

Case Report

Pars Plana Vitrectomy, Lensectomy, Stalk Transection with Capsulectomy for the Primary Treatment of Persistent Fetal Vasculature Cataract

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Abstract

Purpose: The purpose of this report is to present an alternative surgical approach to persistent fetal vasculature.

Case: A 9 month old male presented with leukocoria in the right eye and was found to have lenticular opacity with posterior dysplastic mass with complex exudative tractional retinal detachment and fibrovascular stalk. Surgical approach consisted of fibrovascular membrane transection anterior to termination of the stalk. This was performed without endodiathermy despite vascularization. The posterior capsule was opened and stalk was removed. Lensectomy was performed to remove lens nucleus and cortex. The patient was left aphakic and the both anterior and posterior capsules were opened widely. One month follow up of the patient showed a clear visual axis and refractive error of +5.00 spheres OD

Discussion: Surgical morbidity remains a major hurdle in the management of persistent fetal vasculature. The use of endodiathermy has gained popularity, but has the disadvantage of introducing intraocular energy. There has been a lack of literature regarding the management of persistent fetal vasculature with the presence of posterior involvement, and this case presents an alternative approach without the need for endodiathermy despite vascularization.

Keywords: Persistent fetal vasculature; Persistent hyperplastic primary vitreous; Pediatric ophthalmology

Introduction

Persistent Fetal Vasculature (PFV), a pathology first described by Reese as persistent hyperplastic primary vitreous [1], results from failure to involute the primary vitreous and hyaloids vasculature during the 2nd trimester. The most classic presentation results in a persistent vascular stalk extending from the optic disc to the posterior lenticular capsule. However, significant variability in the spectrum of this disease has been reported [2-5]. PFV is most often unilateral and is associated with decreased axial length, hyperopia, cataract, and glaucoma [6-9].

Surgical management of patient with PFV remains a challenge. Significant complications including posterior synechiae, retinal detachment, phthisis, posterior capsular opacity, inflammatory pupillary membrane, glaucoma, intraocular lens displacement, and vitreous hemorrhage have been reported [8,10-12]. Despite these complications, post-operative improvement in visual acuity has been reported as high as 83% [8]. Various surgical techniques have been documented [3,8,10,11,13-18], however the literature regarding posterior segment surgical approach remains scarce. We report a patient that underwent pars plana vitrectomy, lensectomy, and posterior capsulectomy without the use of endodiathermy as the primary treatment for cataract associated to PFV.

Case Report

A nine-month-old boy born at term via uncomplicated cesarean

section was evaluated due to leukocoria in his right eye (OD). The mother had routine prenatal care with an uneventful pregnancy, and no other pertinent medical or surgical history.

Ophthalmologic examination was remarkable for decreased visual acuity as evidenced by poor fixation and exotropia OD. There was no relative afferent pupillary defect and intraocular pressure (IOP) was 9 mmHg in both eyes (OU). Anterior segment

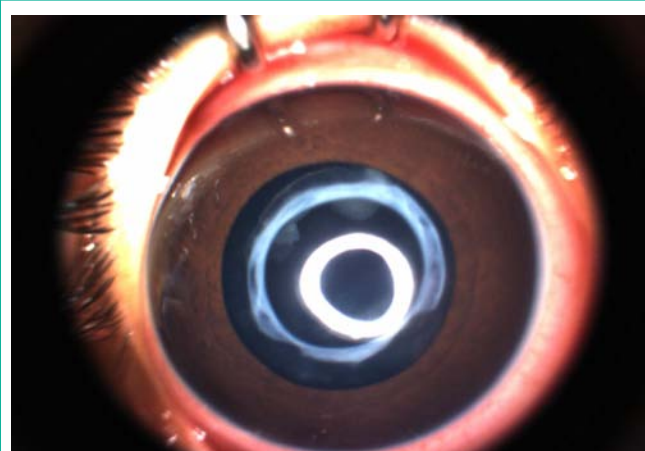


Figure 1: Sectoral Lenticular Cataract, right eye, in patient with persistent fetal vasculature.

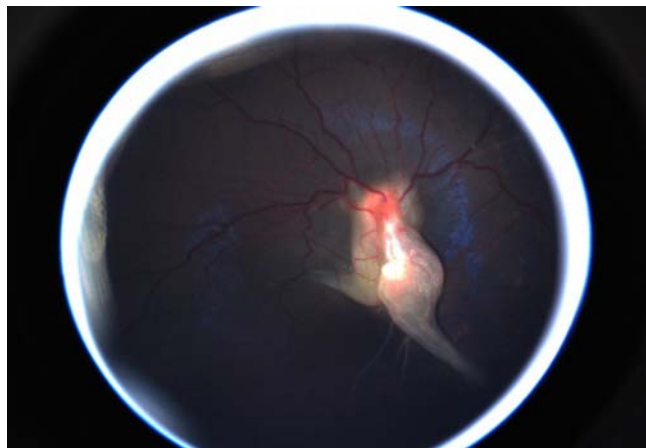


Figure 2: Fundus photo, right eye, showing preretinal posterior dysplastic mass with complex exudative tractional retinal detachment and fibrovascular stalk.

