

Bilateral Persistent Hyperplastic Primary Vitreous

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Abstract

Persistent hyperplastic primary vitreous (PHPV) is a congenital developmental anomaly of the eye caused by the failure of regression of primary vitreous with the abnormal persistence of hyaloid vasculature. Here we present a case of bilateral persistent hyperplastic primary vitreous (PHPV) which is a rare entity, in a 2 months old infant who presented in our department with history of bilateral microphthalmia and leukocoria. In this regard ultrasound doppler, CT scan and MRI findings of PHPV will be discussed. We suggest that this entity, although rare, should be considered in the differential diagnosis while evaluating bilateral leukocoria.

Key words: Bilateral, Persistent hyperplastic primary vitreous, Retinoblastoma.

Introduction

Bilateral persistent hyperplastic primary vitreous (PHPV) is an uncommon entity which usually presents as leukocoria, microphthalmia, and cataract.¹ Most cases are unilateral and sporadic, with no difference of incidence between males and females. It is the second most common cause of acquired cataract during the first year of life.² It is a developmental disorder of the eye which occurs due to abnormal persistence of fetal intraocular vessels in the anterior or posterior segments of the eye. Primary vitreous forms around 7th week of intra-gestation life and starts involuting around 20th week and nearly always disappears at the time of birth. Failure of regression of primary vitreous results in many of the abnormalities seen in PHPV.¹

Case report

A 3-month-old male infant presented with bilateral leukocoria. Birth history of the child was unremarkable. The child was referred for contrast enhanced MRI with a

clinical suspicion of retinoblastoma. On magnetic resonance imaging (MRI) abnormal signals were seen in bilateral vitreous, appearing heterogeneous to hypointense on T2WS and isointense on T1WS, representing organized vitreous hemorrhage. (Figure 1) A tubular structure was seen extending from the posterior surface of the lens through the vitreous canal to the posterior and inner surface of the globe bilaterally and showed enhancement on post contrast T1WS on the left. Total axial length of both eye balls in the present scan was approx 17mm which is normal for the age. However anterior segment length was slightly decreased bilaterally measuring approximately. 6mm in right eye and 5mm in left. Optic nerve, optic tract and optic chiasma appeared normal. Diagnosis of bilateral persistent hyperplastic primary vitreous was made.

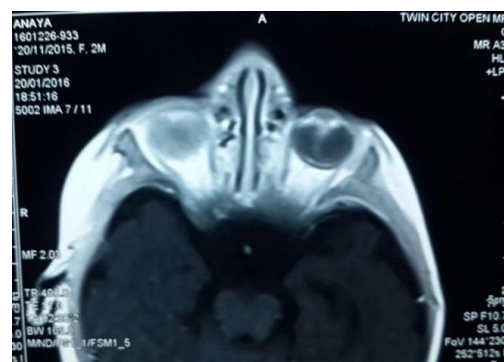


Figure 1: MRI: T1 Post contrast image showing bilateral vitreous detachment and retrovitreous haemorrhage more so on the right. An enhancing tubular structure seen in the left eye suggestive of persistent hyaloid artery.

CT and grey scale ultrasound orbit of this patient had already been done at another center. CT or MRI images showed vitreous detachment and diffusely hyperdense attenuation of vitreous in both globes, suggesting hemorrhage. No mass or calcification was seen.

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Grey scale ultrasound evaluation revealed vitreous detachment and retrovitreal haemorrhage.(Figure 2)



Figure 2: Grey scale ultrasound showing vitreous detachment and vitreous haemorrhage.

Colour doppler examination revealed presence of blood flow in a tubular structure extending from the posterior surface of the lens to posterior surface of globe bilaterally, representing persistent hyaloid artery in the cloquet's canal. (Figure 3) Spectral analysis of this blood vessel showed arterial waveforms. There was no evidence of calcification confirming our diagnosis of bilateral PHPV. PHPV is differentiated from retinoblastoma by the absence of a calcified mass, artery running through Cloquet's canal, and absence of typical signal characteristics of retinoblastoma on MRI, i.e., hyperintense on T1-weighted images and hypointense on T2-weighted images. Differentiation from advanced retinopathy of prematurity (ROP) can be difficult on imaging alone. History of a premature, low birth weight infant undergoing prolonged supplemental oxygen therapy helps to distinguish it from bilateral PHPV. A patent hyaloid artery, as noted in this case, is not a feature of retinopathy of prematurity and vitreoretinal dysplasias.

