Polskie Towarzystwo Okulistyczne
www.pto.com.pl

KLINIKA OCZNA 2022, 124, 1: 36–39
Received: 22.03.2021 Accepted: 4.04.2021

CASE REPORT

Access this article online

Website:
www.klinikaoczna.pl

DOI:
https://doi.org/10.5114/ko.2022.114993

Congenital membranous cataract surgery in an 11-week-old child with persistent fetal vasculature: a case report

Wojciech Lubiński[1], Marta Kirkiewicz[2], Monika M. Modrzejewska[1], Ewelina Soszka-Przepiera[2]

[1]II Clinic of Ophthalmology, Pomeranian Medical University, Szczecin, Poland
[2]Independent Public Clinical Hospital No. 2 in Szczecin, Poland

ABSTRACT

The report presents the surgical management of membranous cataract associated with persistent fetal vasculature. A routine ophthalmological examination in a 3-month-old infant revealed total cataract in the left eye. Persistent hyperplastic vitreous body was visualized on B-scan ultrasound. Under general anesthesia, the cataract was removed through a 1.2 mm corneal microincision, combined with the removal of the fibrovascular retrolenticular membrane and excision of the persistent vitreous artery, anterior vitrectomy, and iridectomy. Early diagnosis and surgical management allow further diagnosis and rehabilitation of the eye with persistent fetal vasculature.

KEY WORDSE: congenital cataract, membranous cataract, persistent fetal vasculature

INTRODUCTION

Persistent fetal vasculature (PFV) occurs as a result of prolonged involution of fetal blood vessels supplying the lens, retinal and choroidal structures, and the optic nerve (CN II) during embryonic development [1]. Residual PFV is observed in over 90% of infants born below 36 weeks' gestation and in 95% of infants with a birth weight below 2,300 g [2]. Complications associated with PFV occur as a result of eye growth rather than vascular progression and proliferation, and include glaucoma, cataract, intraocular hemorrhages, retinal detachment, and optic atrophy [3]. PFV is the second most common cause of white pupillary reflex (leukocoria) after retinoblastoma [4]. It represents a major cause of visual impairment and visual disability [3].

Most cases of PFV are sporadic, however autosomal recessive (AR) and autosomal dominant (AD) patterns of familial transmission have been documented in approximately 10% of patients. The causative gene responsible for the majority of diagnosed cases has not yet been identified. The genes known to be implicated in the pathogenesis of PFV include ATOH7 (AR PHPV) and NDP (AD PHPV) [3]. Bilateral PFV may be linked to systemic diseases (Norrie disease, Aicardi syndrome, and Walker-Warburg syndrome) [5].

Based on the adopted classification, PFV is divided into anterior, posterior and combined (anterior and posterior) subtypes. Anterior PFV accounts for approximately 25% of cases and is associated with the best visual outcomes. Characteristic clinical findings in anterior PFV include a shallow anterior chamber, dilated iris vessels, narrow pupil, microphthalmia, leukocoria, a varying degree of cataract, elongated or drawn-in ciliary processes, retrolental fibrovascular membranes causing traction on the peripheral retina, hemorrhages, secondary glaucoma, strabismus, ectropion uvea, and coloboma iridis. Posterior PFV, accounting for approximately 12% of cases, is associated with poorer outcomes and involves primarily the vitreous and retina. Fundus findings in the posterior subtype have been reported to include an area of glial tissue extending from the CN II to the lens, macular pigmentary disruption with hypoplastic macula, retinal folds, dysplasia, detachment, and CN II hypoplasia. Other abnormalities may include leukocoria, microphthalmia, and strabismus. The lens remains clear. The combined subtype accounts for approximately 60% of all cases of PFV [6], with about 40% of patients in this group losing light perception if surgery is not undertaken [7]. This type of PFV may be complicated by corneal opacity, angle-closure glaucoma, and spontaneous intraocular bleeding [7].

Since membranous cataract (*cataracta membranacea* – CM) associated with PFV is a rare occurrence, there is no algorithm for surgical management. Consequently, each individual case is challenging and requires an anticipatory management approach with possible departures from the recognized procedures. Below, we report on the treatment of a patient with CM associated with PFV at the authors' center.

