Myopic Peripapillary Sinkhole: Prolapse of Retinal Nerve Fiber Layer and Posterior Vitreous Into a Sclerochoroidal Hollow Causing Peripapillary Choroidal Thickening and Cavitation

The pathogenesis of peripapillary choroidal thickening and cavitation, a yellow-orange, dome-shaped lesion inferotemporal to the myopic conus, is unknown. Some investigators believe the anomaly is congenital in origin owing to the presence of a cleft-like communication between the retina and choroid with vitreous prolapse and anomalous vessels. However, we observed a case that developed similar findings but due to a different cause, the gradual sinking of peripapillary retinal tissue into a sclerochoroidal cavity associated with retinal hole formation and posterior vitreous prolapse, newly termed myopic peripapillary sinkhole.

Report of a Case. A 63-year-old myopic man was first evaluated in 1984 for pigment dispersion syndrome and suspicious optic discs. Owing to the appearance of the disc and a visual field defect in his left eye, topical anti-glaucoma therapy was initiated with betaxolol hydrochloride, 0.25%, twice daily and the intraocular pressure remained between 13 and 16 mm Hg over several decades. Serial disc photographs in the left eye between 1984 (baseline) and 1994 revealed the gradual collapse and ultimate disappearance of a peripapillary retinal vessel associated with an enlarging retinal hole, adjacent disc hemorrhages, and the development of a yellow-orange peripapillary lesion (Figure 1).

Results of a 3-dimensional topographic analysis of the peripapillary tissue (Figure 2) were normal in the right eye. However, the left eye revealed a broad and deep inferotemporal peripapillary depression. Additional views showed collapsed retinal tissue with hole formation and an underlying optically empty space, likely representing vitreous prolapse into a sclerochoroidal cavity (video, http://www.archophthalmol.com).

Comment. We believe that prior to the development of myopia, the retinal nerve fiber layer is in contact with underlying sclera. We postulate that as the eye elongates, ectatic sclera pulls away from the overlying retina, creating a hollow or cavern. The roof of the cavern (the retinal nerve fiber layer) gradually collapses, possibly due to excessive overlying pressure, weakened underlying sclerochoroidal architecture, and/or malnutrition from absence of the choroid in the conus. As the retinal nerve fiber layer and accompanying vessels collapse (a telltale sign of the sinkhole), axons and retinal nerve fiber layer capillaries kink, with resulting visual field loss, disc hemorrhages, and retinal hole formation. At a crucial point, the retinal hole facilitates the escape of liquid vitreous into the underlying ectatic sclerochoroidal hollow, completing the sinkhole process. We observed 3 other cases of peripapillary thickening and cavitation that manifested in patients older than 55 years. Although we were not able to witness the development of the sinkhole, we suspect that we missed the initial retinal prolapse phase and witnessed only the end of the sinkhole process.

The definition of a sinkhole is a depression in the ground communicating with a subterranean passage and formed by collapse of a cavern roof. Our patient’s series of events seems to resemble this natural phenomenon. The entity known as peripapillary thickening and cavitation may be part of a constellation of acquired peripapillary findings as evidenced by the chance long-term observation of a series of events culminating in a find-
Progression of an Acquired Vitelliform Lesion to a Full-Thickness Macular Hole Documented by Eye-Tracking Spectral-Domain Optical Coherence Tomography

Vitelliform lesions (VLs), classically seen in young patients with autosomal dominant Best disease, are also seen as acquired lesions in entities such as adult-onset foveomacular vitelliform dystrophy, cuticular drusen, and central serous chorioretinopathy. These lesions appear as round, yellowish deposits of material exhibiting hyperautofluorescence with fundus autofluorescence imaging. Spectral-domain optical coherence tomography (SD-OCT) shows hyperreflective material in the subretinal space, often with focal thickening at the level of the retinal pigment epithelium. The natural course of these lesions is often a gradual reduction in lesion size with fragmentation and resorption of the vitelliform material and eventual photoreceptor disruption and atrophy.

Figure 2. Cirrus high-definition optical coherence tomography 3 (Carl Zeiss Meditec) analysis of a myopic peripapillary sinkhole. A, Topographical 3-dimensional analysis of the right eye shows normal surface peripapillary anatomy with the upper and lower pole of the disc indicated by arrows. B, The curved arrow in the left eye indicates a deep inferotemporal peripapillary depression; the optic nerve poles are located between the straight arrows. The depression may also extend inferiorly between the arrowheads, indicating localized loss of the retinal nerve fiber layer. C, Cross-sectional analysis reveals prolapsed retinal nerve fiber layer tissue (arrows), between which is the retinal hole. Diamond indicates the optically empty sclerochoroidal cavity. The inferior margin of the disc (lower arrowhead) appears to be on a much lower plane than the rest of the disc margin (upper arrowhead). D, The enhanced choroidal view of the left eye reveals the dark area below the disc (arrow), an area with loss of reflectivity, that likely represents prolapsed liquid vitreous. E, Horizontal raster line analysis through the inferotemporal sinkhole. The broad depression shows a hole (arrow) in the prolapsed tissue that communicates to the underlying sclerochoroidal cavity occupied by an optically empty substance, presumably liquid vitreous.

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