How Do You Diagnose and Treat Malignant Glaucoma?

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Malignant glaucoma, also known as aqueous misdirection syndrome, is a rare but serious medical emergency that usually occurs after intraocular surgery in patients with a history of narrow- or closed-angle glaucoma. The rise in IOP and shallowing of the anterior chamber, highly suggestive of this process, is thought to be due to a buildup of aqueous behind the ciliary body, posterior lens capsule, and anterior hyaloid membrane. The blockage of regular aqueous flow through the zonules and into the anterior chamber causes fluid to build up in the vitreous cavity, with the vitreous acting as a sponge for sequestered fluid. The lens/iris diaphragm is pushed forward, causing closure of the angle and an acute rise in IOP.

Several theories have been introduced regarding the pathogenesis of malignant glaucoma. Shaffer hypothesized that the complication was a consequence of aqueous misdirection into or around the vitreous body. Levene proposed that elevated IOP in malignant glaucoma resulted from direct closure of the angle upon the lens’ forward movement after intraocular surgery. Studies using perfused human cadaveric eyes revealed that the volume of the vitreous body increases under high pressure with the anterior hyaloid face pressing on the lens and ciliary body, possibly simulating the mechanism of malignant glaucoma. The investigators suggested that factors other than a diversion of aqueous humor might be important in creating malignant glaucoma. Alternatively, Quigley theorized that hydrated vitreous altered the movement of aqueous fluid and eventually led to malignant glaucoma. There is no universally accepted theory, and many of these mechanisms likely coexist, leading to the typical picture of elevated IOP that frequently resists medical intervention or peripheral iridotomy (PI).

DIAGNOSTIC TECHNIQUES

Clinical diagnosis requires a high index of suspicion. In affected eyes, the anterior chamber typically becomes progressively shallower (Figure 1), and the IOP suggested that factors other than a diversion of aqueous humor might be important in creating malignant glaucoma. Alternatively, Quigley theorized that hydrated vitreous altered the movement of aqueous fluid and eventually led to malignant glaucoma. There is no universally accepted theory, and many of these mechanisms likely coexist, leading to the typical picture of elevated IOP that frequently resists medical intervention or peripheral iridotomy (PI).

Figure 1. This patient developed malignant glaucoma after undergoing the implantation of a Baerveldt drainage device (Advanced Medical Optics, Inc., Santa Ana, CA). The slit-lamp examination revealed a shallow chamber and corneal edema.
suddenly rises (but can be normal after filtering surgery) without a clear posteriorly pushing force such as a choroidal effusion or suprachoroidal hemorrhage.

The practitioner should perform a B-scan to rule out masses in the posterior segment or choroidal hemorrhages or effusions. On ultrasound biomicroscopy, an anterior rotation of the ciliary processes is considered pathognomonic for malignant glaucoma (Figure 2).

TREATMENT

Clinicians should treat malignant glaucoma in a step-wise fashion. After diagnosis, the practitioner attempts a short course of cycloplegic therapy to deepen the anterior chamber and cause the posterior movement of the lens/iris diaphragm. Miotics should be avoided due to possible exacerbation of anterior chamber shallowing.

Angle-closure glaucoma may mimic malignant glaucoma. It is necessary to ensure that there is a patent PI to eliminate angle-closure glaucoma as a possible etiology for high IOP with a shallow anterior chamber. The ophthalmologist should, when possible, place a laser PI at the time of diagnosis if one is not already present.

Nd:YAG disruption of the anterior vitreous face is thought to allow aqueous to flow forward and thus permit the deepening of the anterior chamber and a decrease in IOP. The surgeon may perform laser disruption of the hyaloid membrane through the PI in phakic patients.

OUTCOMES

Elevated IOP and a shallow anterior chamber may persist despite medical and laser treatment. Pars plana vitrectomy may then be required and often resolves the process. Tsai and colleagues investigated the success rate of surgery in treating malignant glaucoma in 25 patients. Four of them experienced improvement after laser treatment alone. Core vitrectomy was successful in four of six pseudophakic patients and in one of four phakic patients. Posterior capsulotomy at the time of cataract extraction and vitrectomy improved success rates (83% vs 25% success rate). The investigators concluded “that surgical vitrectomy in the presence of an intact posterior capsule may preclude the surgical resolution of aqueous misdirection.”

Byrnes and colleagues reviewed the medical records of 21 patients post pars plana vitrectomy for refractory malignant glaucoma. The researchers noted that all patients had preexisting glaucoma and recent intraocular surgery. Pars plana vitrectomy was successful in 14 of 20 eyes (70%). Five of six eyes failing initial vitrectomy were phakic. Additional vitrectomy surgery to relieve ciliary block was required in three (60%) of five phakic patients who failed initial vitrectomy. Complications during the treatment of ciliary block included cataract formation, retinal detachment, bleb failure, and serous choroidal detachment.

Harbour and colleagues also reviewed the results of pars plana vitrectomy for malignant glaucoma in both phakic and aphantic patients. Of 24 individuals, 14 were phakic, and 10 were pseudophakic. In the phakic group, 100% (seven of seven) of the eyes that underwent cataract extraction at the time of pars plana vitrectomy achieved successful outcomes compared with 71% (five of seven eyes) of phakic eyes that did not undergo cataract extraction at the time of pars plana vitrectomy. Nine of 10 pseudophakic eyes were considered treat-
ment successes after initial pars plana vitrectomy. The investigators concluded that lensectomy at the time of pars plana vitrectomy improved success rates in phakic patients.

CONCLUSION

Proper diagnosis and prompt intervention are essential to the successful resolution of malignant glaucoma. By recognizing the risk factors of previous narrow- and closed-angle glaucoma in patients who have undergone intraocular surgery, clinicians will be more likely to identify the complication. In both the laser and surgical treatment of malignant glaucoma, disrupting the anterior hyaloid face and posterior lens capsule are the keys to success.

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