angle anatomy (Shaffer Grade 0). Dilated examination and macular and optic nerve optical coherence tomography (OCT) were normal, and there was nothing obvious on examination to explain the anterior displacement of the IOL.

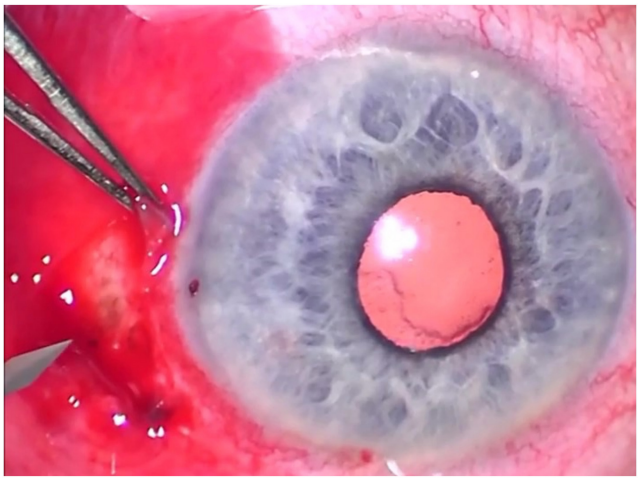

FIGURE 1 A peritomy is performed and light cautery applied

Atropine eyedrops were prescribed for the left eye which successfully deepened the anterior chamber and the myopic shift improved, but the effect was only temporary and disappeared upon cessation of the drops. The dramatic response to the atropine drops along with the closed angle anatomy, axial shallowing of the anterior chamber, persistent anterior displacement of the IOL with myopic shift, and borderline elevated IOP suggested that this could be aqueous misdirection/malignant glaucoma. The decision was made to proceed with a laser peripheral iridotomy with hyaloidotomy of the left eye in the attempt to create a unicameral eye. However, this only caused a focal area of iris to bulge forward in the area of PI, which we surmise was from anterior prolapse of vitreous through the hyaloid face. Despite this maneuver, the patient's uncorrected visual acuity did not improve and remained 20/400, and the myopic refractive error persisted with a manifest refraction of −3.25 + 1.00 × 168 with 20/50-1. Moreover, the IOP worsened to 35 mmHg despite being on timolol/dorzolamide BID and latanoprost qhs in the left eye. The worsening IOP despite maximum topical therapy was further evidence that this was most likely malignant glaucoma.

Since the patient was traveling from over an hour away, we felt the patient needed urgent and definitive intervention. Therefore, the decision was made to proceed with an irido-zonulo-hyaloido-vitrectomy (Figures 1–4) through the peripheral iridectomy site. The procedure was performed uneventfully, and on postoperative day one, the patient noticed dramatic improvement in vision. Uncorrected visual acuity improved from 20/400 to 20/40, and the manifest refraction went from −3.25 + 1.00 × 168 to −0.25 + 0.50 × 166. Intraocular pressure also improved to 17 mmHg, and slit-lamp examination showed a clear cornea, trace cells, and a much deeper and more appropriate anatomical position of the IOL within the capsular bag.

Gonioscopy revealed open to ciliary body band (Shaffer Grade 4). One week postoperatively, uncorrected visual acuity further improved to 20/20-2 with manifest refraction of −0.50 spherical, the IOP was 18 mmHg on timolol/dorzolamide BID and latanoprost qhs, and the remainder of the examination was stable.

The patient was happy with his left eye vision but complained of cataract-related visual symptoms in the right eye. We were concerned that the right eye could behave similarly to the left eye and develop aqueous misdirection; as a result, the decision was made to proceed with a combined cataract extraction with IOL and zonulo-hyaloido-vitrectomy through the iridotomy site. Postoperatively, uncorrected visual acuity was 20/30+2, manifest refraction was plano +0.50 × 100 with visual acuity of 20/25, and intraocular pressure was 19 mmHg. Slit-lamp examination was otherwise normal with an appropriate IOL position within the capsular bag. Gonioscopy revealed open to ciliary body band (Shaffer Grade 4).

