

Ologen in Axenfield-Rieger Anomaly with secondary Glaucoma: A case report with review of literature

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Abstract

We report a case of Axenfield-Rieger anomaly with secondary glaucoma which was managed with trabeculectomy with adjunctive mitomycin-C and Ologen implant. A 27-year old female who was a known case of glaucoma and was on anti-glaucoma medications for last 24 years, presented with heaviness over left eye. A diagnosis of bilateral Axenfield-Rieger anomaly with secondary angle closure glaucoma was made with right eye controlled on topical anti-glaucoma medications and left eye uncontrolled (30 mm Hg) with anti-glaucoma medications. Patient underwent the above-mentioned surgery in her left eye. The IOP was controlled and bleb was well formed at one year of follow-up. This case report highlights the management of Axenfield-Rieger anomaly with secondary glaucoma with the use of Ologen implant.

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Keywords: Axenfield-Rieger anomaly, Secondary glaucoma, Ologen, Trabeculectomy, Mitomycin-C

Introduction

Axenfield-Rieger anomaly (ARA) is characterized by anterior segment dysgenesis which encompasses various irido-corneal anomalies like prominent and anteriorly displaced Schwalbe's line, atrophic holes in iris, iridocorneal adhesions, ectropion uveae, corectopia and polycoria. Over half of patients with ARA have risk of glaucoma which may develop from infancy to any age.¹ The management of secondary glaucoma associated with ARA is primarily surgical. There have been few reports of various surgical procedures like trabeculotomy, combined trabeculotomy-trabeculectomy, and trabeculectomy with variable results in developmental glaucoma including secondary glaucoma associated with ARA.²⁻⁵ The controversy exists regarding the ideal procedure for the management of glaucoma associated with ARA.

Ologen collagen matrix (Aeon Astron Europe BV, Leiden, The Netherlands) is a biodegradable implant which has been proposed to reduce the scar formation after trabeculectomy which is the major cause of failure of trabeculectomy.⁶ There is paucity of literature regarding the use of Ologen implant in trabeculectomy for patients with ARA.⁷ We report the one year treatment outcome of a case of secondary glaucoma associated with ARA managed with trabeculectomy with mitomycin-C and Ologen implant.

Report of Case

A 27-years-old female presented with the complaints of heaviness over her left eye. She was a known case of glaucoma diagnosed at the age of 3 years. She was on anti-glaucoma medications for last 24 years. There was no report of similar problem in any of the family member.

On ophthalmic evaluation, her best-corrected-visual-acuity (BCVA) was 6/9 both eyes on Snellen chart. Intra-ocular pressures (IOP) measured with Goldmann Applanation Tonometry (GAT) were 14 mm Hg in the right eye and 30 mm Hg in the left eye on brimonidine (0.2%) and timolol (0.5%) combination with latanoprost (0.005%) in both eyes. Slit lamp examination revealed diffuse corneal

haze associated with patches of iris atrophy, variable iris pigmentation, corectopia, and peripheral anterior synechiae. The increased vertical cup disc ratio of 0.7 in right eye and 0.8 in left eye was observed. Visual field examination (Humphrey Field Analyzer, Carl Zeiss Meditec, Inc., Dublin) showed two scotomas in right eye and arcuate defect in left eye characteristic of glaucoma which were persistent after surgery (Figure 1). Synechial angle closure of at least 180 degree were seen in both eyes on gonioscopy.

On the basis of clinical features, the diagnosis of bilateral Axenfield-Rieger anomaly with secondary angle closure glaucoma was made with left eye uncontrolled on topical 3 anti-glaucoma medications. The patient was planned for trabeculectomy with mitomycin-C (0.02%) with Ologen implant in left eye and for right eye anti-glaucoma medications was continued.

The surgery was performed with fornix based conjunctival flap, partial thickness scleral flap and subconjunctival application of mitomycin-C (0.02%) away from the scleral flap for 3 minutes then thorough wash with normal saline. The Ologen (6 mm × 2 mm) was placed over the apex of triangular scleral flap. Postoperative regime included moxifloxacin (0.5%) and dexamethasone combination 4-6 times per day tapered over 1 month. After surgery, patient experienced an episode of hypotony, shallowing of anterior chamber which was associated with the blurring of vision on 3rd day of surgery. The bleb was diffuse and elevated with absent bleb leak confirmed with Siedel's test. After the formation of anterior chamber with air, patient IOP was 14 mm Hg with well formed anterior chamber with visual acuity of 6/9 on Snellen chart.

