

Management of Late Onset Sequential Pseudophakic Malignant Glaucoma: A Case Report and Review of Literature

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Abstract

Introduction: Malignant glaucoma or aqueous misdirection syndrome (AMS) can present in the immediate post-operative period or even many years after cataract surgery. We report the clinical course and management of a patient who developed the disease in both eyes at different time intervals after uneventful cataract surgery.

Patient and Clinical Findings: A 76-year-old man developed diffusely shallow anterior chamber (AC) and high intraocular pressure (IOP) in his right eye after 2 years of pseudophakia, which did not resolve with laser peripheral iridotomy. A similar episode happened in left eye after 3 years of cataract surgery in the presence of a patent laser iridotomy.

Diagnosis, Interventions, and Outcome: Shallow anterior chamber in the presence of a patent PI and normal posterior segment confirmed the diagnosis. The right eye was successfully managed with Nd:YAG laser capsulo-hyaloidotomy and pharmacotherapy, whereas in left eye, pars plana vitrectomy (PPV) with irido-zonulo-anterior hyaloidectomy (IZH) was done. Recurrence was noted after PPV also, owing to blockage of surgical iridectomy, which was treated with Nd:YAG laser.

Conclusion: Pseudophakic malignant glaucoma can present many years after cataract surgery. Pars plana vitrectomy with IZH is an effective mode of treatment for refractory cases. Laser capsulo-hyaloidotomy may be more useful in the acute phase of the disease.

Keywords: Malignant glaucoma, Pseudophakia, Aqueous misdirection, Secondary glaucoma, Nd: YAG laser capsulo-hyaloidotomy, Pars plana vitrectomy, Iridozonulohyaloidectomy.

INTRODUCTION

Malignant glaucoma is a rare vision-threatening disorder of unconfirmed etiology seen after incisional eye surgery.¹ However, it has been reported to occur spontaneously after noninvasive procedures.² The three essential diagnostic features for the disease include a diffuse shallowing of the anterior chamber (AC), a patent peripheral iridotomy (PI) and no evidence of suprachoroidal hemorrhage or ciliary effusion.¹ The pathogenesis is not clearly understood but according to one of the theories, an anomalous anatomical relationship between the ciliary body, lens and anterior hyaloid with anterior rotation of ciliary processes blocks the forward flow of aqueous humor and redirects it into or behind the vitreous cavity causing a forward displacement of lens-iris diaphragm and secondary angle closure glaucoma.^{3,4} Intraocular pressure (IOP) levels may range from normal to very high based on the

rapidity of disease development, presentation time and angle closure. Primary angle closure glaucoma patients and suspects have a higher risk of developing this disorder after a surgical procedure. For reasons unknown, the involvement of the fellow eye is common.⁵ Management of malignant glaucoma is difficult because of frequent relapses and need of prolonged treatment with cycloplegics. Laser and surgical treatment aim to make the eye unicameral by removing the ciliary block and connecting the vitreous cavity with the anterior chamber.⁶ The

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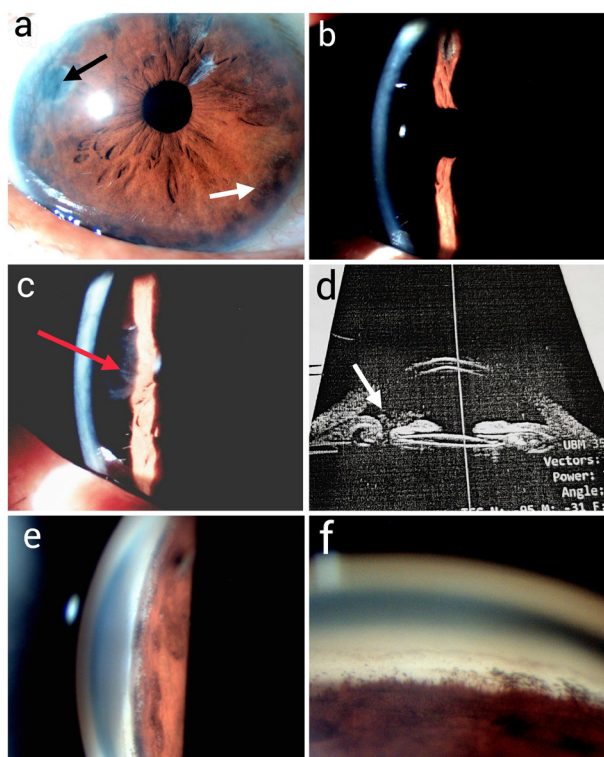


Figure 1: Right eye (a) Diffuse illumination with large PI at 9:30 clock hours (black arrow) and peripheral iridoplasty marks (white arrow). (b) Oblique slit illumination post capsulo-hyaloidotomy showing deep central AC (c) Oblique illumination showing vitreous in AC (red arrow). (d) Ultrasound biomicroscopy showing patent irido-capsulo-hyaloidotomy with vitreous in anterior chamber (white arrow). (e) Gonioscopic view of nasal angle showing open angle and patchy pigmentation suggestive of previous angle closure. (f) Inferior angle gonioscopy shows peripheral anterior synechiae formation with pigment deposits in patches and anterior to schwalbe's line.

time of intervention plays a significant role in the response of eye to therapy and the eventual outcome, which we discuss in this case.

