# A 65-Year-Old Female with a Blind Spot in Her Right Eye

Abdallah M. Jeroudi, MD; Kevin J. Blinder, MD

## Introduction:

A 65-year-old myopic female presented to The Retina Institute for evaluation of a blind spot in her right eye. She reported noticing an area of blurred and absent vision on the temporal side of her visual field in her right eye which she became aware of for the last 2 months. She denied a prior history of trauma, headaches, nausea, vomiting, tinnitus, transient loss of vision, weakness, numbness, or preceding viral syndrome. The patient had a past medical history of hyperlipidemia for which she was taking rosuvastatin and low-dose aspirin. She was a -6.00D myope. She denied a family history of blindness, glaucoma, or age-related macular degeneration.

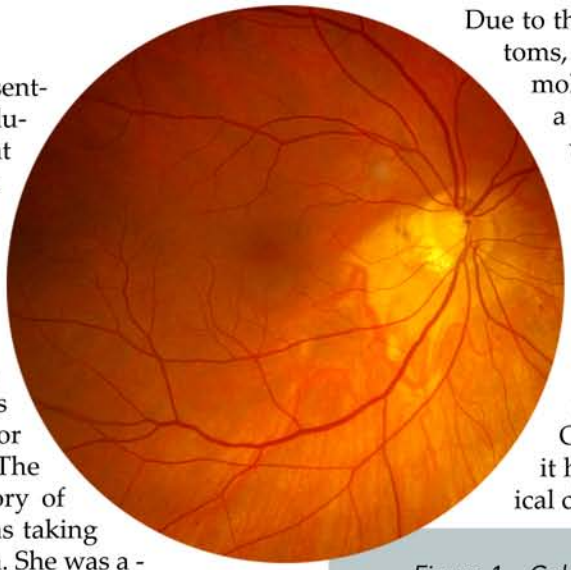

*Figure 1 – Color Photograph showing yellow-orange discoloration around the optic nerve head.*

Visual acuity measured 20/25-1 OD and 20/25+1 OS. There was no relative afferent pupillary defect. Extraocular muscles were intact. Intraocular pressure (IOP) measured 15 OD and 14 OS by applanation tonometry. The anterior segment exam was notable for 2+ nuclear sclerotic cataract OU.

Dilated fundus examination revealed a cup to disc ratio of 0.4 with sloped temporal margins in both eyes with peripapillary atrophy (PPA) more prominent OD > OS (Figure 1). The vitreous was clear. The macula, vasculature, and periphery appeared normal.

Review of Humphrey Visual Field (HVF) testing from the referring physician showed an enlarged blind spot in the right eye with superior paracentral visual field changes (Figure 2). Ocular coherence tomography (OCT) in both eyes showed normal foveal contour and retinal thickness (Figure 3). Fluorescein Angiography (FA) was performed to evaluate for a potential vascular cause of her symptoms and exhibited normal flow in both eyes.

Due to the relative acute-onset of her symptoms, she was referred to neuro-ophthalmology for evaluation which included a normal MRI of the brain and orbits with and without contrast. She was diagnosed with presumed Normal Tension Glaucoma (NTG) and started on nightly prostaglandin analogue drops in both eyes.

On re-presentation 2 months after her evaluation, she did not report a change in her symptoms. Repeat OCT was unchanged, but did exhibit hyporeflectivity and loss of the typical choroidal markings in the peripapillary area OD. OCT obtained through the right optic nerve using enhanced depth imaging (EDI) highlighted this peripapillary choroidal cavity and choroidal schisis (Figure 4). In reviewing the images, this choroidal schisis was seen on multiple rasters of the OCT located under the areas of yellow-orange discoloration seen on color photography OD (Figure 5).

