Persistent Hyperplastic Primary Vitreous: Ultrasound and Magnetic Resonance Imaging Findings

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Purpose

-To describe the types of PHPV using ultrasound and MRI
-To analyze the various presentations of PHPV and identify the typical imaging appearances
-To facilitate the differential diagnosis from other intraocular abnormalities

Methods and Materials

Were retrospectively reviewed cases of PHPV, who were evaluated using ultrasound and MRI and collected over a period of 17 years (1990-2007).

Results

We collected 12 patients with PHPV (20 eyes). The anterior form was observed in 4 eyes, the posterior form in 12 cases and the mixed form in 4 cases.

*The ultra-sound findings:

-microphtalmos

-hyperechoic, inhomogenous structure in the vitreous chamber extending from the lens till the optic nerve head and retina, vascularized on color Doppler.

*The MRI findings:

-The anterior type of PHPV: collapsed anterior chamber, an anterior segment anomaly, and a retrolental vascular membrane which demonstrated hyperintensity after contrast enhancement.

-The posterior type: microphthalmos; a tubular image, representing the hyaloid vessel; a funnel-shaped retinal detachment, with the subretinal fluid hyperintense on both T1- and T2-weighted images; the fluidfluid level, which was hypointense on both T1- and T2-weighted images and probably corresponded to the presence of hemorrhage in the subretinal space; a retrolental mass; and vitreous hemorrhage.

DISCUSSION

The fetal intraocular vascular system can be divided into anterior and posterior systems.
The anterior system supplies the iris anterior to the lens, while the posterior system consists of 3 components:

- the main hyaloid artery which mainly supplies the central primary vitreous;
- the vasa hyaloidea propria, which supplies the peripheral portion of primary vitreous;
- the tunica vasculosa lentis which supplies the iris and lens.

The primary vitreous lies between the lens and the retina. It contains the hyaloid vessels and fibrillar ectodermal tissues, then gradually disappears in the fifth to sixth month of gestation and is replaced by the secondary vitreous, which is the final vitreous.

The anterior and posterior hyaloid vascular systems regress independently.

If they fail to regress normally, then PHPV occurs.

PHPV, therefore, may be clinically divided into a pure (anterior or posterior) type or a combined type.

Most cases of PHPV are unilateral. Bilateral cases are usually accompanied by systemic diseases such as Norrie's disease, Warburg's syndrome, or other intraocular abnormalities.

The most common clinical presentations of PHPV are leukocoria and microphthalmos.

The other signs include retinal detachment, cataracts, glaucoma, an elongated ciliary process, and a shallow anterior chamber.

PHPV are divided into anterior and posterior types, with the majority of PHPV being the mixed type.

**US IMAGING FINDINGS**

- microphthalmos

- hyperechoic, inhomogenous structure in the vitreous chamber extending from the lens till the optic nerve head and retina, vascularized on color Doppler.

It is noted in that B-scan ultrasonography often made a limited contribution to a diagnosis of PHPV.

**MRI IMAGING FINDINGS**

The MRI findings of the anterior type include:

- a shallow or collapsed anterior chamber,
- a retrolental vascular membrane appearing hyperintense after contrast enhancement.

The MRI finding of the posterior type include:

- Microphthalmos: is an important finding to differentiate the posterior type from a retinoblastoma, because a microphthalmos is absent from eyes with a retinoblastoma.

- A tubular image represents the hyaloid vessel or Cloquet's canal.

- A funnel-shaped retinal detachment had subretinal fluid which appeared hyperintense on both T1- and T2-weighted images with respect to the vitreous of the contralateral eye. The high signal intensity was related to the presence of protein.

- The fluid-fluid level was revealed to be hypointense on both T1- and T2-weighted images compared with vitreous of the contralateral side and may correspond to subretinal hemorrhage. The appearance of hemorrhage depends on the level of oxygenation of the red blood cells.

- The retrolental mass appearing hypointense on T1- and T2-weighted images.

- Vitreous hemorrhaging may be present.

The retrolental mass and funnel-shaped retinal detachment are the most common MRI findings of PHPV.

THE DIFFERENTIAL DIAGNOSIS of PHPV includes retinoblastomas, Coats' disease, presumed ocular toxocariasis, and some conditions contributing to subretinal fluid or hemorrhage.

Coats' disease and retinopathy of prematurity (ROP) may have the same MRI findings as PHPV due to retinal detachment and subretinal fluid; however, retinoblastomas usually demonstrate hyperintensity on T1-weighted images and hypointensity on T2-weighted images.

MR IMAGING VS ULTRASOUND IN THE DIAGNOSIS OF PHPV:

MR imaging is superior to US in the diagnosis of PHPV:

- by revealing intraocular details and gravitational effects of the intravitreal fluid,

- identifying tissue components such as melanin, methemoglobin, deoxyhemoglobin, and proteinaceous fluid.
- has a higher sensitivity and specificity in evaluating intraocular abnormalities responsible for leukocoria, such as Coats' disease, Toxocara endophthalmitis, or retinal and vitreal disorders when B-scan ultrasonography do not offer sufficient information.

- has an important role in differentiating PHPV from noncalcified retinoblastomas

However, B-scan ultrasonography scans is easier to perform and less expensive, the detection rate of calcifications on MR images is not as sensitive as on CT scans.

MRI still plays and other intraocular abnormalities.

**Images for this section:**

![Fig. 1: MRI PHVP](image1)

**Fig. 1: MRI PHVP**

![Fig. 3: MRI RETINOBLASTOMA](image3)

**Fig. 3: MRI RETINOBLASTOMA**
Fig. 4: MRI retinopathy of prematurity (ROP)

Orbital MRI T1 FatSat Gadolinium 3 plans in space: the process is enhanced moderately and intralesional calcifications are more pronounced. No extension to the optic nerve and periorbital soft tissue.

Fig. 2: MRI. hyaloid artery
Conclusion

The presentation of PHPV at different stages was variable; the ultrasound and specially the MRI features of PHPV are able to facilitate the differential diagnosis from other intraocular abnormalities.

Unilateral involvement, microphthalmos, and a shallow or collapsed anterior chamber are the most important clinical clues in making the differential diagnosis of PHPV. When PHPV is suspected, a retrolental mass and a funnelshaped retinal detachment are the most common MRI findings.

References

Persistent Hyperplastic Primary Vitreous: Magnetic Resonance Imaging and Clinical Findings
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Personal Information