Phacomorphic Glaucoma

As illustrated by a challenging case, the proper diagnosis and treatment of this form of glaucoma are difficult but essential to improving patients' visual prognosis.

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CASE PRESENTATION

A 72-year-old white woman was referred to us 9 days after she experienced an episode of acute angle-closure glaucoma in her right eye. She had presented to the referring ophthalmologist with a complaint of a right-sided periorbital headache, nausea, and blurry vision upon awakening. Her right eye's IOP was in the 50s and could not be medically controlled. The referring ophthalmologist could not perform a laser iridotomy in the patient's right eye due to corneal edema, but he successfully completed a prophylactic laser iridotomy in her left eye. The next day, he performed a surgical iridectomy in her right eye.

Despite a patent iridectomy, the IOP in her right eye remained in the 40s on timolol 0.5% and acetazolamide 500 mg b.i.d. The ophthalmologist referred her to us at that point for further management. The patient had no other ocular history but had several medical problems, including diabetes mellitus, hypothyroidism, congestive heart failure, sleep apnea, chronic obstructive pulmonary disease, and osteoarthritis.

Upon initial evaluation, the patient's UCVA was 20/250 (pinhole correction, 20/100) OD and 20/60 (pinhole correction, 20/30) OS. She had never worn distance glasses. Her IOP measured 34 mm Hg OD and 9 mm Hg OS with applanation tonometry. Her right pupil was partially dilated and minimally reactive without an afferent pupillary defect. The slit-lamp examination of her right eye revealed a hazy cornea with mild microcystic edema, a shallow anterior chamber, a 1.7-mm layered hyphema, a patent superior iridectomy, and a dense nuclear sclerotic cataract (Figure 1). Nylon sutures from the surgical iridectomy were visible superiorly at the limbus. The slit-lamp examination of her left eye revealed a clear cornea, a patent superior iridotomy, a shallow anterior chamber, and a dense nuclear sclerotic cataract.

Performing gonioscopy on her right eye was difficult due to the corneal edema. Even with compression, the angle appeared to be closed without any identifiable angular structures. Gonioscopy of her left eye revealed an appositionally closed angle, which opened to the anterior trabecular meshwork upon compression. The fundus examination showed a cup-to-disc ratio of 0.3 OD and 0.2 OS. The rest of the fundus examination was unremarkable, without evidence of a mass or choroidal hemorrhage.

A-scan ultrasound biometry showed an axial length of 22.9 mm, an anterior chamber depth of 1.6 mm, and a lens thickness of 5.5 mm OD. The patient's left eye had an axial eye length of 22.5 mm, an anterior chamber depth of 1.8 mm, and a lens thickness of 5.3 mm. A B-scan ultrasound of the anterior segment using an immersion chamber showed a thick lens pushing the iris forward and an

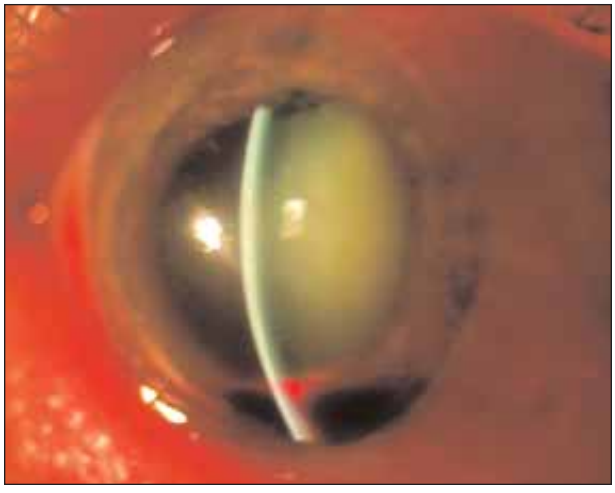

Figure 1. A slit-lamp photograph shows an edematous cornea and a shallow anterior chamber, with a layered hyphema after surgical iridectomy superiorly. Note the dense nuclear sclerotic cataract.

