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Case Report

Spontaneous malignant glaucoma: Case report and review of the literature

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Abstract

Malignant glaucoma usually occurs after anterior segment surgery (typically after glaucoma surgery). The aim of this article is to report a case of spontaneous malignant glaucoma (SpMG), which required phacovitrectomy for resolution and to review the cases of SpMG reported in modern literature. Only nine cases were identified. SpMG has no gender predilection and age at onset seems to be lower (mean age 47 years) than in secondary malignant glaucoma (SeMG). Nearly in half of the reported patients (4 out of 9) the condition had a bilateral presentation. The risk factors that have been identified for SeMG (nanophthalmos, shallow anterior chamber, iris plateau, zonular laxity) are underrepresented in SpMG.

Keywords: Malignant glaucoma, Ciliary block glaucoma, Aqueous misdirection, Acute glaucoma, Ocular hypertension

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Introduction

Malignant glaucoma was first described by von Graefe in 1869. It is a rare condition characterized by an acute intraocular pressure (IOP) rise with a very shallow anterior chamber in the presence of a patent peripheral iridotomy. Malignant glaucoma usually occurs after anterior segment surgery. Most cases of secondary malignant glaucoma (SeMG) are reported after filtration surgery.[1]

The mechanism leading to malignant glaucoma is poorly understood. The most accepted theory suggests that in an anatomically predisposed eye, the anterior rotation of the ciliary body induces misdirection of aqueous flow into or behind the vitreous body, increasing vitreous volume, resulting in anterior displacement of the iris–lens diaphragm, axial and peripheral anterior chamber flattening, and secondary angle closure.[2,3]

Only a few cases of spontaneous malignant glaucoma (SpMG) have been reported in the literature.[4–12] The aim of this article is to describe a case of SpMG, to review if the risk factors associated with SeMG are present in patients with SpMG and if the therapeutic approaches that are useful in SeMG are also valid in SpMG.

Case report

A 57 year-old male patient was seen in our hospital after a ten-day-history of intermittent pain on his left eye (LE). During the previous day, the patient had also noticed severe visual loss and the pain had increased and become constant.

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He had a past medical history of high blood pressure, hiatal hernia, chronic renal failure, hyperuricemia and renal litiasis and was on treatment with valsartan and allopurinol.

Visual acuity was hand movements in his LE. Anterior chamber examination revealed the presence of intense corneal edema and a very shallow anterior chamber (Fig. 1) and Goldman tonometry was 55 mmHg in his LE. A shallow anterior chamber was also present in his right eye (RE) without significant cataract. Fundus examination, although hindered by corneal edema, revealed no abnormalities. The diagnosis of acute angle closure glaucoma was made and the patient was treated with two cycles of intravenous 20% manitol, topical 0.5% timolol, and topical brimonidine. After four hours IOP remained higher than 50 mmHg. Oral acetazolamide and topical dexamethasone were added and a laser peripheral Nd-YAG iridotomy was attempted. As laser treatment was unsuccessful, a surgical iridectomy was performed two hours later.

On the following day, despite the presence of a patent iridectomy, IOP remained higher than 45 mmHg. Posterior segment ultrasonography showed no choroidal or vitreous abnormalities. However, anterior segment ultrasound biomicroscopy (UBM) found an anterior displacement of the iris-lens diaphragm (Fig. 2). The initial diagnosis was switched to malignant glaucoma. Ocular biometry found a *normal* axial length (RE 23.40 mm; LE 23.51 mm) with shallow anterior chambers (RE: 2.02; LE 1.69 mm). A central phacovitrectomy (with the implantation of a 21.5 diopter intraocular lens) normalized IOP. Postsurgical UBM showed a repositioning of the iris-lens diaphragm (Fig. 2). After two months corneal edema resolved in the LE and sixteen months later uneventful phacoemulsification was performed in the RE. In the last examination, two years after presentation, VA was 0.9, IOP was 20 mmHg without treatment and a severe visual field defect (DM −15.56 dB) was present in the patient's LE.

Discussion

Malignant glaucoma is among the most challenging ophthalmologic problems.(1) Some cases of SeMG have been reported after miotic therapy, non-glaucomatous anterior segment surgery or anterior segment laser procedures.[1–3,13]. However SeMG typically occurs after glaucoma filtration surgery in eyes with angle closure. The incidence after glaucoma surgery has been reported to be as high as 2%.[2] In contrast, SpMG is a very rare condition, and only a few cases have been reported (Table 1).[4–12] It is interesting to note that a significant proportion of these spontaneous cases had a bilateral presentation.

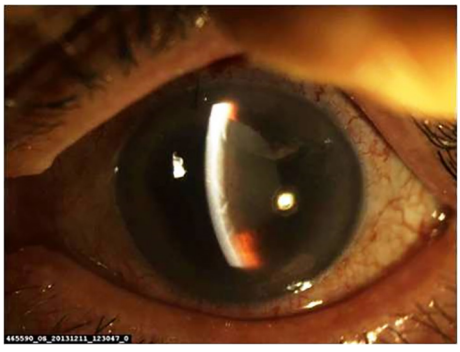

Fig. 1. Anterior segment of the left eye at presentation, with a very shallow anterior chamber and severe corneal edema.