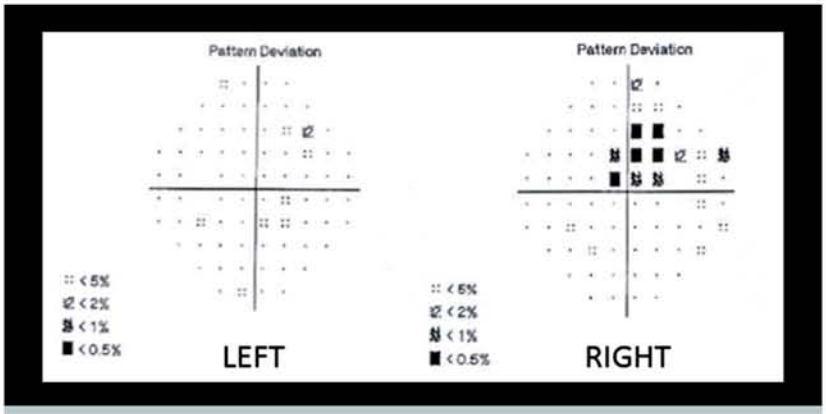


*Figure 2 – Humphrey Visual Field OU, 30-2 SITA standard protocol with enlarged blind spot and superior paracentral scotoma.*

## Diagnosis:

The differential diagnosis of this choroidal schisis seen on OCT imaging is narrow and includes coloboma and Peripapillary Choroidal Cavitation (PCC). She did not have a typical colobomatous appearance. The patient was diagnosed with Peripapillary Choroidal Cavitation (PCC).

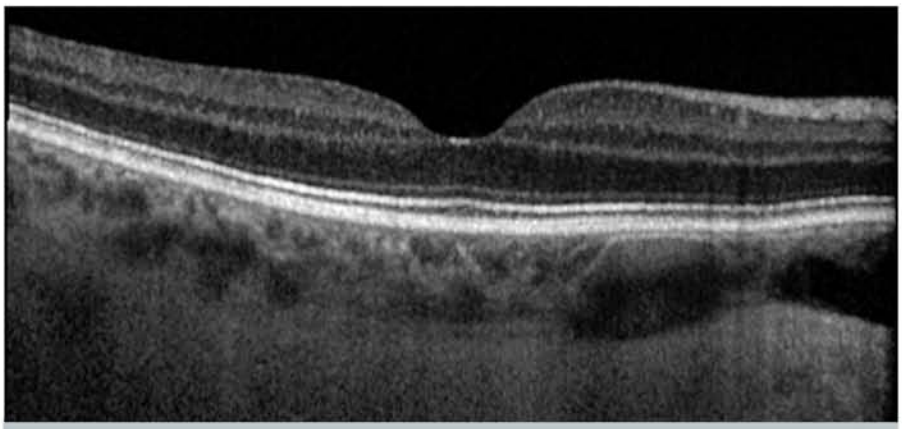

*Figure 3 – OCT macula right eye on initial presentation.*

## Discussion:

Initially described by Dr. Fruend in 2003, PCC was originally known as Peripapillary Detachment in Pathologic Myopia (PDPM) [1]. In the 20 eyes of 17 patients with this condition, all patients exhibited a yellow-orange peripapillary lesion which was located primarily inferior to the myopic conus. This lesion was either semicircular or triangular with the apex of the lesion pointing inferiorly. Time-domain OCT in this area exhibited what appeared to be a retinal pigment epithelium (RPE) detachment in the area of involvement. There did not appear to be an effect on visual acuity in their series of patients with stability of these lesions demonstrated on multi-year follow up [1].

With more advanced OCT technology, in 2011 Dr. Freund proposed renaming the entity Peripapillary Choroidal Thickening and Cavitation (PCC) due reports noting the OCT appearance of intrachoroidal hyporeflectivity below the normal plane of the RPE rather than what appeared to be a pigment epithelial detachment with older OCT technology [2,3]. Further studies on this entity have further characterized the clinical aspects of this condition.

A large study from Taiwan identified 122 eyes of 83 patients diagnosed with PCC and noted that this condition was identified at an average age of 48 ± 13 years with a slight female predominance [4]. Of the cases, 74% were detected in high myopes (worse than -6.00D), 20% in low myopes (better than -6.00D), and 6% among emmetropes and hyperopes. All patients demonstrated choroidal thickening, cavitation, and schisis on OCT imaging, but only 53% exhibited the classic funduscopic appearance of a yellow-orange lesion inferior to the myopic conus. Although the authors could not identify the clinical significance of PCC, they did note that 63% of these patients had glaucoma or were considered glaucoma suspects [4].