AUTOR DO KORESPONDENCJI
Prof. Wojciech Lubiński, II Katedra i Klinika Okulistyki, Pomorski Uniwersytet Medyczny w Szczecinie, al. Powstańców Wlkp. 72, 70-111 Szczecin, e-mail: lubinski@pro.onet.pl

CASE REPORT

An 11-week-old infant was admitted to the 2nd Department of Pediatric Ophthalmology at the Independent Research and Teaching Hospital No. 2 in Szczecin, Poland, for left eye cataract surgery. The medical history showed that the caregivers observed white pupillary reflex in the infant's left eye at the age of three weeks. The baby was born by caesarean section at 38 weeks' gestation due to maternal causes (status post caesarean section in previous pregnancy), with a body weight of 3,190 g, in good general condition.

On admission, routine ophthalmological examination was performed, confirming the reflex of following the light stimulus in both eyes, and normal anterior and posterior segments in the right eye. The findings in the left eye included narrow pupil and total cataract. The clinical picture was consistent with cortical cataract. B-scan ultrasound of the left eye showed hyperechoic shadows extending from the posterior lenticular surface to the optic disc, likely consistent with persistent vitreous artery.

The cataract removal surgery was performed under general anesthesia. Shallow anterior chamber and dilated iris vessels were found. Mydriasis could not be obtained. The anterior chamber was opened by performing a clear corneal incision at 2:00 and 10:00 o'clock with a 1.2 × 1.4 mm knife. Adrenaline and viscoelastic (sodium hyaluronate 1%) were administered. Since an attempt to apply the needle capsulorhexis technique was unsuccessful, anterior diathermic high-frequency capsulorhexis was performed, revealing that the lens had been resorbed to a considerable extent. The residual cortical masses were removed with an irrigator (Figure 1A). In the next step, a diathermic high-frequency capsulorhexis probe was applied to create a 5 mm circular opening in the fibrovascular membrane (Figure 1B). The vitreous artery, adjacent to the posterior portion of the membrane, was visualized. Using diathermy, it was coagulated and severed with microvitrectomy scissors (Figure 1C-D). Subsequently, triamcinolone-assisted anterior vitrectomy was performed (with triamcinolone administered into the anterior chamber) (Figure 1E), and Miostat (carbachol) was applied to achieve pupillary constriction (Figure 1F). In the next step, parabasal iridectomy was carried out. The procedure was completed with corneal hydration, injection of air, and administration of antibiotic to the anterior chamber.

The child was discharged home on day 3, and the caregivers were instructed to administer dexamethasone, levofloxacin, and mydriatics to the conjunctival sac. A follow-up examination was scheduled in three weeks. After this period, medical recommendations included appropriate contact lens selection and occlusion of the healthy eye for most of the child's active time (i.e. uncovering the healthy eye for 1-2 hours).

DISCUSSION

This rare case of membranous cataract associated with PFV highlights a number of important issues related to the diagnosis, treatment, and subsequent therapy of patients with this condition. PFV is not always diagnosable before cataract surgery. It is common for fetal vascularization to be found during the procedure [1]. In the case reported here, PFV was suspected on the basis of ophthalmic ultrasound findings during initial diagnostic workup. The prevalence of PFV in patients with congenital cataract reaches up to 44% [1]. Therefore, the surgical team should be prepared to perform a non-standard procedure in every patient diagnosed with this pathology.

Membranous cataract associated with PFV has a low prevalence. In their study of 31 patients undergoing surgery for congenital cataract associated with PFV, Müllner-Eidenböck *et al.* [8, 9] did not identify any cases of CM. In another paper, Haddad *et al.* [10] described two cases of CM among 62 patients with PFV. There are also isolated case reports of patients with membranous cataract associated with PFV [11]. CM associated with PFV develops as a result of pulling on and eventually damaging the lens capsule [12, 13], which is followed by the absorption of lenticular masses. The pulling effect arises in the process of growth of the eyeball. This pathomechanism seems highly likely in the patient reported here. The small amount of cortical masses found during the procedure and the presence of the vitreous artery seem to confirm this hypothesis. Removal of the fibrovascular membrane by means of thermal capsulotomy device accompanied by posterior capsulorhexis with excision of the vitreous artery, as in the case reported here, seem to be of key significance for fundal assessment and further rehabilitation [11].