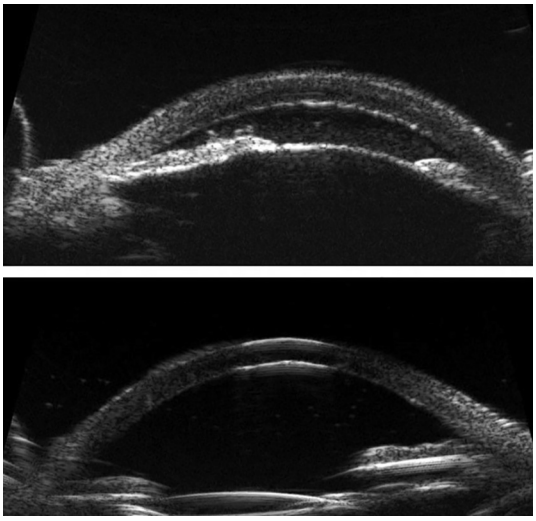

Fig. 2. Anterior segment ultrasound biomicroscopy (UBM) showed anterior displacement of the iris-lens diaphragm. After combined phaco-vitrectomy UBM showed repositioning of the iris-lens diaphragm.

SeMG seems to be more prevalent in women: the location of the lens is more anterior than in men, resulting in a shallower anterior chamber and a narrower space between the lens equator and the ciliary body. Indeed, in Zarnowski's series nine out of ten patients were women. However SpMG seems to have a similar incidence in both genders (Table 1). Mean age of onset of the reported SpMG cases (47 years) seems to be lower than in SeMG (63 years in Zarnowski's series).[14] SeMG is rarely bilateral; however nearly half of the reported cases of SpMG (4/9) had a bilateral presentation. Other reported risk factors are a previous history of primary angle closure, pseudoexfoliation syndrome (lax zonular fibers might facilitate the fluid passage), hyperopia, and a previous history of malignant glaucoma in the contralateral eye.[1,15] All of them seem to be less frequent in patients with SpMG.

Although some cases can be controlled with medical treatment, malignant glaucoma usually requires surgical procedures. The aim of treatment is to disrupt misdirection and to restore normal aqueous flow. The classical intervention, described by Chandler in the sixties, was the aspiration of vitreous with an 18 gauge needle through an incision in the pars plana.[1] Since then, capsulotomy, laser iridotomy and hyaloidotomy, vitrectomy and transscleral cyclophotocoagulation have been reported to be useful in the treatment of this condition.[2,3]. These techniques are usually applied in a step-wise approach.[1] Vitrectomy, if necessary, should be total with surgical disruption of the anterior hyaloid and zonule to break the primary mechanism of aqueous misdirection. It is equally useful in malignant glaucoma secondary to filtration and non filtration surgery[13] and seems to be the most effective intervention.

The nine cases identified of SpMG span over four decades. The table reflects the important changes that have taken place in the approach to malignant glaucoma. Our patient represents the third published case in which vitrectomy was used to treat SpMG.[4,12]

Table 1. Summary of the published cases of spontaneous malignant glaucoma.

Author, year	Clinical information	Gender	Age at onset	Pseudoexfoliation	Hyperopia	Treatment
Schwartz, 1975	Unilateral	M	85	No	No	Intracapsular cataract surgery and vitreous aspiration
Fanous, 1983	Unilateral	F	45	No	No	Medical: cycloplegia
Manku, 1985	Bilateral	M	37	No	No	Medical (pilocarpine, acetazolamide, manitol)
McClellan, 1988	Unilateral Down syndrome Keratoconus, acute hydrops	F	47	No	No	Intracapsular cataract surgery and vitreous aspiration
Gonzalez, 1992	Hyperuricemia Bilateral	M	55	No	No	Trabeculectomy and iridectomy Sclerotomy
Amini, 2005	Bilateral	M	37	No	No	LE: 1.Trabeculectomy and extracapsular cataract extraction 2.PPV and Hyaloidectomy RE: Atropine
Park, 2012	Aphakia, hypotony, traumatism Unilateral	M	38	No	No	Medical: cycloplegia, timolol, manitol
Premsenthil, 2012	Previous iridotomy	F	56	No	No	Phacoemulsification and PPV
Jarade, 2014	Bilateral	F	24	No	No	Trabeculectomy and PPV
Present case	Unilateral Hyperuricemia	M	57	No	Yes	Phacoemulsification and PPV

M male; F female; PPV pars plana vitrectomy.

In conclusion, SpMG has a younger age of onset than SeMG, it is more frequently bilateral and pars plana vitrectomy seems to be the best procedure to achieve resolution when medical treatment is unsuccessful.

Conflict of interest

None.

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