Case Report
The successful therapy of intravenous steroid on spontaneous and simultaneous bilateral malignant glaucoma post monocular phacoemulsification

Rui Hua, Hong Ning, Yizhou Sun, Jingnan Han, Xue Bai
Department of Ophthalmology, First Hospital of China Medical University, Shenyang, China

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Abstract: We describe a rare case of bilateral malignant glaucoma (MG) after phacoemulsification in a 59-year-old woman with binocular cataracts, pulmonary fibrosis and hypertension. Phacoemulsification and intraocular lens implantation was performed for the left eye, and ten hours later, the patient complained of binocular pain due to recent bilateral intraocular pressure (IOP) elevation associated with a severe drop in vision and a shallow anterior chamber with no posterior segment anomalies detected by ocular ultrasound in both eyes. A bolus of steroid therapy was initially used to handle such lesions, and it was found to be effective. The potential underlying mechanism may be related to choroidal expansion, communication between the two eyes along the cerebrospinal fluid pathways, and the bilateral simultaneous immune response in choroids due to pulmonary fibrosis. Steroid therapy effectively reduced choroidal edema and anti-inflammatory activity to control MG.

Keywords: Phacoemulsification, intraocular pressure, malignant glaucoma, steroid therapy

Introduction

In 1869, malignant glaucoma (MG) was first coined by von Graefe to describe an aggressive form of postoperative glaucoma that is resistant to treatment and leads to blindness [1]. It is also known by alternative names such as ciliary block glaucoma, aqueous misdirection syndrome, and direct lens-block glaucoma [2-4], and it has been reported in different situations such as following cataract surgery [5]. Traditionally, the proposed underlying pathogenic mechanisms include ciliolenticular block presumably induced by anterior movement of the lens-iris diaphragm, poor vitreous conductivity, and choroidal expansion [6]. Ultrasound biomicroscopy (UBM) also confirmed that, during an attack, the ciliary processes lie parallel to the longitudinal muscle, and in some cases the processes indent the posterior iris [7]. Following resolution, the ciliary body and processes rotate back to their anatomic position [8] with the deepening of the anterior chamber (AC) [9]. In addition, it is plausible that MG may arise in some of these situations following ciliary body swelling and the formation of an inflammatory barrier in the zonular-capsular region, which impedes the anterior flow of the aqueous humour [10]. However, the current etiological hypothesis that is supported by the most evidence is the forward movement of the vitreous via choroidal expansion [6]. Inward expansion of the choroid is normally opposed by intraocular pressure (IOP). Some inciting event, such as intraoperative inflammation or hypotony, could lead to breakdown of the blood-choroid barrier and to protein exudate in the extravascular choroid. Choroidal expansion and the diverted aqueous will induce poor vitreous conductivity or vitreous block. Vitreous block appears to be more common in female patients [11], potentially in association with a more anterior lens and smaller anterior chamber volume [12]. Moreover, risk factors for the development of vitreous block include a history of vitreous block in the fellow eye, nanophthalmos [13], plateau iris [14], zonular laxity predisposing to an anterior lens-iris diaphragm [7], larger phakic lenses [15], preoperative shallow anterior chamber, and preoperative high IOP [16]. A short axial length is a common finding, although vitreous block may occur in eyes with a range of
axial lengths [11]. For the diverted aqueous to be another cause of vitreous block, a one-way valve allowing the entry of aqueous into the vitreous, forming vicious circle, would have to be present.

Only two cases of bilateral simultaneous malignant glaucoma have been reported in the literature [5, 17]. Here, we report a case of post-monocular cataract surgery and simultaneous bilateral MG in an elderly female with pulmonary fibrosis. We also investigate the therapeutic effect of intravenous steroid for the control of MG.