The patient's final visual acuity depends on several factors including the duration of clinically significant cataract, progression of PFV lesions, clarity of the optical media, postoperative complications, and the patient's cooperation throughout further treatment of visual impairment. Patients undergoing surgery before 77 days of age have been found to be 13 times more likely to have visual acuity of counting fingers or better [14]. Therefore, early diagnosis and treatment of such patients are critical to prognosis. Cooperation with parents is another factor determining the child's final visual acuity outcomes. The duration of occlusion of the dominant eye in unilateral cataract cases should be adjusted according to the degree of lens opacity [15]. In total cataract cases, the eye must be patched for most of the day. After cataract surgery, children require regular monitoring. At each follow-up visit, intraocular pressure measurement and fundus assessment should be performed. These precautions are necessary, as secondary glaucoma is expected in 4% of children with unilateral and 9% of children with bilateral cataract associated with PFV [1] at one-year follow-up.

In summary, prompt diagnosis, surgical procedure performed by an experienced team, as well as regular follow-up and cooperation with parents may contribute to achieving useful visual acuity in patients with membranous cataract associated with PFV.

OŚWIADCZENIE

Autorzy deklarują brak konfliktu interesów.

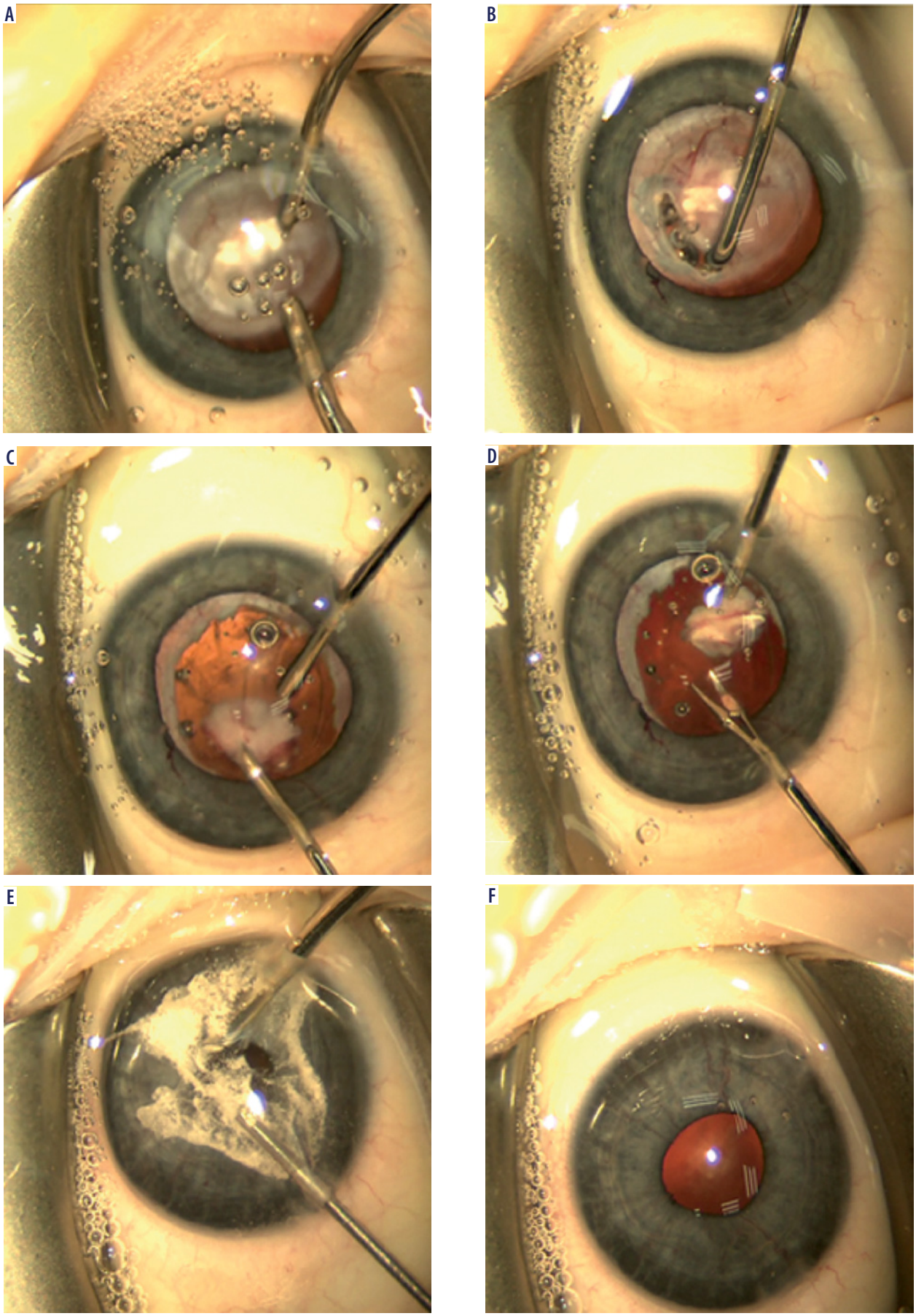

Figure 1. Consecutive steps in membranous cataract removal surgery: A) aspiration-irrigation of residual cortical masses, B) posterior capsulotomy performed with a diathermic high-frequency capsulorhexis probe, C) coagulation of the vitreous artery, D) excision of the vitreous artery, E) triamcinolone in the anterior chamber, F) image of the anterior segment on completion of surgery

References

1. Solebo AL, Russell-Eggitt I, Cumberland P et al. Congenital cataract associated with persistent fetal vasculature: findings from Io-Lunder2. Eye (Lond) 2016; 30: 1204-1209.
2. Singh S, Hasanreisoglu M, Pointdujour R et al. Stunning Visualization of Fetal Hyaloid Artery. Retina Today 2015: 44-46.
