Arquivos Brasileiros de
Oftalmologia

Phacoemulsification and 1% atropine eye drops for treatment of antimetropic congenital microcoria associated with cataracts

Facoemulsificação e colírio de atropina a 1% para tratamento de microcoria congênita antimetrópica associada a catarata

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ABSTRACT | A rare case of bilateral congenital microcoria associated with antimetropia in a 47-year-old man is here described. The patient presented with a chief complaint of progressive vision loss in his right eye over the past five years. A slit-lamp examination and ultrasound biomicroscopy confirmed congenital microcoria and cataracts. Phacoemulsification was performed using an iris expansion device and the anterior capsule was stained using the "trypan down under" technique. Preoperative considerations, the surgical approach, and postoperative management are discussed.

Keywords: Pupil/abnormalities; Cataract; Phacoemulsification; Anisometropia; Atropine

RESUMO | Um caso raro de microcoria congênita bilateral associada à antimetropia em um homem de 47 anos de idade é descrito aqui. O paciente queixava-se de perda visual progressiva em seu olho direito nos últimos 5 anos. Um exame com lâmpada de fenda e biomicroscopia ultrassônica confirmaram microcoria congênita e catarata. A facoemulsificação foi realizada usando dispositivo de expansão iriana, e a cápsula anterior foi corada através da técnica "trypan down under". Considerações pré-operatórias, abordagem cirúrgica e manejo pós-operatório são discutidos.

Descritores: Pupila/anormalidades; Catarata; Facoemulsificação; Anisometropia; Atropina

INTRODUCTION

Congenital microcoria (CM) is a rare ocular entity defined as a pupil diameter of <2 mm[1], with reduced iris thickness on ultrasound biomicroscopy (UBM)[2], and incomplete development of the dilator muscle, as confirmed by histopathological analysis[3].

This condition may be associated with many ocular disorders, although its association with antimetropia has not previously been reported.

CASE REPORT

A 43-year-old man presented with a chief complaint of progressive blurred vision in both eyes over the past five years. He was diagnosed with bilateral amblyopia, markedly of the right eye, and had been wearing a -10.00D contact lens on his left eye for a decade. Previous best corrected visual acuity (BCVA) was 20/400 and 20/30. Yttrium-aluminum-garnet-laser iridotomy was performed because of a bilateral shallow anterior chamber (AC). There was no other remarkable medical history or use of medications.

The subjective refractive errors of the right (OD) and left (OS) eyes were +13.00 + 2.00 × 35° and -7.75 + 2.25 × 90°, with BCVA of hands motion and 20/100, respectively. Keratometry (K1 and K2) were as follows: 38.25 × 53° and 40.25 × 143° for OD and 41.00 × 178° and 43.25 × 88° for OS. The AC was uniformly shallow. Pinhole pupils (about 1.6 mm in diameter) with an absence of crypts, and iridotomies at 9 o'clock in the right eye and at 2 o'clock in the left eye were observed (Figures 1A and E). Pupil light reflexes were poor. Intraocular pressure by applanation tonometry of the OD and OS was 6 and 12 mmHg, respectively. Significant

Submitted for publication: July 24, 2018
Accepted for publication: January 29, 2019

Funding: No specific financial support was available for this study.

Disclosure of potential conflicts of interest: None of the authors have any potential conflicts of interest to disclose.

