Secondary Glaucoma Due to Congenital Ectropion Uveae

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ABSTRACT

Here, we present a case with secondary glaucoma due to congenital ectropion uveae. A 6-year-old girl was referred to our clinic with the diagnosis of congenital ectropion uveae and secondary glaucoma in her right eye. In the ophthalmic examination, visual acuity as measured by Snellen charts was 0.4 (with correction of -2.50 D) in the right eye and 1.0 (uncorrected) in the left eye. In the slit-lamp examination, congenital ectropion uveae was present in the right eye and the left eye was normal. Intraocular pressure was 35 mmHg (with maximum medical treatment) in the right eye and 19 mmHg in the left eye. In addition, there was also mild ptosis in the right eye. There was no systemic abnormality in the patient. The patient was treated with Ahmed glaucoma valve implantation in the right eye. Glaucoma may develop due to angle dysgenesis in patients with congenital ectropion uveae. They should be examined in a periodical manner for the early detection of glaucoma.

Key words: Ectropion uveae, Glaucoma, Ahmed glaucoma valve.

INTRODUCTION

Ectropion uveae (EU) is a clinical presentation where iris pigment epithelium is seen on anterior surface of iris. There are two subtypes as congenital and acquired EU. The acquired form generally occurs as a result of membranous tractions secondary to conditions such as ischemia and inflammation.[1] Congenital ectropion uveae (CEU) is a neural crest-originated disorder which characterized by non-progressive proliferation of iris pigment epithelium on anterior surface of iris. It was first described by Wicherkiewicz in 1891 and by Spiro in 1896.[2, 3] Glaucoma development is associated to iridotrabecular dysgenesis. In patients with CEU, glaucoma can be present upon birth; however, it may also develop at infantile period or older ages. In cases with CEU, glaucoma is generally refractory, requiring aggressive treatment.[3]

Here, we aimed to present a pediatric case with glaucoma secondary to CEU and treated with Ahmed glaucoma valve (AGV) implantation surgery.

CASE REPORT

A 6-year-old girl was referred to our clinic as CEU and glaucoma in the right eye from another facility where she presented with redness in her right eye. In initial ophthalmological examination in our clinic, visual acuity as measured by Snellen charts was recorded as 0.4 (with correction of -2.50 D) in the right eye and as 1.0 (uncorrected) in the left eye. In the slit-lamp examination, no pathological finding other than CEU was detected in the right eye (Figure 1) and the left eye was normal. Intraocular pressure (IOP) as measured by Goldmann applanation tonometry was 35 mmHg (under fixed combination of topical brinzolamide plus timolol with travoprost therapy) in the right eye and 19 mmHg in the left eye. In fundus examination, cup: disc ratio was 0.9 (Figure 2). On optical coherence tomography, there was thinning in peripapillary retinal nerve fiber layer in the right eye (Figure 3). In addition, there was congenital ptosis not interfering with visual axis in the right eye (Figure 4); however, levator palpebrae superioris function was normal. In her history, there was no systemic disease or medication. In systemic examination by pediatrics department, no abnormal finding was detected.

The patient was diagnosed as secondary glaucoma due to CEU and AGV implantation was performed in the right eye (Figure 5). At postoperative period, antiglaucomatous agents were withdrawn and topical prednisolone acetate

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Received: 28.02.2022
Accepted: 17.09.2022

