Angle-closure Glaucoma Associated With Occult Annular Ciliary Body Detachment

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Objective: To evaluate the role of annular ciliary body detachment in the development of postoperative angle-closure glaucoma.

Design: Case series.

Setting: Tertiary care glaucoma referral center.

Methods: High-resolution, anterior segment ultrasound biomicroscopy, ophthalmoscopy, and B-scan ultrasonography were performed on 6 eyes of 6 patients with a clinical diagnosis of postoperative malignant glaucoma.

Results: Each eye had an elevated intraocular pressure, a shallow anterior chamber, 1 or more patent iridotomies, and no ophthalmoscopic or B-scan ultrasound evidence of serous or hemorrhagic ciliochoroidal detachment. Ultrasound biomicroscopy revealed annular ciliary body detachment in each eye. In 4 eyes, observation with topical medical treatment was associated with deepening of the anterior chamber, reduced intraocular pressure, and resolution of the detachment. Drainage of the supraciliary fluid was performed in 2 eyes.

Conclusion: Occult, annular, serous detachment of the ciliary body may cause postoperative angle-closure glaucoma. This entity is clinically indistinguishable from malignant glaucoma. Ultrasound biomicroscopy facilitates the diagnosis.

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Malignant glaucoma (ciliary block glaucoma or aqueous misdirection) is an incompletely understood and serious complication of intraocular surgery. First described by von Graefe in 1869, this secondary angle-closure glaucoma is characterized by elevated intraocular pressure (IOP), shallow anterior chamber, patent iridectomy, and normal posterior segment anatomy by ophthalmoscopy or B-scan ultrasonography. Although a relatively uncommon postoperative complication, the need for reoperation is frequent and the long-term visual outcome is often poor. A spectrum of disease severity exists.

Little is known regarding the anatomy and pathophysiology of this disorder. Shaffer was the first to propose that aqueous flow is diverted into the posterior segment in malignant glaucoma. He attributed this to a blockage of normal anterior aqueous flow by an abnormal vitreociliary relationship, and later coined the descriptive term ciliary block glaucoma. Epstein et al. later proposed forward displacement of the vitreous into apposition with the iris and ciliary body as the cause of the posterior diversion of aqueous. They theorized that this might be associated with thickening of the anterior hyaloid and were able to demonstrate an impedance to flow across the intact anterior hyaloid in enucleated human and animal eyes with artificially elevated IOP. The accumulation of aqueous within the posterior segment forces the ciliary body and anterior hyaloid anteriorly, shallowing the anterior chamber and causing a secondary angle-closure glaucoma.

Although most cases of malignant glaucoma develop after filtration surgery in eyes with chronic angle-closure glaucoma, the condition can follow a wide variety of ophthalmic procedures. These include intracapsular cataract extraction, extracapsular cataract surgery with posterior chamber intraocular lens implantation, Nd:YAG transscleral cyclophotocoagulation, laser iridotomy, and laser release of scleral flap sutures. Malignant glaucoma has also been reported in association with central retinal vein occlusion, retinopathy of prematurity, and in eyes that had previously undergone filtration surgery months or years following initiation of pilocarpine therapy.

Medical therapy for malignant glaucoma involves the use of topical steroids to decrease inflammation, intensive cyclopentolate to reverse the anterior rotation of the ciliary body, aqueous suppressants, and hyperosmotic agents to decompress the vitreous. If the presentation is acute and medical treatment to lower IOP and open the angle proves inadequate, argon laser peripheral iridoplasty may be used to break the attack.

Surgical correction of malignant glaucoma involves rupture of the anterior hyaloid face to eliminate the vitreociliary block.
PATIENTS AND METHODS

All patients were evaluated within 2 weeks of trabeculectomy. The clinical diagnosis of malignant glaucoma required the presence of a shallow anterior chamber, 1 or more patent iridectomies, and increased IOP. Sltlamp biomicroscopy, ophthalmoscopy, B-scan ultrasonography, and ultrasound biomicroscopy were performed on each eye.