At last follow-up (1 year), her IOP was 12 mm Hg without any anti-glaucoma medications and bleb was formed (Figure 2).

Discussion

Angle surgeries (goniotomy and trabeculotomy) have been the mainstay of treatment for primary congenital glaucoma. However, it has been suggested that the angle surgery may

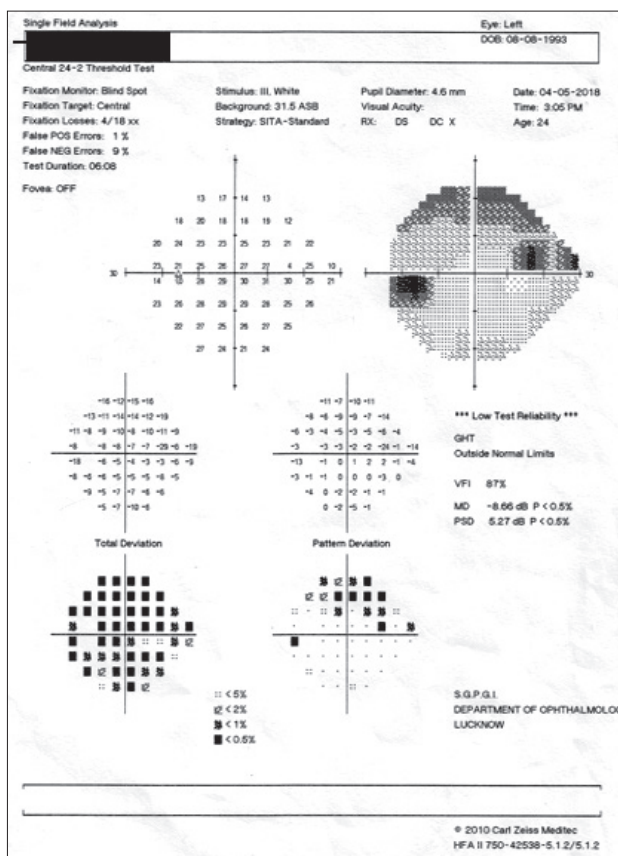
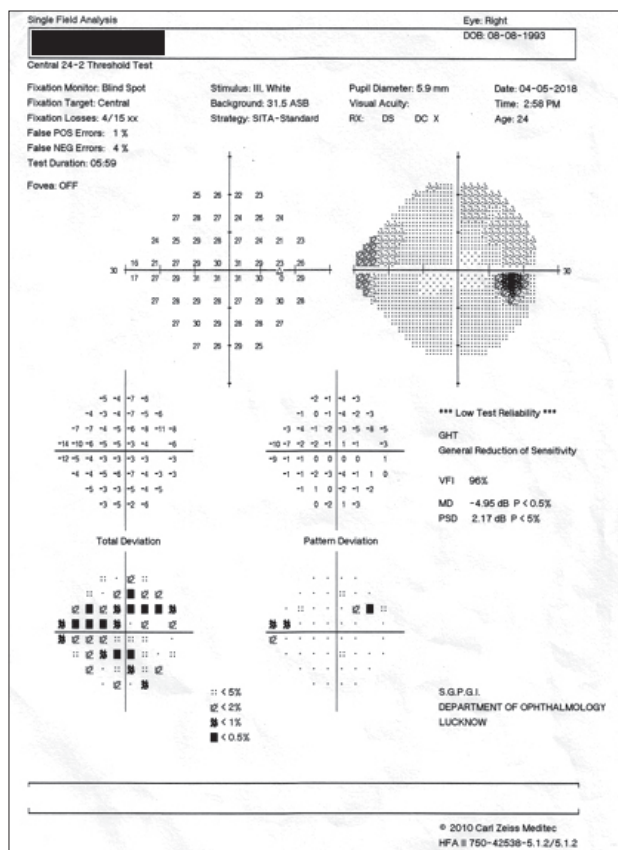


Figure 1: Visual field (Humphrey Field Analyzer, Carl Zeiss Meditec, Inc, Dublin) print-out showing two scotomas in right eye and arcuate defect in left eye, both in superior part of field.