J Glau-Cat 2022; 17: 200-203

DOİ: 10.37844/glau.cat.2022.17.33

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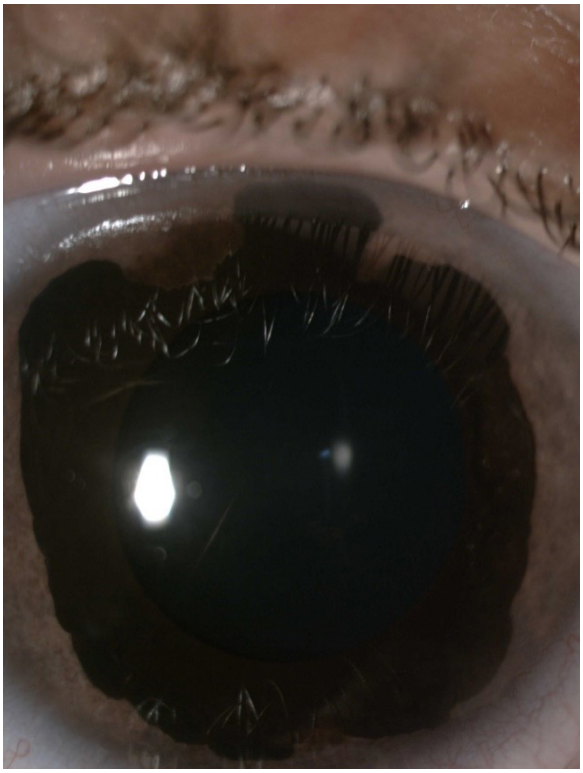

Figure 1: *Ectropion uveae on anterior segment image of the right eye obtained in initial examination.*

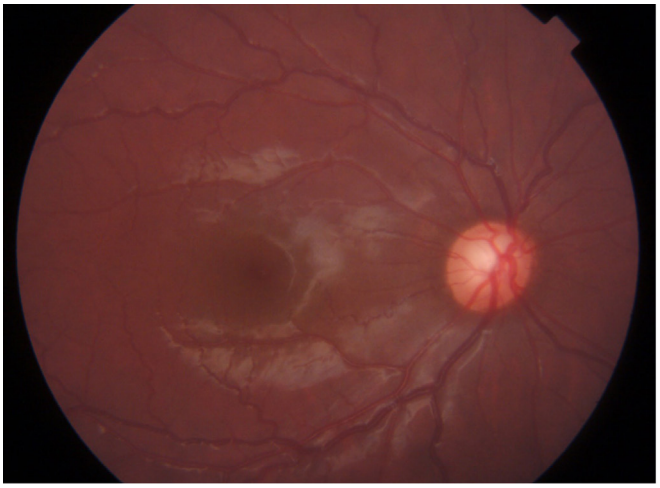

Figure 2: *Cup: disc ratio of 0.9 on fundus image.*

(6x1) and ofloxacin (6x1) eye drops were prescribed. The topical ofloxacin was discontinued after 2 weeks. The IOP was normalized at early postoperative period (<18 mmHg), however, it was measured as 30 mmHg on postoperative month 1, implying onset of hypertensive phase. Thus, antiglaucomatous agents (latanoprost with timolol plus brinzolamide combination) were added to treatment and ocular massage was recommended. The IOP was controlled with above-mentioned therapy (<18 mmHg). Currently, the patient is attending regular control visits.

DISCUSSION

Ectropion uveae is a clinical presentation where iris pigment epithelium is seen on anterior surface of iris and there are two subtypes as congenital and acquired EU. The inflammatory, ischemic or neoplastic processes play role

