

Quiz Section

Answer to Quiz No 2 (PMR Vol 1 No 02 June 2009)

1. *What is the diagnosis?*

Persistent Hyperplastic primary vitreous (PHPV).

The B-scan of right eye reveals echogenic band extending from optic disc to posterior aspect of right eye lens. On Colour Doppler, this echogenic band shows vascular colour flow in the persistent hyaloid artery. The right eye is microphthalmic. There is also co-existent retinal detachment in right eye. Left eye is normal and shows normal axial length. The photograph of the patient reveals presence of leukocoria of right eye.

Persistent hyperplastic primary vitreous is a serious unilateral disorder of childhood presenting as leukocoria¹. It is the result of failure of regression of embryonic hyaloid artery resulting in abnormal lenticular development and secondary changes in retina and globe^{1,2}. The involved eye is frequently microphthalmic^{1,2}. On Colour Doppler ultrasound, a vascular structure is seen running from the lens to the optic disc and there is presence of fibrovascular retrolental tissue^{2,3}. There may be co-existent retinal detachment¹ as in this case. The anterior chamber is shallow and frequently there is development of

glaucoma³. The condition can be rarely bilateral⁴. It is divided into anterior and posterior subtypes however; usually the both types are co-existent.

The treatment consists of visual rehabilitation by various surgeries including cataract surgery, IOL placement, treatment of glaucoma, vitrectomy and removal of embryonic remnants⁵.

References:

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- 2) Ismail Mihmanli et al: Persistent Hyperplastic Primary Vitreous and von Hippel-Lindau disease. *J Ultrasound Med* 2002; 21: 565-568
- 3) Alex v. Levin, MORINformation, Winter 2003, Volume X: Issue 1
- 4) Sanghvi DA et al, *Australas Radiol.* 2005; 49(1): 72-74
- 5) Irene Anteby et al: Unilateral persistent hyperplastic primary vitreous: course and outcome. *JAAPOS* 2002; 6(12): 92-99

