Short communication

Bilateral persistent hyaloid artery. A case report

A.M. Borbolla-Pertierra a,*, C.K. Martínez-Hernández b, J.C. Juárez-Echenique a

a Instituto Nacional de Pediatría, Mexico City, Mexico
b Hospital General Manuel Gea González, Mexico City, Mexico

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A B S T R A C T

Case report: A 5-year-old male presented with bilateral poor vision, esotropia and a previous diagnosis of cataract since he was 1 year old. The physical examination revealed bilateral posterior paracentric capsule opacification, vitreous cavity with a permeable pulsatile blood filled hyaloid artery in both eyes. He was kept under observation.

Discussion: Persistent hyaloid artery is an uncommon faulty primary vitreous regression, often unilateral (although it may be bilateral) and sporadic, associated with microphthalmos. It may be complicated with glaucoma and phthisis bulbi. Vitrectomy plus lensectomy or simple observation is the accepted treatment option.

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P A S T E N C l a i r e:

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P S E R T E N C i a b i l a r d e l a a r t e r i a h i a l o i d e a .   R e p o r t e d e u n c a s o

RESUMEN

Caso clínico: Varón de 5 años de edad con mala visión bilateral, endotropía y diagnóstico previo de catarata desde el año de edad. En la exploración, se observa en ambos ojos opacidad capsular posterior paracentral, cavidad vitrea con arteria hialoidea permeable, ocupada por sangre y con pulso. Se dejó en vigilancia.

Discusión: La persistencia de la arteria hialoidea es infrecuente y es una falla en la involución del vitreo primario. Comúnmente es unilateral (aunque hay casos bilaterales), esporádico y asociado a microftalmos. Puede complicarse con glaucoma y phthisis bulbi. Vitrectomía más lensectomía u observación son opciones de tratamiento según el caso.

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* Corresponding author.
E-mail address: ambp09@gmail.com (A.M. Borbolla-Pertierra).

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Case report

Male patient, age 5, whose mother reported having chickenpox in the fifth month of pregnancy, consulted for poor vision, inward ocular deviation and bilateral cataract since he was 12 months old.

Physical examination produced a visual acuity of no light perception in right eye (RE) and 20/200 in left eye (LE). The following were observed: Refraction: RE +2.00 with −7.25 × 90, LE +1.00 with −5.25 × 70, without achieving visual improvement; endotropia of 25 prismatic dipters, normal duction and version; ocular posterior capsular opacity in both lenses and retrolental tissue in RE (Fig. 1); open iridocorneal angles and intraocular pressure of 15 mmHg in both eyes (BE); vitreous cavity of BE with functional vascular canal from the papilla to the lens, presence of pulse, small and pale optic nerves with coloboma-type defect, hypopigmented macular chorioretinal scar of about 2 papillar diameters in RE (Figs. 2 and 3).

Fluorangiography: fetal vessels filled with fluorescein, demonstrating functionality (Figs. 4 and 5).

Magnetic resonance: hypotensive central hyaloid canal in T2 projection (Fig. 6). It was decided to maintain the patient under observation.

Discussion

The persistence of fetal vessels, also known as persistence of hyaloid artery or persistent hyperplastic primary vitreous (PHPV), is a rare congenital anomaly generally of unknown cause. Most cases exhibit retrolental plate in a microphthalmic eye. It was initially described as a syndrome by Reese in 1955.

The defect consists in failed primary vitreous regression. The hyaloid artery is formed on the basis of mesenchymatous cells which gained access through the optic fissure. This artery surrounds the lens contributing to form the tunica vasculosa lentis. Complete primary vitreous regression must take place between the seventh and eighth month of gestation. It is anatomically classified on the basis of anterior or posterior primary vitreous persistence.

PHPV is the second most common cause of acquired cataract during the first year of life. It is most frequently diagnosed during the first year of life and no relevant familial history has been identified.

With greater frequency it is unilateral although it can express bilaterally in up to 11% of cases and is associated to microphthalmos in 2/3 of patients.

It expresses mainly with leukocoria, which requires differential diagnosis with cataracts, retinoblastoma, retinopathy of prematurity, posterior uveitis and Coats disease, among others.

Natural history is toward recurring vitreous hemorrhage, retina detachment, uncontrollable secondary glaucoma and phthisis bulbi, for which reason its only treatment on some occasions is enucleation.

Diagnostic can be clinical. However, A-B mode echography is essential particularly in the presence of opacity, exhibiting a stem from the posterior pole to the lens, in addition to discarding or confirming the presence of microphthalmos or associated retina detachment. Color Doppler echography helps to confirm the vascular nature of the lesion.
Computerized tomography is useful for discarding calcification although this is extremely rare in PHPV. Magnetic resonance (MR) enables a clearer image of PHPV and is excellent for differential diagnostics. In MR, the retrolental membrane is hyperintense after the application of contrast and shows a tube-shaped image which represents the hyaloid vessel.

Treatment is controversial and has three objectives: (1) to prevent glaucoma, (2) pupil esthetics and (3) visual improvement. Pollard and Haddad reported long series including vitrectomy and lensectomy in some cases, concluding that the patients who only exhibited the anterior primary vitreous persistence variety have good results, in contrast with those having the posterior variety who, despite all efforts, did not obtain visual improvements and exhibited complications with greater frequency. Mittra concluded that good results are obtained with the latest vitreoretinal techniques for surgery and intensive treatment for amblyopia. Sisk treated 81 cases with surgery and observed greater incidence of glaucoma in operated patients.

There are very few reported bilateral cases. Hunt reported five patients with bilateral anomaly out of 55 with PHPV, and reported only two patients with isolated hyaloid artery as sole expression of PHPV, who were managed conservatively. Said author also recommended avoiding surgery when the visual axis is free, anatomic anomalies are not progressive and the chamber angle is not compromised. In turn, Kumar reported 11 cases of bilateral fetal vessel persistence and indicated this could represent a separate clinical entity.

In the case reported herein it was decided to treat the patient conservatively due to the presence of blood flow in the remaining vessel (vitreous hemorrhage risk), absence of progression during the observation period, lack of alteration of iridocorneal angles and the certainty of poor visual prognosis (RE macular scar, LE amblyopia).

**Conflict of interest**

No conflict of interest was declared by the authors.
REFERENCES