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Case Report

Persistent Hyperplastic Primary Vitreous Found at the Time of Cataract Surgery in an Adult

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Abstract

Purpose: To report a rare case of PHPV found at the time of cataract surgery in an adult.

Methods: Observational case report.

Results: A 37-year-old woman presented complaining of longstanding left leukocoria and poor vision. She was found to have a mature, white cataract in the setting of a microphthalmic left eye, and a left afferent pupillary defect. During cataract extraction, she was found to have a dense, opaque, fibrotic retrolental membrane, adherent to the ciliary body. Intraocular scissors and anterior vitrectomy were used to clear away the membrane. Follow up funduscopic exam revealed the presence of fibrovascular tissue extending anteriorly from the inferonasal optic disc, confirming a diagnosis of adult persistent hyperplastic primary vitreous (PHPV). Postoperatively, the patient had normal intraocular pressure, regained some peripheral vision and had a good cosmetic outcome.

Conclusion: PHPV is a congenital anomaly resulting from failed regression of embryonic structures. We report an unusual case of PHPV found in an adult eye, involving both anterior and posterior segments. This is the first reported case of adult PHPV in Hawaii, and the tenth reported case found by our literature search.

Keywords: *Cataract; Complicated Cataract Surgery; Congenital Cataract; Persistent Fetal Vasculature; PFV; Persistent Hyperplastic Primary Vitreous; PHPV*

Introduction

The embryonic vasculature structures, the vasa hyaloidea propria and the tunica vasculosa lentis, reach peak prominence at the 9 week or 40 mm stage. A gradual state of involution follows until the seventh month, at which point blood flow in the hyaloid artery ceases [1]. Persistent hyperplastic primary vitreous (PHPV), also known as primary fetal vasculature (PFV) is a rare congenital developmental anomaly of the eye that results following failure of the embryologic primary vitreous and hyaloid vasculature to follow its normal course of regression [2,3].

PHPV is typically unilateral and is rarely associated with systemic findings [4]. The anatomical description of the disease can be subdivided in to anterior and posterior manifestations, often patients demonstrate a combination of anterior and posterior disease. Although surgical intervention does have the potential to improve vision, and is associated with a low rate of complication [5], limiting factors in final visual outcome are advanced disease or posterior involvement at the time of presentation, glaucoma, amblyopia, or noncompliance with amblyopia therapy [6-8].

A high proportion of PHPV patients will develop glaucoma [9]. The mechanisms include secondary open angle glaucoma-such as that induced by recurrent intraocular hemorrhage or post-cataract surgery aphakic glaucoma in children-or angle closure glaucoma due to anterior displacement of the lens-iris diaphragm in the setting of elongated ciliary processes and cataract [9-11].

The differential diagnosis of leukocoria includes cataract (congenital, traumatic, nutritional, toxic, idiopathic), retinoblastoma, Norrie's disease, retinopathy of prematurity, retinal detachment, Coats' disease, and PHPV with cataract. There are few reported cases of PHPV found in an adult eye. Most cases are discovered early in life, worked up and treated promptly, as the child often presents with leukocoria which raises concern for potentially life-threatening retinoblastoma. Dr. Reese in the Jackson Memorial Lecture in 1955 commented that he had never seen a recognizable case in an adult [12].

Case Presentation

A 37-year-old female was referred for leukocoria of the left eye. She had emigrated from the Philippines several years prior and had no known past medical history. The patient complained of longstanding poor vision in the left eye, and the patient's mother corroborated this story, stating that the patient was "blind" in her left eye "since birth". While in the Philippines, the patient had been unable to receive ophthalmologic care due to financial limitations.

Her chief complaint was that her left pupil looked "white." And more than anything she desired surgery to remove the white pupil. Working as a cashier, the patient stated that she was becoming increasingly self-conscious, as customers would often look at her quizzically or point out that her left eye looked "different".

Examination revealed vision OD was 20/20 with -0.75 D sphere correction, and OS was light perception without projection. There was a left afferent pupillary defect. Slit lamp examination demonstrated that the right eye was anatomically normal, but the left eye had a hypermature cataract. Manifest sensory exotropia as well as left sided ptosis and microphthalmia were noted. A fundus examination revealed that her right eye was normal; there was no view of the fundus OS. Intraocular pressure was 19 mm Hg OD and 13 mm Hg OS.

