Case Reports

Intracameral injection of tissue plasminogen activator to treat severe postoperative fibrinous reaction in iridocorneal endothelial syndrome

Bryan K. Hong, MD, and Brian A. Francis, MD, MS

Author affiliations: Department of Ophthalmology, Division of Glaucoma, Doheny Eye Institute, Keck School of Medicine, University of Southern California

Summary

Iridocorneal endothelial (ICE) syndrome is a primary endothelial abnormality that can cause a spectrum of iris changes, corneal edema, and glaucoma. Glaucoma secondary to ICE is difficult to manage because of the inflammatory reaction and fibrosis it can cause. We present a case of postoperative fibrinous reaction following endoscopic cyclophotocoagulation in ICE that caused aqueous tube shunt occlusion and high intraocular pressure. The condition was successfully managed with tissue plasminogen activator.

Introduction

There are three clinical variants of iridocorneal endothelial (ICE) based on changes in iris morphology: iris nevus syndrome, Chandler’s syndrome, and essential iris atrophy.1 Glaucoma affects roughly 50% of eyes with ICE,2 as a result of dysfunctional corneal endothelial cell migration to the periphery, over the trabecular meshwork and anterior iris. These changes result in aqueous outflow obstruction, iris-hole formation, peripheral anterior synchiae, ectropion uvea, and cor-rectopia.3 It has been speculated that the success rate of aqueous shunting procedure in ICE glaucoma is lower than with most other forms of glaucoma because ICE may predispose to a more aggressive inflammatory response;2 or, more commonly, an anteriorly displaced iris may cause malpositioning and blockage of a glaucoma-drainage tube by ICE membranes or by the iris itself, requiring either tube repositioning or other repeat surgical procedure.4,5 We report a case of exuberant fibrinous reaction causing aqueous tube shunt occlusion following endoscopic cyclophotocoagulation in ICE-related glaucoma that was successfully managed with intracameral tissue plasminogen activator injection.

Case Report

A 45-year-old, monocular white woman with a history of iris-nevus variant of ICE as confirmed by specular microscopy presented to the Doheny Eye Institute with a complaint of blurry vision in her right eye of 3 months’ duration. The blurriness was worse in the morning and improved gradually throughout the day. She denied pain and photophobia. Her past ocular history was remarkable for laser peripheral iridotomy and Baerveldt shunt implantation in the right eye 6 years prior to presentation to treat elevated intraocular pressure (IOP) secondary to ICE-related glaucoma as well as dense ambylopia of the left eye secondary to congenital cataract. The patient was on maximum medical therapy, including latanoprost 0.005%, brimonidine tartrate 0.2%, and dorzolamide hydrochloride (20 mg/mL)-timolol maleate (5 mg/mL) ophthalmic solutions.

On examination, best-corrected visual acuity was 20/40 in the right eye and hand motions in the left eye; IOP was 16 mm Hg in the right eye and 20 mm Hg in the left eye. The right eye had a functioning superotemporal Baerveldt implant with a bleb over the plate, microcystic corneal edema with beaten-metal appearance of the endothelium, a well-positioned tube in a deep and quiet anterior chamber, scattered pseudonevi on the iris, peripheral iridotomy scar that was not patent, moderate nuclear sclerotic cataract, cup-to-disc ratio of 0.65 in an otherwise normal fundus, and high, broad-based peripheral anterior synchiae and a sheen over the angle that obstructed a clear view of any angle structures. The left eye had a shallow anterior chamber along with a dense
white cataract that precluded examination of the retina. Humphrey visual fields showed a dense inferior arcuate scotoma in the right eye, which showed worsening compared to previous studies.

In order to achieve better IOP control in the right eye, endoscopic cyclophotocoagulation (ECP) was performed concomitantly with cataract extraction, placement of intraocular lens, and intraoperative injection of intracameral dexamethasone. In addition to routine postoperative antibiotic eyedrops, prednisolone acetate 1% was prescribed to be taken every hour while awake.

