



Case Report

Surgical Treatment of Persistent Hyperplastic Primary Vitreous: A Case Report

Min Zhuang, Tao Sun*

Department of Ophthalmology. The Yancheng Clinical College of Xuzhou Medical University, The Fourth Affiliated Hospital of Nantong University, The First People's Hospital of Yancheng, Yancheng 224000, Jiangsu Province, China

***Corresponding author:** Tao Sun, Department of Ophthalmology. The Yancheng Clinical College of Xuzhou Medical University, The Fourth Affiliated Hospital of Nantong University, The First People's Hospital of Yancheng, Yancheng 224000, Jiangsu Province, China

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Abstract

Introduction: Persistent Hyperplastic Primary Vitreous (PHPV) due to incomplete regression of the embryonic hyaloid vasculature is a rare congenital developmental ocular disorder. Here we present a case of PHPV disguised as exotropia.

Case Presentation: A four-year-old boy was diagnosed with unilateral mixed type PHPV in the left eye. It manifested with a perceptual exotropia and a hyaloid artery connected to the optic papilla. The patient was treated with a vitrectomy to prevent complications.

Conclusions: Early surgical intervention for PHPV can avoid long-term complications.

Keywords: Case report; Perceptual exotropia; Persistent fetal vasculature; Persistent hyperplastic primary vitreous; Surgery

Introduction

Primary Hyperplastic Persistent Vitreous (PHPV) is a congenital disease resulting from ocular dysplasia during the embryonic development of the primary vitreous. Its most common complications include tractional retinal detachment, secondary glaucoma, strabismus, and amblyopia [1]. Here, we present the first case of unilateral anterior PHPV posing as perceptual exotropia.

Case presentation

A four-year-old Chinese boy was admitted to our department with a major complaint of exotropia for one year. The patient previously underwent strabismus surgery to correct the issue without any additional intervention. He was a full-term baby, and his mother was 34-year-old at the time of birth. There is no family history of similar complaints. His physical and mental developments are within normal ranges, with a best-corrected visual acuity of 20/25 in the right eye and 20/125 in the left eye. The right eye examination

was unremarkable, while the left eye showed a 30° exotropia with normal eye movement. The slit-lamp examination revealed a normal anterior segment and a transparent lens, while the fundus examination revealed a fibrous vascular membrane originating from the optic disc and extending forward to the rear segment of the lens (Figure 1). An A-scan indicated axial lengths of 22.3mm and 22.2mm in the right and left eyes, respectively. A B-scan, performed for posterior segment assessment, revealed a funnel-shaped echo band in the vitreous cavity extending to the anterior segment of the vitreous and adhering to the lower part of the posterior capsule of the lens. It displayed ocular motion-induced movements without posterior shift (Figure 2). The continuation of the cord-like blood flow signal was detected by Color Doppler ultrasonography, which also indicated the presence of the central retinal artery within the lesion (Figure 3). A diagnosis of PHPV with perceptual exotropia in the left eye was made based on these clinical findings and investigations. Promptly after admission, a 23G minimally invasive vitrectomy was performed under general anesthesia. A vitreous cutting head was used to section the preoptic disc fibrovascular column to release it from the posterior part of the lens. Fibrovascular membrane adherent to the optic papilla

was electrocoagulated to avoid active hemorrhage, and laser photocoagulation was used to seal the adjacent retina. The one-week (Figure 4) and two months (Figure 5) post-surgery follow-up visits showed no postoperative complications.

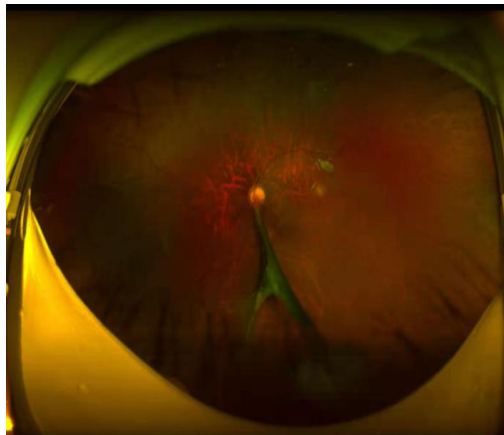


Figure 1: Fundus examination revealing the fibrous vascular membrane from the optic disc extending forward to the rear of the lens.