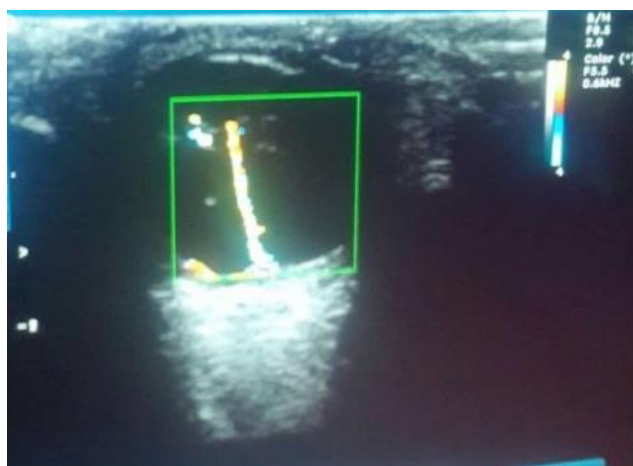


Figure 3: Doppler ultrasound showing persistent hyaloid artery in cloquet's canal

Discussion

PHPV occurs because of an incomplete regression of the embryonic vitreous and hyaloid vasculature. The primary vitreous is formed during the first month of development and contains branches of the hyaloid artery. This hyaloid artery begins to regress during the formation of the avascular secondary vitreous at 9 weeks. By the third month, the secondary vitreous, which ultimately forms the adult vitreous, fills most of the developing vitreous cavity. The primary vitreous becomes condensed into a narrow band (Cloquet's canal), running from the optic disc to the posterior aspect of the lens.³

PHPV is usually isolated and unilateral. Bilateral lesions are usually rare and associated with systemic or syndromic conditions such as trisomy 13, 15 and 18, Norrie disease and Warburg disease.¹

The most common presenting signs and symptoms are leukocoria, poor vision, small eye and strabismus. PHPV is classified into three types: Anterior, posterior, or combination of anterior and posterior. Clinically leukocoria, strabismus and a small eye may be present in all types.⁴ Anterior PHPV has the best prognosis for vision, but approximately half of the patients also have an associated posterior component. The anterior type of PHPV includes a shallow or collapsed anterior chamber, a retrolental vascular membrane, cataract and anterior chamber anomalies. Abnormalities of lens and anterior chamber are signs of combined anterior and posterior variant of PHPV.¹ Typical imaging findings of posterior PHPV includes the demonstration of cloquet canal which contains the hyaloid artery.⁴

Diagnosis of PHPV can be made on imaging. Ultrasound reveals a hyperechoic, inhomogenous linear structure bilaterally in the vitreous chamber extending from the posterior wall of the lens till the optic nerve head and retina.¹ It may also show echogenic vitreous due to vitreal hemorrhage and hyperechoic band extending from papilla to ora serrata presenting retinal detachment. Doppler may show arterial flow within this band representing a persistent hyaloid artery.²

CT findings of PHPV are absence of calcification, increased density of the entire vitreous, tubular intravitreal density (Cloquet's canal or nonattached retina), decubitus positioning showing a gravitational effect on fluid-fluid level, microphthalmia, enhancement of abnormal intravitreal tissue, and small or irregular lens.² MRI findings of PHPV consist of a tubular structure representing hyaloid artery, a funnel-shaped retinal detachment, with the subretinal fluid hyperintense on both T1- and T2-weighted images; fluid-fluid level due to the presence of hemorrhage in the subretinal space; a retrolental mass; microphthalmia, and vitreous hemorrhage.²

Differential diagnosis of PHPV includes retinoblastoma, Coat's disease, vitreoretinal dysplasia, ocular toxocariasis, and a condition contributing to subretinal fluid or hemorrhage and retinopathy of prematurity. It is important to exclude retinoblastoma in all cases of leukocoria. PHPV has a typical imaging appearance which allows reliable differentiation from retinoblastoma.

Complications of PHPV include rupture of the lens capsule, cataract formation, intraocular hemorrhage, secondary glaucoma, tractional retinal folds, and subsequent phthisis bulbi. The friability of the hyaloid vasculature predisposes to vitreous hemorrhage, which was noted in both globes in the present case. Majority of patients with posterior PHPV never obtain useful vision.²

Typical imaging features of ultrasound, CT, and MRI can be helpful in the diagnosis.¹ However a rational sequence of imaging modalities could prove cost effective and prevent

unnecessary radiation exposure, as in this case a definitive diagnosis could have been made with the help of Color Doppler without proceeding to CT and MRI.

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