

A Note on Pathologic Myopia

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Perspective

Pathologic myopia is a type of myopia that affects up to 3% of the world's population. Pathologic myopia-related vision loss is of great clinical importance because it can be progressive, irreversible, and affects people during their most productive years. A refractive error of at least -6.00 D or an axial length of 26.5 mm or more is considered high myopia. In the early studies, the definition of pathologic myopia was inconsistent and mostly revolved around a combination of refractive error and axial length, which may simply reflect a high degree of myopia. Furthermore, there was no clear evidence to support the cutoff values chosen. Pathologic myopia is now defined as "the presence of myopic maculopathy equal to or more severe than diffuse chorioretinal atrophy." Diffuse chorioretinal atrophy, patchy chorioretinal atrophy, lacquer cracks, myopic choroidal neovascularization (myopic CNV), and CNV-related macular atrophy are all symptoms of myopic maculopathy.

The overall global prevalence is estimated to be 0.2-3.8 percent with regional variation, but differences in pathologic myopia definitions used in early epidemiological studies may limit the comparability of findings. Pathologic myopia-related visual impairment has been reported to affect 0.1 percent to 0.5 percent of people in European studies and 0.2 percent to 1.4 percent of people in Asian studies.

The main factors implicated in the development of pathologic myopia are axial length elongation and posterior staphyloma. Biomechanical forces associated with axial elongation of the eye cause ocular layer stretching and progressive thinning of the retina, choroid, and sclera.

Risk factors

Environmental and genetic factors both play a role in the development of myopia, as discussed further in the corresponding article. Currently, the roles of known myopia-associated genetic variants in the development of pathologic myopia are not well established. Older age, greater axial length, and a higher myopic spherical equivalent are primary risk factors for pathologic myopia. Other possible risk factors include female gender, a larger optic disc area, and a family history of myopia. Currently, the role of education level in the development of pathologic myopia is unknown.

Signs and symptoms

During the gradually progressive attenuation of the RPE and choroid, patients may be asymptomatic. When central CNV or foveal schisis develops, the patient may notice a focal area of blurring, metamorphopsia, or scotoma, which can lead to a rapid decline in central vision. Peripheral CNV has the potential to go undetected.

Progressive retinal pigment epithelial (RPE) thinning and attenuation develops throughout the fundus at various clinical stages. Even in young

patients with high myopia, a tessellated appearance corresponding to irregular distribution of RPE atrophy and variable light reflection can be appreciated. This hypo pigmented finding is known as peri-papillary atrophy when RPE attenuation surrounds the optic disc.

The optic disc is commonly referred to as a tilted disc because it has an oval appearance en-face. The optic nerve appears to insert at an angle into the elongated globe. The tilted appearance is caused by temporal flattening of the disc, which is caused in part by peri-papillary scleral expansion. As a result, where the sclera is directly visible, a hypopigmented myopic crescent or myopic cone is seen. Choroidal vessels will be more visible beneath the atrophic RPE in intermediate disease. However, as the disease progresses, the choroid atrophies and the choroidal vessels become less visible.

Lacquer cracks are irregular yellow-appearing bands found in the posterior pole of 4.2 percent of eyes with an axial length of at least 26.5 mm. These are Bruch's membrane breaks that could lead to choroidal neovascularization in the future (CNV). It has been reported that 29.4 percent of patients with lacquer cracks eventually develop CNV. Over time, these breaks can expand and stretch, resembling the appearance of geographic atrophy seen in advanced non-neovascular Age-related Macular Degeneration (AMD).

Fuchs spots (also known as Forster-Fuchs spots) are areas of RPE hyperplasia that are thought to be the RPE's response to previously regressed CNV. Myopic CNV is the most common cause of vision loss in high myopia, accounting for 5% to 10% of all cases of pathologic myopia.

Staphyloma development, characterised by scleral tissue outpouching typically involving the optic disc or macula, is a common occurrence, estimated to occur in 35% of eyes with high myopia. This is difficult to see with biomicroscopy but is visible with Optical Coherence Tomography (OCT) or B scan ophthalmic ultrasound. Lacquer cracks, RPE attenuation, epiretinal membrane, and macular or foveal schisis are all common features of staphylomata.

Diagnosis

Fluorescein Angiography can be used to screen myopic patients for the development of CNV. Early images may reveal a transmission defect in patches or areas of RPE atrophy around the macula and/or the optic disc. Lacquer cracks in the early and transit phases can be identified using angiography due to the linear distribution of the transmission defect. When compared to AMD, the development of CNV in pathologic myopia is smaller and less exudative. Myopic CNV may appear as a hyper fluorescent focus with a hypo fluorescent rim corresponding to hyperpigmentation at the lesion's border. Any associated haemorrhage will cause fluorescence to be blocked. Late images show leakage with or without blurring of the pigmented rim. Myopic CNV leakage is more subtle than AMD CNV leakage, and it is common for the CNV leakage to be partially or completely obscured by overlying sub-retinal haemorrhage.

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