Pseudocapsulorrhexis in a Patient With Iridocorneal Endothelial Syndrome

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We describe a patient with Chandler's syndrome variant of the iridocorneal endothelial syndrome in whom ectopic Descemet's membrane was found intraoperatively on the anterior surface of the lens. Initially, the membrane was confused with the anterior lens capsule during extracapsular cataract extraction, leading to the performance of a pseudocapsulorrhexis. Electron microscopy disclosed that the epilenticular membrane was composed of multiple layers of abnormal basement membrane consistent with the iridocorneal endothelial syndrome.

Iridocorneal endothelial (ICE) syndrome includes 3 closely related entities (iris nevus or Cogan-Reese syndrome, Chandler's syndrome, and essential iris atrophy) that have characteristic abnormalities of the corneal endothelium, progressive closure of the anterior chamber angle, and a variable spectrum of iris abnormalities.1-3 These 3 entities are distinguished by the degree of iris and corneal involvement and by the temporal relationship of the development of clinical signs. Glaucoma, either associated with broad-based peripheral anterior synechiae or not, is a common complication of ICE syndrome.4

It has been hypothesized that this condition is caused by a posterior corneal abnormality that results in overgrowth of corneal endothelium and Descemet's membrane over the anterior chamber angle and anterior aspect of the iris. A unique case characterized by extension of an ectopic Descemet membrane onto the lens, which interfered with cataract extraction, is presented here.

REPORT OF A CASE

A 70-year-old white woman had glaucoma diagnosed in her left eye 2 years prior to our initial examination. Several topical medications (0.5% levobunolol hydrochloride twice daily, 0.1% dipivefrin hydrochloride twice daily, and 1% pilocarpine hydrochloride 4 times daily) failed to control her intraocular pressure (IOP). Family and medical histories were noncontributory. On examination her best-corrected visual acuities were 20/25 OD and 20/40 OS. The right eye was normal. The nasal corneal endothelium of her left eye had a hammered-silver appearance and the stroma was mildly edematous. The anterior chamber was deep and clear. The pupil was minimally reactive and displaced slightly inferiorly. There were no iris holes or nodules. Intraocular pressures were 17 mm Hg OD and 37 mm Hg OS. Gonioscopy of her left eye revealed scattered broad-based peripheral anterior synechiae closing most of the angle. Ophthalmoscopy revealed marked disc asymmetry, with a cup-disc ratio of 0.3 OD and 0.7 OS.

Chandler’s syndrome was diagnosed clinically. Specular microscopy of the corneal endothelium showed polymegathism and demarcated islands of abnormal cells with dark-light reversal, compatible with ICE syndrome. The addition of timed-release oral acetazolamide, 500 mg twice daily, failed to control the IOP and a trabeculectomy with mitomycin was performed. Although the procedure initially controlled the IOP, it abruptly increased to 39 mm Hg 2 years later. At that time a large elevated diffuse ischemic bleb was observed and, on gonioscopy, the scle-
rostomy appeared to be open. Medical treatment did not lower the IOP. Neodymium:YAG laser internal revision of the sclerostomy successfully controlled the IOP for 1 year. The IOP rose again, and a second Nd:YAG internal revision was performed, which controlled the IOP for 6 months. At this time visual acuity was 20/300 OS (with pinhole, 20/80 OS). Slitlamp examination disclosed a thin avascular cystic bleb, moderate stromal edema, and clinically relevant (3+) nuclear sclerosis. The IOP was 43 mm Hg, and the optic disc cupping had enlarged (cup-disc ratio of 0.9).

The patient underwent cataract extraction by phacoemulsification, posterior chamber intraocular lens implantation, and trabeculectomy with mitomycin. During the procedure, which was performed by 1 of us (R.P.W.), a continuous circular capsulorhexis was apparently performed. Surprisingly, after this maneuver the surgeon realized that the anterior lens capsule was still intact and that he actually had excised a membrane that was covering the anterior aspect of the lens. This tissue was sent for pathologic studies and the procedure (with a true capsulorrhexis) was finished without complications.

Transmission electron microscopy performed on deparaffinized tissue confirmed that the membrane was not anterior lens capsule. The membrane was composed of multiple layers of abnormal basement membrane material that varied in caliber and incorporated foci of iris pigment. The findings were consistent with the ICE syndrome (Figure). Four months postoperatively visual acuity was 20/50 OS and IOP was in the lower teens without antiglaucoma treatment.

In this case the clinical diagnosis of Chandler’s syndrome was based on the identification of abnormal corneal endothelium (by biomicroscopy and specular microscopy) and the presence of broad anterior synchiae in a patient with unilateral closed-angle glaucoma and mild corneal edema. The normal appearance of the contralateral eye and the negative family history excluded posterior polymorphous endothelial dystrophy. Pathologic examination of the membrane on the anterior aspect of the lens was consistent with the diagnosis.

To our knowledge, this is the first time that ectopic Descemet membrane material has been found intraoperatively on the anterior lens capsule in a patient with the ICE syndrome. It is important to recognize this possibility to avoid confusion and complications during cataract surgery.

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REFERENCES