**Case report**

A 59-year-old female was diagnosed with bilateral age-related cataracts, pulmonary fibrosis (rheumatoid factor 113.0 IU/mL, C-reactive protein 31.3 mg/L, IgM 2.6 g/L) and hypertension in our hospital and received phacoemulsification plus intraocular lens implantation in her left eye. The best-corrected visual acuity was 0.5 OD and 0.1 OS (pre-surgery), while the IOP was 20 mmHg OD and 18 mmHg OS. The axial length of the left eye was 23.65 mm. In addition, the anterior chamber depths and angle configurations was normal. Moreover,
Bilateral malignant glaucoma and therapy

there was no psuedoexfoliation, iridodonesis, phacodonesis, glaucomatous optic neuropathy, as well as trauma history. Ten hours post-surgery, the patient complained of binocular swelling pain associated with a severe drop in vision, and clinical examination showed corneal edema, aqueous flare (++), weak pupil reflection (diameter, R: 6 mm; L: 5 mm) and high IOP (T+2, >60 mmHg) correspondingly. The deepest region of the anterior chamber in the left eye and the position of the IOL were normal. In contrast, the deep region of the AC in the right eye was flat. UBM of the right eye revealed a flat AC (1.26 mm), forward rotation of the ciliary body, disappearance of the ciliary sulcus and a small idiopathic supraciliary effusion (Figure 1). Bilateral MG was diagnosed according to the shallowing of the central (axial) AC in association with an increase in IOP and the UBM results.

An intravenous injection of 250 mL 20% manitol was immediately delivered, and oral aqueous suppressants (100 mg methazolamide mixed with 100 mL glycerol) were taken orally. Dexamethasone (2 mg) was injected into the left eye subconjunctivally. Topical cycloplegics (1% atropine eye drops) were used to dilate the bilateral pupils. Unfortunately, the IOP and corneal edema were not relieved the following day. Additional medical management included topical steroid eye drops (0.1% fluorometholone) and intravenous steroid (0.5 g methylprednisolone). The third day post-surgery, the patient had no complaints and presented a binocular clear cornea, dilated pupils (6 mm), and a decrease in the IOP to 14 mmHg. Aqueous flare (+++) was still observed in the left eye to a greater extent than in the right eye. The study adhered to the tenets of the Declaration of Helsinki and was approved by the Medical Research Ethics Committee of the First Hospital of China Medical University, and a written informed consent was obtained from the patient.

Discussion

It is well known that the normal configuration of the ciliary body and iris-lens diaphragm in the pseudophakic eye results in normal aqueous flow toward the AC. In contrast, a malrotation of the ciliary body results in aqueous misdirection. Subsequently, the aqueous becomes trapped in the vitreous fluid, resulting in an elevation of the pressure in the posterior segment and a forward displacement of the iris-lens diaphragm [18]. During cataract surgery, AC decompression may contribute to anterior rotation of the ciliary body by inducing separation of the vitreous base from the pars plana, resulting in aqueous misdirection [19]. The use of cycloplegics has been thought to improve the aqueous flow by tightening the lens zonules and subsequently pulling the lens posteriorly to reverse the shallowing of the AC.

Another cause of the increase in vitreous pressure is choroidal expansion [20], and the initial compensatory outflow of aqueous along the posteroanterior pressure gradient leads to shallowing of the AC. Choroidal expansion has been detected by UBM in eyes with MG, and choroidal effusion secondary to angiooeedema has also been shown to result in MG [21]. A 50-µm choroidal expansion leads to a large increase in IOP [20], and in the presence of high pressures (40-60 mmHg), the vitreous is significantly more resistant to outflow than the AC [22]. The pressure differential leads to vitreous expansion [23] and shallowing of the AC. Choroidal expansion could also explain the high incidence of uveal edema in vitreous block [24]. Indirect support includes the observation of choroidal effusions in 43% of 70 eyes in acute angle-closure glaucoma and in 9% of eyes with narrow angles and no history of angle closure, suggesting a possible link between uveal edema and angle closure [25].