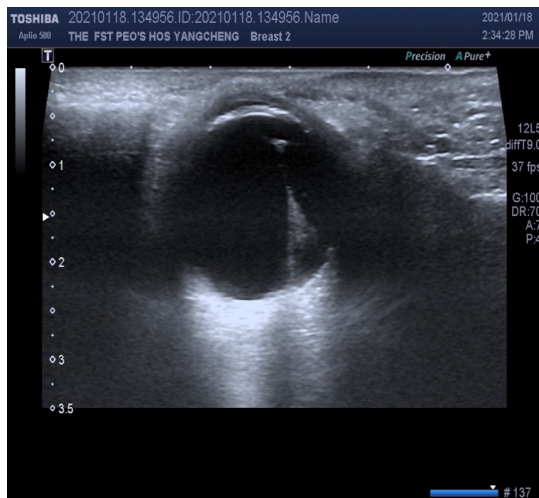


Figure 2: B-scan showing a funnel-shaped echo band in the vitreous cavity extending to the anterior segment of the vitreous and adhering to the lower part of the posterior capsule of the lens.

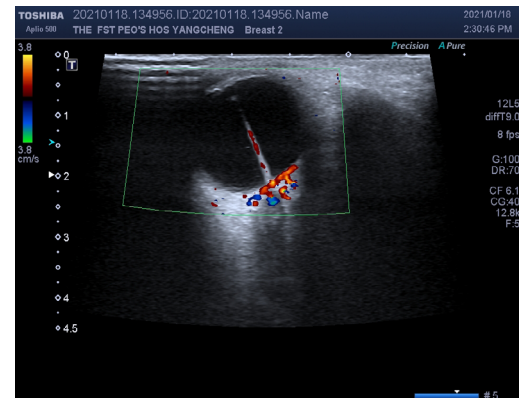


Figure 3: Color Doppler ultrasonography showing the continuation of the cord-like blood flow signal with the central retinal artery located within the lesion.

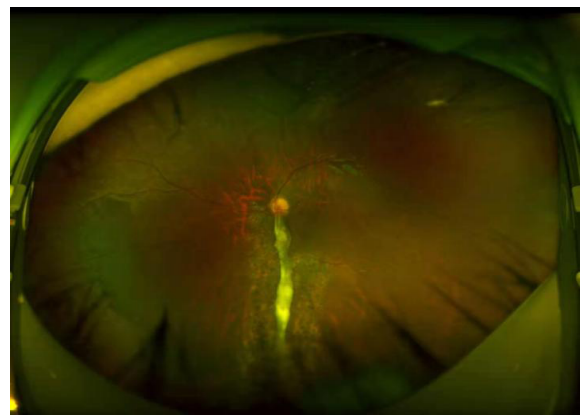


Figure 4: A follow-up visit one week after surgery.

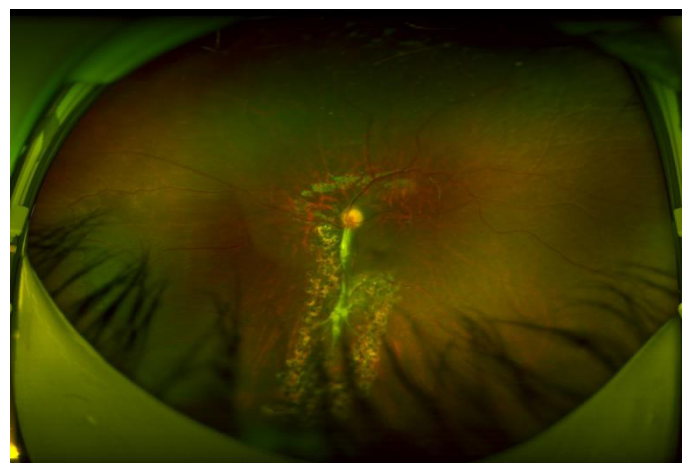


Figure 5: A follow-up visit two months after surgery.