Cataract surgery was performed under topical anesthesia. Despite the use of trypan blue dye to highlight the anterior capsule, the capsulorrhexis was difficult to perform. Due to the soft nucleus, irrigation and aspiration was used instead of phacoemulsification. A thick, white fibrotic membrane (engulfing the posterior, equatorial and part of the anterior capsule) remained after cataract removal.

The white fibrotic membrane caused the patient pain when the surgeon attempted gentle pulling directed away from the ciliary body. Intraocular scissors were then used to cut open the membrane, and vitrectomy cutting was used to remove as much of the fibrotic membrane behind the pupil as safely possible. An intraocular lens was not inserted due to the poor zonular support, and because recovery of vision was not the primary goal of this case. The wound was hydrated and closed tightly. At the end of the case, the media was clear. Video of the surgery is available at http://www.youtube.com/.watch?v=KNFbGxlyE4U&sns=em.

A two week postoperative follow up revealed that vision OS vision had improved to hand motion with projection. Intraocular pressure was 15 mm Hg with clear cornea and media. Mild ptosis, exotropia and microphthalmia remained. See figure 1 for preoperative and postoperative photos. The patient was happy with the results and has since returned to work without issue.

Postoperative retina consultation re-demonstrated the presence of widespread pigmentary changes, abnormal vasculature emanating from the inferonasal disc and partially regressed vessels traversing the vitreous toward the anterior hyaloid face (Figure 2). Fluorescein angiography, transit left eye, demonstrated the presence of a hyaloid artery at the inferonasal disc, which filled in the early arterial phase (Figure 3). Spectral domain optical coherence tomography (OCT) demonstrated inferior displacement of the macula (Figure 4).

