

Bilateral Persistent Hyperplastic Primary Vitreous: A Rare Entity

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Abstract: Persistent hyperplastic primary vitreous (PHPV) is a rare congenital, nonhereditary malformation of the eye which results from persistence of the foetal fibrovascular primitive hyaloid system which should normally regress otherwise. Most commonly seen is unilateral and sporadic while bilateral is rare. We report a case of bilateral PHPV in a 10 year old male patient who presented with bilateral leukocoria and dimness of vision along with microphthalmos. Child was referred for ultrasound, CT and MRI imaging with a clinical suspicion of retinoblastoma which is a close differential of PHPV. The imaging findings are as described in the article which confirms the diagnosis of bilateral PHPV.

1. Introduction

Persistent hyperplastic primary vitreous (PHPV) is an uncommon condition, presenting clinically as leukocoria (white pupillary reflex), micro-ophthalmia, and cataract. Bilateral PHPV is rare. Most important differential diagnosis is retinoblastoma, which can be differentiated by imaging features.

2. Case Report

A case of bilateral persistent hyperplastic primary vitreous (PHPV) in a 10 year old male child, who had bilateral leukocoria, is presented. Birth history of the child was unremarkable. On initial examination, the size of the cornea looked smaller. The child was referred for ocular ultrasound and later it was followed up with CT and MRI.

3. Imaging Findings

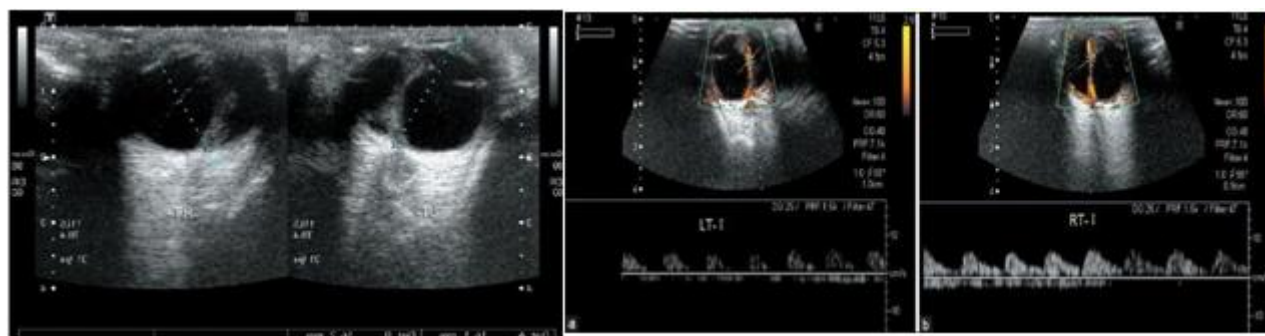
An ultrasound study was performed with a high-frequency transducer operating at 10 MHz. Gray-scale ultrasound evaluation revealed echogenic band in the posterior segment

of both globes, extending from the posterior surface of the lens capsule to the optic disc. The axial length of right and left globes were 16 mm and 15.7 mm respectively which was suggestive of microphthalmos. No calcification was seen.

Doppler examination revealed the presence of arterial flow in the band in both globes. Spectral doppler of hyaloid artery shows continuous, low-resistance biphasic forward flow.

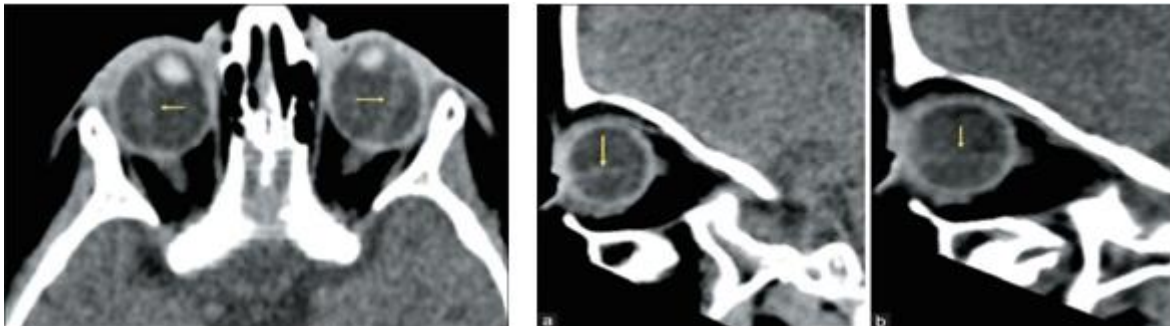
CT imaging of bilateral globes showing linear hyperdensity extending from optic nerve head to the posterior surface of the lens representing hyaloid artery in Cloquet's canal (arrows). No mass or calcification seen.