The fluid behind the vitreous gel, which also appeared to be more dense than normal, was believed to prevent the forward flow of aqueous [2]. Schwartz et al. reported that, in cases of vitreous block, the anterior vitreous is abnormally compressed intraoperatively [26], causing positive pressure [27]. Furthermore, it is densely adherent to the ciliary body [28]. The abnormally dense anterior vitreous can push the lens anteriorly into the iris, resulting in a type of phacomorphic [29] angle closure. In aphakic cases, it is thought that the anterior hyaloid directly pushes the ciliary body and iris forward [26]. Both cases lead to anterior movement of the lens-iris diaphragm [30]. Regardless of the mechanism, the final common pathway is the establishment of a vicious cycle in which the transvitreal pressure cannot be equalized by the outflow of the aqueous humour.
After an episode of MG in one eye, there is a high risk of the occurrence of this complication in the fellow eye following the surgical intervention [31]. MG after phacoemulsification in one eye appears to increase the risk for a similar occurrence post-surgery in the fellow eye [32]. The authors proposed a mechanism to explain the fellow eye with MG simultaneously based on the following three lines of evidence.

First, the intraocular fluid and pressure can be transmitted or conducted inversely into the optic nerve. Papp A et al. found that in silicone oil eyes, silicone oil infiltration sites in the optic nerve, in the central retinal artery, and in the subarachnoidal spaces could be located by light microscopy and confirmed by energy-dispersive X-ray analysis. An important factor in this migration process appears to be an elevated intraocular pressure [33]. Second, the intraocular fluid and pressure can be transmitted or conducted into the sella cistena superioris and cerebral ventricles inversely via the optic nerve. Fangtian D used CT imaging, MRI, and OCT to reveal the migration of ocular silicone oils into the sella cistena superioris and the cerebral ventricles due to elevated IOP post-surgery. The silicone may have entered the atrophic optic disk due to an elevated IOP. Communication of the subarachnoidal spaces that surround the optic nerve with the intracranial subarachnoidal spaces would then permit the passage of silicone into the ventricles [34]. Finally, the intraocular fluid and pressure can be transmitted or conducted into the fellow optic nerve and eyes to induce choroidal expansion from the cerebral ventricles under high pressure. Computed cisternography with a contrast agent in idiopathic intracranial hypertension and asymmetric papilloedema cases has revealed a lack of contrast-loaded cerebrospinal fluid (CSF) in subarachnoidal spaces of the optic nerve despite its presence in the intracranial subarachnoidal spaces, suggesting compartmentation of the subarachnoidal spaces of the optic nerve. The finding is further supported by a concentration gradient of brain-derived L-PGDS between the spinal CSF and the CSF from the optic nerve subarachnoidal spaces in the same patients [35].

For treatment, cycloplegia widens the ciliary body diameter, increasing the forward diffusional area for fluid to leave the posterior vitreous cavity. Osmotic agents remove fluid from the eye via the extravascular space of the choroid. A bolus of hyperosmotic fluid would initially pass through the choroidal vessels, permitting the generation of a high osmotic gradient between the vessels and the choroidal stroma [6]. MG with annular uveal swelling has been observed by UBM [36]. Topical steroids can help to reduce inflammation [37]. In the present special case with pulmonary fibrosis, however, uveal swelling and inflammation would be aggravated. We used intravenous steroid (methylprednisolone) to dry the choroidal effusion and an anti-inflammatory to treat the uveal (ciliary body and choroids) swelling.

This study had several limitations. For example, because the situation was an emergency, few imaging materials were recorded. Conduction of the pressure and fluid during bilateral MG may result in increased intracranial pressure, and simultaneously, intravenous mannitol may further reduce the increase in intracranial pressure. These limitations will be improved in future studies.

To the best of our knowledge, this is the first report of a very rare case report of bilateral MG post-monocular phacoemulsification in China. The potential underlying mechanism may be related to choroidal expansion, the presence of communication between the two eyes along the CSF pathways, and the bilateral simultaneous immune response in the choroids due to pulmonary fibrosis. Steroid therapy has been shown to be effective for reducing choroidal edema and providing anti-inflammatory effects for the control of MG. Finally, we prefer to term this lesion as “uvea related MG”, due to the association of uvea in both pathogenesis and treatment.

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Disclosure of conflict of interest

None.

Address correspondence to: Hong Ning, Department of Ophthalmology, First Hospital of China Me-
Bilateral malignant glaucoma and therapy

dical University, No. 155, Nanjingbei Street, Heping District, Shenyang, Liaoning Province, China. Tel: +86-13840392288; E-mail: ninghong2015cmu@126.com

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Bilateral malignant glaucoma and therapy