Juvenile Uveitic Glaucoma

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CASE PRESENTATION

In October 2006, an 11-year-old white male was referred to the glaucoma service at the Wills Eye Institute in Philadelphia for nonspecific uveitis with uncontrolled IOP. His BCVA was 20/40 OD and 20/80 OS, and his IOPs measured 33 mm Hg OD and 28 mm Hg OS with Goldmann applanation tonometry. His current medications included brimonidine 0.15% t.i.d. and a fixed combination timolol/dorzolamide b.i.d. in both eyes. He was also using prednisolone acetate 1% t.i.d. in both eyes and receiving weekly injections of methotrexate for his uveitis.

An ocular examination revealed visually significant band keratopathy and +1 cell and flare in his left eye, trace flare and rare cells in his right eye, and bilateral posterior subcapsular cataracts (3+ in his left eye and 1+ in his right eye). We also observed a few peripheral anterior synechiae. His optic nerves were pink, sharp, and flat with cup-to-disc ratios of 0.35 bilaterally.

Humphrey visual field testing (24-2 SITA-Fast; Carl Zeiss Meditec, Inc., Dublin, CA) showed nonspecific changes in both eyes and general depression in the left eye. Imaging with the HRT3 (Heidelberg Engineering GmbH, Heidelberg, Germany) was within normal limits for his right eye and could not be obtained for his left eye.

Our diagnosis was bilateral uveitic glaucoma for which we started the patient on oral acetazolamide 125 mg. We also tapered his current dose of prednisolone 1% to once daily. He was unable to tolerate the oral acetazolamide, however, and his IOPs remained elevated in the high 20s and low 30s. Within the first 2 months after presentation, his rheumatologist added systemic infliximab (Remicaide; Centocor, Inc., Horsham, PA) to his medical regimen to help control the ocular inflammation.

Over the next 6 months, the patient's uveitis was controlled on this regimen. His IOPs continued to measure approximately 30 mm Hg, his optic nerves seemed stable, and his cataracts were still present.

HOW WOULD YOU PROCEED?

1. Would you perform a trabeculectomy? If so, would you use an antifibrotic agent?
2. Implant a glaucoma drainage device? If so, what kind?
3. Extract the patient's cataracts and implant IOLs?
4. Perform combined or staged cataract and glaucoma surgery?
5. Administer intravitreal steroids?
6. Prescribe oral steroids?

SURGICAL COURSE

After the patient's uveitis was quiescent, he underwent combined cataract extraction, IOL implantation, and the placement of a Baerveldt 250 drainage device (Advanced Medical Optics, Inc., Santa Ana, CA) under a Tutoplast scleral patch graft (IOP, Inc., Costa Mesa, CA) in his left eye only.

We combined several treatment strategies, because we felt that performing cataract surgery alone would put this patient, who was currently on maximal medical therapy, at risk of high postoperative IOP spikes. Although his optic nerves appeared stable, we were concerned that the poor view through the cataracts could be masking progression.

We prescribed 30 mg of oral prednisolone for 5 days pre-

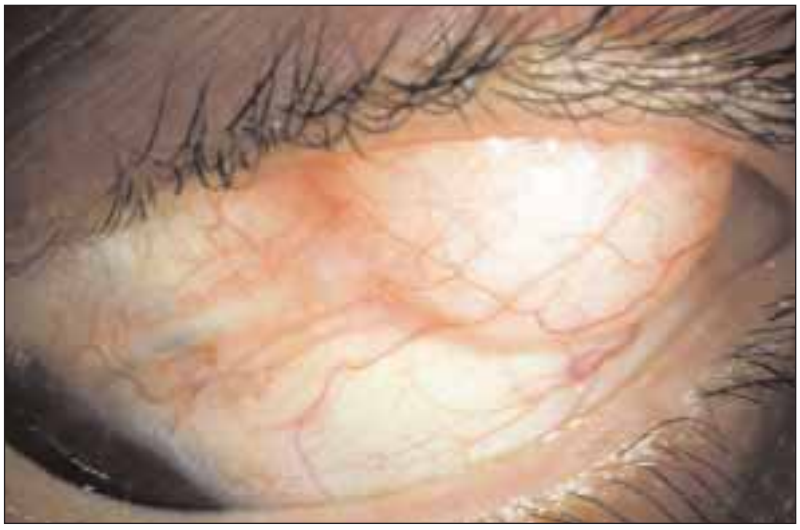

Figure 1. Unlike the mature capsule shown here, the one that developed around the shunt in the patient's left eye had thin walls and was poorly delineated.

operatively and 5 days postoperatively and slowly tapered the dosage over the month following the uneventful surgery. During the combined surgery, we fenestrated the tube in one spot and tied a 7–0 vicryl suture tightly around it to prevent early postoperative hypotony.