At 6 months follow-up, optic nerve OCT showed a new area of focal loss in the superior quadrant only in the left eye, with a corresponding inferior arcuate defect with formal visual field testing. The patient has since been followed over the next 4 years and has been stable. At his last visit, the uncorrected visual acuity was 20/25 in the right eye and 20/30 in the left eye. His IOP was 19 mmHg in the right eye and 18 in the left eye on latanoprost qhs and dorzolamide/timolol BID in both eyes. His optic nerve OCT and formal VF also continue to be stable and

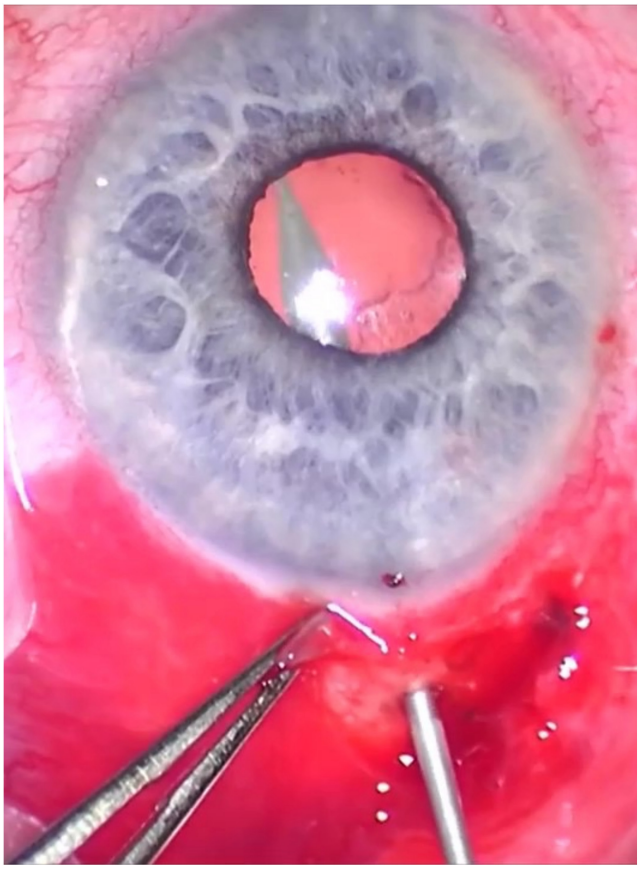

FIGURE 2 A 23 g MVR blade is used to place a sclerotomy 3 mm posterior to the limbus taking care not to contact the posterior capsule

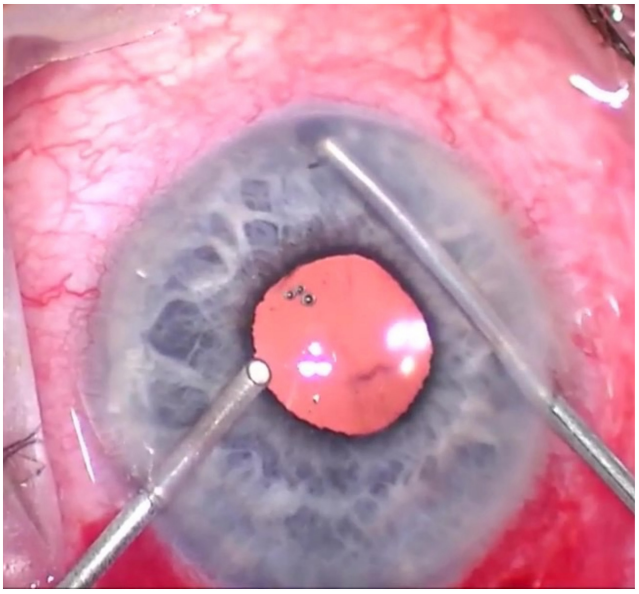

FIGURE 3 A 23 g irrigation cannula and vitrector are placed through limbal incisions for a vitrector-assisted peripheral iridectomy

