

Persistent hyperplastic primary vitreous – The martini glass sign

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Persistent hyperplastic primary vitreous (PHPV) is a congenital lesion due to incomplete regression of the embryonic ocular blood supply (hyaloid vasculature).¹ It represents 28% of childhood presentations of leukocoria and is almost always accompanied by poor vision, microphthalmia and often retinal detachment.^{1,2,3} The absence of ocular calcifications helps distinguish PHPV from the more common retinoblastoma.⁴ The appearance of PHPV has been likened to that of a martini glass. The martini glass is represented by triangular retroretinal fibrovascular tissue and a central tissue stalk of hyaloid remnant extending to the optic disc in Cloquet's canal (see Figure 1a).⁵ The retroretinal fibrovascular tissue and stalk-like hyaloid remnant are hypointense to isointense on T1- and T2-weighted images and show enhancement post contrast administration. The globe may be hyperintense on T1-weighted images; this may represent either subhyaloid or subretinal fluid with blood degradation products (methaemoglobin) or high protein content (see Figure 1b).¹

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Competing interests

The author declares that he has no financial or personal relationship(s) that may have inappropriately influenced him in writing this article.

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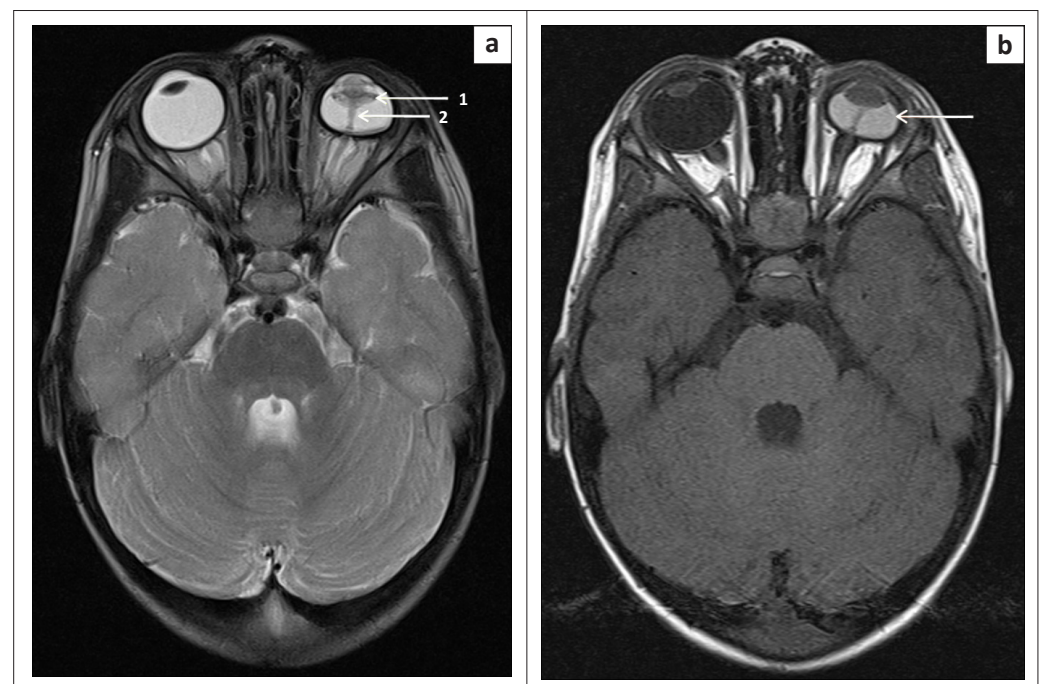


FIGURE 1: Axial T2-weighted (a) and axial T1-weighted (b) images show the triangular retroretinal fibrovascular tissue (arrow 1) and a central tissue stalk of hyaloid remnant in Cloquet's canal (arrow 2) representing the 'martini glass' sign; (b) Microphthalmia and hyperintense vitreous (arrow) on T1-weighted image due to accumulation of blood degradation products or high protein content compared to normal right globe.