Discussion and Conclusions

Primary Hyperplastic Persistent Vitreous (PHPV) is a congenital disease resulting from ocular dysplasia during the embryonic development of the primary vitreous [1] with clinical symptoms including small globes, small corneas, ciliary body elongation towards the center, posterior fibrous membranes within the lens, and persistent vitreous arteries. The primary vitreous is formed during the 1st month of intrauterine life and starts regressing around the 9th week during the formation of the secondary vitreous. The secondary vitreous fills most of the vitreous cavity by the end of the 3rd month, and the primary vitreous condenses into a narrow band (Cloquet's canal) running from the optic nerve to the posterior aspect of the lens [2]. The remnant vessel connects the optic nerve's head posteriorly to anterior ocular structures, such as the lens and ciliary process, and its manifestation varies among patients. Based on the affected ocular structures, it can be divided into three types: anterior, posterior, and combined (or mixed) types [3] with a distribution of 25, 12, and 63%, respectively, among all affected subjects [4]. Anterior PHPV is usually associated with an opacity of the lens or posterior capsule cortex, continuous fibrovascular membrane hyperplasia behind the lens, as well as elongation of the ciliary body. Posterior PHPV, on the other hand, is associated with connections between the vitreous vascular membrane, the optic disc, eyeball shrinkage, and immature growth of the optic disc, macula, and retina. Meanwhile, Mixed PHPV includes features of both the anterior and posterior types and has the highest clinical occurrence.

PHPV was previously considered a rare condition. However, a recent study about childhood blindness and visual loss in the United States revealed that PHPV accounts for about 5% of all cases of blindness [5]. Another similar pilot study of ocular disease screening for neonates in China indicated a PHPV percentage of about 0.351% among 15398 assessed newborns [6]. PHPV is more common in full-term babies, and most cases are monocular and sporadic, with no significant difference in the incidence between males and females [7]. Primary and secondary complications due to PHPV include angle-closure glaucoma, vitreous hemorrhage, and retinal detachment. These complications are most likely to be seen at birth or within a few years after. PHPV is also one of the most common causes of infantile leukocoria [8], with a retrospective study demonstrating that strabismus represents not only a common complication of PHPV but also the cause of the initial diagnosis, accounting for 15.5% of all cases [9]. The diagnosis of PHPV is typically made based on the results of the B-mode ultrasound, optical coherence tomography (OCT), or Doppler ultrasound [10,11]. The B-mode ultrasonography of PHPV is characterized by a short axial length, an abnormal lens shape, a conical or funnel-shaped hyperechoic mass posterior to the lens, and a narrow

posteriorly to the optic disc in a cable shape. Additionally, Color Doppler blood flow imaging often reveals a ribbon-like hyperecho in the vitreous body with a blood flow signal extending from the optic papilla to the posterior part of the lens. The blood flow signal often continues with the arteries and veins in the central optic papilla, and a flocculent hyperecho appears in the vitreous body when there is bleeding [12-14].

The treatment of this disease remains controversial, and some researchers recommend conservative treatment for posterior PHPV [15], with the only absolute indication for surgery being intractable high intraocular pressure [16, 17]. However, if left untreated, PHPV can cause blindness due to resultant complications. A study found that 70 % of patients with posterior type had retinal folds, which may be formed by fibrous proliferations that develop backward along the Cloquet tube and adhere to the retina, potentially leading to tractional retinal detachment [18]. More and more researchers nowadays believe that surgery can significantly ameliorate patient prognosis and even improve vision, particularly with current advances in microsurgical techniques [10,19]. Early surgical intervention can salvage vision and achieve acceptable cosmetic outcomes. Recent evidence indicated that the prognosis of surgical intervention is related to the time window of visual development in children. In young children, surgical treatment followed by refractive correction and treatment for amblyopia is very likely to restore vision to some degree [20,21]. Lu Jiao et al. [22] found that vitrectomy through the keratoscleral margin approach could effectively deal with the severe complications of mixed type PHPV by controlling the disappearance of the anterior chamber, corneal opacity, and the development of some lesions in the posterior segment of the eye, therefore, improving vision through reconstruction of visual access and improvement of eye appearance. A retrospective study demonstrated that some favorable outcomes were achieved in a relatively large number of patients with cataracts and PHPV through different choices of surgical methods and the introduction of sophisticated microsurgical techniques combined with aggressive anti-amblyopic therapy [23].

In this case, the patient was diagnosed with mixed PHPV due to exotropia of the left eye after a thorough examination. A posterior approach vitrectomy was performed to prevent retinal detachment, and the retina remained flat two months post-surgery. However, close follow-ups are needed to monitor phase and long-term disease changes. Additionally, amblyopia training also represents an important method to improve future vision.

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