Phacolytic Glaucoma and Homocystinuria

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CASE PRESENTATION
A 58-year-old man was referred for an evaluation of presumed aphakic glaucoma in both eyes. During a routine examination at the referring doctor’s office, the patient’s IOP had measured 38 mm Hg OD and 40 mm Hg OS. He had no ocular complaints but stated that he had needed a very strong glasses prescription since childhood. Upon further discussion, he denied any additional notable ocular history, specifically prior ocular surgery or trauma. In general, the patient was a poor historian with a very mild cognitive delay. He reported that he had suffered a heart attack approximately 10 years earlier and that he was on several medications, but he was uncertain of which ones.

Our initial examination revealed a visual acuity of 20/80 OD and 20/100 OS with a current correction of +10.00 D OU. The IOP was primarily unchanged in the mid-30s. An examination of the anterior segment found trace conjunctival injection and 1+ anterior chamber cell in both eyes. The patient appeared to be aphakic in both eyes on slit-lamp examination. Gonioscopy was open to the ciliary body band for 360º in both eyes. The patient appeared to be aphakic in both eyes on slit-lamp examination. Gonioscopy was open to the ciliary body band for 360º in both eyes. A dilated fundus examination showed trace cells in the vitreous. The optic nerve exhibited slightly enlarged cupping to approximately 0.5 OU. The macula and peripheral retinal examination was within normal limits, with the exception of the far inferior periphery. At this location in both eyes, a dislocated crystalline lens was seen with dense and opaque nuclear sclerotic changes. We diagnosed the patient with phacolytic glaucoma.

HOW WOULD YOU PROCEED?
- Would you initiate a topical ocular hypotensive and steroid medication?
- Would you proceed with surgical intervention? If so, would you refer the patient for a vitrectomy and lensectomy, perform filtering surgery, or use a combination of both?
- Would you pursue a further workup?

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SURGICAL COURSE
Because the patient had no known history of trauma or evidence of pseudoexfoliative material, a syndromic condition predisposing him to lens dislocation was suspected. Moreover, the patient’s history of a heart attack at a young age pointed toward the possibility of homocystinuria. Elevated homocystine was confirmed with quantitative blood testing.

We started the patient on topical glaucoma and steroid medications. With the assistance of a retinal specialist, the patient subsequently underwent sequential vitrectomy and lensectomy in combination with the implantation of an anterior chamber IOL.

OUTCOME
Postoperatively, his BCVA improved to 20/40 OU. The IOP returned to normative values around the high teens in both eyes without a need for topical medication. The patient continues to be observed by his primary care provider for risk factor management and is doing well.

DISCUSSION
Phacolytic glaucoma (or lens protein glaucoma) was first described in 1900. The typical symptoms include ocular pain, red eyes, and significantly decreased vision. An examination often reveals a grossly normal anterior chamber angle with accompanying heavy flare. In some cases, hyperrefringent particles may be visible and offer a diagnostic clue. These particles are thought to represent calcium oxalate or cholesterol crystals. A less common
presentation of phacolytic glaucoma, as was seen with our patient, occurs when the lens dislocates into the vitreous. In these cases, the glaucoma and inflammation tend to be more subacute. Several theories have been posed to describe how the leaking of soluble lens protein through microscopic defects in the lens capsule may lead to elevations in IOP. These theories include a blocking of the trabecular meshwork by macrophages filled with phagocytosed lens material and the possibility that the protein itself is directly obstructing outflow.

Phacolytic glaucoma is usually considered a surgical emergency. Medication is initiated prior to urgent lens removal to reduce IOP and inflammation. Thorough irrigation of all lens material should be performed. Patients with phacolytic glaucoma usually experience a return of good vision and an improvement in IOP after surgery.

A few studies have analyzed outcomes for phacolytic glaucoma after surgical management and whether or not a trabeculectomy may be necessary in these cases. In a retrospective case series involving 135 eyes, 89 eyes underwent trabeculectomy combined with cataract surgery if the patient was symptomatic for more than 7 days or if IOP was uncontrolled on maximum tolerated medical therapy. The remaining eyes underwent cataract surgery alone. The initial IOP control was significantly better in the combined cases during the initial postoperative period. Six months postoperatively, however, the IOP and visual acuity were similar in both groups. Another study of 45 eyes with phacolytic glaucoma demonstrated that all patients who underwent cataract surgery alone had an IOP less than 21 mm Hg without any antiglaucoma medication postoperatively. Based on the literature to date, it would be reasonable to proceed with cataract extraction alone for the treatment of phacolytic glaucoma, particularly in patients with recently developed symptoms or with subacute findings or pressure elevation.

An interesting aspect of this challenging case was the evident lens dislocation in both eyes related to homocystinuria. This disorder is inherited in an autosomal recessive fashion and results from one of several enzyme deficiencies in homocysteine metabolism.

Patients with homocystinuria frequently experience a progressive downward dislocation of the lens (and in some cases, complete dislocation into the vitreous) as well as corresponding early myopia. Other ocular manifestations include optic atrophy, iris atrophy, anterior staphylomas, and lenticular and corneal opacities.

Glaucoma is a common complication of lens dislocation, but it is typically caused by the lens’ displacement into the anterior chamber. In addition to ectopia lentis, other physical manifestations of homocystinuria include Marfanoid habitus, pes excavatum, mental retardation, seizures, and thromboembolic events.

There is a wide range of phenotypic variance and broad variation in the severity of signs and symptoms with homocystinuria, depending on the enzymes involved and whether or not the condition responds to vitamin B₆ (pyridoxine). Recently, newborn screening has led to earlier diagnosis. Treatment with vitamin supplementation and/or restriction of methionine in the diet can slow disease progression and reduce the risk of ocular complications. In many cases, any surgical intervention requires general anesthesia due to the patient’s age and mental status. These patients are also known to have an increased risk of thromboembolic events and hypoglycemia associated with anesthesia. A similar case to the one I described required prophylactic measures, including the use of aspirin, compression stockings, and dextrose infusion for the surgeon to safely perform a vitrectomy under general anesthesia. Fortunately, our patient’s level of cooperation allowed for a successful surgery under a retrobulbar blockade.

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