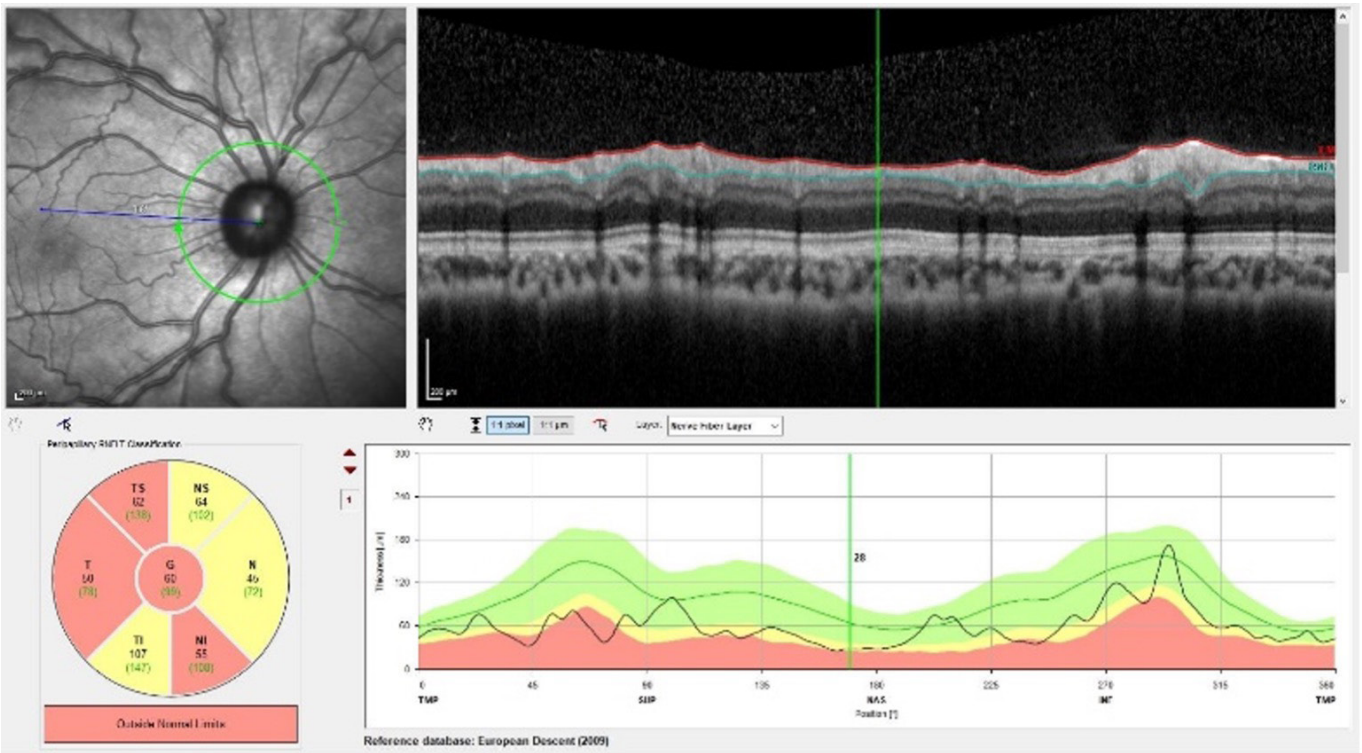

Figure 3: *Thinning in peripapillary retinal nerve fiber layer of the right eye on optical coherence tomography.*

Figure 4: *Ptosis not interfering with visual axis in the right eye.*

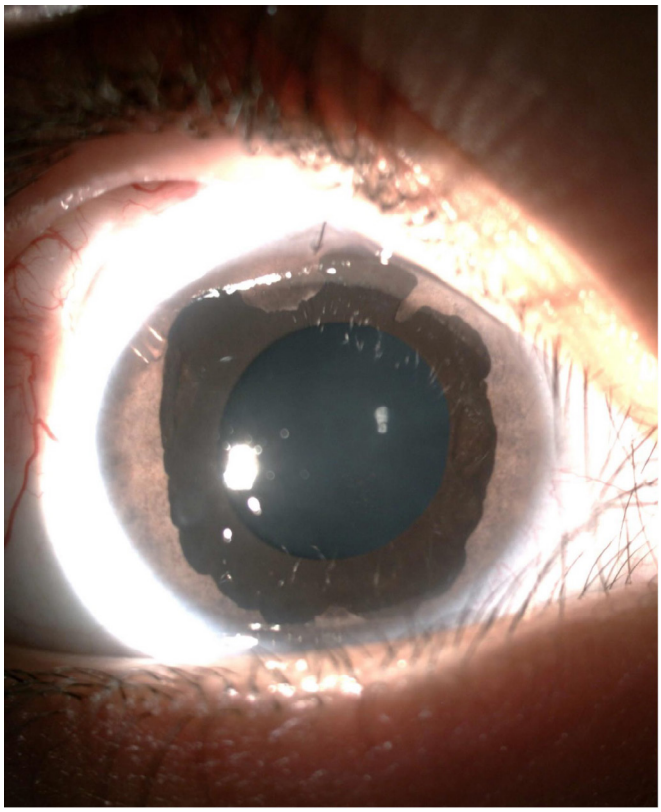

Figure 5: *Ahmed glaucoma valve tube is seen at superior temporal quadrant on postoperative anterior segment image of the right eye.*