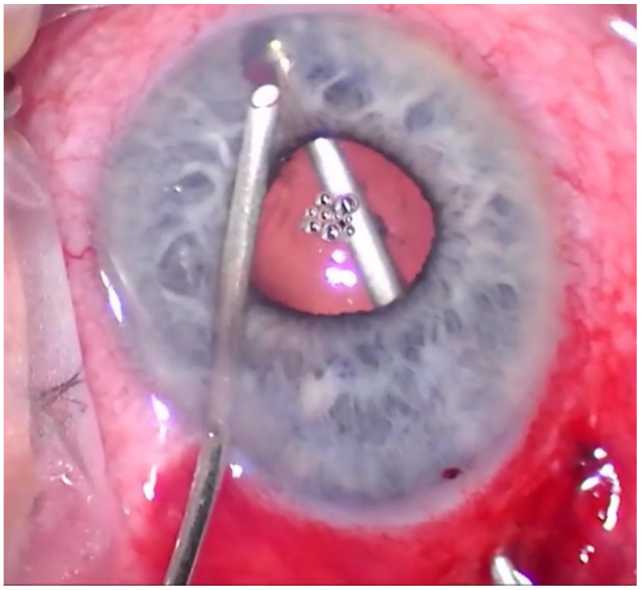

FIGURE 4 The vitrector is then switched to a pars plana approach through the sclerotomy to perform the zonulo-hyaloido-vitrectomy. The irrigation cannula is placed over the iridectomy site to help the vitrector tip penetrate into the anterior chamber to ensure a unicameral eye

unchanged with a persistent dense inferior arcuate defect only in the left eye.

3 | DISCUSSION

We present a seemingly typical case of bilateral angle closure with elevated intraocular pressures which responded to laser peripheral iridotomy in the right eye; however, after routine cataract surgery, developed worsening angle closure with axial shallowing, escalating intraocular pressure, anterior displacement of the IOL, and myopic shift in the left eye.

After a thorough examination, there was nothing to explain the persistent angle closure and axial shallowing of the left eye after cataract surgery. Both the slit lamp and posterior segment examinations were unremarkable. Utilization of ultrasound biomicroscopy (UBM) would have been helpful to rule-out pathology such as suprachoroidal hemorrhage, choroidal effusion, or ciliary body tumor. Even in the absence of such pathology, UBM would have helped to support the diagnosis of malignant glaucoma by showing anterior rotation of the ciliary body, thereby explaining the cause for anterior displacement of the IOL, but unfortunately, we did not have access to this technology. Although we did not have UBM, the use of atropine was a reasonable diagnostic and therapeutic maneuver to see whether it would help to deepen the anterior chamber, which would then support the diagnosis of malignant glaucoma. Moreover, focal anterior displacement of the iris after LPI in the left eye was highly unusual and increased the suspicion for malignant glaucoma.

Not only was the patient's IOP progressively climbing despite maximal topical glaucoma therapy, this was complicated by the fact that the patient lived over an hour away and had difficulty returning for his appointment. One could argue that we should have simply performed bilateral LPI at the onset rather than performing cataract surgery in the left eye which would most likely have prevented the malignant glaucoma; however, the patient was having sufficient cataract-related symptoms and wished to proceed with cataract surgery. In this context, it would have been unnecessary and more costly for the patient to perform the LPI if he was going to undergo cataract surgery anyway. In addition, since the LPI failed to resolve the issue, urgent intervention with irido-zonulo-hyaloido-vitrectomy was a bold but reasonable next step. Dramatic resolution after irido-zonulo-hyaloido-vitrectomy was both diagnostic and therapeutic confirmation that this was most likely malignant glaucoma. By creating a unicameral eye, normal aqueous fluid flow was restored; therefore, the IOP improved, the chamber deepened, and the IOL shifted back into a more normal anatomic position, while resolving the myopic shift.