The equipment and technique of high-frequency, high-resolution ultrasound biomicroscopy (Humphrey Instruments Inc, San Leandro, Calif) have been described in detail elsewhere. Our commercially available units use a 50-MHz transducer, achieve a resolution of approximately 50 µm, and have a tissue penetration of 4 to 5 mm. All scanning was performed with the patient in the supine position under standardized room lighting conditions. Accommodation and fixation were held constant by having the patient fixate with the fellow eye on a ceiling target. After topical anesthesia, a 20-mm eyecup was inserted and filled with saline solution. Images were obtained at the 12-, 3-, 6-, and 9-o’clock positions. Cross-sectional images were taken through the cornea, iris, ciliary body, anterior chamber angle, and peripheral sclera to demonstrate structural relationships.

The treatment of choice in aphakic or pseudophakic eyes is rupture of the anterior hyaloid with the Nd:YAG laser to eliminate it as a fluid barrier. This allows equalization of pressure between the posterior and anterior segments. Brown et al reported successful use of this technique in a phakic eye. Rupture of the anterior hyaloid face using the technique described by Chandler et al are also options.

The differential diagnosis of postoperative anterior chamber shallowing associated with elevated IOP includes aqueous misdirection, pupillary block, choroidal hemorrhage, and choroidal effusion with anterior rotation of the ciliary body and secondary angle closure. We evaluated a group of patients with annular supraciliary effusion detectable only by ultrasound biomicroscopy and secondary angle-closure glaucoma whose presentations were clinically indistinguishable from that of malignant glaucoma.

REPORT OF CASES

CASE 1

A 68-year-old, phakic, Hispanic woman with chronic angle-closure glaucoma and patent laser iridotomies in both eyes underwent an uncomplicated trabeculectomy with peripheral iridectomy of the right eye. On the first postoperative day, the anterior chamber was moderately deep and the IOP was 9 mm Hg. Postoperative topical medications, consisting of 0.25% scopolamine twice daily, tobramycin 4 times daily, and 1% prednisolone acetate every 2 hours, were begun in the right eye. Four days later, the anterior chamber had shallowed and iridocorneal apposition was present for 360°, almost to the collarette. The incisional and laser iridectomies were patent. The bleb was low and without leak, and the IOP was 18 mm Hg. Indirect ophthalmoscopy was not possible owing to hazy media. Contact B-scan ultrasonography was performed and the posterior segment was unremarkable. A diagnosis of malignant glaucoma was made and the frequency of scopolamine dosing was increased to 4 times daily. A trial of 2% cyclopentolate did not alter the anterior chamber depth. Ultrasound biomicroscopy revealed supraciliary fluid (Figure 1, Figure 2, and Figure 3). Pupillary block was not present. Two days later, the anterior chamber remained formed only centrally. The IOP was 24 mm Hg while the patient was receiving a regimen of 0.25% scopolamine 4 times daily, 1% prednisolone acetate every hour, 0.5% timolol twice daily, and sustained-release acetazolamide. There was no change in the appearance of the ciliary detachment. Oral prednisone therapy, 80 mg once daily, was begun and the patient was admitted to the hospital. Two days later, the examination and IOP were unchanged. Because of an extremely shallow anterior chamber, the patient underwent drainage of serous fluid from the supraciliary space approximately 3.5 mm posterior to the limbus in the inferonasal and inferotemporal quadrants and the anterior chamber was reformed using viscoelastic. Postoperatively, the anterior chamber remained deep after the medications were tapered. The IOP, which was low following reoperation, gradually rose to approximately 12 mm Hg, where it has remained.