3. Madison Duff S, Ramasubramanian A. Persistent hyperplastic primary vitreous. Dostępne na: URL https://eyewiki.aao.org/Persistent_hyperplastic_primary_vitreous.
4. Payabvash S, Anderson JS, Nascene DR. Bilateral persistent fetal vasculature due to a mutation in the Norrie disease protein gene. Neuroradiol J 2015; 28: 623-627.
5. Apushkin MA, Apushkin MA, Shapiro MJ et al. Retinoblastoma and simulating lesions: role of imaging. Neuroimaging Clin N Am 2005; 15: 49-67.
6. Vasavada AR, Vasavada SA, Bobrova N et al. Outcomes of pediatric cataract surgery in anterior persistent fetal vasculature. J Cataract Refract Surg 2012; 38: 849-857.
7. Chen C, Xiao H, Ding X. Persistent Fetal Vasculature. Asia Pac J Ophthalmol (Phila) 2019; 8: 86-95.
8. Müllner-Eidenböck A, Amon M, Moser E, Klebermass N. Persistent fetal vasculature and minimal fetal vascular remnants: a frequent cause of unilateral congenital cataracts. Ophthalmology 2004; 111: 906-913.
9. Müllner-Eidenböck A, Amon M, Hauff W i wsp. Surgery in unilateral congenital cataract caused by persistent fetal vasculature or minimal fetal vascular remnants: age-related findings and management challenges. J Cataract Refract Surg 2004; 30: 611-619.
10. Haddad R, Font RL, Reeser F. Persistent hyperplastic primary vitreous, a clinic opathologic study of 62 cases and review of the literature. Surv Ophthalmol 1978; 23: 123-134.
11. Zong-Duan Z, Li-Jun S, Jia Q. Congenital membranous cataract associated with persistent fetal vasculature. Int J Ophthalmol 2010; 3: 370-371.
12. Goldberg MF. Persistent fetal vasculature (PFV): an integrated interpretation of signs and symptoms associated with persistent hyperplastic primary vitreous (PHPV). The Edward Jackson Memorial Lecture. Am J Ophthalmol 1997, 124: 587-626.
13. Pollard ZF. Persistent hyperplastic primary vitreous: diagnosis, treatment and results. Trans Am Ophthalmol Soc 1997; 95: 487-549.
14. Hunt A, Rowe N, Lam A, Martin F. Outcomes in persistent hyperplastic primary vitreous. Br J Ophthalmol 2005; 89: 859-863.
15. Wang J. Compliance and patching and atropine amblyopia treatments. Vision Res 2015; 114: 31-40.