SHORT REPORT

Persistent hyperplastic primary vitreous versus retinal detachment

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Magnetic resonance imaging (MRI) of the orbits occasionally demonstrates retinal detachment (RD); this may be as an incidental finding, as a separate entity related to orbital pathology with differing local causes, or as a part of other congenital systemic or metabolic diseases. One congenital ocular disease which could be misdiagnosed as RD is persistent hyperplastic primary vitreous (PHPV), which itself can be associated with RD. MRI scanning features of retinal detachment and associated causes can assist in differentiating both entities effectively, allowing the radiologist to reach the proper conclusions with resultant benefits in subsequent management.

Persistent hyperplastic primary vitreous (PHPV)

This is a congenital ocular lesion due to incomplete regression of embryonic ocular blood supply. The primary vitreous is supplied by the embryonic hyaloid vasculature, including the hyaloid artery (a branch of the ophthalmic artery of the developing globe), and should normally have disappeared by the time of birth.[1,2] PHPV is usually isolated and unilateral; bilateral lesions tend to be associated with systemic or syndromic conditions. The most common presenting signs and symptoms are leukocoria, poor vision and small eye.

Anterior PHPV has the best prognosis for vision, but approximately half of these patients also have an associated posterior PHPV. Abnormalities of the lens and anterior chamber are signs of a combined anterior and posterior variant of PHPV; the isolated forms are roughly equal in incidence.[1] Repeated episodes of intravitreal haemorrhage in patients with PHPV can lead to retraction of the posterior hyaloid membrane and retina by intravitreal fibrovascular tissue. This can then cause detachment of the posterior hyaloid membrane and retina.[3] Retinal detachment is common and may be associated with layering of blood or debris (subretinal hemorrhage) (Fig. 1).[1, 2]

MRI findings

PHPV typically appears as a triangular, retrolental vascular soft tissue mass, often with a central tissue stalk of hyaloid remnant connected to the optic disc. The overall shape of PHPV has been likened to a martini glass. The globe is usually small, with the vitreous typically abnormally hyperintense on both T1 and T2-weighted sequences. The signal intensity of layered haemorrhage in vitreous can vary with the age of blood (Fig. 1).

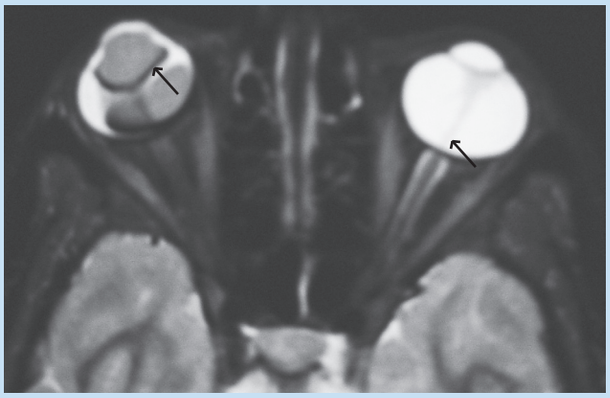

Fig. 1. Axial T2-weighted image demonstrates bilateral anterior and posterior PHPV. Left eye: (Posterior PHPV) most of the globe content gives a very high signal that could be due to intraocular haemorrhage accompanying retinal detachment. Note canal of Cloquet (hyaloid canal) connecting the optic disc to the lens. Right eye: the displaced retina appears as a thin black line (arrow). The low signal represents subretinal haematoma that extends to the anterior chamber.

Retinal detachment (RD)

Separation of the inner sensory retina from the pigmented retinal epithelium is referred to as retinal detachment. The sensory retina is part of the central nervous system, so that if there is a tear, the sensory retina cannot heal.[3] Patients typically will present with symptoms such as light flashes, floaters, peripheral visual field loss and blurred vision. RD should be considered in the differential diagnosis of any visual loss.

