• Letter to the Editor •

Ahmed glaucoma implant and augmented MicroPulse transscleral cyclophotocoagulation in a monocular Axenfeld-Rieger syndrome patient with glaucoma

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Dear Editor,

We report the case of severe mixed-mechanism glaucoma observed in a monocular Axenfeld-Rieger syndrome (ARS) patient who was successfully managed with an ahmed glaucoma implant (AGI) followed by a novel technique described as augmented MicroPulse transscleral cyclophotocoagulation (MP-TSCPC)[1]. ARS encompasses a heterogeneous spectrum of diseases involving anterior segment dysgenesis and systemic developmental anomalies[2]. The ocular features of ARS are postulated to arise from developmental arrest of neural crest derived cells in gestation, leading to retention of endothelial tissue on the iris and anterior chamber[3]. Other characteristics include incomplete maturation of the trabecular meshwork and Schlemm's canal as well as high insertion of the iris, all of which predispose patients to elevated intraocular pressure (IOP) and glaucoma, which has been reported in as many as 50% of ARS patients[3-4]. However, optimal surgical management of elevated IOP and downstream complications is unclear in patients with ARS. The present case describes successful surgical management of severe mixed-mechanism glaucoma in a patient with ARS. We obtained the written informed consent from the patient, and this case study is in accordance with the tenets of the Declaration of Helsinki.

A 30-year-old monocular man with severe-stage mixed-mechanism glaucoma (ARS, multiple ocular surgeries, history of steroid response) was referred to our glaucoma clinic after presenting to the emergency department in November 2019 with an IOP of 46 mm Hg in his seeing left eye (OS). His ocular history was notable for trabeculectomies in both eyes (OU) in 1989 and 1993, goniotomies OU in 1990, pars plana vitrectomy (PPV) with endolaser right eye (OD) on 3 occasions for recurrent retinal detachments between 2017 and 2018, iris prosthesis and lens implantation OD in 2018, and 1 PPV with endolaser for retinal detachment OS in February 2019. His family history was significant for glaucoma of unclear etiology in his mother.

On exam, his visual acuity (VA) was long-standing light perception (LP) OD and 20/200 at near, with no improvement on pinhole OS. The patient reported that his left eye vision allowed him to work full time and read up close without difficulty. His IOP measurements were 4 mm Hg OD and 33 mm Hg OS on 6 medications: dorzolamide-timolol and brimonidine twice daily (BID), netarsudil-latanoprost once nightly (QHS), and oral acetazolamide 500 mg BID.

Given the patient's monocular status, subsequent discussion is focused primarily on his functional left eye. Central corneal thickness in this eye was 556 microns. Slit lamp examination OS revealed a scarred superior bleb, iris atrophy with corectopia and polycoria, endothelial pigment deposits centrally, and phakic lens status with trace nuclear sclerosis. His optic nerve was diffusely thin with extensive peripapillary atrophy and a cup-to-disc ratio of 0.9. No Drance hemorrhages were noted. His last reliable 24-2 Humphrey visual field stimulus V revealed superior arcuate and inferior paracentral defects (Figure 1). Disc photographs and optical coherence tomography were unobtainable due to the patient's nystagmus and corectopia.

Given his monocular status and uncontrolled IOP on maximally-tolerated medical therapy, the patient underwent superotemporal Ahmed-FP7 (New World Medical, Rancho Cucamonga, CA, USA) drainage implant OS in November

2019 (Figure 2). The surgery proceeded without complications. On postoperative day 1, IOP decreased from 33 mm Hg to 4 mm Hg OS off glaucoma medications. However, by day 10, his IOP rebounded to 24 mm Hg. Preservative-free (PF) dorzolamide-timolol three times a day (TID), tafluprost QHS, netarsudil QHS, and brimonidine TID were successively reintroduced over the next month to achieve IOP control. At postoperative week 4, his IOP remained elevated at 19 mm Hg. Augmented MP-TSCPC with IRIDEX's Generation 1 MicroPulse P3 glaucoma device (IRIDEX, Mountainview, CA, USA) was subsequently performed in December 2019 in the left eye for AGI-associated hypertensive phase (HP). In this novel cyclodestructive procedure (validated by Nirappel *et al*[5]), a higher-than-usual 2200 mW laser setting was applied to the patient's superior and inferior hemisphere for 180 seconds each. This approach also utilized a combination of sweeping and stop-and-continue techniques instead of the sweeping technique alone, where 90 seconds of the sweeping technique were followed by 90 seconds of the stop-and-continue technique in each hemisphere. The stop-and-continue technique was performed by dividing each hemisphere along the limbus into 9 equal sections and applying a 10-second continuous treatment to each section. One day after the augmented MP-TSCPC procedure, the patient's IOP was 12 mm Hg on tafluprost QHS only. Preservative free dorzolamide-timolol BID was restarted in the left eye to accompany a loteprednol etabonate 0.5% taper given his history of steroid response. At his last follow up on postoperative month 18 after MP-TSCPC (19mo after AGI), his IOP was stable at 11 mm Hg on PF dorzolamide-timolol TID and tafluprost QHS. His VA remained unchanged and no inflammation was noted at this time.

