

## Congenital unilateral ectropion uveae with refractory glaucoma

Kamala. S<sup>1</sup>, Rashmi N R<sup>2\*</sup>

<sup>1</sup>Consultant, <sup>2</sup>Glaucoma Fellow, Dept. of Ophthalmology, Sankara Eye Hospital, Shimoga, Karnataka, India

\*Corresponding Author: Rashmi N R

Email: [dnrrashmi@gmail.com](mailto:dnrrashmi@gmail.com)

### Abstract

An eight year old healthy female child was referred for the management of raised intraocular pressure (IOP) in left eye. Examination of left eye revealed 360° ectropion uveae, trabecular dysgenesis and glaucomatous disc. A diagnosis of congenital ectropion uveae with glaucoma was made. As IOP became refractory to maximal topical therapy, trabeculectomy with adjunctive Mitomycin-C had to be performed. The IOP normalized after surgery and is well maintained without any anti glaucoma medication. Early recognition of this rare anomaly and close monitoring of IOP is very crucial. Refractive nature of raised IOP needs to be promptly addressed at the earliest with trabeculectomy.

**Keywords:** Congenital glaucoma, Ectropion uveae, Mitomycin-C, Trabecular dysgenesis, Trabeculectomy.

### Introduction

Congenital ectropion uveae (CEU) is a rare congenital anomaly characterized by iris pigment epithelium on the anterior surface of the iris. Majority of cases are isolated but few will have associated other ocular and systemic comorbid conditions.<sup>1</sup>

Most of these eyes with ectropion uveae eventually develop glaucoma.<sup>3</sup> We have described a similar case of CEU in young who presented with refractory glaucoma, its surgical management and clinical course over 5 years of follow-up.

### Case Report

An eight year old girl was seen at glaucoma clinic for management of uncontrolled raised intraocular pressure (IOP) in her left eye. On examination, right eye (RE) BCVA was 6/6 and left eye (LE) was 6/12p. The IOP recorded with applanation tonometer was 14 and 38mmHg in right and left eye respectively.

RE was essentially normal on slit lamp examination. LE showed 360° ectropion uveae without neovascularization of iris (Fig. 1), slightly larger round pupil reactive to light. Gonioscopy revealed featureless angles with dense pigment deposition in LE and normal angles in RE. Fundus examination showed advanced cupping in LE and 0.3 cupping in RE. Patient denied any history of trauma or previous surgical intervention.

A diagnosis of CEU with secondary glaucoma was made and patient was started on 3 antiglaucoma medications including oral, topical carbonic anhydrase inhibitors and beta blockers along with amblyopia management. The IOP remained high in 30's even after 6 months of follow-up. Later, LE underwent trabeculectomy with adjunctive Mitomycin-C (0.2mg/ml). IOP normalized to 18mmHg in LE post-surgery and continued to remain the same during

follow-up over 5 years. The development of posterior subcapsular cataract was managed by clear corneal phacoemulsification with foldable lens implantation retaining BCVA at 6/12 vision.

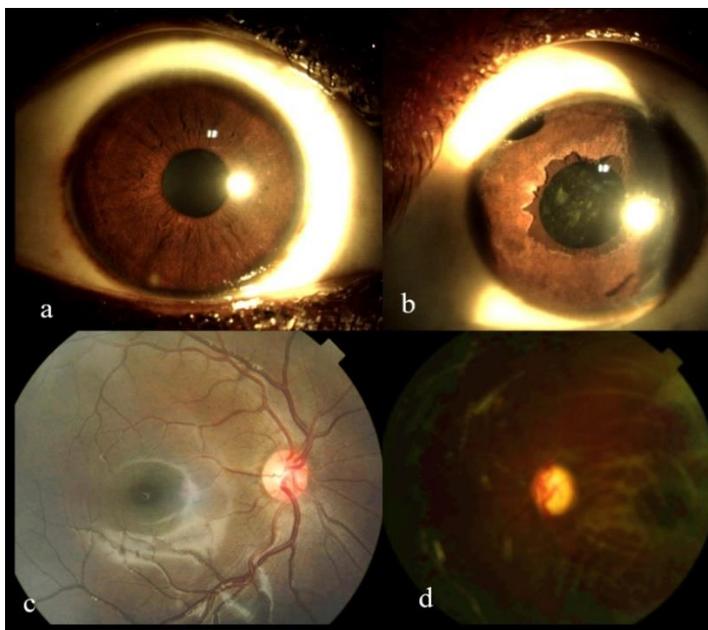
### Discussion

Congenital ectropion uveae was described first by Wicherkiewicz B<sup>4</sup>. A rare congenital condition manifesting at younger age with unilateral ectropion uveae and glaucoma. Development of glaucoma was noted in 80-90% of affected individuals. Patients need comprehensive evaluation as CEU may be associated with other ocular anomalies (e.g. Riegers anomaly, coloboma, and ptosis) and systemic diseases (e.g. neurofibromatosis type 1, Prader-Willi Syndrome and facial hemi hypertrophy).<sup>1</sup>

The major concern with this condition is high chances of development of glaucoma as noted by Ritch R et al where 7/8 eyes developed glaucoma in his series.<sup>3</sup> Majority of these cases are refractory to conventional anti glaucoma medications. Dowling JL Jr et al., observed poor control of IOP with topical medication necessitating surgical intervention in his all 10 cases.<sup>2</sup>

The poor control of IOP with topical anti glaucoma medication appears to be secondary to drainage angle dysgenesis and incomplete development of trabecular meshwork and Schlemm's canal. Harasymowycz PJ et al., observed presence of a fibrovascular membrane covering the anterior aspect of the iris stroma probably pulling posterior pigment layer anteriorly creating ectropion.<sup>5</sup>

Although glaucoma can occur from birth to middle age, majority get diagnosed between 3-15 years of age. CEU is mostly associated with refractory glaucoma. Hence, early surgical intervention should be considered in such cases.



**Fig. 1**

Composite picture showing normal anterior segment of right eye (a) and 360° ectropion uveae in left eye (b) {post-trabeculectomy picture}. Normal right eye fundus (c) and advanced cupping in left eye (d).

### Conclusion

Visual outcome in patients with CEU depends on prompt diagnosis and treatment. Management of raised IOP needs early surgical intervention to prevent permanent glaucomatous damage. Usually trabeculectomy with MMC application appears to give adequate IOP control in majority of cases.

**Conflict of Interest:** None.

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