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Short communication

Unilateral persistent fetal vasculature coexisting with anterior segment dysgenesia☆

B. Cañizares, I. Yago, Á. Piñero, M. Ruiz*

Unidad de Oftalmología Infantil, Servicio de Oftalmología, Hospital Virgen de la Arrixaca, Murcia, Spain

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ABSTRACT

Case report: A case is presented of a 4 week-old female neonate with Peters anomaly (PA) and unilateral persistent foetal vasculature (PFV) referred to our centre due to esotropia. At 12 weeks of age, a penetrating keratoplasty and vitrectomy were performed without major complications in the immediate post-operative period. The patient is currently under an intensive treatment for amblyopia and secondary glaucoma.

Discussion: Surgical treatment of PFV is controversial, with prevention of amblyopia, phthisis, and glaucoma being the main reasons for it. Patients with unilateral PFV and type II PA could be good candidates for this combined surgical procedure.

Vasculatura fetal persistente unilateral asociada a disgenesia del segmento anterior

Palabras clave:
Persistencia vasculatura fetal
Anomalía de Peters
Queratoplastia penetrante
Vitrectomía

RESUMEN

Caso clínico: Presentamos un caso clínico de una neonata de 4 semanas con anomalía de Peters (PA, por sus siglas en inglés) y persistencia de vasculatura fetal unilateral (PFV, por sus siglas en inglés), remitida a nuestro servicio con el diagnóstico de esotropia. A las 12 semanas de vida se realizó una queratoplastia penetrante y vitrectomía, sin complicaciones relevantes en el postoperatorio inmediato. Actualmente la paciente se encuentra bajo tratamiento intensivo de ambliopía y glaucoma secundario.

Discusión: El tratamiento quirúrgico de la PFV es controvertido, siendo la prevención de ambliopía, *phthisis* y glaucoma las principales razones que lo motivan. Los pacientes afectos de PFV unilateral y PA tipo II, podrían ser buenos candidatos para un procedimiento quirúrgico combinado como el descrito.

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* *Corresponding author.*
E-mail address: belen.nse@gmail.com (B. Cañizares).

Introduction

Persistent foetal vasculature (PFV) is a common congenital anomaly in the development of the eyes derived from failed regression of the primary vitreous and the hyaloid vasculature during the embryonary period.[1] It is classified as anterior, posterior and mixed depending on the location of ocular involvement,[2] and is characterized by the presence of a vascular stem between the optic disc and the posterior capsule of the lens. Peters anomaly (PA) is a rare congenital malformation of the anterior segment, characterized by the presence of a central corneal opacity having variable size and density.[3] PA is classified in 3 types: types I, II and Peters plus depending on the involved structures in the anterior segment and/or the association of extraocular involvement.[4] The association of PFV with PA is extremely rare and significantly worsens prognosis. Surgical treatment and post-operative of an PFV case associated to PA type II is presented.

Clinic case report

Newborn, 4 weeks old, referred to Ophthalmology due to microphthalmos and convergence strabismus in the right eye (RE). Relevant personal antecedents included the presence of chromosome 22 deletion (22q11).

In the examination, the patient exhibited microphthalmos and fixed endotropia in the RE, the biomicroscopy of which revealed the presence of central adherent leukoma with iris-corneal-lenticular synechiae (Fig. 1). Funduscopic examination was not possible due to medium opacity. Mode-B echography showed athalamia and hyper-ecogenic central mass from the optic nerve to the posterior pole of the lens (Fig. 2), without vascular structures inside. Flash evoked potentials and flash electroretinogram studies did not show significant asymmetries between both eyes. Examination of the opposite eye produced normal results. A diagnostic of unilateral type II PA was established.

It was decided to perform penetrating keratoplasty at 12 weeks of life. After surgery, a new ecograph-Doppler showed an arterial structure within the hyper-ecogenic mass (Fig. 3) that confirmed the PFV diagnostic. A 23-gauge vitrectomy was performed. The fibrovascular stem was separated from its adhesion to the elongated ciliary processes (Fig. 4) and, after the application of diathermy, vitrectomy was completed (Fig. 5). No relevant complications appeared in the postop.

