

Management of a Bilateral Post-Uveitic Complex Glaucoma with Pupillary Block, Rupture of the Anterior Lens Capsule, and Malignant Glaucoma following Laser Peripheral Iridotomies: Case Report and Literature Review

Khaled El Matri^{1,2,3}, Dhouha Gouider^{1,3}, Rim Limaïem^{1,2,3}, Ahmed Chebil^{1,2,3}, Meher Henchiri^{1,3}, Yousra Falfoul^{1,2,3}, Leila El Matri^{1,2,3}

¹Department B, Institut Hedi Rais d'ophtalmologie de Tunis, Tunis, Tunisia, ²Oculogenetic Laboratory, LR14SP01, Tunis, Tunisia, ³Faculté de Médecine de Tunis, Université Tunis – El Manar, Tunis, Tunisia

Abstract

Purpose: To report a case of a bilateral complex uveitic glaucoma (UG) with pupillary block, rupture of the anterior lens capsule, and malignant glaucoma in a young high-myopic patient and to report anterior segment optical coherence tomography (AS-OCT) findings initially and following surgery.

Methods: A 21-year-old high-myopic woman who had a history of anterior uveitis with extensive posterior synechiae, presented with acute bilateral ocular pain, redness, and blurred vision following bilateral Nd: YAG laser peripheral iridotomy (LPI).

Results: Visual acuity was limited to light perception in both eyes (OU), with a flat anterior chamber (AC) and anterior luxation of lens fragments. Intraocular pressure (IOP) was over 60 mmHg OU. AS-OCT showed closed angles and hyperreflective heterogeneous material within the flat AC. The iris and lens fragments were plated against the corneal endothelium OU. We performed an urgent pars plana vitrectomy associated with lensectomy. It was uneventful in OU. Repeated AS-OCT revealed a deep AC, widely open angles, and aphakia. IOP was lowered to 9 mmHg and visual acuity improved to 5/10 in OU.

Conclusion: Performing LPI might be harmful in the presence of UG with extensive posterior synechia, resulting in complex mechanism glaucoma with aqueous misdirection syndrome associated with a pupillary block due to anterior lens luxation, even in high-myopic eyes. Nd: YAG LPI should not be performed simultaneously in OU, especially in pathologic eyes, to prevent bilateral vision-threatening complications. AS-OCT was of great help, allowing easy and detailed ultrastructural assessment of the ACs, and iridocorneal angles before and after surgery.

Keywords: Anterior segment optical coherence tomography, High-myopia, Laser peripheral iridotomy, Lens luxation, Lensectomy-vitrectomy, Malignant glaucoma, Pars plana vitrectomy, Pupillary block, Uveitic glaucoma

Address for correspondence: Khaled El Matri, Department B, Institut Hedi Rais d'ophtalmologie de Tunis, Boulevard du 9 Avril 1938, 1006 Tunis, Tunisia.
E-mail: khaled.elmatri@gmail.com

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INTRODUCTION

Malignant glaucoma (MG) is a rare and potentially devastating condition. Its characteristics are uniform shallowing of the anterior chamber (AC) and elevated intraocular pressure (IOP), with a patent peripheral iridotomy.¹ It is alternatively called

“ciliary block glaucoma” or “aqueous misdirection syndrome” based on its presumed pathogenic mechanisms.¹ In general, it is observed following trabeculectomy in hyperopic patients. Cataract surgery and iridotomy (laser or surgery) represent

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other incriminated etiologies.²⁻⁴ Spontaneous MG is also possible.⁵

Uveitic glaucoma (UG) is a clinical challenge in diagnosis and management,⁶ as it can be caused by one or more mechanisms. These include acute uveitic angle closure due to extensive posterior synechiae responsible for pupillary seclusion and uveal effusion by forward rotation of the ciliary body and iris root.¹ Other accounted for mechanisms are corticosteroid-induced ocular hypertension and mixed-mechanism ocular hypertension caused by chronic damage to the trabecular meshwork. Studies have shown that complex UG with pupillary block, rupture of the anterior lens capsule, and MG remains a rare condition,⁶ hence the topic of our research.

In the framework of the present case study, we report a case of bilateral complex UG associating pupillary block, rupture of the anterior lens capsule, and MG in a young high-myopic patient, assessed with anterior segment optical coherence tomography (AS-OCT), before and after surgery.

