Peripapillary choroidal cavitation as a feature of pathological myopia

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DESCRIPTION
A man in his 40s was referred for evaluation of peripapillary orange pigmentation in the left eye. His best-corrected (−6.00 D) visual acuity was 6/6 in each eye. Anterior segments and intraocular pressures (IOP) were normal. He had a posterior vitreous detachment in his right eye with a treated retinal break and multiple bilateral myopia-related features (axial length (AL) 25.5 mm). These included white without pressure, tilted optic nerve head (ONH) and peripapillary atrophy without myopic maculopathy. The most striking feature was unilateral (left) inferior peripapillary orange chorioretinal discolouration (figure 1). Spectral domain optical coherence tomography (OCT) demonstrated the absence of normal choroidal vasculature within this area, consistent with peripapillary choroidal cavitation (PCC, figure 2). Overlying outer retinal lamination was generally uninterrupted other than a focal vitreo-choroidal ‘sink-hole’ connection. Visual field revealed a superior hemifield/arcuate scotoma consistent with a tilted ONH.

PCC (formerly peripapillary detachment in pathological myopia or intrachoroidal cavitation) is an anatomical choroidal modification (53% unilateral) typically associated with axial myopia (−9 to −12.5 D, AL ~27 mm), although emmetropia/hyperopia accounts for −7% 2−4. High myopia (≤−6 D) is associated with younger PCC presentation. 3

The classic en face appearance is a non-elongated yellow-orange peripapillary area most often inferior to a tilted ONH. 3−5 PCC may affect up to 50% of high myopia and 10% of low myopia. 3−6 OCT has been instrumental in delineating the anatomical layer responsible for the en face appearance. 3 A hyporeflective appearance is seen at the level of the choroid, with posterior scleral deformation and/or a full-thickness neurosensory retina/retinal pigment epithelium defect contiguous between the choroidal and vitreous cavities (16%−25%). 2,3 Multiple choroidal schises can be noticed alongside cavitations potentially representing a precursor lesion. 3 Early hypofluorescence on fluorescein angiography confirms absence of choroidal tissue/circulation unless choroidal neovascularisation is present where OCT angiography may help delineate the neovascular complex. 4,5 Other pathological myopia features may be present, including tilted ONH (69%), peripapillary atrophy (98%), posterior staphylomata (40%), myopic maculopathy (14%−37%), peripheral retinal degenerations/breaks, PVD and glaucoma (38%−70%). 3−5

Clinically, these cases may be referred for concern of amelanotic choroidal nevus/melanoma; however, multimodal imaging helps to clarify their ‘cystic’ rather than solid nature. Although thought to be acquired rather than congenital lesions, PCC development is gradual (no change over 6-year follow-up). 8

The pathogenesis may be related to myopia (ie, increased AL influences angle of optic nerve entry) and ageing (few PCC cases described <30 years of age). 3 Loss of choroidal adhesion (ie, tissues detached from the inner choroid) is often associated with pathological myopia (≥−6 D). 3−5 OCT angiography may help confirm absence of choroidal vasculature (figure 2). Angiography may also confirm absence of choroidal new vessels. 4,5 Clinical cases in our cohort have been followed for up to 6 years, with occasional progression of choroidal cavitation.

Figure 1 Colour fundus photographs (TRC-50DX, Topcon, Tokyo, Japan) of the right (A) and left (B) optic nerve heads (ONH) demonstrating myopic changes on the left including tilted ONH, peripapillary atrophy and orange discolouration at the inferior disc margin. Widefield fundus autofluorescence (‘California’, Optos, Dunfermline, UK) of the right (C) and left (D) eyes showing no gross abnormalities of the retinal pigment epithelium.

Figure 2 Optical coherence tomography images (Spectralis, Heidelberg Engineering, Germany) of the left optic nerve head (ONH) and peripapillary retinal choroid. (A) Normal retina/choroid superior to ONH. (B−F) Choroidal cavitation at different planes in the inferior peripapillary region. Note intact overlying retinal layers. (D) Direct communication between the choroidal and vitreous cavities through a ‘sinkhole’.
Peripapillary choroidal cavitation (PCC) is prevalent in those with high myopia (up to 50% in some series).

Novel studies show PCC development is influenced by tractional (optic nerve sheath, dura mater, vitreous) forces on predisposed areas (eg, thinned choroid/sclera at borders of peripapillary staphyloma, tilted optic nerve head, gamma-peripapillary atrophy).

It is a benign lesion, but may be associated with choroidal neovascularisation or treatable features of pathological myopia.

REFERENCES