MRI imaging of the brain and orbit revealed bilateral microphthalmos. Axial T2W image shows iso to hypointense triangular retroretinal fibrovascular tissue and a central tissue stalk of hyaloid remnant in Cloquet's canal extending up to the optic disc representing the 'Martini glass' sign. Post contrast T1W image shows enhancement of the retroretinal tissue.



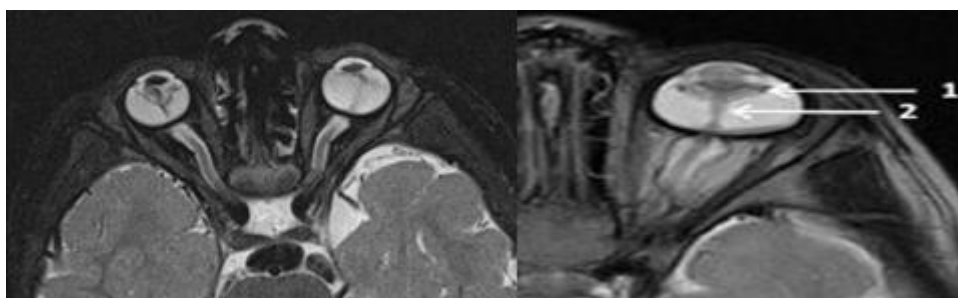
Gray-scale ultrasound evaluation revealed echogenic band in the posterior segment of both globes, extending from the posterior surface of the lens capsule to the optic disc. The axial length of right and left globes were 16 mm and 15.7 mm respectively which was suggestive of microphthalmos.

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Axial CT imaging of bilateral globes showing linear hyperdensity extending from optic nerve head to the posterior surface of the lens representing hyaloid artery in Cloquet's canal (arrows). No mass or calcification seen.

Sagittal CT of (a) right and (b) left globe showing linear hyperdensity extending from optic nerve head to the posterior surface of the lens representing hyaloid artery in Cloquet's canal (arrows).



Axial T2W image shows iso to hypointense triangular retrolental fibrovascular tissue (arrow 1) and a central tissue stalk of hyaloid remnant in Cloquet's canal (arrow 2) representing the 'Martini glass' sign.

A patent hyaloid artery, as noted in this case, is not a feature of vitreoretinal dysplasia. 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.

4. 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.

Complications of PHPV can be rupture of lens capsule, cataract, intraocular hemorrhage, secondary glaucoma, traction retinal fold, and subsequent phthisis bulbi.

Differential diagnosis of PHPV includes retinoblastoma, vitreoretinal dysplasia, retinopathy of prematurity, ocular toxocariasis.

Retinoblastoma can be differentiated from PHPV on ultrasound by presence of solid, irregular masses with heterogenous echogenicity. CT scan shows typical calcifications in most cases and typical signal characteristics on MRI, i.e., hyperintense on T1-weighted images and hypointense on T2-weighted images. In contradistinction to PHPV, in which there is usually microphthalmos, the eye typically is normally sized or enlarged in patients with retinoblastoma.

The imaging features in this case point toward the diagnosis of bilateral PHPV. This entity, although rare, should be considered in the differential diagnosis of retinoblastoma while evaluating bilateral leukocoria. Typical imaging features of ultrasound, CT and MRI can be helpful in diagnosis of bilateral PHPV.

References

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