CASE REPORT

A 21-year-old high-myopic woman (−8.50 diopter [D]) presented to the emergency department with acute bilateral ocular pain, redness, and blurred vision following bilateral Nd: YAG laser peripheral iridotomy (LPI).

The patient reported being treated a few years prior for bilateral anterior uveitis of unknown etiology with topical steroids. According to her physician, she presented extensive posterior synechiae resulting in a secondary pupillary block, with ocular hypertension refractory to medical treatment (around 25–30 mmHg under local treatments). Her physician performed Nd: YAG LPI on both eyes (OU) in the same day. We do not have any data related to her initial IOP or her best-corrected visual acuity (BCVA) before laser procedures.

On presentation 1 day following LPI, the patient's BCVA was limited to light perception in OU. She had bilateral conjunctival hyperemia and corneal edema. AC was flat in the periphery while central AC was completely filled with lens fragments, preventing a detailed examination of the anterior segment. Hence, the fundus examination was similarly inaccessible. Goldmann IOP was 69 mmHg in the right eye (RE) and 64 mmHg in the left eye (LE).

AS-OCT (Topcon DRI OCT Triton, Tokyo, Japan) showed anterior iris displacement with closed angles, in OU. A hyperreflective heterogeneous material was present in the ACs corresponding to luxated lens fragments. Peripheral iris and lens fragments were plated against the corneal endothelium. Iris defects with posterior hypertransmission were noted within peripheral iridotomies localizations that were blocked with lens fragments [Figure 1a and b]. Central corneal thickness was 558 μm in OU. B-mode ultrasonography ruled out posterior segment abnormalities as there was no retinal detachment in either eye, and vitreous and choroid were

unremarkable. A-mode ultrasonography found an axial length of 27.14 mm in the RE and 27.43 mm in the LE. Unfortunately, ultrasound biomicroscopy (UBM) was not immediately available in the department to eventually confirm the presence of ciliary rotation.

We urgently admitted the patient for acute ocular hypertension secondary to bilateral complex UG with pupillary block, MG, and anterior lens fragments luxation. Accordingly, she was initially treated with topical antiglaucoma eye drops and intravenous mannitol 20% to eventually reduce IOP before surgery. However, we did not observe any pressure-lowering. We decided to urgently perform 23G pars plana vitrectomy (PPV) associated with lensectomy–vitrectomy under general anesthesia. Corneal edema, posterior synechiae, and lens material filling the AC prevented us from having intraoperative visibility initially. After inserting the trocars, we started with a central vitrectomy while the infusion was stopped to help reduce the severely elevated IOP and prevent a sudden increase in IOP. Central vitrectomy was followed by posterior lensectomy and removal of lens fragments within the AC, using the vitrectomy cutter. As AC remained flat, extensive PPV was completed under fluid infusion, and we did observe intraoperative AC deepening only after extreme peripheral vitrectomy and vitreous base shaving. We first performed surgery in the RE. Then, the LE was operated on the following day using the same surgical technique, as the IOP did not lower without surgery. The procedures were uneventful in OU.

On postoperative day 1, IOP was normalized to 9 mmHg in OU, and postoperative BCVA on the 7th day after the second procedure was 5/10 (20/40) in OU. Ocular examination revealed resolution of corneal edema, deepening of the AC, and reopening of iridocorneal angles. Both peripheral iridotomies were patent and moderately peripheral. Fundus examination showed bilateral myopic staphylomas and tilted disc syndromes without optic disc excavation nor color change, and ruled out the presence of peripheral retinal lesions. AS-OCT has been performed again in each eye one day following the respective procedures. Each exam revealed a normally deep anterior chamber with widely-open iridocorneal angles, and aphakia (no crystalline lens) [Figure 1c].

BCVA remained stable 12 months postoperatively. Secondary implantation with scleral-fixated intraocular lenses is planned.

The patient has consented for the submission and the publication of this case report and our local ethics committee approved it.

