Axenfeld-Rieger Syndrome and a Post-Trabeculectomy Presentation of Angle Closure

Steven Kane, MD, Will Griffeth, MD, Charles Blake, MD
From Department of Ophthalmology, University of Florida, Gainesville

ABSTRACT
One rare form of anterior segment dysgenesis, Axenfeld-Rieger syndrome, can be associated with glaucoma in half of the cases. We present an illustrative case of a patient managed with trabeculectomy complicated by angle closure.

Key words: Anterior segment dysgenesis; Axenfeld-Rieger syndrome; glaucoma.

INTRODUCTION
Axenfeld-Rieger syndrome is a rare, inherited dysgenesis of the anterior segment that is associated with glaucoma approximately 50% of the time. This case illustrates a unique presentation of angle closure despite adequate trabeculectomy in an Axenfeld-Rieger patient that was initially managed with dynamic gonioscopy.

CASE REPORT
A 24 year-old female with a past medical history of congenital glaucoma and trabeculectomy surgery in the right eye (OD) presented to the Emergency Department (ED) with acute onset of boring right eye pain, nausea, vomiting, and blurred vision OD of 1 hour duration. She reported having had a trabeculectomy OD for similar symptoms 4 years prior, with stable...

Photo 1 – Right eye 1 week after presentation
postoperative intraocular pressure (IOP) in the low single digits despite multiple procedures to correct bleb overfiltration. She denied any symptoms relating to hypotony maculopathy. Exam revealed visual acuity of OD 20/250 (pinhole 20/70) and normal 20/20 vision in the left eye (OS). IOP was 58mmHg OD and 12 mmHg OS by applanation. She had extensive iris atrophy and corectopia OD with no relative afferent pupil defect. Slit lamp exam OD showed a low lying superior conjunctival bleb, corneal edema, extensive irido-corneal adhesions, extensive iris atrophy and corectopia, and a trace nuclear sclerotic cataract. The left eye also showed superior-nasal irido-corneal touch and superior-nasal iris atrophy. Her slit lamp exam was consistent with the diagnosis of Axenfeld-Rieger syndrome (Photo 1 and 2). Gonioscopy of the right eye showed no visible angle structures secondary to obstructing iris remnants and gonioscopy of the left eye showed irido-corneal touch in the superior temporal quadrant (Photo 3 and 4). Initial treatment in the ED included multiple administrations of topical antihypertensive glaucoma medications and dynamic gonioscopy. The dynamic gonioscopy was successful in breaking the acute glaucoma attack and her IOP OD dropped to 02 mmHg. She was discharged without glaucoma medications. The following day, her vision OD had improved to 20/80 (pinhole 20/40) and her IOP OD remained low at 02 mmHg. The trabeculectomy site OD appeared to be functioning well, however the lens-zonule complex appeared excessively loose and mobile. An anterior segment ocular coherence tomography (OCT) of the right eye revealed a flat anterior chamber with significant iridocorneal touch (Photo 5).

We suspected the mechanism of her acute IOP elevation was migration of the lens-zonule complex into the trabeculectomy site causing mechanical obstruction which was acutely alleviated by dynamic gonioscopy.

To maintain posterior positioning of the lens-zonule complex, we initiated 1% atropine TID OD which was weaned to every other day over 4 weeks. She continued to have good control of her IOP and no additional angle closure attacks.

We discussed removal and replacement of the crystalline lens. Considering the complexity of the surgery, the patient chose observation. In conclusion, dynamic gonioscopy represents a useful method to lower IOP in cases of angle closure and other forms of obstructive outflow.

Photo 2 – Left eye 1 week after presentation
Postoperative intraocular pressure (IOP) in the low single digits despite multiple procedures to correct bleb overfiltration. She denied any symptoms relating to hypotony maculopathy. Exam revealed visual acuity of OD 20/250 (pinhole 20/70) and normal 20/20 vision in the left eye (OS). IOP was 58mmHg OD and 12 mmHg OS by applanation. She had extensive iris atrophy and corectopia OD with no relative afferent pupil defect. Slit lamp exam OD showed a low lying superior conjunctival bleb, corneal edema, extensive irido-corneal adhesions, extensive iris atrophy and corectopia, and a trace nuclear sclerotic cataract. The left eye also showed superior-nasal iridocorneal touch and superior-nasal iris atrophy. Her slit lamp exam was consistent with the diagnosis of Axenfeld-Rieger syndrome (Photo 1 and 2). Gonioscopy of the right eye showed no visible angle structures secondary to obstructing iris remnants and gonioscopy of the left eye showed irido-corneal touch in the superior temporal quadrant (Photo 3 and 4).

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Photo 3 – Gonioscopy of the right eye superior view showing iridocorneal adhesion and severe corectopia.

Photo 4 – Gonioscopy view of the left eye showing superior temporal iridocorneal adhesion. Angle structures were identifiable elsewhere.

Photo 5 – Anterior Segment OCT of the right eye demonstrating significant areas of irido-corneal adhesion and flat anterior chamber.

REFERENCES