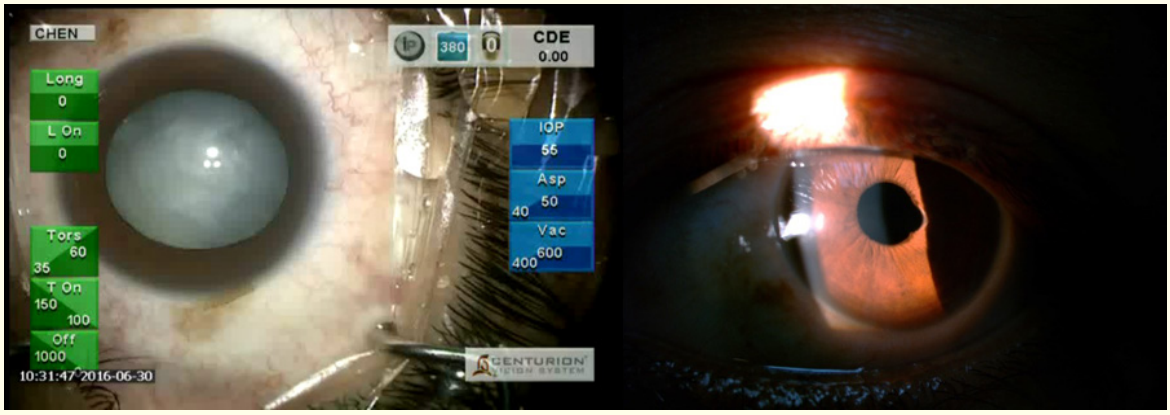

Figure 1: *Preoperative (left) and postoperative (right) images.*

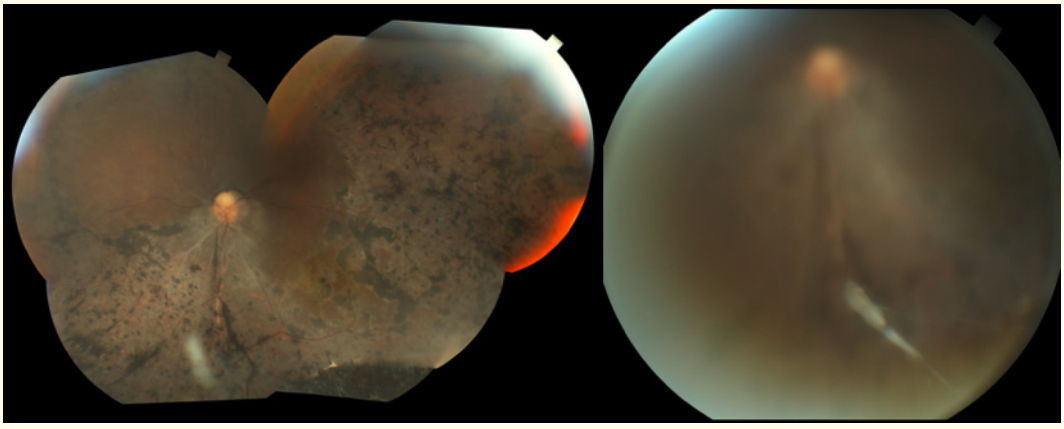

Figure 2: *Fundus photos of left eye showing pigmentary changes, persistence of fetal circulation (left image, montage) and remnants of these vessels in the anterior vitreous.*

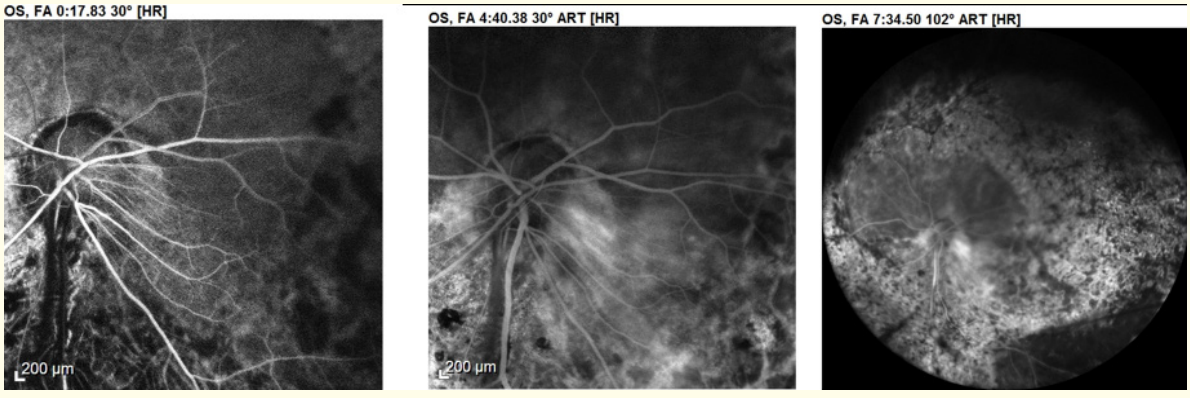

Figure 3*: Fluorescein angiogram of the left eye (left = early arterial phase, middle = venous phase, right = wide angle recirculation phase). The early arterial phase image shows early filling of the persistent hyaloid artery at the inferior disc (at approximately 6:30 clock hours). The recirculation phase also shows filling of the persistent hyaloid artery.*

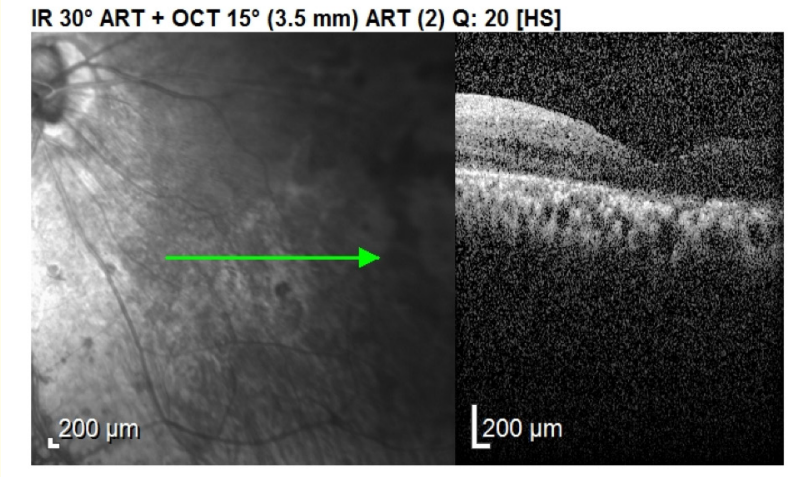

Figure 4: *Spectral domain OCT of the left macula showing inferiorly displaced fovea*

