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VISION ACADEMY VIEWPOINT

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Pachychoroid: Current Concepts on Clinical Features and Pathogenesis

Background

The term "pachychoroid" refers to a newly described phenotype in which functional and structural choroidal changes are thought to play a key pathogenic role in a spectrum of related retinal disorders.[1] Several disorders, such as central serous chorioretinopathy, polypoidal choroidal vasculopathy, pachychoroid neovasculopathy, and pachychoroid pigment epitheliopathy, overlap in the pachychoroid spectrum. However, the lack of consensus on various phenotypes can lead to these disorders being categorized together as neovascular age-related macular degeneration. In some cases, polypoidal choroidal vasculopathy is included in clinical trials for neovascular age-related macular degeneration, despite having different clinical characteristics, natural history, and response to treatment.[2] In order to avoid such miscategorization, an understanding of how the choroid is involved in this spectrum and a better knowledge of the most relevant clinical signs of the pachychoroid phenotype are important to differentiate these disorders from other chorioretinal conditions.

A review of the literature and available evidence was conducted to[3]:

- Provide consensus on the definition of pachychoroid and the commonalities that may be present in the pathologies included in this spectrum
- Provide details on the examination, monitoring, and management of these disorders

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Variations in opinion

Viewpoint

Pachychoroid: quantitative and qualitative features

Comparisons of anatomical differences between polypoidal choroidal vasculopathy, central serous chorioretinopathy, and age-related macular degeneration in epidemiological studies have suggested differences in the pathophysiology of the phenotypes, where increased choroidal thickness appears to be a prevalent factor.[4,5] However, several choroidal changes other than increased choroidal thickness are believed to play an important pathogenic role in the development of the clinical manifestations of pachychoroid.

Quantitative features	Qualitative features
• Congestion as manifested by thickening on optical coherence tomography • Choroidal hyperpermeability • Blood-flow signal attenuation in the choriocapillaris and inner choroid with optical coherence tomography angiography	• Pachyvessels or dilated choroidal vessels in Haller's layer • Retinal pigment epithelial layer alterations and thinning of outer nuclear layer • Choroidal neovascularization • Pachydrusen

The presence of hypertrophic or congested vessels in the choroid (pachyvessels), not thickened choroid *per se*, under an area of reduced or absent choriocapillaris in the posterior pole, appears to be the most salient feature of pachychoroid. Other quantitative and qualitative features are indocyanine green angiography hyperfluorescence, blood-flow signal attenuation in the choriocapillaris and inner choroid by optical coherence tomography angiography, retinal pigment epithelial layer alterations and thinning of the outer nuclear layer, and the presence of pachydrusen.[3]

This document was prepared on behalf of the Vision Academy by Veronica Castro-Navarro, Francine Behar-Cohen, Woohyok Chang, Antonia M. Joussen, Timothy Y. Y. Lai, Rafael Navarro, Ian Pearce, Yasuo Yanagi, and Annabelle A. Okada.

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Clinical spectrum of disorders

Pachychoroid constitutes a common pathogenic process, and overlapping features[6] and progression from one disorder to another are frequently observed in the clinical spectrum. Several disorders are currently included in the spectrum and, according to their main form of presentation, can be divided into three subtypes.

Disorders with atrophic changes	Disorders with exudative changes	Disorders with neovascularization
• Pachychoroid pigment epitheliopathy	• Focal choroidal excavation • Central serous chorioretinopathy • Peripapillary pachychoroid syndrome	• Pachychoroid neovasculopathy • Polypoidal choroidal vasculopathy

Differential diagnosis

Other disorders involving uveitis and infiltrative diseases that also manifest as thickening of the choroid need to be included in the differential diagnosis. However, the presence of other pachychoroid features in the absence of systemic conditions and signs of intraocular or scleral inflammation would support the diagnosis of pachychoroid disease.

Clinicians should be aware of the different disorders included in the spectrum and of their main forms of presentation, which can be divided into those with exudation, those with neovascularization, and those with retinal pigment epithelial and chorioretinal atrophic changes.[3]

Further considerations

Consensus on pachychoroid terms is needed. Choroidal features would be useful in the management of patients, and in terms of prognosis, characteristic features of pachychoroid may serve as a predictive factor, but these are still to be confirmed in a large study.[3]

Vision Academy Viewpoints are intended to raise awareness of a clinical challenge within ophthalmology and provide an expert opinion to engage in further discussion.
They can be downloaded from https://www.visionacademy.org/resource-zone/resources/all
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