Update on Malignant Glaucoma

Treating and managing this complication of ocular surgery.

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Initially described in 1869 by von Graefe, malignant glaucoma is a postoperative glaucoma that, historically, was challenging to manage. Other names for this condition, based on its proposed mechanisms, include aqueous misdirection and ciliary block glaucoma. The clinical features of malignant glaucoma include shallowing of the anterior chamber and elevated IOP with the confirmed absence of either pupillary block glaucoma or suprachoroidal hemorrhage. Pupillary block is ruled out by the presence of a patent peripheral iridotomy (PI), whereas suprachoroidal hemorrhage is confirmed either by direct examination or a B-scan if an evaluation of the posterior segment is impeded.

Despite the many mechanisms proposed for malignant glaucoma, it is essentially thought to be caused by a buildup of aqueous posterior to the ciliary body and anterior hyaloid face that causes the forward displacement of the lens-iris diaphragm, leading to angle closure and a rise in IOP. Vitreous perfusion studies have shown that the resistance to flow increases as the IOP rises. The resistance is felt, in part, to be secondary to compression of the vitreous face against the ciliary body, lens, and iris, thus reducing the surface area by which fluid can diffuse through this barrier. This creates a vicious circle that historically been difficult to break.

EPIDEMIOLOGY

Malignant glaucoma is rare. It has been reported in 0.4% to 6% of surgical cases for the treatment of primary angle-closure glaucoma. Eyes that have had a previous, acute angle-closure attack or in which the angle closed at the time of surgery are at increased risk. Other risk factors include a previous attack in the fellow eye, abrupt cessation of cycloplegic therapy, and the initiation of miotic drops.

DIAGNOSIS

Although the combination of a shallow anterior chamber and elevated IOP in the absence of pupillary block or choroidal hemorrhage/effusion suggests diagnostic simplicity, malignant glaucoma is often subtle at its inception. Initial findings may be as understated as a mild myopic shift when the lens begins to move forward.

When differentiating pupillary block glaucoma from malignant glaucoma, a careful examination of the anterior chamber can offer some clues. In pupillary block, the central chamber is of moderate depth, and there is peripheral, anterior bowing of the iris. In malignant glaucoma, however, in addition to a patent PI, the central and peripheral chamber shallows as the entire lens-iris diaphragm rotates forward. Ultrasound biomicroscopy in particularly subtle cases can be diagnostic if anterior rotation of ciliary processes can be documented.
INITIAL TREATMENT

Treatment begins with medical management. The initiation of cycloplegic therapy facilitates the posterior rotation of the ciliary body and the lens-iris diaphragm. Oral and topical aqueous suppressants should also be employed. Medical management is reported to be curative in 50% of patients.3

If not already present, a PI should be performed to ensure that the problem is not simply pupillary block glaucoma. An Nd:YAG laser capsulotomy can be used to rupture the anterior hyaloid face in aphakic or pseudophakic eyes. In phakic eyes, the surgeon can target the hyaloid face through the PI.

SURGICAL MANAGEMENT

If medical therapy fails to resolve the process, a pars plana vitrectomy is the next step in the management of malignant glaucoma. The development of pars plana vitrectomy has greatly improved outcomes compared with the days when the term *malignant glaucoma* was coined. It is crucial that, during the vitrectomy, the anterior hyaloid face be removed or broken. Core vitrectomy without disruption of the anterior hyaloid—in order to spare the crystalline lens—resolves malignant glaucoma in only 25% to 50% of phakic eyes compared with 65% to 90% of pseudophakic eyes.5 Combining cataract surgery and vitrectomy with a posterior capsulotomy can increase the success rate from 25% to 83% in phakic eyes.6

Additionally, an iridozonulohyaloidectomy (IZH) can be performed at the time of vitrectomy.7 By first measuring the distance from the pars plana incision to the intended location of the IZH, marking this distance on the shaft of the vitrector, and then inserting the vitrector through the pars plana incision to the mark, the surgeon can ensure that the IZH will be properly located. The goal of IZH with or without vitrectomy is to form a unicameral eye by removing any blockage of flow from the posterior segment to the anterior chamber.

CONCLUSION

Malignant glaucoma can be difficult to manage, but medical therapy resolves up to 50% of cases. If pharmaceutical agents are ineffective, surgical therapy with a laser or vitrectomy with disruption of the anterior hyaloid face can successfully treat the majority of cases.

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