Persistent hyperplastic primary vitreous – The martini glass sign

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Dates

Received: 07 Jan. 2014 Accepted: 16 Feb. 2014 Published: 20 June 2014

How to cite this article:

Bezuidenhout AF. Persistent hyperplastic primary vitreous – The martini glass sign. S Afr J Rad. 2014;18(1); Art. #597, 1 page. http://dx.doi. org/10.4102/sajr.v18i1.597

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Scan this QR code with your smart phone or mobile device to read online. Persistent hyperplastic primary vitreous (PHPV) is a congenital lesion due to incomplete regression of the embryonic ocular blood supply (hyaloid vasculature). It represents 28% of childhood presentations of leukocoria and is almost always accompanied by poor vision, micropthalmia and often retinal detachment. The absence of ocular calcifications helps distinguish PHPV from the more common retinoblastoma. The appearance of PHPV has been likened to that of a martini glass. The martini glass is represented by triangular retrolental fibrovascular tissue and a central tissue stalk of hyaloid remnant extending to the optic disc in Cloquet's canal (see Figure 1a). The retrolental 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).

Acknowledgements

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