On postoperative day 1, the patient had visual acuity of 20/400, IOP 23 mm Hg, a vigorous fibrin reaction, 4+ anterior chamber cell, and small layering hyphema. The patient was started on oral prednisone 40 mg daily and atropine 1% twice daily in the right eye. On postoperative day 8, visual acuity was 20/150, IOP was 22 mm Hg, and the inflammatory reaction had organized into a large fibrinous membrane (Figure 1A), which by anterior segment optical coherence tomography was confirmed to be occluding the lumen of the glaucoma drainage tube (Figure 1B). Because there was no longer a bleb overlying the plate, the decision was made to inject tissue plasminogen activator (tPA) into the anterior chamber. After releasing approximately 0.1 mL of aqueous humor from the anterior chamber, approximately the same volume of tPA (10 μg/0.1 mL) was injected. One hour after injection the membrane had dissolved completely and IOP measured 17 mm Hg (Figure 1C) and a shallow bleb had reappeared. The oral prednisone was tapered over 2 weeks, whereas the topical steroid was slowly tapered over 8 weeks. On postoperative day 30, visual acuity was 20/30 and IOP was 9 mm Hg with trace anterior chamber inflammation. At one year’s follow-up, IOP remained below 10 mm Hg off of all glaucoma medications.

**Discussion**

Glaucoma secondary to ICE syndrome is notoriously difficult to treat due to a predisposition to aggressive inflammatory response and the continued growth of the endothelial membrane over the sites of filtration surgery, which may be apparent only on pathologic examination, or contraction and synechial closure of the ostium.\(^2\) Trabeculectomies are prone to fail as a result of marked subconjunctival fibrosis.\(^2,5\) More success has been found with glaucoma drainage implants for this very reason,\(^5\) yet it seems that tubes may require repositioning to maintain patency in some cases.\(^4\) According to Doe et al,\(^5\) the success rate of trabeculectomy with antimetabolite at 5 years is 29% and with glaucoma drainage implant is 53%.

In this case, the senior author (BAF) decided that further IOP reduction was necessary in the setting of a function-
ing glaucoma drainage implant (GDI) and maximum medical therapy because of a worsening visual field. It was determined that other interventions at the angle or anterior chamber would have a higher chance of failure due to corneal endothelial cell migration as compared to ECP’s targeted destruction of ciliary processes. It was decided that ECP could safely treat this yet-uncontrolled glaucoma with a prior aqueous tube shunt, as shown by Francis et al\textsuperscript{6} in 2011.

Despite aggressive steroid treatment, the patient developed a vigorous inflammatory response. With the aid of anterior segment optical coherence tomography, we were able to observe that the previously functioning tube, likely the only route of aqueous egress, was occluded. In keeping with previous observations of using intracameral tissue plasminogen activator after glaucoma surgery,\textsuperscript{7} we observed complete resolution of fibrin membranes with long-term patency of the glaucoma drainage device and excellent IOP control.

Post-tPA hyphema has been reported at higher doses (25 μg) in post-trabeculectomy eyes, and there is a risk of endophthalmitis with any intraocular injection\textsuperscript{7}; however, we observed no complications. Apart from the relatively low potential for inciting recurrent hyphema, low doses of preservative-free tPA, ranging from 6 μg to 12.5 μg, are generally well tolerated.

As a result of observations based on this case, the glaucoma surgeons at our institution generally prescribe patients with ICE undergoing ECP at least 1 day of oral prednisone (20–40 mg) preoperatively, administer 10 mg of intravenous dexamethasone intraoperatively, inject 600 to 700 μg of preservative-free dexamethasone intracameraly, and prescribe a short postoperative course of oral prednisone (20–40 mg per day) based on age, weight, and other comorbidities followed by a taper.

As this case demonstrates, postoperative inflammation in ICE is sufficient to occlude the lumen of a previously functioning aqueous tube shunt. To our knowledge, this is the first report to describe the injection of intracameral tPA as a quick and effective method of dissolving inflammatory fibrin membranes in ICE syndrome.

**Literature Search**

The authors conducted a MEDLINE search using the PubMed database (National Library of Medicine) through March 11, 2013. A combination of key words were used, including the following: \textit{iridocorneal endothelial syndrome, glaucoma, fibrinous reaction, tissue plasminogen activator, and endoscopic cyclophotocoagulation}. Words such as \textit{eye, ophthalmic, and intracocular} were cross-referenced with various key words to retrieve the specific articles. The search results were confined to articles written in English and publications with an English abstract. Inclusion and exclusion were based on relevance to the subject.

**References**