Case Report

A Rare Case Report of Unilateral Congenital Ectropion Uveae (CEU) with Iridotrabecular Dysgenesis, Juvenile Glaucoma and Ptosis

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Abstract

Congenital ectropion uveae is a rare disorder resulting from proliferation of iris pigment epithelium on the anterior surface of iris. We describe a case of a 28-year-old female presenting with diminution of vision in the left eye. She was using topical drugs for high intraocular pressure. On examination, visual acuity was 20/60. There was mild ptosis present along with ectropion uveae, 360 degrees around the pupillary border. Intraocular pressure was 32 mm Hg. On fundoscopy, there was glaucomatous cupping with superior notching of the disc. Gonioscopy revealed an open angle with anterior insertion of iris over trabecular meshwork. Visual field analysis showed inferior arcuate scotoma. Examination of the right eye revealed no abnormality and visual acuity was 20/20. No systemic illness was found. The fall in IOP in the left eye following medical treatment remained unsatisfactory. Hence, trabeculectomy was performed. Following a period of 3 months post trabeculectomy, control of IOP was achieved (15 mm Hg). It is important to evaluate for glaucoma in congenital ectropion uveae cases to prevent blindness in young patients and improve their prognosis.

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Introduction

Congenital ectropion uveae (CEU) or congenital iris ectropion(CIE) is a rare, non-progressive disorder resulting from proliferation of the iris pigment epithelium on the anterior surface of iris. It is considered as a disorder of neural crest cells.[1-5] Usually unilateral, the condition is frequently associated with ipsilateral glaucoma due to iridotrabecular dysgenesis.[6] Neurofibromatosis is the most commonly associated systemic disorder.[3] Filtration surgery often becomes the treatment modality for necessary reduction of IOP.[2] This report describes one such case but without any associated systemic disease.

Case Presentation

A 28-year-old female presented with gradual diminution of vision in her left eye over the past 5 years. She was using topical timolol maleate, dorzolamide and brimonidine eye drops twice daily at presentation. There were no signs of any systemic illness.

On examination, the patient's corrected Snellen acuity was 20/20 in the right eye and 20/60 in the left eye. Ocular motility was full in all cardinal directions of gaze in both eyes. Evaluation of the anterior segment of the left eye showed mild ptosis with good levator function and no mass palpable on the lids (Figure 1). The cornea was clear with normal cell morphology. Iris evaluation showed a loss of normal pattern with ectropion uveae present in the entire 360 degrees around the pupillary border. This ectropion uveae extended to the mid periphery of the iris and was separated from an otherwise smooth and cryptless rest of iris by a definite demarcation border. Also, the rest of iris showed no nodules. The pupil was circular but reacted sluggishly to light. Anterior chamber was normal in depth (Figure 2). Anterior segment evaluation of the right eye showed no abnormalities.

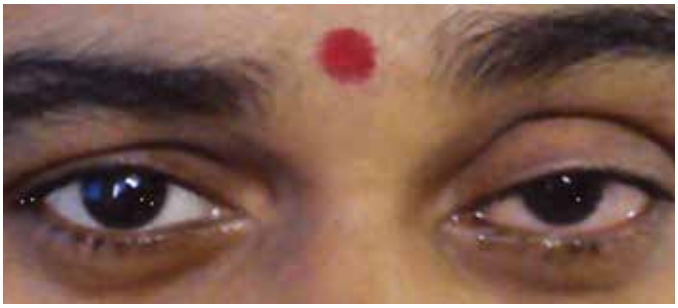

Figure 1: *Mild ptosis of the left eye*

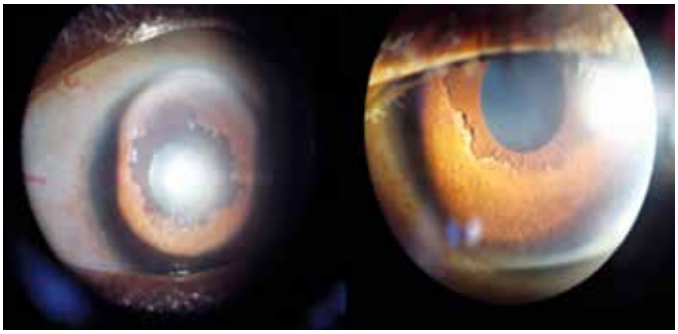

Figure 2: *Ectropion uveae present all around the pupillary border with definite demarcation from rest of the iris*

IOP recorded by applanation tonometry was 18 mm Hg in right eye and 32 mm Hg in left eye. On fundoscopy, the left eye showed glaucomatous cupping with superior notching of the disc, with rest of the fundus being within normal limits. Fundus was normal in the right eye.

Gonioscopy of the left eye revealed an open angle with anterior insertion of the iris over the trabecular meshwork impairing the aqueous drainage at the angle (Figure 3).

Humphrey visual field analysis showed inferior arcuate scotoma in the left eye (Figure 4).

The fall in IOP following medical treatment remained unsatisfactory and hence trabeculectomy was performed on the left eye. Following a period of 3 months post trabeculectomy, the IOP was recorded to be 15 mm Hg.

Figure 3: *Gonioscopy: Open angle with anterior insertion of iris over the trabecular meshwork*

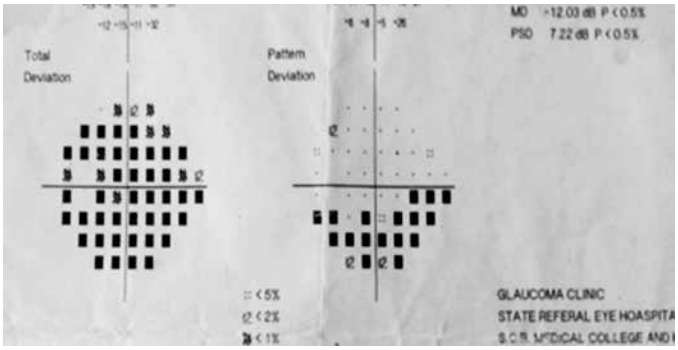

Figure 4: *Humphrey visual field analysis: Inferior arcuate scotoma in the left eye*

Discussion

Ritch et al[3] found glaucoma in seven out of eight cases of CEU, Dowling et al[2] in eight of nine cases, highlighting the frequent association of glaucoma with CEU and necessitating the evaluation of the same in patients of CEU. Ipsilateral mild ptosis with good levator function was present in six of nine cases reported by Dowling et al and one case each reported by Ritch et al and Hertzberg.[6]

The most common association is neurofibromatosis though other ocular and systemic associations have been described, including a chromosomal abnormality but in this case, no systemic association was found.

Conclusion

CEU is a benign ocular abnormality, however its frequent association with glaucoma which can lead to a possible vision threatening irreversible optic neuropathy calls for a detailed evaluation and necessary treatment. Glaucoma is usually refractory to medical treatment and filtration surgery may be necessary. Blepharoptosis is also one of the common associations. Not all cases have systemic associations.

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