Malignant glaucoma is a rare, yet serious form of glaucoma which typically arises after intraocular surgery in the setting of pre-existing angle closure glaucoma. Its onset has been noted after trabeculectomy, cataract surgery, pars plana vitrectomy, laser capsulotomy, and laser iridotomy.[4] Rarely, malignant glaucoma has been observed after miotic agent use or even spontaneously with no prior operation or medical intervention,[5] but these cases are exceptionally rare. The overall incidence of malignant glaucoma has been reported roughly between 1% and 3% postoperatively.[6] When incisional surgery is performed in the case of bilateral primary angle closure glaucoma, the incidence of malignant glaucoma increases slightly to approximately 2–4%. Patients most typically present with myopic shifts noted to range from −2.5 to −8 diopters due to anterior displacement of the lens.

Timely intervention with selective therapy is often necessary as response to conventional glaucoma therapy is poor. Treatment revolves around lessening anterior displacement of the lens-iris diaphragm and reducing vitreous volume. Medical therapy includes cycloplegic agents such as atropine and hyperosmotics such as mannitol and acetazolamide, alleviating ciliary block and shrinking the vitreous body, respectively.[7] Pars plana vitrectomy when combined with pharmacotherapy has proven to be effective as a conservative measure, especially in pseudophakic eyes.[8] However, due to the complex nature and pathogenesis of malignant glaucoma, in many cases, conservative treatment may prove to be inadequate. In retrospective studies, relapse proved to be as high as 100% after medical therapy and up to 75% after vitrectomy.[8] However, with continuous advancements in surgical techniques, prognosis is relatively good in most cases. In patients where further intervention is required, often a pragmatic goal is to achieve a unicameral eye and allow for communication of aqueous fluid from the posterior to anterior segment.[8] Treatment modalities can vary slightly depending on whether the patient presents with phakic or pseudophakic eyes based on suggested algorithms. In treatment-resistant phakic eyes, lens or cataract extraction, posterior capsulotomy and anterior vitrectomy are typically advisable treatments.[9] In pseudophakic or aphakic eyes, posterior capsulotomy and laser hyaloidotomy are acceptable procedures performed.[10] As such, our pseudophakic patient underwent laser peripheral iridotomy and hyaloidotomy. This procedure unfortunately proved to be unsuccessful despite common uses reported in literature.[11] Further refractory cases have seen benefit with transscleral cyclophotocoagulation.[5] Additional surgical procedures, if not yet performed, including goniosynechialysis and peripheral iridectomy, often adjunctively, have shown varying

success in treating malignant glaucoma.[12] Surgical interventions should be promptly planned to prevent permanent damage and reduce long-term complications. Our patient was stabilized after irido-zonulo-hyalodo-vitrectomy, which has also proven to be a successful procedure in prior cases of pseudophakic malignant glaucoma and also refractory cases.[10]

In patients diagnosed with malignant glaucoma unilaterally, there is often an increased risk of developing malignant glaucoma in the contralateral eye. As such, it can be prudent to perform a prophylactic iridotomy or vitrectomy in the opposing eye.[7] This was the case in our patient because despite a patent peripheral iridotomy in the right eye, he had a visually significant cataract which needed surgery. If his left eye was any indication, the right eye was at risk for developing malignant glaucoma after the cataract surgery. Therefore, he underwent prophylactic zonulo-hyaloido-vitrectomy in the right eye, and he did well. Even after surgical correction and prophylaxis, however, there remains a risk of recurrence. As such, patients should be followed closely with long-term follow-up.

Malignant glaucoma is likely multifactorial since it can present after a wide variety of intraocular procedures or sometimes even without surgery and the IOP may not even be dramatically elevated.[13] This patient also had a 3.50 diopter myopic refractive miss which is highly unusual with optical biometry and modern IOL calculation formulas. All this is to say a high index of suspicious is needed to make the correct diagnosis.

AUTHOR CONTRIBUTIONS

AX and DBK involved in drafting and revising the manuscript. AX and DBK contributed to analysis and interpretation of data. DBK Involved in clinical management of patient. Both authors involved in review of the final version of the manuscript and agree to be accountable for all aspects of the manuscript.

CONFLICT OF INTEREST

The authors have no conflicts of interest to disclose.

DATA AVAILABILITY STATEMENT

The data are available from the corresponding author upon reasonable request.

ETHICAL APPROVAL

None.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

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