Patient Consent Statement

A written informed consent was taken from the patient for publication of report and de-identified pictures.

Case report

A 76-year-old-man with no significant ocular and medical history presented two years after an uneventful cataract surgery with severe pain and a vision of counting fingers in right eye (RE). Examination showed circumcorneal congestion with aqueous flare grade 2+ in RE and shallow anterior chambers in both eyes with more axial shallowing in RE (central AC depth 1.16 mm). IOP was 40 mm in right eye and 9 mm in left eye (LE). Gonioscopy showed closed angles in RE, whereas the phakic LE showed narrow angles. The patient was administered systemic and topical anti-glaucoma therapy, along with four hourly steroid drops in RE steroid drops in RE. Nd:YAG laser PI and argon laser iridoplasty was done in right eye but the AC remained shallow. Nd:YAG laser assisted capsulo-hyaloidotomy was then done through

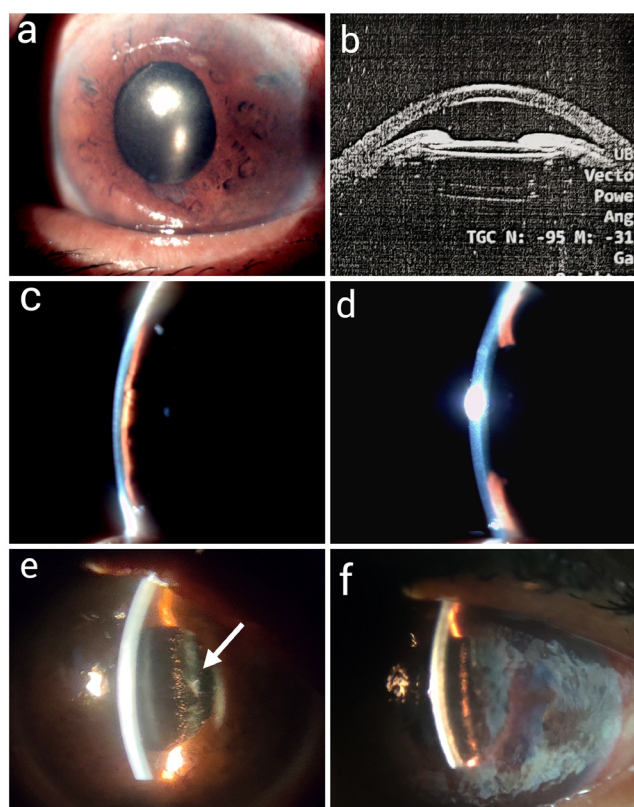


Figure 2: Left eye (a) Diffuse illumination showing shallow AC with PI openings at first visit (b) Ultrasound biomicroscopy picture at first visit showing anterior rotation of ciliary processes and anterior shift of IOL and iris diaphragm. (c) Obliterated peripheral anterior chamber after atropine withdrawal. (d) Extremely shallow central AC after atropine withdrawal. (e) Reformation of AC after central posterior capsule-hyaloidotomy (white arrow) (f) Another recurrence with re-shallowing of AC and diffuse corneal edema just before PPV and IZH.

the PI opening at 9:30 clock hours [Figure 1a], leading to vitreous extrusion through the PI and the anterior chamber was formed instantaneously. Nd: YAG laser vitreolysis of prolapsed vitreous was also done to prevent future blockage. Patient was put on anti-glaucoma drugs and atropine sulfate 1% drops which was slowly withdrawn in 3 months. Fundus examination showed a cup:disc ratio of 0.3:1 in both eyes. YAG laser PI was done in LE after a week. A follow up after two years showed a deep AC with small vitreous herniation without any endothelial touch and IOP controlled on one drug in RE [Figures 1-d]. Gonioscopy revealed peripheral anterior synechiae (PAS) formation in two quadrants and open-angle with patchy pigment in the remaining two quadrants in RE [Figures 1e and f], whereas LE angles were open. The patient had an uneventful cataract surgery in LE at this time.

After three years, the patient revisited with a history of severe headaches and pain in LE for 5 months. On examination, his RE was stable with CDVA of 6/9 and IOP, 15 mm on one drug. His LE had a vision of hand movements and IOP, 55 mm. Slit lamp examination showed diffuse shallowing of the anterior chamber with a patent PI in LE [Figure 2a]. In ultrasound biomicroscopy (UBM) the ciliary