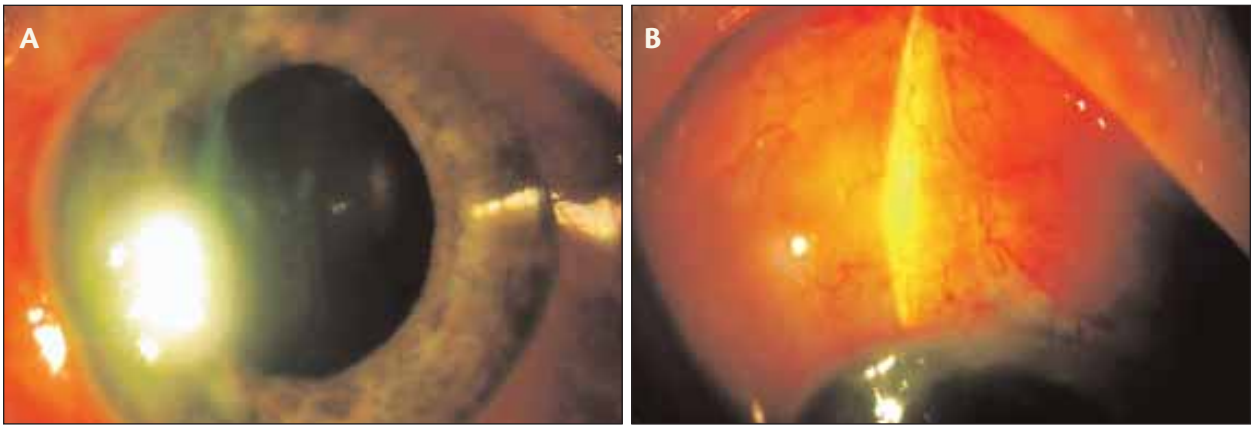

Figure 2. These slit-lamp photographs were taken 1 day after the patient underwent combined cataract surgery with implantation of a posterior chamber IOL and trabeculectomy with mitomycin C in her right eye. A clearer cornea, a deep anterior chamber, and a well-positioned posterior chamber IOL were visible (A). The layered hyphema resolved. A filtering superior bleb was apparent (B).

appositionally closed angle consistent with the diagnosis of phacomorphic glaucoma.

HOW WOULD YOU PROCEED?

1. Would you manage this patient medically or consider a surgical intervention?
2. If you were to perform cataract surgery, would you perform glaucoma surgery at the same time or later?
3. If you elected to perform a combined cataract surgery and trabeculectomy, how might you improve the surgical view for cataract extraction, particularly concerning her corneal edema?

SURGICAL COURSE

After a discussion with the patient, we performed a combined procedure of cataract surgery and trabeculectomy with mitomycin C in her right eye. We administered intravenous mannitol 60 g intraoperatively to lower the IOP and help clear the corneal edema. First, we performed the trabeculectomy with mitomycin C (0.15 mg/mL for 4 minutes) in the superior quadrant. By the end of this procedure, the cornea was sufficiently clear for us to visualize the lens for the subsequent cataract surgery. We made a separate temporal corneal incision for phacoemulsification and inserted a foldable, acrylic, posterior chamber IOL in the capsular bag. We completed the combined surgery without complication.

OUTCOME

On the first postoperative day, the patient's UCVA was 20/125 OD, and her IOP was 17 mm Hg with a deep anterior chamber and an elevated, superior bleb (Figure 2). She received topical prednisolone acetate 1% and atropine sulfate 1%. We performed laser suture lysis 2 weeks postoperatively to increase filtration (Figure 3). Two months after the surgery, her right eye had a BCVA of 20/20 OD and an untreated IOP of 14 mm Hg. She had a well-functioning, superior filtering bleb. Repeat gonioscopy revealed multiple areas with peripheral anterior synechiae (collectively covering approximately 90°), with the remainder of the angle open to the anterior trabecular meshwork.

The presence of a patent iridotomy in the patient's left eye prevented the development of pupillary block angle closure. However, she was still at risk for developing phacomorphic glaucoma because of a thick cataractous lens and a narrow angle configuration. For this reason, we discussed cataract surgery relatively early with her to prevent the future development of phacomorphic glaucoma in her left eye. She eventually decided to delay the surgery and returned to the referring ophthalmologist.

DISCUSSION

Primary angle-closure glaucoma occurs from pupillary block or plateau iris syndrome. Secondary angle-closure glaucoma is due to an anterior pulling mechanism (eg, iridocorneal endothelial syndrome, neovascular glaucoma) or a posterior pushing mechanism (eg, phacomorphic glaucoma, posterior segment tumor). Phacomorphic glaucoma can have a component of primary pupillary block and a secondary angle-closure mechanism. Patients with this condition may present with decreased vision, eye pain, and headache—much as in primary angle-closure glaucoma.