Risk factors for RD include advancing age, previous cataract surgery, myopia, and trauma.[4] Mostly, RD is the result of separation caused by a mass (neoplastic, such as melanoma), a fibroproliferative disease in the vitreous such as vitreo-retinopathy, often either due to prematurity or diabetes, or an inflammatory process (such as uveitis). RD may also result from retinal vascular leakage (lipoproteinaceous exudates), as seen in patients with Coats' disease, a vascular anomaly of retina (telangiectasis), or due to congenital diseases such as PHPV.[3]

MRI findings of RD

As the retina itself is beyond the limits of resolution of MRI scanning, it is only seen when outlined by the significant contrast differences between the signal intensities of subretinal effusions and of the vitreous cavity. The MRI appearance of RD varies depending on the amount of exudate and the organisation of subretinal changes. MRI can differentiate exudative RD (rich in protein; hyperintense on both T1 and T2-weighted sequences) from subretinal fluid formation as in a rhegmatogenous RD (transudate fluid; hyperintense on T2 and hypointense on T1-weighted imaging sequences). In the case of haemorrhagic RD, the MRI signal

depends on the age of the haematoma. Total RD characteristically has a V-shaped appearance, with its apex at the optic disk and its extremities toward the cilliary body (Fig. 2). Retinal detachment in axial sections obtained above or below the lens will appear as a homogeneous increase in density of the globe.[3]

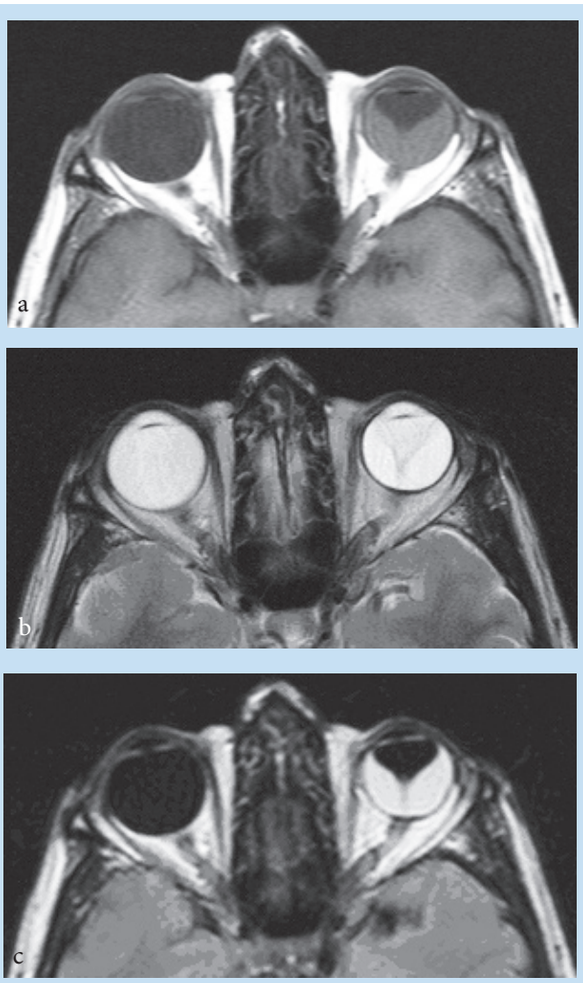

Fig. 2. (a) T1-weighted, (b) T2-weighted and (c) FLAIR sequences through the orbit.
A 'V'-shaped abnormal signal is present posteriorly in the left eye with its apex at the optic disc and the arms of the 'V' angled towards the ciliary body. Note signal increase in the space outside the 'V' on both T1-weighted and FLAIR sequences, in keeping with haemorrhage.

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2. Harnsberger HR. *Diagnostic Imaging, Head and Neck*. 1st ed. Manitoba, Canada: Altona, 2004; 11:1-10 to 11:1-11
3. Mafee MF, Peyman GA. Retinal and choroidal detachments: role of magnetic resonance imaging and computed tomography. *Radiol Clin North Am* 1987; **25**; 487-507.
4. McGehee BE, Chaljub G, Shah RK, *et al.* Bilateral retinal detachment in a patient with Vogt-Koyanagi-Harada syndrome. *Am Soc Emerg Radiol* 2005; **11**: 366-371.