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Persistent hyperplastic primary vitreous

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Introduction :

Persistant hyperplastic primary vitreous (PHPV) was described in detail as a clinical entity by Reese1 in 1955 in his Jackson Memorial Lecture. Goldberg in his 1997 Jackson Memorial Lecture2 renamed PHPV as persistent fetal vasculature (PFV).
PHPV is a pathologic entity resulting from abnormal persistence of the fetal fibrovascular primitive stroma (hyaloid system) of the eye3,4 which should disappear by the time of birth. The primary vitreous forms around the seventh week of life and begins involuting by 20 weeks.5 Persistence and hypertrophy of these vessels can result in PHPV in the anterior and/or posterior chambers. It can give rise to leukocoria, retinal detachment and subretinal hemorrhage.6

Case Report
A male child of 2 years and 5 months of non-consanguineous parents, presented with a fall from the table while playing at home which resulted in no serious injuries. However the parents noted a right sided white eye. Neurological examination showed leukocoria and poor vision of the right eye. An ultrasound of the eye done elsewhere revealed vitreous hemorrhage. The elder sibling was healthy.

An MRI was performed on a 1.5 tesla system with a head coil, due to findings of leukocoria and vitreous hemorrhage. Pre and post contrast scan of the orbits was performed in sagittal, axial and coronal planes. The MRI shows flattening of the right eyeball anterioposteriorly. The right eye anterior chamber was shallow due to anterior displacement of the lens. An enhancing tubular structure extending from the retrolental tissue on the way towards optic disc and nerve representing a persistent hyaloid canal containing hyaloid vessels (Cloquet's canal) was noted. Debris with a fluid level was seen in vitreous indicative of hemorrhage. The above mentioned findings in the right eyeball were consistent with persistant hyperplastic primary vitreous (PHPV) with associated vitreous hemorrhage. The parents refused any surgery as there was not much hope for restoration of the eyesight.

Discussion
Leukocoria and intraocular hemorrhage in children require imaging to establish the diagnosis as treatment depends on the etiology. Retinoblastoma is the most frequent intraocular malignancy and one of the most frequent causes of ocular hemorrhage in children. In addition to Coat's disease, persistent hyperplastic primary vitreous must be included in the differential diagnosis4,7 before embarking on aggressive therapy.
In the fetus, the primitive lens and vitreous, receive their blood supply mainly via the hyaloid system, which gains entry to the developing eye via the choroidal fissure. The

anterior portion of this system involutes at 8 months of life and is usually present in premature infants. The posterior portion of the arterial system normally regresses by seven months of life, but is also occasionally present in premature infants. The anterior and the posterior hyaloid vascular systems may persist independently or together. The pathogenesis of the hypertrophy of any portion of the primary vitreous is unknown.8

Most patients with PHPV have a combination of both anterior and posterior types. In one large series 28 of 48 patients had both types, 12 had the anterior type and 8 had the purely posterior type.9 Clinically leukocoria, strabismus and a small eye may be present in all types. The typical imaging findings of posterior PHPV include the demonstration of Cloquet's canal and a small eye. Cloquet's canal in the fetal eye contains the hyaloid artery. In a study by Howard and Ellsworth of 500 children with leukocoria, PHPV accounted for 51 of the 265 nonretinoblastoma cases. PHPV is other than retinoblastoma the most frequent cause of leukocoria in childhood.10
PHPV is often associated with severe malformation of the optic nerve and retina. Ocular malformation is usually a manifestation of more extensive disease such as Norrie's disease, Warburg's syndrome, primary vitreoretinal dysplasia or other congenital defects.

A and B scan ultrasonography may assist in precise measurement and in accurate diagnosis of PHPV.11 In some cases retrolental and intravitreal components of PHPV have been imaged only by ultrasound, but not by CT.11 Nonetheless the functional abnormalities visualized by post infusion enhancement and the simultaneous imaging of both eyes in several body positions remain advantages of the CT and MR techniques.

The diagnosis of anterior PHPV is usually obvious clinically and therefore these patients are not routinely imaged. In posterior PHPV the globe is small and contains retinal detachments that are hyperdense on CT scan and of increased signal intensity on T1 and T2 weighted MR images.12 A fine linear structure extending from the head of the optic nerve to the posterior surface of the lens represents Cloquet's canal and when seen is considered typical of PHPV. After contrast administration the vitreal abnormalities may enhance which is believed to reflect a persistent hyperplastic vitreous.

In cases of anterior PHPV the anterior chamber can be shallow and lens small and irregular.12-14 The vitreous chamber may be normal. Both clinically and by imaging the main differential diagnosis is infantile cataracts. Surgical management of PHPV depends on its type.1,6,8,9 In patients with anteroposterior PHPV in which vision is unsalvageable a lensectomy may be done and when rehabilitation is deemed possible a vitrectomy may be performed. Treatment of posterior and combined anteroposterior PHPV has a favorable outcome with most patients attaining perception of motion or light. Conversely in purely anterior PHPV a good visual outcome is achieved when aphakic correction (contact lenses) and amblyopia therapy are successful.

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