in the etiology of acquired EU (e.g. neovascular glaucoma, iridocorneal endothelial syndrome).[4] The CEU is a rare cause of secondary glaucoma, which is characterized by more anterior insertion of iris root, smooth iris surface without crypt, incomplete formation of trabecular network and Schlemm canal, and iridotrabecular angle dysgenesis.[5] It is thought these findings originate from abnormal migration of neural crest cells during intrauterine period. In these patients, the proliferation of iris pigment epithelium is observed on anterior surface of iris.[4] It is thought that iris pigment epithelium hyperplasia is induced by failure in the regression of primordial endothelial cells in anterior chamber due to arrest in the development of neural crest cells during intrauterine period.[6] In histopathological examinations, it was suggested that there is a fibrovascular membrane over anterior stroma of iris and this membrane causes ectropion development by pulling iris pigment epithelium towards anterior surface of iris.[1] In cases with CEU, the age at onset of glaucoma findings and variability in the severity of glaucoma are associated with the degree of arrest in iris migration and malformation of trabecular network.[7]

Glaucoma is a common complication in cases with CEU. Anterior displacement of iris tissue leads incomplete formation of Schlemm canal and trabecular network as well as impaired drainage in iridotrabecular angle.[1] In a study by Dowling et al.,[7] it was observed that 9 of 10 patients developed glaucoma during follow-up. Of these, glaucoma responded to medical therapy in 2 cases while remaining cases were treated with goniotomy, filtration surgery and posterior deep sclerotomy alone or in combination. In a study by Ritch et al.,[8] it was reported that glaucoma was developed in 7 of 8 cases with CEU. There was neurofibromatosis in 3 cases, Prader-Willi syndrome in 1 case and facial hemihypertrophy in 1 case. In addition, it was reported that there was Horner syndrome in contralateral eye in 1 case while Rieger Anomaly was accompanied in another case. Laser trabeculoplasty and trabeculectomy were performed in refractory cases.

In majority of cases with CEU, IOP can be initially

controlled by medical therapy; however, filtration surgery or tube shunt surgery is needed in most cases.[9] The success rate of goniotomy is lower in patients with CEU when compared to primary glaucoma cases.[10, 11] In a previous study, 3 cases with glaucoma secondary to CEU were treated with goniotomy but IOP could not be controlled and trabeculectomy was required. In our case, AGV implantation was preferred as surgery and it was seen that hypertensive phase was developed during follow-up. The hypertensive phase is highly common in AGV patients and IOP reaches the peak value between postoperative month 1 and 3; however, it is recovered within 6 months in general. In the hypertensive phase, the IOP elevation primarily results from capsular fibrosis around AGV plate at postoperative period.[12]

Congenital ectropion uveae may be seen in association with systemic disorders such as neurofibromatosis type 1 or Prader-Willi syndrome. Although genetic inheritance cannot be confirmed, R1748X mutation was detected in NF-1 gene in CEU patients with diagnosis of neurofibromatosis.[13] Ocular malformations such as micro-cornea, microphthalmia, melanosis, multiple telangiectasia and corneal dysplasia can accompany to CEU.[7] In addition, Rieger anomaly, facial hemihypertrophy and ptosis can be seen in CEU cases. It has been proposed that ptosis is caused by neuroectodermal origin of Muller muscle in CEU cases and that levator palpebrae superioris function is normal.[14] In our patient, there are no systemic disorders such as neurofibromatosis.

In conclusion, CEU can lead refractory, secondary glaucoma. These patients should be monitored regularly for glaucoma development.

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