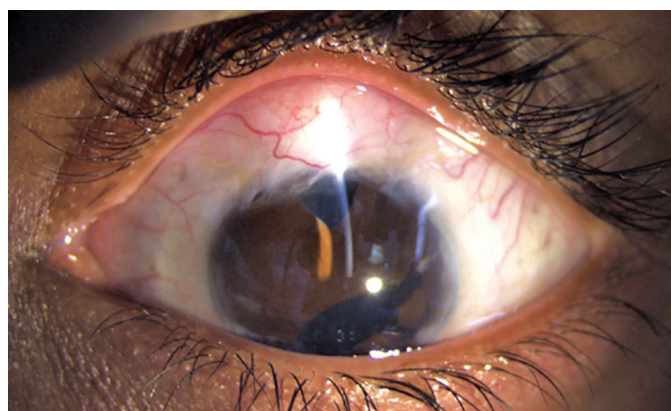


Figure 2: The postoperative photograph of left eye after trabeculectomy with mitomycin-C and Ologen implant in a patient with Axenfield-Rieger anomaly with secondary angle closure glaucoma.

not be a viable option for the treatment of ARA as they have different pathological mechanism of glaucoma due to the arrested development of anterior chamber angle and incomplete maturation trabecular meshwork and Schlemm’s canal.⁸ Shields suggested the possible role of trabeculectomy with antimetabolites for these patients.⁸ Trabeculectomy has been found to be an effective option in developmental glaucoma over a long time by Ikedal, et al but 3 out of 15 eyes with ARA with one or more trabeculotomies needed further trabeculectomy and/or cyclocryotherapy to control IOP.² They advocated the possible role of vigorous wound healing and exuberant scarring behind the failure of these cases.

Ologen implant has been found to have the comparable efficacy and safety as with mitomycin-C over the longer period of time in earlier studies including a meta-analysis of randomized controlled trial.⁹ We performed surgery using low dose mitomycin-C and Ologen implant to prevent the exuberant fibrosis associated with the failure of trabeculectomy. Similarly, Ologen implant has been found to be an effective option in the management of secondary glaucoma associated with ARA in a single case report.⁷ We reported shallow anterior chamber and hypotony which was resolved with anterior chamber formation. No specific complication regarding the use of Ologen and mitomycin-C was reported. This case report may provide a useful clinical information regarding the management of secondary glaucoma associated with ARA. However, further studies involving large number of patients with possible randomization are needed to prove the role of Ologen implant in the management of secondary glaucoma associated with ARA.

References

1. Shields MB, Buckley E, Klintworth GK, Thresher R. Axenfield-Rieger syndrome. A spectrum of developmental disorders. *Surv Ophthalmol*, 1985; 29:387-409.
2. Ikeda H, Ishigooka H, Muto T, Tanihara H, Nagata M. Long term outcome of trabeculectomy for the treatment of developmental glaucoma. *Arch Ophthalmol*, 2004; 122:1122-1128.
3. Susanna R Jr, Oltrogge EW, Carani JC, Nicoletta MT. Mitomycin as adjunct chemotherapy with trabeculectomy in congenital and developmental glaucomas. *J Glaucoma*, 1995; 4:151-157.

4. Mandal AK, Prasad K, Navduvilath TJ. Surgical results and complications of mitomycin C-augmented trabeculectomy in refractory developmental glaucoma. *Ophthalmic Surg Lasers*, 1999; 30:473-480.
5. Mandal AK, Pehre N. Early-onset glaucoma in Axenfield-Rieger anomaly: long-term surgical results and visual outcome. *Eye (Lond)*, 2016; 30:936-942.
6. Chen HS, Ritch R, Krupin T, Hsu WC. Control of filtering bleb structure through tissue bioengineering: an animal model. *Invest Ophthalmol Vis Sci*, 2006;47:5310-5314.
7. Radhakrishnan OK, Pahuja K, Patel K, Chandna S. OLOGEN implant in the management of glaucoma in an unusual case of Axenfield-Rieger syndrome. *Oman J Ophthalmol*, 2014; 7:90-92.
8. Shields MB. Axenfield-Rieger syndrome: a theory of mechanism and distinctions from the iridocorneal endothelial syndrome. *Trans Am Ophthalmol Soc*, 1983; 81:736-784.
9. Ji Q, Qi B, Liu L, Guo X, Zhong J. Efficacy and safety of ologen implant versus mitomycin-C in primary trabeculectomy: a meta-analysis of randomized clinical trials. *J Glaucoma*, 2015; 24:e88-94.

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