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Case Report: Unilateral Developmental Cataract with Persistent Hyperplastic Primary Vitreous

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Abstract: ***Persistent Fetal Vasculature (PFV), previously termed Persistent Hyperplastic Primary Vitreous (PHPV), is a congenital ocular anomaly caused by the failure of the hyaloid vascular system to regress during fetal development. A common secondary manifestation of PFV is cataract formation, which may result from lenticular invasion by persistent vascular remnants, lens swelling, intralenticular hemorrhage, or calcific changes. Cataract surgery in eyes with PFV carries an elevated risk of intraoperative hemorrhage due to the presence of persistent vascular tissue. A characteristic feature of PFV is the presence of a fibrovascular plaque adherent to the posterior lens capsule. In this case report, we describe a female child of 2years old, weighing 12kg presenting with unilateral leukocoria, who on further evaluation discovered to have PHPV. Patient underwent lensectomy via limbal approach with diathermy prior division of stalk and vitrectomy. IOL implantation was deferred as WTW distance was inadequate. On follow up visit contact lens was adviced as visual rehabilitation based on objective refraction.***

Keywords: Persistent hyperplastic primary vitreous, Congenital cataract, Persistent fetal vasculature, Developmental cataract

1. Introduction

Persistent fetal vasculature (PFV) comprises a spectrum of clinical features caused by failure of the normal process of regression or apoptosis of the fetal intraocular vasculature, which usually occurs during the second trimester. PFV was previously termed persistent hyperplastic primary vitreous (PHPV), due to the characteristic feature of a plaque of remnant tissue sitting on the posterior face of a cataractous lens, attached to a persistent stalk of hyaloid vasculature. However, any part of the fetal vasculature can persist, resulting in iris, pupillary, or optic nerve vascular remnants. Congenital pupillary fibrovascular membranes, rather than being a separate entity, are histopathologically similar to the retrolenticular manifestations of PFV.

Cataract is a frequent secondary event in PFV due to invasion of the vascular remnants, lenticular swelling, intralenticular haemorrhage, or calcification. PFV features have been found in a fifth of cases of unilateral congenital or infantile cataract, but are much less commonly noted in bilateral cataract. Cataract surgery in children with PFV is associated with a higher incidence of adverse events and worse outcomes. In addition, bilateral cases of PFV may be associated with systemic abnormalities, which impact on a child's global development. In this case report, we describe a female child of 2years old, weighing 12kg presenting with unilateral leukocoria, who on further evaluation discovered to have PHPV. Patient was operated for the same.

2. Case Report

A 2yr old female baby weighing 12kg presented to opd with chief complaints of white reflex of right eye since 7 months of age, informants being parents. They noticed that the white reflex was gradually progressive to the current state. On examination baby was fixating and following light (OU). Ocular examination showed leukocoria with divergent squint of 15 degrees of right eye. Anterior segment examination showed mature cataract. Baby was diagnosed to have developmental cataract and was advised for surgery. Media haze was noted on fundus evaluation. So, B-scan was performed, on which a linear echogenic band was observed extending from posterior surface of lens to optic disc, which represents a fibrovascular stalk. This led to a probable diagnosis of persistant fetal vasculature (PFV) or persistant hyperplastic primary vitreous (PHPV). Preoperative evaluation was performed under sedation. (Table.1)

Patient underwent lensectomy (limbal approach) with PPC (Primary Posterior Capsulotomy) with diathermy followed by division of stalk. (Figure.1) Anterior and posterior capsulotomy was performed using 25G vitrector. Anterior and core vitrectomy performed to remove surrounding vitreous to clear visual axis. IOL implantation was deferred as WTW (White-to-White) distance was short of 10mm. Postoperatively retina was stable. On follow up visit, patient was adviced soft contact lens for the very same eye as visual rehabilitation based on objective refraction values.

3. Discussion

PFV is classically categorized as anterior, posterior, or combined disease. Anterior disease features a vascularized retrolental membrane, elongated ciliary processes and cataract; posterior disease presents with a stalk from the optic disc to the posterior lens capsule, retinal fold, dysplasia, and tractional or rhegmatogenous retinal detachment (RD). Combined PFV is common and carries a worse prognosis, especially when associated with microphthalmia. (1)

In our case, it is complete or combined PFV with inadequate WTW diameter. These factors are repeatedly linked to poor visual outcome. (2)

When media opacity obscures the fundus, B-scan ultrasonography helps demonstrate a thin hyaloid/vascular stalk coursing from disc to posterior lens, supports biometry for microphthalmia, and guides IOL calculations. MRI aids differentiation from retinoblastoma when leukocoria is present. In this case report B scan imaging showed a vascular stalk and ruled out associated RD, which is consistent with recommended work-up. (3)

In cases of anterior PFV with cataract: Most centers perform limbal lensectomy with primary posterior capsulotomy and anterior vitrectomy to clear the axis and reduce vitreous traction. Hemostasis can be challenging because of a persistent tunica vasculosa lentis; careful endodiathermy of vascularized membranes/stalk is described, while some surgeons try to minimize cautery to limit bleeding and iatrogenic traction. In our case need for diathermy mirrored these reports. (4)

Because PFV eyes are often small with higher risks (visual axis opacification, inflammation, glaucoma), many guidelines avoid routine primary IOL in children <2 years, favouring aphakia with contact lens or spectacles and amblyopia therapy. Selected series, however, report acceptable outcomes with carefully selected primary or secondary IOLs in older infants/children or in less complex anterior PFV. In our case IOL implantation was deferred taking into account of patient's age, disease extend and inadequate WTW distance. (5, 6)

4. Conclusion

Persistent fetal vasculature is a rare but important cause of childhood visual impairment, with clinical outcomes largely determined by the extent of disease, presence of posterior segment involvement, ocular size, and associated complications. Our patient's presentation and management closely parallel strategies described in major case series and reviews, emphasizing early diagnosis, appropriate surgical planning, and vigilant amblyopia therapy. While anatomic success is achievable in most cases, long-term visual prognosis depends on minimizing postoperative complications—particularly retinal detachment and secondary glaucoma—and ensuring sustained adherence to visual rehabilitation. This case reinforces existing evidence that individualized surgical timing and technique, combined with comprehensive follow-up, offer the best potential for preserving vision in PFV.

Disclosure
The Authors report no conflict of interest in this work.
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Table 1: Biometry parameters

	OD	OS
White To White	9.8	11
Axial Length	20.80	20.64
Anterior Chamber Depth	2.85	2.90

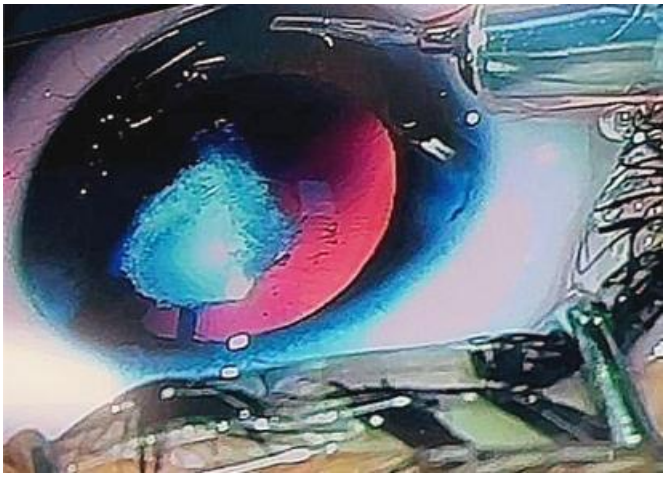

Figure 1: Intraoperative picture of PFV