

# Spontaneous simultaneous bilateral malignant glaucoma of a patient with no antecedent history of medical or surgical eye diseases

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**Purpose:** Malignant glaucoma, or aqueous misdirection syndrome, is a condition characterized by sudden intraocular pressure (IOP) elevation, and it is usually unilateral and induced by ocular surgical intervention or by medical therapy. Here, we report a case of simultaneous bilateral malignant glaucoma in a young patient with no history of any ocular diseases.

**Case report:** A case of a 24-year-old female with no apparent previous history of ocular medical or surgical conditions was referred to our hospital because of recent bilateral IOP elevation associated with a severe drop in vision and shallow anterior chamber with no posterior segment anomalies detected by ocular ultrasound in both eyes. Yttrium aluminum garnet (YAG) laser iridotomy dropped the IOP only temporarily and the patient received topical atropine treatment with combined trabeculectomy and anterior vitrectomy.

**Results:** In this case, the patient had a typical presentation of bilateral malignant glaucoma and her IOP dropped only temporarily following laser iridotomy to rise again shortly thereafter. Also, deepening of the anterior chamber and IOP decrease after topical atropine was very supportive of the diagnosis of malignant glaucoma. Successful management with trabeculectomy and limited vitrectomy also affirmed the diagnosis of malignant glaucoma.

**Conclusion:** This is a very rare case of bilateral malignant glaucoma in a young adult without any prior eye conditions; only one similar case has been reported in the literature. We propose our own theory regarding this simultaneous occurrence of the pathology based on previously published studies about the presence of communication between the two eyes along the cerebrospinal fluid pathways.

**Keywords:** intraocular pressure, iridotomy, malignant glaucoma, young adult

## Introduction

Malignant glaucoma, or aqueous misdirection syndrome, was first described by Von Graefe in 1869 as an aggressive form of postoperative glaucoma that is resistant to treatment and can result in blindness.<sup>1</sup> This entity is characterized by intraocular pressure (IOP) elevation, with shallowing to complete flattening of the anterior chamber (AC) in the presence of a patent peripheral iridectomy and in the absence of posterior segment anomalies, particularly suprachoroidal hemorrhage.<sup>2</sup> Malignant glaucoma is thought to occur in anatomically predisposed eyes,<sup>2</sup> with one of the proposing mechanisms being posterior misdirection of the aqueous humor within or behind the vitreous substance. This produces a continuous expansion of the vitreous cavity and increased posterior segment pressure, leading to anterior displacement of the lens–iris diaphragm in phakic and pseudophakic eyes, or forward displacement of the anterior hyaloid face in aphakic eyes.<sup>3</sup>

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This generates a vicious cycle, in that the higher the pressure in the posterior segment, the more firmly the lens is held forward. The IOP is classically markedly increased, but it may also be normal.<sup>4</sup> Malignant glaucoma has been reported in different situations, such as following cataract surgery, laser iridotomy, neodymium:yttrium aluminum garnet (Nd:YAG) cyclophotocoagulation or sclerotomy; following glaucoma drainage device implantation, viscoelastic use, intravitreal injection of triamcinolone acetonide; with *Aspergillus flavus* intraocular infection; and with the ingestion of topiramate or sulfa drugs.<sup>5,6</sup> Only a few cases of isolated malignant glaucoma have been reported in eyes without any antecedent eye diseases or surgery, and only one case of bilateral simultaneous idiopathic malignant glaucoma has already been reported in the literature.<sup>7</sup>

Here, we report a case of spontaneous, simultaneous, bilateral malignant glaucoma in a young female with no systemic or eye diseases and no prior medical or surgical eye treatments. We also propose a postulated mechanism that might explain the simultaneous bilaterality of this case.

