


# Peripapillary choroidal cavitation as a feature of pathological myopia

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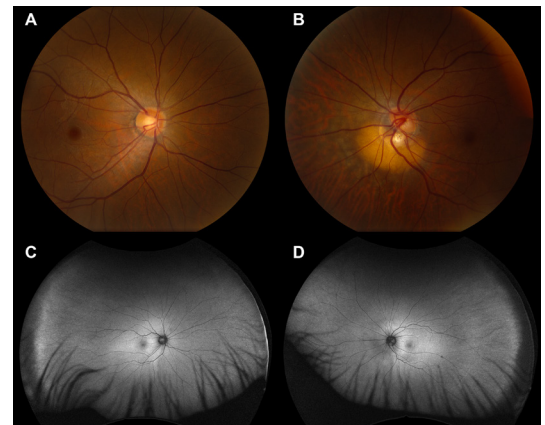
## DESCRIPTION

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 discoloration (**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.<sup>1</sup>

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  $\sim 27$  mm), although emmetropia/hyperopia accounts for  $\sim 7\%$ .<sup>2-4</sup> High myopia ( $\leq -6$  D) is associated with younger PCC presentation.<sup>3</sup>

The classic *en face* appearance is a non-elevated yellow-orange peripapillary area most often inferior to a tilted ONH.<sup>2,3</sup> PCC may affect up to 50% of high myopia and 10% of low myopia.<sup>3,5,6</sup> OCT has been instrumental in delineating the anatomical layer responsible for the *en face* appearance.<sup>3</sup> 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%).<sup>2,3</sup> Multiple choroidal schises can be noticed alongside cavitations potentially representing a precursor lesion.<sup>5</sup> 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.<sup>5,7</sup> 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%).<sup>3,5</sup>

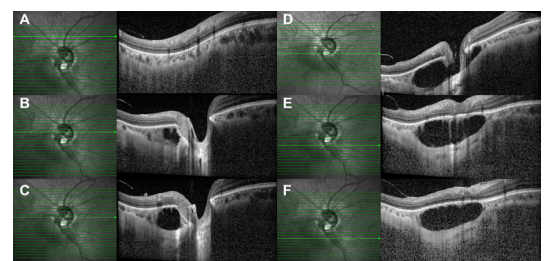
Clinically, these cases may be referred for concern of amelanotic choroidal nevus/melanoma;



**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 discoloration 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.

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).<sup>8</sup>

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).<sup>3</sup> Loss of choroidal adhesion (ie, tissues



**Figure 2** Optical coherence tomography images (Spectralis, Heidelberg Engineering, Germany) of the left optic nerve head (ONH) and peripapillary retina/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'.



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## Learning points

- ▶ 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 staphylomata, 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.

of Jacoby/Elschnig) to underlying sclera/ONH may be exacerbated by tractional/liquefactive effects of vitreous humour and increased susceptibility to physiological IOP leading to PCC.<sup>2,3</sup> Other factors include dural traction causing peripapillary suprachoroidal detachment in areas of staphyloma with tilted ONH which is promoted by adduction (ie, reading).<sup>9</sup>

Similarities to coloboma-adjacent choroidal changes have been suggested with potential genetic links (eg, *ABCB6*, *PAX2*, cadherin family), although myopia-associated genes are equally implicated (eg, *ABCC6*, *PLOD1*, collagen family).<sup>10</sup>

Although PCC may cause concern to the referring clinician, it is a benign lesion requiring no treatment; however, associated features of myopia warrant careful assessment ± treatment.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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