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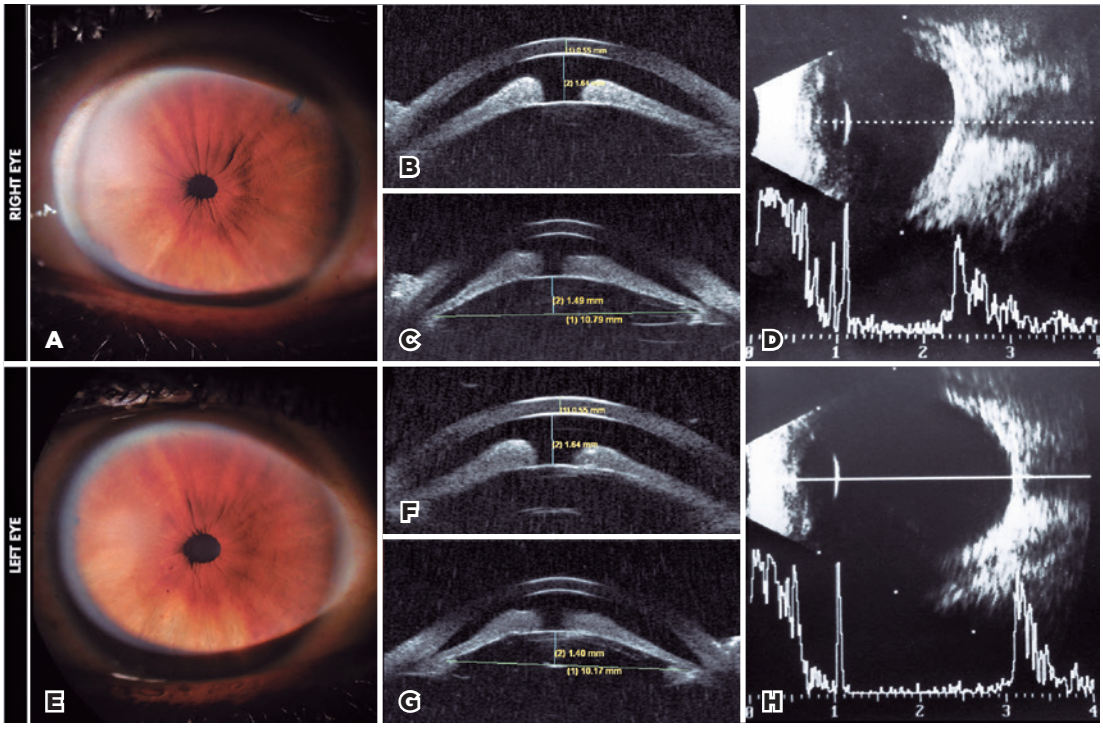

Figure 1 Preoperative assessment of both eyes. (A and E) Slit lamp evaluation showing microcoria (pupil size <2 mm). AC and iris thickness were observed on UBM, which highlights (B and F) AC depth and (C and G) lens measurements. (D and H) Ocular US showing differences in axial lengths

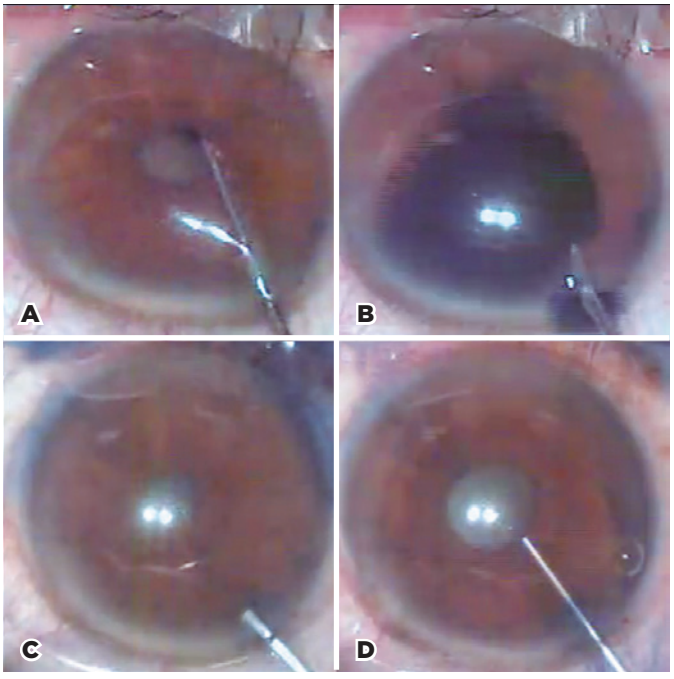

Figure 2. Steps of the TDU staining technique. (A) Cannula placement just under the iris; (B) trypan blue injection; (C) dye washout; (D) finally, viscoelastics are injected into the AC

cataractous lenses were observed in both eyes, thus an ecography study was performed.

UBM showed small pupil diameters, hypoplastic irises (stromal thickness of 550 µm), and shallow ACs (Figures 1B and F). Crystalline lens rise (CLR) of the right and left eyes was 1,490 and 1,400 µm, respectively (Figures 1C and G). On posterior segment ultrasound (US), the left eye had a larger axial length (AL) than the right eye (Figures 1D and H). Partial coherence interferometry (IOL Master®) showed an axial length of the right and left eyes of 20.78 and 29.49 mm, respectively, and an intraocular lens (IOL) predicted power for emmetropia of +36.00 and +9.00, respectively. At this point, a diagnosis of CM associated with cataract was made, and phacoemulsification was indicated.