On examination, patients with phacomorphic glaucoma usually have elevated IOP, an edematous cornea, a shallow anterior chamber, and an advanced cataract. By definition, these individuals have closed anterior chamber

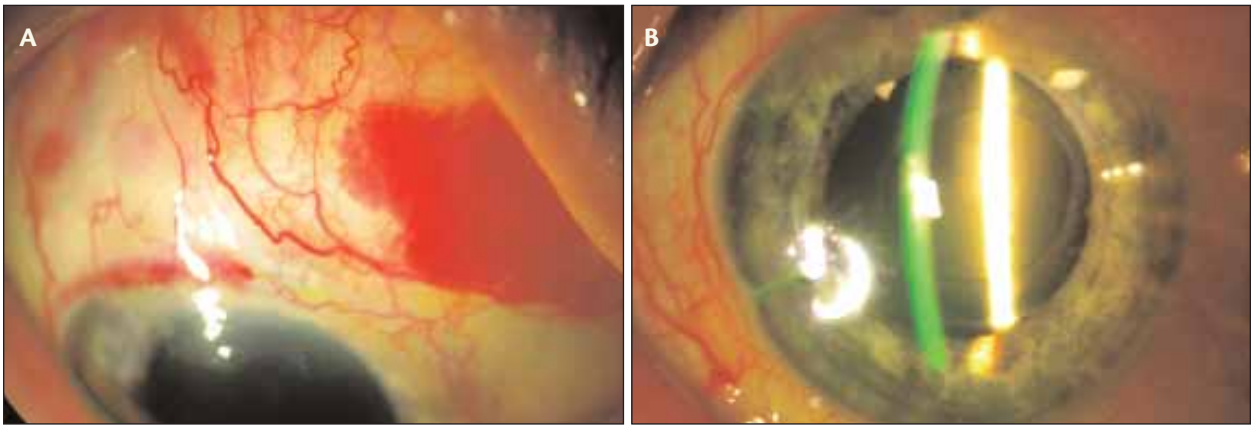

Figure 3. The patient had a diffusely elevated superotemporal bleb 2 weeks postoperatively (A). The two nylon sutures, visible subconjunctivally at the 12-o'clock position, are from the previous iridectomy surgery. A residual subconjunctival hemorrhage was visible superonasally. During the visit, one of the authors performed a single laser suture lysis, which reduced the IOP to 7 mm Hg. The patient had a clear, compact cornea and a deep anterior chamber with a well-positioned posterior chamber IOL in the capsular bag 4 weeks postoperatively. A 10–0 nylon corneal suture for the cataract incision is visible temporally (B).

angles. Unfortunately, visualization of the anterior chamber angle and fundus may be challenging due to the corneal edema. If the clinician cannot see the fundus well, he or she can use ultrasonography or other imaging techniques to rule out intraocular tumors.[1] To assist the diagnosis of phacomorphic glaucoma, the physician may use A-scan biometry and ultrasound biomicroscopy to determine the depth of the anterior chamber and the thickness of the crystalline lens.

The treatment of phacomorphic glaucoma may require the use of medical therapy or laser and incisional surgery. The definitive treatment for phacomorphic glaucoma is cataract extraction, but it is easier and safer to perform cataract surgery after first lowering the IOP with medication. Glaucoma medications that reduce aqueous production are usually employed first. Cholinergic medications such as pilocarpine can be associated with a paradoxical increase in IOP, because they cause the iridolenticular diaphragm to shift forward and further close the angle, and they should be avoided. Laser iridotomy is recommended in the initial management of phacomorphic glaucoma to relieve any potential components of pupillary block.[2]

A recent study suggested that laser peripheral iridoplasty may also be effective in initially lowering the IOP.[3] In many cases, however, a filtration procedure and cataract extraction are necessary for long-term IOP control. Performing trabeculectomy without cataract surgery poses an increased risk of a flat anterior chamber and aqueous misdirection. Cataract surgery in patients with phacomorphic glaucoma can be challenging. A high preoperative IOP is often associated with a cloudy, edematous cornea and thus a poor surgical view. Moreover, a shallow anterior chamber and increased anterior capsular convexity can present a technical challenge during cataract surgery.[4] A high preoperative IOP can also increase the chance for a choroidal hemorrhage during or after surgery.

The proper diagnosis and treatment of phacomorphic glaucoma, although difficult, can significantly improve the patient's visual prognosis. ■

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