There is little consensus regarding optimal management of glaucoma in patients with ARS. It has been previously observed that medical therapy alone often fails to control IOP and prevent progression of glaucoma in these patients, perhaps due to developmental malformations of the trabecular and post-trabecular meshwork and uveoscleral outflow tracts[6]. In refractory cases where surgery is necessary, there is controversy regarding which procedural option is indicated. Some have suggested that goniotomy and alternative angle surgery (or *ab externo* trabeculotomy) are inappropriate for ARS-associated glaucoma[3,7]. Others have documented IOP improvement with combined trabeculotomy-trabeculectomy and one case report described short term IOP control with AGI, although longer-term efficacy was not ascertained due to tube migration after 6mo[6,8].

Optimal approaches to AGI-associated HP are similarly understudied. One group suggests that sub-Tenon's injection of triamcinolone acetonide during tube implantation may decrease the occurrence of HP[9]. Others recommend waiting

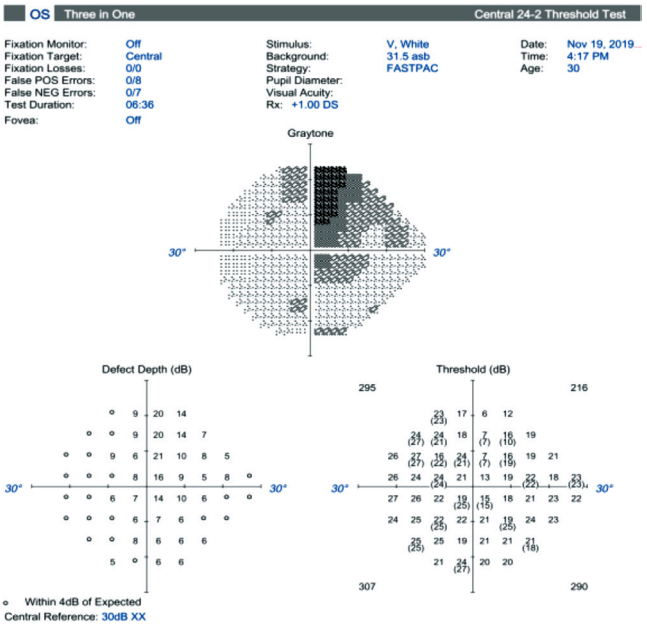

Figure 1 Humphrey visual field (left eye) showing superior arcuate and inferior paracentral defects.

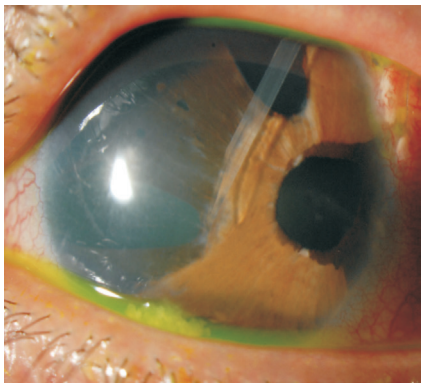

Figure 2 Slit lamp image showing Ahmed tube draining the anterior chamber in patient with Axenfeld-Rieger syndrome There is also evidence of notable anterior segment dysgenesis, including iris atrophy, corectopia, and polycoria.

based on evidence of spontaneous resolution[10]. Given this patient's monocular status, a more aggressive approach such as TSCPC was desirable.

Studies of MP-TSCPC have demonstrated its ability to achieve significant reductions in IOP (by 6.9-12.6 mm Hg at 1y) and medication burden (by 0.5-1.6 at 1y) across a variety of glaucoma types[11]. However, defining optimal laser settings like power and duration of treatment is difficult. This may partly be due to differences between patient populations and variability in laser settings used in prior MP-TSCPC studies. Nirappel *et al*[5] showed that augmented MP-TSCPC can improve the longevity of treatment effect without increased risk of postoperative complications (*i.e.*, prolonged anterior chamber inflammation, hypotony or visual decline). However, the use of this approach in a patient with ARS has not yet been reported.

Finally, it is worth noting that this patient has an extensive history of recurring retinal detachments. Retinal detachments do not belong to the list of characteristic clinical signs that are typical of this disorder. However, other studies of ARS patients with large or bilateral retinal detachments have raised the question of whether ARS is also a disease of the posterior segment of the eye[12].

Using AGI followed by augmented MP-TSCPC for HP, we show substantial reductions in IOP (24 to 11 mm Hg) and medication burden (6 to 3 agents) that have persisted without complications at least 18mo since the last intervention. Further studies should be performed to help elucidate optimal surgical management in ARS-associated glaucoma.

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