Cataract surgery was performed in both eyes on different days through a 2.75 mm temporal incision using the "trypan down under" (TDU) staining technique (Figures 2A-D). Afterwards, a 6.25-mm Malyugin Ring® (Micro-Surgical Technology, Redmond, WA, USA) was inserted

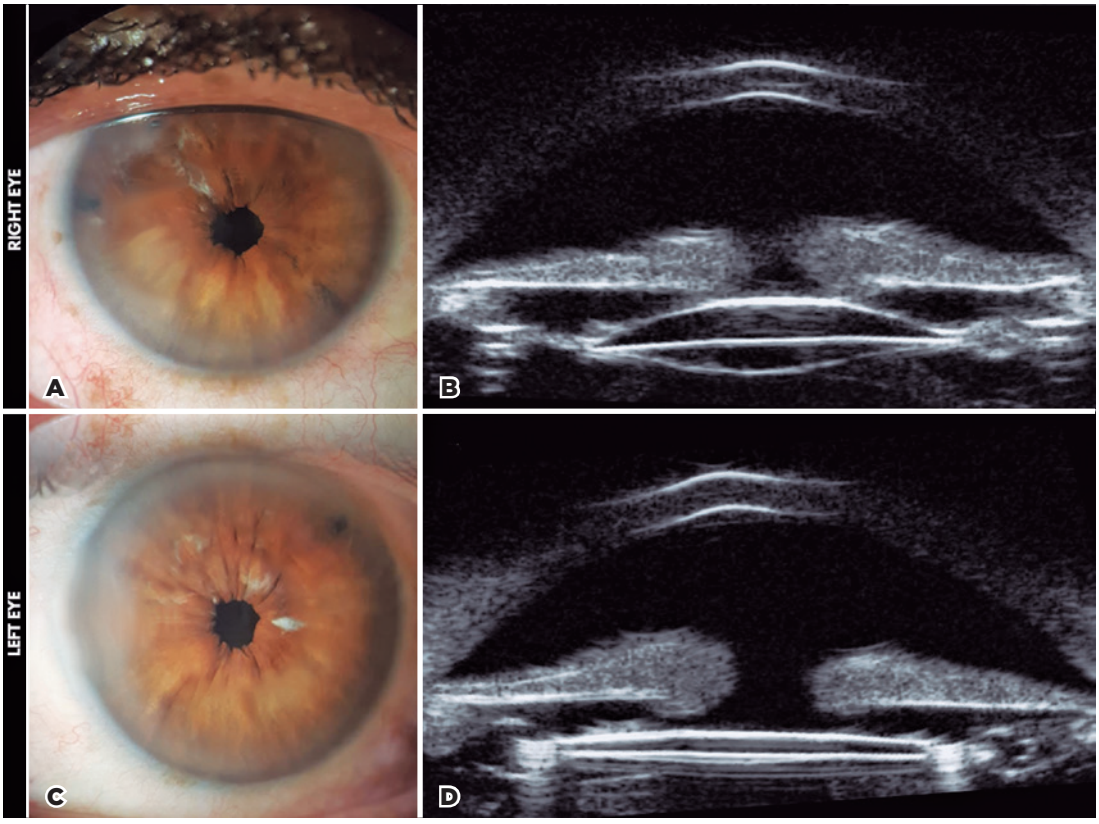

Figure 3. Postoperative assessment of both eyes. (A) On slit lamp, focal areas of iris atrophy can be observed; (B) UBM shows an increase in AC depth, although pupil diameter remained small.

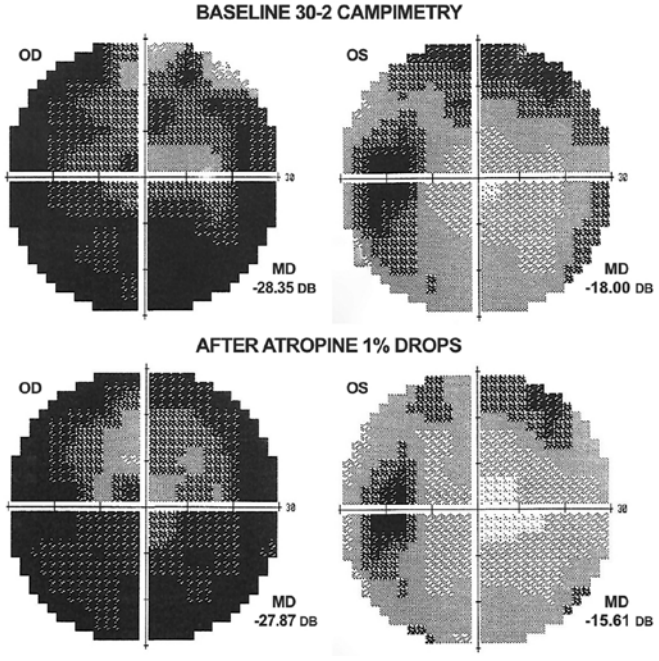

Figure 4. Postoperative Humphrey VF of both eyes. The use of 1% atropine eye drops improved the VF with a reduction in peripheral scotomas.