DISCUSSION

MG is a rare condition, usually observed in hyperopic or even nanophthalmic eyes. Risk factors predisposing to ciliary block include short axial length, narrow angles, and plateau iris configuration.⁷ However, angle-closure has been reported in high-myopic eyes, with few cases of MG.^{8,9}

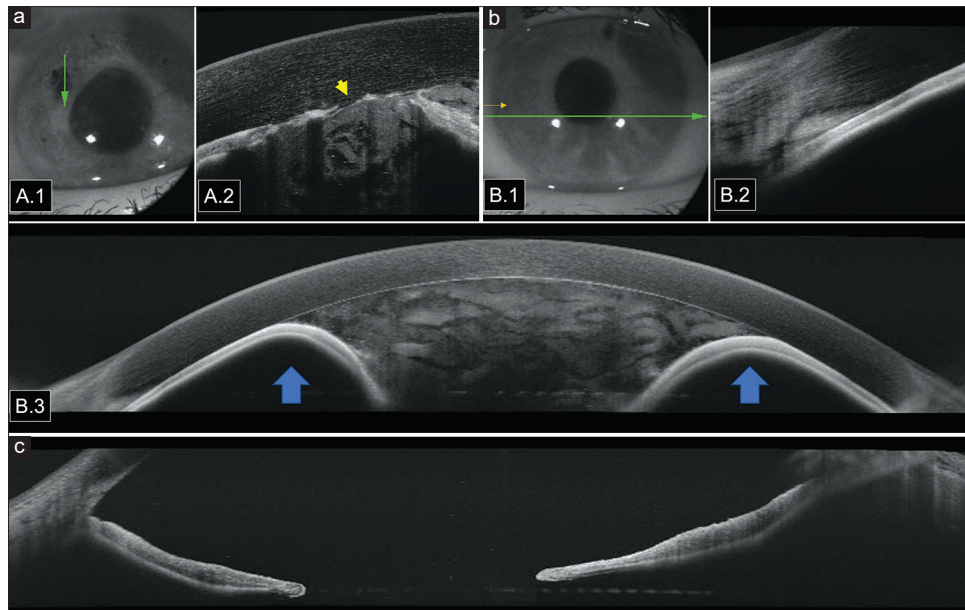


Figure 1: Anterior segment optical coherence tomography (AS-OCT) of malignant glaucoma and anterior lens luxation, initially (A, B) and after surgery (c). (a) Initial imaging of the right eye: Infrared anterior segment photography of the right eye (A.1) showing a mid-dilated pupil and a superior temporal moderately peripheral large iridotomy. Green arrow corresponds to the level of AS-OCT passing through the peripheral iridotomy (A.2) showing an extensive iris defect (yellow arrowhead) with posterior hypertransmission presence of lens material within the iridotomy. (b) Initial imaging of the left eye: Infrared AS photography of the left eye (B.1) showing a mid-dilated pupil and a superior smaller peripheral iridotomy. Orange arrow corresponds to the level of AS-OCT passing through the nasal iridocorneal angle (B.2) showing a completely closed angle. Green arrow corresponds to the level of wide-angle AS-OCT (B.3) showing hyperreflective heterogeneous material within the anterior chamber (AC) (luxated lens fragments), flat AC with iris and lens fragments plated against the corneal endothelium, and anterior displacement of the iris (blue arrows) due to vitreous pressure central corneal thickness was normal (558 μm). (c) Postoperative wide-angle AS-OCT of the left eye: Widely-open iridocorneal angles, absence of crystalline lens (aphakia), and deep AC

MG is commonly reported in the aftermath of filtering surgery in angle-closure glaucoma. Equally common causes are cataract extraction and implantation of several glaucoma drainage devices.³ The study of a few cases has shed the light on additional associations with LPI, such as surgical peripheral iridotomy, capsulotomy, and the use of miotic agents, as well as spontaneous occurrence.^{5,7,10} Yet, MG following LPI is considered rare.

The mechanisms of UG are not fully understood. While angle-closure UG is usually caused by a pupillary block secondary to posterior synechiae, association with anterior lens rupture and MG is quite rare.⁶ Explanations of this condition differ. On the one hand, the anterior lens capsule rupture could be secondary to the abnormal fragility of the capsule and the extensive posterior synechiae, but on the other hand, it could also be induced by incidental laser impact following LPI.⁶

In the present case, the young patient was highly myopic (-8.50 D), with an axial length of 27.14 mm in the RE and 27.43 mm in the LE, and bilateral tilted disc syndromes with myopic staphylomas. According to the data gathered, she presumably developed bilateral MG following LPI for extensive posterior synechiae with refractory ocular hypertension. The diagnosis of MG, in this case, was established based on a range of factors, mainly the presence of flat ACs, very high IOP over 60 mmHg, patent peripheral iridotomies, and the absence of choroidal effusion or suprachoroidal hemorrhage. Unfortunately, we

could not perform UBM to confirm the presumed mechanism of ciliary rotation. Indeed, the pathogenesis consists of the ciliary body anterior rotation preventing the aqueous humor from following its normal pathway. It results in its accumulation behind the iris–lens diaphragm. The aqueous humor gets trapped in the vitreous fluid, leading to an anterior iris–lens diaphragm displacement, AC flattening, and secondary angle closure. As the vitreous pressure rises, a one-way valve leads the anterior vitreous to a forward movement.^{2,11}