Discussion

Several details of this surgical case described herein are worth noting. First, the surgeon was surprised intra-operatively by the presence of a retrolental membrane, and was forced to change the intraoperative plan. Had B scan been performed preoperatively, this may have helped establish the diagnosis [13,14] and design a better surgical plan; and this certainly would have allowed for more accurate informed consent. (Postoperative B scan is shown in Figure 5). Had the diagnosis been more clear preoperatively, a vitreoretinal specialist would have been consulted for lensectomy [15,16]. And given the myriad complications associated with PHPV, the preoperative consent also would have also included the possibility of intraoperative hemorrhage, postoperative glaucoma, phthisis and enucleation.

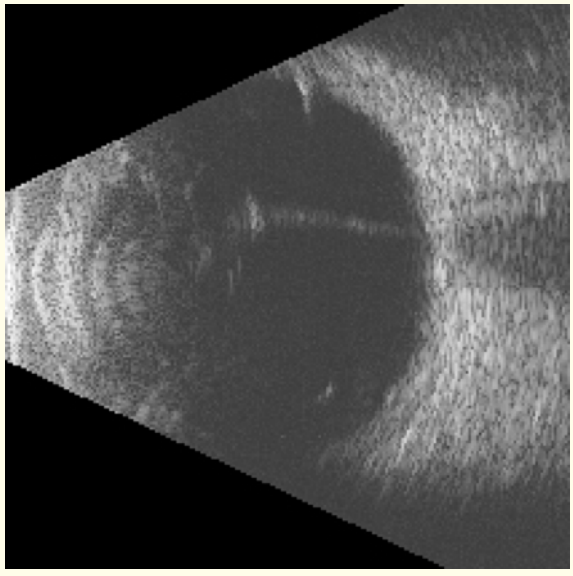

Figure 5: *B scan ultrasound of the left eye demonstrating persistent hyaloid vasculature extending to anterior, retrolental fibrotic tissue.*

At the 2016 ASCRS Film Festival, Jagat Ram presented a strategy for operative management of anterior PHPV in infants (http://ascrs2016.conferencefilms.com/). The surgical footage showed that intraocular cautery was performed on visible vasculature of the retrolental membrane, intraocular scissors were used to remove this membrane, and an intraocular lens was inserted to prevent amblyopia. Notably, in his cases, a significant amount of whitish fibrotic capsule remained at the pupil.

The case described herein is an adult mature cataract with PHPV involving both anterior and posterior segments. Fortunately, no vascular structures were encountered during surgery. Unlike with infant PHPV, visual recovery was not an expected outcome of this case. The left afferent pupillary defect indicated likely amblyopia. And so the main goals of cataract extraction were to reduce the risk of inflammation and glaucoma, and to improve the patient's appearance and lifestyle.

This was the surgeon's first adult hypermature cataract case associated with PHPV. In retrospect, careful history taking and preoperative B scan ultrasound on this young female patient could have helped with diagnosis and preparation of a more accurate preoperative informed consent form. There are important risks of cataract surgery in the setting of PHPV that must be disclosed. The aberrant vasculature obscured by a mature cataract may be damaged inadvertently and may thus induce massive hemorrhage and glaucoma, leading possibly to phthisis and subsequent enucleation. The importance of accurate preoperative diagnosis thus cannot be overemphasized. As such, another key teaching point in this paper is that all patients with an unusual white cataract of this sort should undergo B scan ultrasonography preoperatively. If there is any doubt about what might be encountered intraoperatively, consultation with a vitreoretinal surgeon for possible lensectomy should be considered.

Summary Statement

A 37-year-old woman was found to have persistent hyperplastic primary vitreous (PHPV) at the time of cataract surgery. This is one of the few reported cases of PHPV discovered in an adult.

Conflicts of Interest

None.

Proprietary Interest in Products Discussed

None.

Sources of Financial Support

None.

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