CASE 2

A 77-year-old, phakic, black man with chronic angle-closure glaucoma and patent laser iridotomies underwent uncomplicated trabeculectomy with peripheral iridectomy of the left eye. Postoperatively, there was progressive IOP elevation and anterior chamber shallowing. Four days postoperatively, the IOP was 20 mm Hg and extensive iridocorneal apposition was present. Ophthalmoscopy and B-scan ultrasonography results were normal. Ultrasound biomicroscopy revealed an annular ciliary body detachment. Medical therapy with cycloplegics and aqueous suppressants resulted in gradual deepening of the anterior chamber to normal depth and a reduction in IOP to 10 mm Hg.

CASE 3

A 66-year-old, phakic, Hispanic woman with primary open-angle glaucoma underwent a trabeculectomy with peripheral iridectomy of the left eye. On the first postoperative day, the anterior chamber was reported to be moderately shallow and the IOP 12 mm Hg. Nine days later, her status was unchanged and the patient was referred for evaluation. Examination revealed a shallow anterior chamber with extensive iridocorneal contact sparing the collarette, patent iridectomy, flat bleb without leak, and an IOP of 19 mm Hg while the patient was receiving a topical regimen of 1% atropine 3 times daily, 1% prednisolone acetate 4 times daily, and tobramycin. Indirect ophthalmoscopy and B-scan ultrasonography revealed normal posterior segment anatomy. Ultrasound biomicroscopy revealed a supraciliary effusion for 360°. The atropine therapy was continued and the frequency of the prednisolone acetate was increased to every hour. During the next week, the anterior chamber gradu-
ally deepened, the IOP fell, and a bleb formed. The topical medications were discontinued without sequelae.

CASE 4

A 70-year-old, phakic, white woman with chronic angle-closure glaucoma underwent trabeculectomy with peripheral iridectomy in the right eye. The early postoperative course was notable for ocular hypotony, shallow anterior chamber, and ciliochoroidal detachment requiring drainage and anterior chamber re-formation on day 12. Five days later, however, the anterior chamber shallowed, the IOP rose, and the patient was referred for evaluation. Ocular medications were 0.25% scopolamine twice daily and 1% prednisolone acetate every 2 hours. Slit-lamp biomicroscopy revealed a shallow anterior chamber, raised bleb without leak, and IOP of 18 mm Hg. Indirect ophthalmoscopy and B-scan ultrasonography indicated the apparent resolution of the choroidal effusions. However, ultrasound biomicroscopy revealed an annular ciliary detachment, anterior ciliary body rotation, and angle closure. Two percent cyclopentolate twice daily was added to her ocular medication regimen. The anterior chamber progressively deepened during the following days and the remainder of her postoperative course was uneventful.

CASE 5

A 74-year-old, phakic, white woman with primary open-angle glaucoma underwent trabeculectomy with peripheral iridectomy in the right eye. Postoperatively, the anterior chamber was shallow, the bleb was moderately elevated, and the IOP ranged from 17 to 22 mm Hg. The only ocular medication was 1% prednisolone acetate 4 times daily. The patient was referred for evaluation. Slit-lamp biomicroscopy revealed the anterior chamber to be formed only centrally, with a low bleb without leakage. Ophthalmoscopy and B-scan ultrasonography revealed a normal posterior segment. Treatment with aqueous suppressants in addition to the previously described medications did not change the clinical findings. Ultrasound biomicroscopy revealed an annular supraciliary effusion and angle closure. Surgical drainage of the supraciliary effusion combined with reformation of the anterior chamber with viscoelastic permitted restoration of normal ocular anatomy and bleb function; these remained normal after cessation of the topical medications.

COMMENT

Accurate assessment of the posterior segment is critical to establish the presumptive diagnosis of malignant glaucoma. Although indirect ophthalmoscopy in eyes with
clear media usually suffices to determine the health of the posterior segment, eyes with hazy media require B-scan ultrasonography to make certain that choroidal separation is absent. If pupillary block is absent and the posterior segment is normal by ophthalmoscopy and B-scan ultrasonography, aqueous misdirection has been a presumptive diagnosis. The presence of fluid accumulation limited to the supraciliary space detectable only by ultrasound biomicroscopy in our study indicates that there is a subgroup of patients with a clinical diagnosis of malignant glaucoma who most likely do not have aqueous misdirection.

