Stabilization of Glaucoma Associated with Microcoria: Case Report

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ABSTRACT

We report a case of glaucoma associated with microcoria, stabilized by surgical treatment.

Case report: A 27-year-old male patient consulted in 2015 for hemeralopia that has been evolving since childhood and which was never taken care of.

An examination of the iris revealed significant miosis in both eyes with the right pupil measuring 1.6mm and the left pupil 1.3mm, temporal iris atrophy with exaggerated iris furrows but no transillumination defect. The sphincter was retracted inside reduced to almost half its size and pupils margin were normal. A persistent pupillary membrane arose from the iris collarette, and extended to the nasal half on the right and on 360° on the left, only seen on pupil dilation (Figure 1).

As a result of iris sphincter dysfunction, the patient had poor pupillary light reflex.

The IOP measurements with the Goldmann Applanation Tonometer were 36 mmHg on the right and 38 mmHg on the left.

Trabeculectomy seemed indicated. The right eye required two surgeries and the left eye required one to lower ocular pressures.

Ten years after the first consultation, best corrected visual acuity was 0.1 in both eyes, IOP without treatment was 13 mmHg in the right eye and 17 mm Hg in the left eye.

Conclusion: Glaucoma associated with microcoria usually appears in adolescence. In this paper trabeculectomy proved to be an effective method of lowering intraocular pressure in glaucoma.

Keywords: Microcoria, Glaucoma, Myopia, Trabeculectomy
Introduction

Congenital microcoria is an autosomal dominant disorder characterized by pupil with a diameter of less than 2 mm when looking at a distant object. It is thought to be due to a maldevelopment of the dilator pupillae muscle of the iris [1-3].

Other lesions associated with this pathology are: trabeculodysgenesis [4,5], persistent pupillary membrane [1,6,7], myopia [1,8], and glaucoma [1,2,4].

A genotypic anomaly on 13q31-q32 on chromosome 13 has been frequently associated with this condition [2,3].

Several other genetic forms are described in the literature, such as: autosomal recessive forms [8] and sporadic forms without family history [9].

We report a case of glaucoma associated with a microcoria, stabilized by surgical treatment.

Case Report

The Patient of 27 years old male who consulted for the first time in 2015 for hemeralopia that has been evolving since childhood and which was never taken care of.

Born of twin pregnancy, whose second twin died just after delivery, the patient was, at the time of the consultation, fatherless and motherless. The family survey among collaterals was found no similar case.

Our patient has no history of medication use and no evidence of drug intoxication.

On ophthalmic examination, uncorrected visual acuity was 0.1 in both eyes and best-corrected visual acuity was 0.6 in the right eye, 0.7 in the left eye, with spherical equivalents of -4,5 diopters and -5.5 diopters respectively.

Corneas were clear with no posterior embryotoxon and deep anterior chambers.

Examination revealed significant miosis in both eyes with the right pupil measuring 1.6mm and the left pupil 1.3mm (Figure 1), temporal iris atrophy with exaggerated iris furrows but no transillumination defect. The sphincter was retracted inside reduced to almost half its size and pupils margin were normal. A persistent pupillary membrane arose from the iris collarette, and extended to the nasal half on the right and on 360° on the left, only seen on pupil dilation (Figure 2).

As a result of iris sphincter dysfunction, the patient had poor pupillary light reflex.

The IOP measurements with the Goldmann Applanation Tonometer were 36 mmHg on the right and 38 mmHg on the left.

Gonioscopy showed a irido trabeculodysgenesis is characterized by an anterior insertion of the iris in the superior region, and a grade III open angle in the inferior region.

Instillation of tropicamide eye drops and neosynephrine 10% did not result in pupillary dilation, as did the institution of atropine 1% eye drops for 5 days. Only a small relaxation of the iris sphincter was noted.

Miosis made it hard to perform an adequate examination of the posterior segment.

- B-Mode ultrasound revealed no opacification of the lens, no vitreous condensation, no retinal detachment. The disc could not be seen.
- A-Mode ultrasound revealed an axial length of 26.62 mm in the right eye and 27.11 mm in the left eye, related to the patient’s myopia. A dual therapy (beta-blocker + carbonase inhibitor) was prescribed but the patient did not have insurance prescription coverage and could not afford medication.

Two years later, due to visualacuity degradation, the patient returned to consultation.

Corrected visualacuity was 0.2 in the right eye and 0.1 in the left eye. Intraocular pressure was 38 on the right and 39 on the left. The dynamic visual field at this time showed a concentric narrowing of the isopters less than 20° temporal and 70° nasal (Figure 3).

Trabeculectomy was indicated. The right eye required two
surgery and the left eye required one to lower ocular pressures (Figure 3).

- Genotyping was not feasible given current available technical facility.

Ten years after the first consultation, best corrected visual acuity was 0.1 in both eyes, IOP without treatment was 13 mmHg in the right eye and 17 mmHg in the left eye.

Discussion

Microcoria is usually familial, although sporadic cases have been found in the literature [9]. Since our patient has no living ascendants, it has been difficult for us to find out medical family history.

The patient consulted for the first time for hemeralopia. A similar motive of consultation had been reported by Bremner [6] with absence of mydriasis in scotopic conditions. Consequently, the narrowing of the isopters would be more related to this phenomenon with pupils slightly off-centered. The patient was clinically diagnosed with microcoria based on of less than 2 mm when looking at a distant object. This diameter did not change under the effect of mydriatics and / or cycloplegic, and remained less than 2mm.

The fundamental lesion in microcoria is iris dilator muscle dysfunction. This dysfunction is often associated with lesions of other layers of the iris [10]. As a result, the clinical manifestations of the iris are variable: some patients have normal aspects of the anterior iris leaflet while others have atrophy with iris thinning, and others have iris perforations. The pupillary light reflex maybe absent [9,11], but sometimes normal [5,8].

Iridotrabeculodys genesis, reported in this issue, has previously been described in the microcoria [1,4,5] but there are also patients with completely normal iridocorneal angle [6,8]. Glaucoma and myopia are often associated with congenital abnormalities of the anterior segment [12]. Glaucoma is often linked to goniodysgenesis, responsible for OHT. Moreover, it can lead to higher axial length, notably on corneal-scleral shells that can be expanded during growth between 2 and 3 years [13,14]. Despite the fact that patients present an increase in axial length, this pathophysiological mechanism can hardly explain myopia in case of microcoria, since the average age of glaucoma diagnosis is 20 years [1]. Tawara et al. [4] toodiscuss the notion of “late-onset congenital glaucoma”. IOPs are usually very high, but less than 60mmHg [1].

Toulement [1] has shown that the association between microcoria and glaucoma is statistically significant.

The impact of these two simultaneous anomalies on visual acuity should be appreciated with great discernment. In our case, myopia is responsible for an initial decrease in far-sightedness, which is aggravated by glaucoma. The persistent pupillary membrane found in our patient is a diagnostic element of the microcoria [1,7], but it is not always present [6,8]. Other authors have reported a maldevelopment of iris and the collarette [4,5].

A particular form of microcoria is that associated with diffuse mesangial sclerosis of the kidneys [9,11,15]. In this clinical form, microcorias associated with absent pupillary light reflex, and shows buphthalmia and hypoplasia of the iris dilator muscle, associated with other lesions such as atrophy of the ciliary muscles.

Conclusion

Glaucoma associated with microcoria usually appears in adolescence. Elevated IOP is a critical risk factor for blindness in Glaucoma. In this issue, trabeculectomy proved to be an effective method of lowering intraocular pressure in glaucoma.
References