## Case report

The patient is a 24-year-old female who was referred to our eye center in Mediclinic Dubai Mall, Dubai, United Arab Emirates, because of recent bilateral IOP elevation with a severe drop in vision. She had been hospitalized elsewhere 1 week earlier because of severe bilateral frontal headache, nausea, and vomiting. Initially, she was investigated for possible central nervous system disease. Blood tests and brain magnetic resonance imaging (MRI) were performed and revealed no abnormalities. During her stay, there was a progressive worsening of the condition. After 5 days in hospital, the patient complained of loss of vision in the right eye, and after approximately 6 hours, also in the left. Another brain MRI was normal. The ophthalmology consultant suspected bilateral acute angle-closure glaucoma as a possible cause for the severe bilateral loss of vision. The patient was thus referred to our eye center for urgent management.

Our examination revealed visual acuity of hand movements alone in both eyes. Goldmann applanation pressure was >60 mmHg in both eyes; the patient presented with severely miotic pupils and flat chambers, with almost iridocorneal touch centrally, cloudy corneas, and severe bulbar conjunctival injection. Bilateral diagnostic ultrasound revealed no posterior segment anomalies. The patient's past history was negative for any medical or surgical diseases and her eye history was also negative. Of note, a routine eye examination had been performed a few months earlier and

was reported to be normal. Initial management at our center consisted of multiple topical antiglaucoma medications and intravenous acetazolamide (500 mg). Ultimately, we managed to perform bilateral Nd:YAG laser peripheral iridotomy despite the corneal clouding. After the peripheral iridotomy, the IOP dropped in both eyes to around 45 mmHg, but it rose back to its original level after less than half an hour. The diagnosis of bilateral malignant glaucoma was raised and the patient was admitted to hospital for surgical management. Topical atropine 1% drops and intravenous mannitol (1 g/kg of body weight) were started, and the patient was put under continuous monitoring. In less than 2 hours, the ACs started to deepen in both eyes, with partial clearing of the corneas. The IOP dropped again to 45 mmHg in both eyes. At that stage, the possibility of bilateral malignant glaucoma as a diagnosis was more likely.

The next day, the patient was scheduled for trabeculectomy with mitomycin-C (0.02% for 2 minutes) combined with pars plana vitrectomy. Informed consent was obtained from the patient prior to surgery. In the meantime, overnight treatment with topical atropine and antiglaucoma medications was continued, along with intravenous acetazolamide. A second dose of intravenous mannitol was repeated 1 hour before the planned surgery. At the time of surgery, the IOP in both eyes was around 25 mmHg. Surgery was performed in the right eye as planned, and the same procedure was carried out in the left eye 1 day later. In brief, the surgical procedure was performed in both eyes as follows: a limbus-based conjunctival flap was performed, followed by topical 0.2% mitomycin-C application for 2 minutes; prior to the dissection of a 3×3 mm half-thickness trabeculectomy flap, one-port limited core vitrectomy was performed through a pars plana approach 4 mm from the limbus. After completion of the vitrectomy, the trabeculectomy was completed by entering the AC through a small sclerotomy opening using a super blade. The trabeculectomy flap was sutured back using two 10-0 nylon titrated sutures, and the conjunctiva was closed using running 8-0 VICRYL® (Johnson & Johnson, New Brunswick, NJ, USA). Postoperatively, the patient was put on a topical combination of tobramycin (3 mg/mL) and dexamethasone (1 mg/mL), and atropine was continued once a day for 2 weeks. Both eyes had well-formed superior blebs and, surprisingly, visual acuity returned to 20/20 without correction, and there was no optic nerve (ON) damage in either eye. The IOP remained <18 mmHg all throughout the postoperative follow-up period, which extended for more than 1 year, with both eyes maintaining deep chambers and reactive pupils.

The study was approved by the review board/ethics committee of the Beirut Eye Specialist Hospital, Beirut, Lebanon. All patients signed an informed consent prior to treatment.