We have previously reported the ultrasound biomicroscopic features of malignant glaucoma in a patient with posterior chamber pseudophakia. In that case, anterior rotation of the ciliary body, without evidence of ciliochoroidal effusion, physically pushed the iris against the cornea and trabecular meshwork and was responsible for shallowing of the anterior chamber. The ultrasound biomicroscopic images were consistent with a posterior diversion of aqueous into the vitreous, but could not identify an abnormality of the vitreous relationship. Hyaloidectomy using the Nd:YAG laser permitted aqueous to move into the anterior chamber, allowing the iris and intraocular lens to move posteriorly, deepen the anterior chamber, and expose trabecular meshwork to aqueous.

In contrast, ultrasound biomicroscopic analysis of iris configuration and ciliary body position in phakic, aphakic, and pseudophakic pupillary block shows the angle closure to be on the basis of a convex iris configuration, without evidence of significant anterior lens movement or ciliary body rotation. Under these circumstances, iridociliary contact does not contribute to angle narrowing. Laser iridectomy relieves the angle closure by eliminating relative pupillary block. In cases of suspected malignant glaucoma, pupillary block should be eliminated as a possible contributory element to the shallow anterior chamber by assessing the size and patency of the iridectomy, when present, or by the creation of a patent iridectomy, if necessary. Axial shallowing of the anterior chamber occurs in pseudophakic malignant glaucoma, but should not occur in pupillary block. Two or 3 laser iridectomies may be necessary to eliminate the possibility of loculated regions of pupillary block contributing to the anterior chamber shallowing.

Ultrasound biomicroscopic evidence of supraciliary effusion in 2 eyes with clinical malignant glaucoma was reported by Trope et al. Given the amount of supraciliary fluid visible in their ultrasound image, one would have expected that fluid would have been obtained on entering the ciliochoroidal space in the eye requiring surgical intervention. The authors attributed the lack of identifiable fluid to loculations in the supraciliary space or the relatively small amount of fluid present. We were able to identify supraciliary fluid at the time of surgical drainage in both our patients who underwent surgery. Our ability to access the supraciliary fluid may have been due to the anterior location of the sclerotomy drainage site and the slow manner in which the supraciliary space was entered.

Occult, annular peripheral choroidal detachment as a cause of postoperative anterior chamber shallowing after glaucoma surgery in eyes without hypotony has been reported by Dugel et al. In their series of 18 eyes with IOPs ranging from 10 to 36 mm Hg, they were able to identify low-lying peripheral choroidal effusions in conjunction with the postoperative shallowing of the anterior chamber. However, they used standard B-scan ultrasonography and could not image the supraciliary space. The IOPs in our patients were less than 25 mm Hg, suggesting that annular ciliary body detachment should be suspected in eyes with clinical malignant glaucoma and IOPs that are not markedly elevated.

Pupillary block and anterior ciliary body rotation due to ciliary body detachment are not mutually exclusive. We examined a patient in whom, following panretinal cryopexy for progressive iris neovascularization in a phakic eye without previous ocular surgery other than panretinal photocoagulation, the central and peripheral anterior chamber shallowed and the angle closed. Ciliochoroidal detachment related to the cryopexy was suspected as the cause of the change in the anterior chamber anatomy. Ultrasound biomicroscopy confirmed the suspected detachment, but also revealed a convex iris configuration consistent with pupillary block. Laser iridectomy relieved the pupillary block component to the angle closure, allowing the angle to open and the pressure to normalize. The chamber remained shallow and spontaneously deepened several days later with spontaneous resolution of the ciliochoroidal separation.

