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Cas Clinique

Stabilization of Glaucoma Associated with Microcoria

Glaucome associé à une microcorie, stabilisé par un traitement chirurgical

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RÉSUMÉ

Nous rapportons un cas de glaucome associé à une microcorie, stabilisé par un traitement chirurgical.

Observation. Patient de 27 ans qui a consulté pour la première fois, à l'âge de 17 ans pour héméralopie évoluant depuis l'enfance et qui n'a jamais été prise en charge. L'examen de l'iris avait trouvé un important myosis au niveau des deux yeux, une pupille droite mesurant 1,6 mm contre 1,3 mm pour la pupille gauche, une atrophie irienne surtout du côté temporal avec des fentes iriennes radiaires profondes mais sans transillumination irienne évidente. Le sphincter était rétracté en dedans réduite presque à la moitié de sa taille, les bords des pupilles étaient normaux. Un reliquat de membrane pupillaire était présent se fixant sur la collerette, sur la moitié nasale à droite et sur 360° à gauche, mieux visible sur la pupille en dilatation. Le reflexe photomoteur faible n'impliquait que la portion sphinctérienne de l'iris. La pression intraoculaire (PIO) prise au tonomètre à aplanation de Goldmann était à 36 mmHg à droite et 38 mmHg à gauche. L'indication d'une trabéculectomie avait été posé. Il a fallu deux interventions à l'œil droit et une intervention à l'œil gauche pour stabiliser la tension oculaire. Dix ans après la première consultation, l'acuité visuelle avec correction était à 0,1 aux deux yeux ; la PIO sans traitement à 13 à l'œil droit et à 17 à l'œil gauche.

Conclusion. Le glaucome dans la microcorie apparait souvent à l'adolescence, il est souvent grave avec une tension élevée, pouvant conduire rapidement à la cécité. La trabéculectomie peut stabiliser ce glaucome, c'est ce qui s'est vu dans notre cas.

ABSTRACT

We report a case of glaucoma associated with a 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. Examination of the iris revealed significant miosis in both eyes with the right pupil measuring 1.6 mm and the left pupil 1.3 mm, 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 The sphincter was retracted inside reduced to almost half its size and pupils margin were normal.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. As a result of iris sphincter dysfunction, the patient had poor pupillary light reflex.

The intraocular pressure (IOP) measurements with the Goldmann Applanation Tonometer were 36 mmHg on the right and 38 mmHg on the left. Trabeculectomy was a good choice. 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 mmHg in the left eye.

Conclusion. Glaucoma associated with microcoria usually appears in adolescence. In our case, trabeculectomy proved to be an effective method of lowering intraocular pressure in glaucoma.

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 [1,2, 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

A male patient ages 27 years consulted for the first time in 2015 for hemeralopia that had 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 found no similar case.

There was no history of medication use and no evidence of drug intoxication.

Ophthalmic examination, findings were as follows: uncorrected visual acuity at 0.1 in both eyes, best-corrected visual acuity at 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.

Th rest of examination found significant miosis in both eyes with the right pupil measuring 1.6mm and the left pupil 1.3 mm (Fig 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 (Fig 2).

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

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

Gonioscopy showed a iridotrabeculodysgenesis 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 dilatation, as did the institution of atropine 1% eye drops for 5 days. Only a small relaxation of the iris sphincter was noted.

Because of miosis, it was difficult to perform an adequate examination of the posterior segment.

B-Mode ultrasound revealed no opacification of the lens, no vitreous condensation and 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 could not afford medication because of financial limitations (no insurance prescription coverage).

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

Corrected visual acuity 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 (fig 3).

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

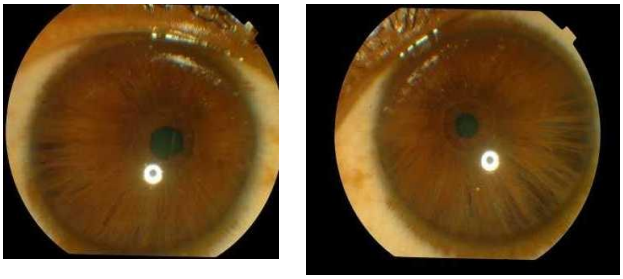

Fig 1. Dilation with atropine eye drops 1%, highlighting the pupillary membrane. Slightly larger dilation in the right eye. Peripheral bilateral and iris atrophy predominantly in the temporal region.

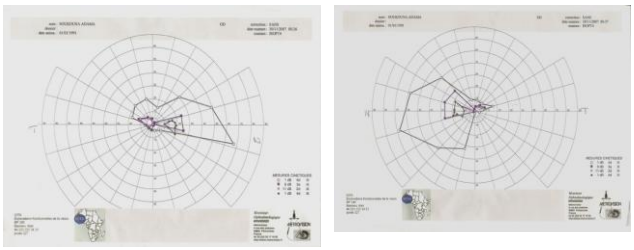

Fig 2. Dynamic visual field shows a concentric narrowing of the isopters less than 20 ° temporal and 70 ° nasal

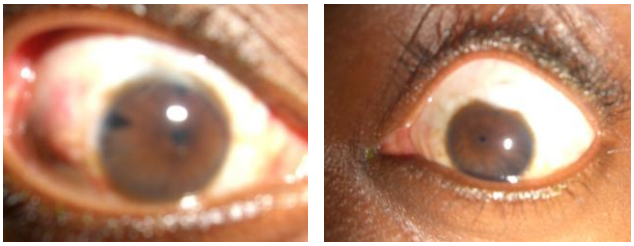

Fig 3. Both iridotomies seen following two trabeculectomies performed in the right eye and one trabeculectomies performed in the left eye.

DISCUSSION

Microcoria is usually familial, although sporadic cases have been found in the literature [9]. Since our patient had no living ascendants, it was 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. On clinical examination, the patient had 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 2 mm.

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 vary: 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 may be absent [9, 11], or normal [5,8].

Iridotrabeculodysgenesis, 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 corneo-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] too discuss 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 abnormal development 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, microcoria is 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.

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