By 3 weeks postoperatively, the patient's BCVA in his left eye improved to 20/25, and his IOP measured 28 mm Hg. Slit-lamp examination showed 1+ cells in the same eye. One month later, his IOP measured 4 mm Hg OS, and his BCVA had decreased to 20/60 with 1+ posterior capsular opacification. We noted diffuse elevation of the conjunctiva in the area of the drainage device's plate but no distinct encapsulation (Figure 1). In addition, the anterior chamber was shallow with rare cells, and no choroidal effusions were present. The clinical impression was that the vicryl ligature suture had dissolved and that the flow to the plate was now established. The patient's medical regimen included prednisolone acetate 1% q.i.d., fluorometholone 0.1% ointment at bedtime and cyclopentolate 1% b.i.d. in his left eye as well as systemic methotrexate and infliximab. Despite increasing the frequency of his topical steroids to every 2 hours while awake, the IOP in the patient's left eye remained low. Given the high bleb, we believed that the hypotony was caused by the inadequate development of scar tissue over the device's plate, a situation that subsequently caused excessive outflow through the drainage tube. During a surgical revision of the shunt, we tied several sutures around the tube to occlude its outflow completely. Intraoperatively, we noted that the scar tissue around the plate and tube was limited, thin, and porous. We also observed that the aqueous continued to flow diffusely from the anterior chamber until the tube was completely occluded by the sutures.

To suppress inflammation that could reduce the production of aqueous or lead to excessive scarring, we injected intravitreal triamcinolone and had the patient take 30 mg of oral prednisolone for 5 days preoperatively and 5 days postoperatively. The drug was rapidly tapered over the following 3 days.

An examination of the patient's right eye revealed IOPs that were persistently in the low-to-mid 30s with no other changes on maximal medical therapy.

OUTCOME

One month after the revision of the patient's tube shunt, his IOPs were stable at 18 mm Hg on anti-inflammatory medications alone. He also had minimal inflammation and an improved BCVA of 20/30. His dose of methotrexate had been reduced based on recent liver function tests, which indicated possible mild hepatotoxicity.

During this same period, the IOP in the patient's right eye had dropped from the low-to-mid 30s to between 16 and 18 mm Hg. The increased use of IOP-lowering eye drops or a reduction in the secretion of aqueous secondary to subclinical inflammation may have led to the improvement in the patient's right eye, but we are not certain of the reason for this change.

Although we did not feel the patient required surgery on his right eye at this time, we predicted he would need cataract extraction and a glaucoma procedure in the future. Given the postoperative course in the first eye, we may have difficulty choosing between trabeculectomy and a tube shunt in his second eye if the IOP is excessively elevated. If the patient's IOP appears to be controlled at that time, we would consider performing cataract surgery only.

DISCUSSION

Persistent hypotony is an uncommon but highly undesirable complication of tube shunts, especially in an active 12-year-old child. Classically, children and patients with uveitis are predisposed to postoperative inflammation and scarring. This case was atypical because the failure of scar tissue to form over the drainage device's plate resulted in the development of a diffuse bleb. We believe that the lack of a plate-delimited bleb contributed to the hypotony and excessive outflow in the patient's left eye.

Lowering IOP in juvenile patients with uveitic glaucoma presents several challenges. The implantation of an IOL and a tube shunt during combined surgery effectively controlled IOP in one small study,[1] and larger trials have proven the efficacy of tube shunts alone in pediatric populations.[2-5] Surgeons have increasingly used glaucoma drainage devices to manage uveitic glaucoma in adults, because trabeculectomy is prone to failure in these patients due to scarring related to the inflammation.

Children and adults with suppressed immune systems are particularly susceptible to bleb-related infections after trabeculectomy.[6] The literature does not provide information about the use of tube shunts in a large series of pediatric patients with uveitic glaucoma, although it suggests that systemic immunosuppressive therapy may improve the success of surgery in uveitic patients. For example, a small study showed that patients who had uveitis secondary to juvenile rheumatoid arthritis achieved favorable outcomes after cataract surgery (with or without trabeculectomy) and the implantation of a posterior chamber IOL when topical and systemic immunotherapy was used.[7] The literature provides little information on the use of tube shunts in patients with this particular type of uveitis. Goniosurgery has also been proposed as an effective treatment for refractory glaucoma in children with chronic uveitis.[8]

We theorize that our patient's use of systemic immunomodulating therapeutic agents reduced the formation of scar tissue around the plate of the tube shunt and thus contributed to his postoperative hypotony. The topical, intravit-

real, and systemic anti-inflammatory drugs administered in conjunction with his glaucoma surgery probably also limited the development of scar tissue postoperatively. The tendency for uveitic patients to produce less aqueous may have exacerbated the low pressure despite maximal anti-inflammatory therapy.

To reverse the hypotony in the patient's left eye, we needed to reduce the flow of aqueous through the drainage device. Our options included (1) removing the tube shunt and replacing it with a valved implant, (2) removing the tube from the anterior chamber and reinserting it after scar tissue formed over the device's plate, and (3) restricting outflow through the device's tube. Given the already complicated management of the patient's uveitis, we decided that the surgical manipulation required to replace his current drainage device would increase his risk for inflammation and other complications. Specifically, the potential for uncontrolled elevations in IOP between the tube's reinsertion and removal made this option undesirable, so we chose to modify the existing tube.

We could revise the tube by (1) placing a ripcord or Latina suture in its lumen or (2) constricting it with a nonabsorbable and/or absorbable suture, either intracamerally or subconjunctivally. We performed the latter procedure with both absorbable and nonabsorbable sutures. To date, this technique has appropriately controlled the patient's IOP, affected his intraocular inflammation minimally, and facilitated his rapid recovery of visual function. ❑

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