## Discussion

The main features of malignant glaucoma (or aqueous misdirection syndrome) include: increased IOP; the presence of shallowing to complete flattening of the AC; the absence of posterior segment anomalies – particularly suprachoroidal hemorrhage; no response to peripheral iridectomy; and adequate response to topical atropine.<sup>2,7</sup> Malignant glaucoma is difficult to treat and characteristically progresses to blindness.<sup>2</sup> In our case, the patient had a typical presentation of malignant glaucoma and the IOP dropped only temporarily after laser iridotomy, to rise again shortly thereafter. Also, deepening of the AC and IOP decrease after topical atropine was very supportive of the diagnosis of malignant glaucoma. Successful management with trabeculectomy and limited vitrectomy also confirmed the diagnosis of malignant glaucoma. Although malignant glaucoma typically occurs in predisposed eyes, especially among those subjected to surgical or medical therapy, only a few cases have been reported where isolated malignant glaucoma occurs without a previous history of any kind of treatment and, to the best of our knowledge, only one case of bilateral simultaneous idiopathic malignant glaucoma has been reported in the absence of any history of previous systemic or eye diseases.<sup>7</sup> As such, our case is considered as the second report of bilateral idiopathic malignant glaucoma.

The rare phenomenon of malignant glaucoma occurring simultaneously in both eyes of young patients who do not have any predisposing factors is highly unusual and warrants exploration. Malignant glaucoma is thought to occur in anatomically predisposed eyes,<sup>2</sup> with one of the proposed mechanisms being posterior misdirection of the aqueous humor within or behind the vitreous body, which leads to anterior displacement of the iris–lens diaphragm and anterior hyaloid face.<sup>3</sup> In our case, the occurrence of malignant glaucoma in one eye (most probably the right eye first, as deduced from the patient's own story) was in an anatomically predisposed eye, and the aqueous entrapped behind the vitreous established a pressure gradient between the ON head and the cerebrospinal fluid (CSF) in the subarachnoid space (SAS) of the ON. This pressure gradient encouraged fluid movement toward the SAS of the ON and subsequently to the SAS of the brain. Migration of fluid to the SAS of the contralateral ON could occur directly through the optic chiasm, or it could be mediated by the elevated CSF pressure

within the SAS of the central nervous system through the phenomenon of bidirectionality of CSF flow.<sup>8</sup> This proposed mechanism of aqueous humor migration from the vitreous cavity to the ON and CSF of the brain is also supported by the observation of silicone oil (SO) migration from the vitreous cavity to the ON and to the brain in vitrectomized eyes.<sup>9,10</sup> Also, Espinosa et al<sup>11</sup> reported the migration of SO into the ON of the contralateral eye causing optic neuritis. This migration is facilitated if there is a gradient of pressure between the vitreous cavity and the CSF, or in a predisposed ON head, such as in the presence of an ON pit.<sup>12</sup>

## Conclusion

In conclusion, one possible explanation for the occurrence of simultaneous bilateral malignant glaucoma is that the entrapment of aqueous humor behind the posterior vitreous face can establish a pressure gradient between the vitreous cavity and the fluid around the orbital ON. This facilitates the migration of fluid to the SAS of the ON, and then to the SAS of the contralateral orbital ON, either directly through the optic chiasm or due to increased intracranial CSF pressure. The fluid subsequently migrates to the vitreous cavity of the contralateral eye and pushes the total vitreous core anteriorly; this precipitates forward movement of the anterior hyaloid face and lens–iris diaphragm, resulting in malignant glaucoma in the contralateral eye. This postulated mechanism could also explain the observed phenomenon of immediate constriction of the contralateral pupil when a strong miotic agent is injected into the AC of the first eye upon completion of the phakic posterior chamber intraocular lens (PIOL) implantation in a planned bilateral PIOL surgery. Given this observed phenomenon, we avoid the injection of miotic agents into the AC upon completion of surgery on the first eye in the setting of bilateral implantation of phakic PIOL. Although it is very rarely indicated in our surgical practice of cataract surgery, the same phenomenon is observed during bilateral cataract surgery. One more lesson to learn from this case is that severe IOP elevation causes symptoms that can mimic those of certain central nervous system disorders. Finally, the diagnosis of idiopathic malignant glaucoma is challenging because, on the one hand, it is an uncommon entity, and on the other hand, early intervention with vitrectomy could possibly maintain good visual acuity and prevent blindness.

## Disclosures

The study did not receive external funding. None of the authors has any proprietary, commercial, or financial interest

in any of the products mentioned. The authors report no conflicts of interest in this work.

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