One year after surgery, the patient exhibits transparent graft without signs of rejection, aphakia and a morning

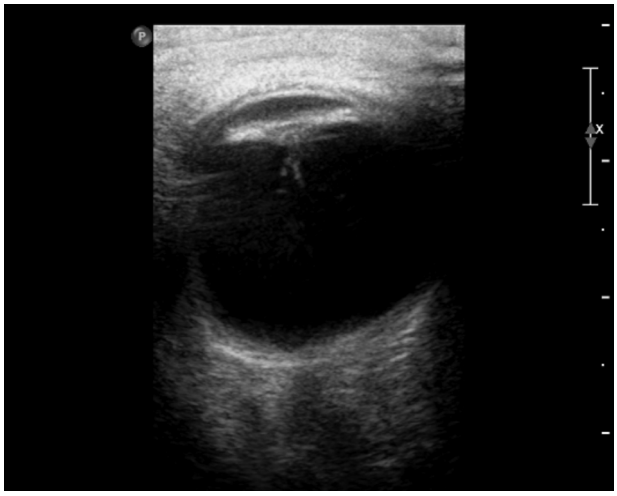

Fig. 2 – Mode-B echography (pre-transplant): athalamia and central hyper-ecogenic mass from the optic nerve to the posterior lens capsule, without evidencing associated vascular structures.

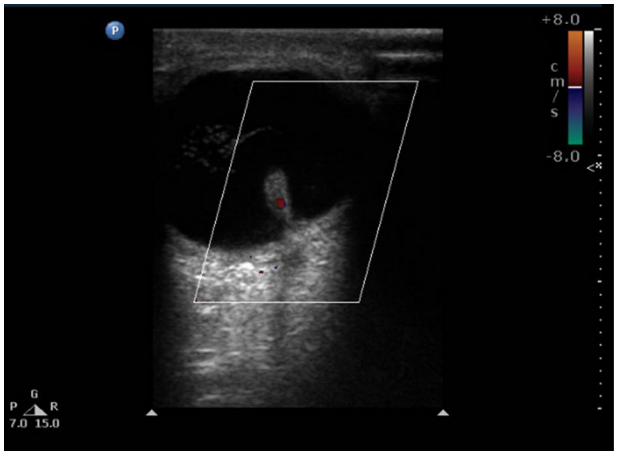

Fig. 3 – Doppler Ecography (post-transplant): vascular structure within hyper-ecogenic mass.

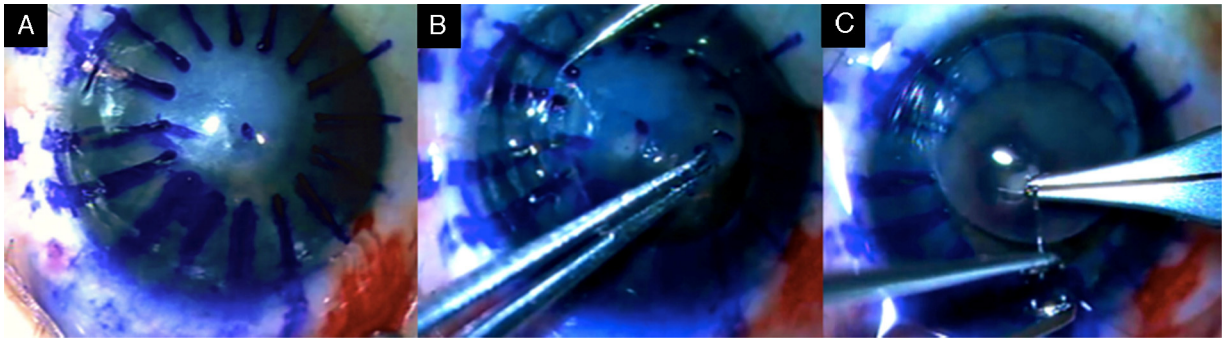

Fig. 1 – (A) Corneal leukoma. (B) Extraction of pathological corneal button and release of iris-corneal-lenticular synechiae. C) Suture of donor corneal button.