into the AC, to maintain an anatomic pupil. These steps were followed by capsulorrhexis, quick-chop phacoemulsification, and implantation of an Alcon Acrysof® MA60AC IOL (OD, +30.00; OS, +9.00). Enlargement of pupil size to 2.00 (OD) and 1.6 mm (OS), and AC deepening (Figures 3A and B) were observed.

Within one month, once-daily 1% atropine eye drops were prescribed, which resulted in an increase in pupil size of 2.5 mm (OD) and 2.0 mm (OS). This anatomical response was followed by visual field (VF) improvement (Figure 4). His BVCA improved to 20/400 (+3.00 + 2.00 × 75) and 20/30 (-0.25 + 2.25 × 50), without symptoms of binocular vision dysfunction.

DISCUSSION

CM is a rare ocular condition, first described by Holth in 1923[4], which is defined as a pupil diameter of less than 2 mm when looking at a distant point. This anomaly is caused by a mutation at 13q31-q32 and usually

has autosomal dominant transmission, although it also can be sporadic, as with our patient. It also has no sex predilection[1].

The etiology of CM includes intermediate filament defects during differentiation of the iris anterior pigmented epithelium, resulting in the absence of stromal myofilaments and dysfunction of the pupil dilator muscle, which leads to a static pupil diameter of less than 2 mm, poor development of the crypts and collarette (displaced toward), peripheral iris hypopigmentation, and transillumination defects. Affected individuals have poor mydriasis associated with nyctalopia, proportional to the pupillary area[5].

UBM allows to confirm CM and evaluate the status of the posterior capsule[2]. Histopathologically, CM is associated with incomplete development of the dilator muscle with a significant reduction in the number of smooth muscle cells, but with no effect on innervation or vasculature[3].

CM is also associated with axial myopia, cataract, astigmatism, goniodysgenesis, and glaucoma (up to 30%)[6]. Our patient had no evidence of glaucoma, even though he presented with amblyopia, cataract, and antimetropia with hyperopic and myopic refractive errors.

Antimetropia is a peculiarity of this case. For the hyperopic eye, a partial coherence interferometry biometer and the Holladay II formula were preferred for IOL power calculation, as they result in a lower predicted postoperative refractive error than ultrasound biometry and other formulas[7]. For the myopic eye, the Barrett II Universal formula was preferred.

CLR is defined as the distance between a straight line joining the iridocorneal angle and the anterior face of the lens, with the use of UBM. This distance is used to predict the risk of pigment dispersion related to intraocular lens: the risk is lower if less than 600 µm, and up to 67% if equal to or greater than 600 µm. Higher CLR values are usually found in hyperopic eyes, which implies a lens component in the angle closure mechanism[8]. Our patient presented with CLR of 1,490 µm (OD) and 1,400 µm (OS) (Figures 1C and F), thereby explaining the unexpected shallow AC in the myopic eye and the angle opening after cataract surgery (Figure 3).

Phacoemulsification with small pupils is challenging. In our case, we decided on the TDU staining technique (Figures 2A-D), during which a dye is injected under the iris and is then washed out just before insertion of viscoelastic and a Malyugin Ring®[9]. These steps can improve intraoperative visualization and the safety of cataract extraction.

Cases in which the pupils are exceptionally small, especially <2 mm, may result in experience of VF loss. Atropine is a competitive antagonist of muscarinic acetylcholine receptors, which can enlarge pupils and improve VF in such cases[4]. As it results in cycloplegia, it is indicated for myopic or presbyopic eyes (for emmetropia or hyperopia, pupilloplasty should be considered). Generally, the long-term use of 1% atropine eye drops is well tolerated by the patients. However, treatment should be changed to 2% scopolamine if drowsiness or red eye occur. In our case, the pupils were enlarged about 25% and the mean deviation increased from -28.35 to -27.87 DB (OD) and from -18.00 to -15.61 DB (OS) (Figure 4).

To the best of our knowledge, this is the first report of antimetropia being associated with CM. Phacoemulsification and the use of atropine eye drops seem to be both safe and feasible in such cases.

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