examination was unremarkable OU, with the exception of a sector cataract OD. (Figure 1). Indirect ophthalmoscopy was remarkable for a preretinal posterior dysplastic mass with complex exudative tractional retinal detachment and a fibrovascular stalk OD (Figure 2). There were no macular holes, tears, or breaks OU. Vascular ectatic alterations, as seen in fluorescein angiography (Figures 3a & 3b), were present OU. Due to the presence of a visually significant cataract, the patient was scheduled for pars plana vitrectomy, and lensectomy with capsulectomy as described below:

The right eye was prepped and draped and a wire lid speculum was inserted. The 25 gauge valved trocars were introduced 1.5 mm posterior to the limbus and posterior to the lens. The infusion line was then attached and IOP was stabilized. The primary 25 gauge vitrector and endoilluminator were introduced and, with contact 130 degree wide field viewing, a primary vitrectomy was performed. There were marked tractional alterations encircling the membranous stalk. There was posterior retinal dysplasia. The termination of the stalk was identified and, anterior to this, a transection of the fibrovascular membrane was performed. There was direct lenticular insertion of

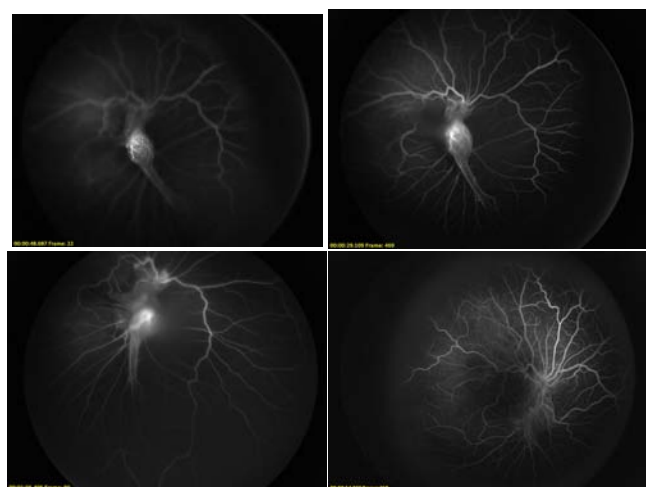


Figure 3: Vascular ectatic alterations with vascularized stalk, right eye, seen on fluorescein angiography.

the stalk with lenticular alterations. The posterior capsule was then opened, and the stalk was removed in its entirety. Lensectomy techniques were utilized to remove all of the lens nucleus and cortex. Due to the patient's age and a crowded anterior segment, it was determined appropriate to leave the patient aphakic. The capsule was then opened both anterior and posterior widely to deal with the potential postsurgical capsular phimosis. After the procedure was completed, the retina was noted to be free of open holes, tears, or breaks. The patient received subconjunctival injection with dexamethasone and gentamicin 180 degrees away from the valved trocars. Topical prednisolone acetate and gentamicin were also applied followed by an occlusive patch and Fox shield.

Follow up evaluation at one month showed a clear visual axis, a refractive error of +5.00 sphere OD, and restoration of the posterior foveal anatomy and vascular sclerotic alterations. There was no active retinal hole, tear, break, epiretinal membrane, or proliferative vitreoretinopathy formation.

Discussion

Surgical complications are a primary concern in the treatment of PFV. While there have been innovations in technology and approach, the majority of complications remain, in large part, direct consequences of the procedure itself. The use of the plasma blade and endodiathermy offers certain advantages, but also introduces significant intraocular energy. Post-operative complications include synechiae, inflammatory pupillary membranes, and increased IOP [7,8], which provide motivation to reduce exogenous intraocular energy to minimal levels.

Literature on surgical technique for PFV tends to focus on anterior management, with a lack of guidance available for posterior segment involvement. While the point of transection of the hyaloid stalk remains controversial, we have had success performing the transection at the most anterior location where the stalk inserts into the lens. We have been able to perform this without the use of endodiathermy despite a vascularized stalk, thereby minimizing unnecessary intraocular energy. Some of the major complications are bleeding after stalk transection, phthisis bulbi, and spontaneous retinal detachment. The use of diathermy, specifically the use of unnecessary power in a child's eye can possibly lead to increasing the risk of all of the aforementioned complications. There are a few other case studies and case series which back our finding that our technique is likely the technique of choice to perform on children with PFV [18,19].

Aphakia, as in our patient, is one of the most common childhood causes of glaucoma [7,20], and close follow up is critically important. Axial length measurement and ultrasound are vital tools to utilize due to the risk of glaucoma associated with aphakia.

Recent studies have shown that vitrectomy with lensectomy in patients with PFV have a significant improvement in visual acuity after surgery [8]. Further studies will elucidate the best management practices for amblyopia, stalk transection, and intra-ocular lens placement in patients with PFV.

Conclusion

We present a surgical technique for removal of the fibrovascular

membrane and cataract related to persistent fetal vasculature. This technique provided a much quicker removal of pathological tissue with no complications. This case adds to the growing literature to aid in the decision of the mainstay of treatment for PFV. Further studies are needed to compare this technique with traditional techniques that include the use of diathermy.

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