Our findings are consistent with at least 2 distinct mechanisms that may be responsible for what is usually called postoperative malignant glaucoma. In the first group, ciliary detachment is absent or, if present, is not sufficient by itself to cause anterior rotation of the ciliary body and secondary angle closure. Aqueous misdirection plays a major role in the forward displacement of the ciliary body in these eyes. In the second group, which is clinically indistinguishable from the first group by slitlamp biomicroscopy, ophthalmoscopy, and contact B-scan ultrasonography, the main factor in the anterior rotation of the ciliary body is a detachment of the ciliary body by supraciliary fluid. Ultrasound biomicroscopy is the only noninvasive technique that can reliably detect the presence of the ciliary body detachment. The only other method to predictably determine which subtype of malignant glaucoma is present is to tap the supraciliary space. The presence of fluid and ease of subsequent anterior chamber re-formation confirm the diagnosis in the latter group. We have observed that annular ciliary body detachment is identifiable in about one half of eyes with clinical evidence of malignant glaucoma at our institutions.

The annular ciliary body detachment present in our cases differs in an important way from the ciliochoroidal effusion that often occurs after filtration surgery. In cases without posterior segment inflammation, the accumulation of fluid in the potential space between the uvea and sclera depends on relative hypotony and transudation of fluid along the hydrostatic gradient. If the annular detachment present in some of our cases of malignant glaucoma were simply a result of hydrostatic forces, the detachment should have remitted as the pressure rose. A second mechanism must be present, either as a result of ciliary body inflammation or abnormal re-
sponse to surgery, to account for the persistence of the supraciliary fluid in these eyes. Although a toxic effect of mitomycin C on the ciliary body has been suggested, there is as of yet no reason to suspect this agent to be related to this phenomenon. Our cases differ significantly from those reported by Fourman, in which angle-closure glaucoma developed in eyes with inflammatory uveal effusion syndrome of differing causes.

The presence of ciliary body detachment does not rule out the possibility that some degree of aqueous misdirection is present in these eyes as well. The anterior position of the ciliary body alters the vitreociliary relationship. The curative effect of fluid drainage in 2 of our cases, however, suggests that aqueous misdirection plays a less prominent role in these eyes. On the other hand, the effect of fluid drainage may sufficiently alter the vitreociliary relationship to eliminate the blockage to anterior aqueous movement, if this indeed is present.

The presence of at least 2 clinical subtypes of clinical malignant glaucoma indicates 2 specific, although similar, approaches to therapy. As with most serious ciliochoroidal separations, the majority of focal ciliary body separations resolve with conservative therapy consisting of intensive cycloplegia, topical steroids, and, occasionally, systemic steroids. Surgical drainage is usually not required. However, because most reports suggest that approximately 50% of all cases of malignant glaucoma remit with intensive cycloplegia, aqueous suppression, and topical steroids, malignant glaucoma caused primarily by aqueous misdirection may require surgical intervention much more commonly than previously thought. We hypothesize that cases of malignant glaucoma that respond to medical therapy are more likely to have had ciliary body detachment rather than vitreociliary block as the primary cause of the malignant glaucoma.

Because the supraciliary and suprachoroidal spaces are contiguous, it may have been possible to detect fluid that accumulated predominantly in the supraciliary space with immersion B-scan ultrasonography. This was not performed in the patients reported here. However, the image quality achieved with the high-frequency transducers used during ultrasound biomicroscopy is superior to traditional B-scan ultrasonography. In addition, the technique is simpler, faster, and does not require an extremely experienced ultrasonographer.

In summary, malignant glaucoma caused by annular ciliary body detachment has a clinical appearance indistinguishable from aqueous misdirection. The only reliable way to differentiate this subgroup of patients from other disorders causing an identical clinical picture is with ultrasound biomicroscopy. Therapy can then be directed toward the anatomic cause of the disease process and the effect of intervention assessed.

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