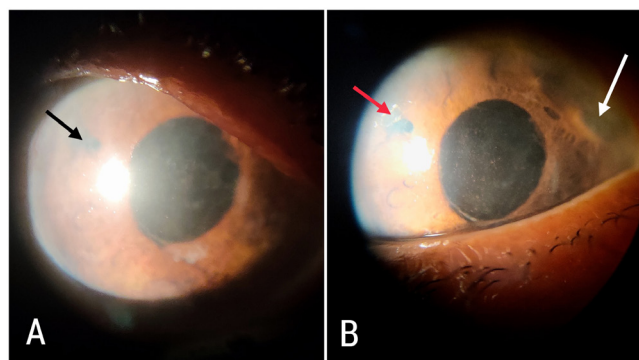


Figure 3: Left eye (a) Recurrence of aqueous misdirection after PPV with blocked surgical IZH opening (black arrow) and corneal edema. (b) Slit lamp picture after YAG laser assisted reopening of blocked fistula (red arrow) showing resolved corneal edema and focal areas of iridocorneal synechiae formation (white arrow).

processes appeared anteriorly rotated [Figure 2b]. He was prescribed systemic and topical ocular hypotensives and atropine sulphate 1% drops, which reformed the anterior chamber in a week. Atropine withdrawal after 3 weeks caused a re-shallowing of AC [Figures 2c, 2d]. Due to the presence of dense arcus senilis, Nd:YAG laser capsulo-hyaloidotomy was attempted through the central posterior capsule which temporarily reformed the anterior chamber [Figure 2e] and improved vision to finger counting at 3 metres. The patient returned after 6 months with a very shallow chamber, corneal edema and IOP of 65 mm in LE [Figure 2f] and gave a history of not having used any medicines for last 3 months. A Pars plana vitrectomy (PPV) with irido-zonulo-hyaloidectomy (IZH) was done in LE, which formed the anterior chamber. We noted a recurrence with raised IOP and diffusely shallow AC after one week of vitrectomy and the surgical IZH opening was found to be blocked with fibrin and pigment [Figure 3A]. Nd: YAG laser was used to open the fistula [Figure 3B]. At three months follow-up visit, the patient was comfortable with a well-formed anterior chamber and a vision of finger counting at 1 meter and IOP of 22 mm in LE on 3 glaucoma drugs.

DISCUSSION

In our patient, there was no reported history of angle closure glaucoma before cataract surgery. However, at the time of acute episode in right eye, the left eye showed signs of a primary angle closure suspect, because of which we did a prophylactic laser PI and the eye remained stable till 3 years after cataract surgery. The axial shallowing of the anterior chamber in the presence of a patent iridotomy in LE and failure of AC reformation after creating a PI in RE confirmed our diagnosis of malignant glaucoma which was supported by the absence of posterior segment pathology and supraciliary effusion in B-scan ultrasonography and UBM.

The therapeutic efficacy of laser hyaloidotomy has been found to be limited in previous studies. Dave *et al.* in their series found complete resolution after laser hyaloidotomy in

46.7% of cases.⁷ Debrouwere *et al.* in their retrospective series of 24 eyes with malignant glaucoma, reported a relapse rate of 75% after YAG capsulotomy-hyaloidotomy.⁸

Although we performed laser capsulo-hyaloidotomy in both eyes, a complete resolution was achieved only in RE in which the procedure was done through the peripheral iridotomy in the acute phase of the disease. No recurrence was noted for 6 years (72 months) thereafter, despite the presence of vitreous in the fistula. To the best of our knowledge, this is the longest-reported success of laser capsulo-hyaloidotomy in malignant glaucoma. However this finding is not in agreement with previous reports in which the blockage of laser hyaloidotomy with vitreous is considered as the reason for relapses.⁸ In our case the complete obstruction of hyaloidotomy fistula may have been prevented by simultaneous laser vitreolysis, the presence of large PI and thin and degenerate vitreous. In LE, laser treatment was done nearly 6 months after the appearance of symptoms through the central posterior capsule as a peripheral hyaloidotomy could not be completed because of the presence of a dense peripheral arcus senilis. The procedure was effective only till the patient was compliant with cycloplegic and glaucoma drugs.

Various surgical techniques have been described for malignant glaucoma through anterior and posterior routes.⁷⁻¹⁰ Debrouwere *et al.* reported 100% success after the vitrectomy tunnel technique comprising of complete PPV with IZH with phacoemulsification (in phakic eyes) and a relapse rate of 66% after anterior vitrectomy and iridectomy-zonulectomy.⁸ They explained that a partial vitrectomy can cause a blockage of the fistula with remaining vitreous and is the reason for frequent recurrences.

Dave *et al.* reported recurrent aqueous misdirection post vitrectomy, which resolved with YAG laser hyaloidotomy and membranotomy.¹¹ In our patient a recurrence was noted in the left eye after one week of vitrectomy caused by blockage of surgical fistula with inflammatory membrane or pigment debris and the anterior chamber was immediately reformed after YAG laser treatment.

CONCLUSION

In this unusual case, we reported sequential spontaneous onset malignant glaucoma in both eyes of a patient after 2 years (RE) and 3 years (LE) of cataract surgery. We also reported successful management with laser hyaloidotomy in the right eye of the patient without any recurrence for 6 years. A more refractory disease in left eye with late presentation was managed with pars plana vitrectomy and irido-zonulo-hyaloidectomy.

CONFLICT OF INTEREST

The authors declare no conflict of interest and no public and private support.

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