Different mechanisms eventually producing ciliary body edema or constriction have been hypothesized for ciliary blocks following iridotomies.¹² In this context, laser iridotomy might be an initiating factor. The association of the miotic agents use, the presence of lens dislocation, and the application of extensive iridotomy are contributing factors to the onset of ciliary block.^{7,12}

In this case, we noted extensive iris defects on the initial AS-OCT. The postoperative assessment showed that both peripheral iridotomies were patent and moderately peripheral. The latter findings might have contributed to MG. Besides, the patient had a history of anterior uveitis with extensive posterior synechiae. In addition to presumed MG in our case, there was a particular association with pupillary block due to anterior lens fragments luxation. Two theories were put forward: it was either due to incidental laser impact with anterior capsule tear and crystalline lens perforation, as previously reported in

extremely rare cases,¹³⁻¹⁵ or forward movement of hydrated vitreous with anterior displacement of the iris–lens diaphragm in the presence of circumferential posterior synechia which could have led to anterior lens capsule ripping with extraction and luxation of lens fragments into the AC.

Once MG is diagnosed, urgent treatment is necessary to relieve misdirection of aqueous humor and restore physiological aqueous circulation. Cycloplegics would improve aqueous flow by tightening the lens zonules and pulling the iris–lens diaphragm posteriorly. Osmotic agents would dehydrate the vitreous volume, and aqueous suppressants would reduce the production of aqueous humor.¹¹ However, vitreous aspiration is necessary in 50% of cases.⁸ Indeed, the mentioned medications were ineffective in lowering IOP in this patient. Besides, the presence of lens fragments in the AC ruled out the medical therapeutic option alone. Therefore, a surgical procedure with PPV and lensectomy was necessary.

PPV is an efficient treatment option in MG cases. It can reduce the vitreous volume and restore AC depth. However, the ciliary block and the aqueous misdirection would persist,¹ unless a surgical disruption of the anterior hyaloid is combined with PPV.¹¹ Some authors recommend the anterior vitrectomy, while some others demonstrated that the remaining vitreous would move forward blocking the created passage and leading to MG recurrences.² Total vitrectomy combined with zonulectomy, iridotomy, and capsulectomy was effective in 100% of cases in a large series of patients.²

In this case, lens extraction and anterior vitrectomy were essential. However, there was a high risk of corneal endothelial injury when operating in a shallow AC and a threatening risk of expulsive choroidal hemorrhage when operating the anterior segment with an IOP over 60 mmHg. Therefore, we performed 23G complete PPV associated with posterior lensectomy and removal of lens fragments within the AC. Infusion was stopped initially during central vitrectomy to prevent the sudden increase in IOP. In our case, we did observe AC deepening during surgery only after extreme peripheral vitrectomy and vitreous base shaving. It consolidated the presumed mechanism of MG with a ciliary block and not only a pupillary block mechanism. We performed the same procedure for OU separated by 1 day. The surgery was uneventful, with satisfactory postoperative results.

We observed the following: IOP decreased to 9 mmHg in OU and BCVA improved dramatically from light perception to 5/10 (20/40) in OU. We also noted a resolution of corneal edema with deepening of ACs. Postoperative AS-OCT revealed deep ACs, widely-open iridocorneal angles, and aphakia.

MG or misdirection syndrome is among the most challenging problems faced by ophthalmologists today, and it is rarely observed following Nd: YAG LPI. However, performing LPI might be harmful in the presence of UG with extensive posterior synechia, resulting in complex mechanism glaucoma with aqueous misdirection syndrome associated with a pupillary block due to anterior lens luxation, even in high-myopic eyes. Nd: YAG LPI should not be performed

simultaneously in OU, especially in pathologic eyes, to prevent bilateral vision-threatening complications.

AS-OCT was of great help in the presence of material such as lens fragments in the AC hampering clinical examination. It allowed easy and detailed ultrastructural assessment of the ACs and iridocorneal angles before and after surgery.

Acknowledgment

This case report has been presented as a poster presentation at the European Society of Cornea and Refractive Surgery (ESCRS) congress held in Amsterdam (Netherlands) in October 2021.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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