In another large series from Japan, the charts of 324 patients with high myopia were reviewed identifying 31 eyes with PCC [5]. All 31 PCC eyes as well as 564 eyes with high myopia without PCC had Goldmann Visual Field data. In their series, 71% of the PCC eyes exhibited glaucomatous visual field defects compared to only 23% of non-PCC eyes. All PCC eyes had normal intraocular pressures (average 13 mmHg, range 10 to 22 mmHg). In some of the patients, the visual field defects did not correspond to the PCC lesion location. Although the visual field defects in these highly myopic eyes may not be purely attributable to glaucomatous damage, the presence of PCC may be a novel marker for glaucoma or visual field defects in patients with high myopia [5].

The patient was counseled that her visual field defect may or may not be related to the PCC seen by OCT. Previous experience in the literature has shown that PCC remains stable over time. Since she presented with symptoms of a visual field deficit and such a high proportion of patients with PCC are considered to have glaucoma, be glaucoma suspects, or have a history of

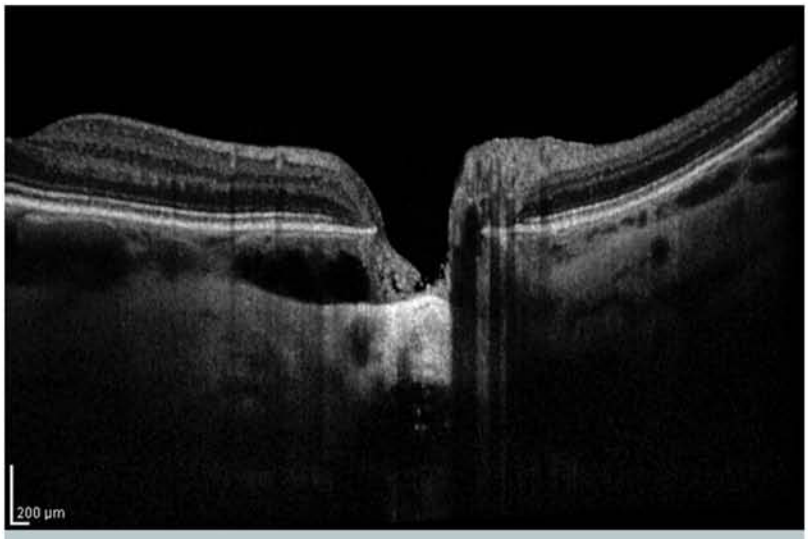


*Figure 4 – OCT of optic nerve head showing intrachoroidal cavitation.*

visual field deficits, she was counseled to continue treatment and surveillance for presumed Normal Tension Glaucoma.

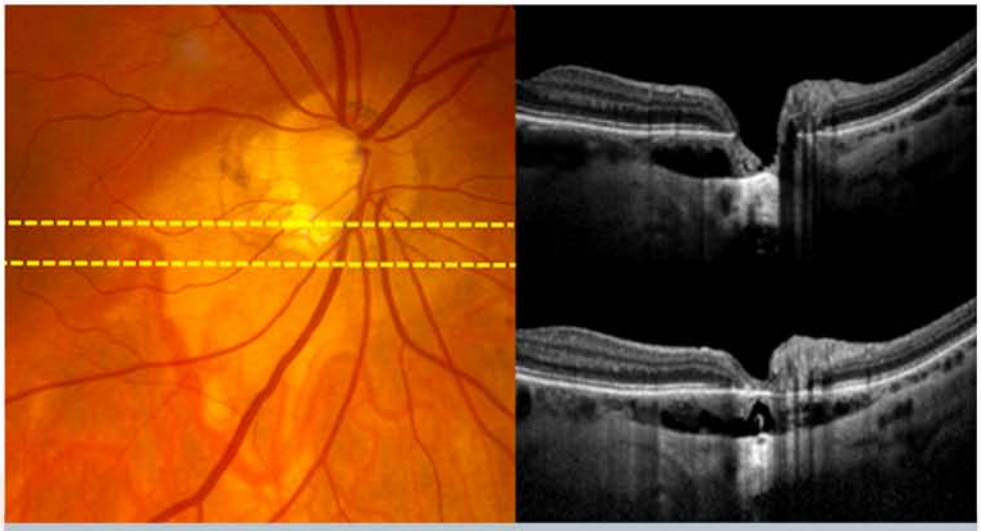

*Figure 5 – Comparison of Color Photograph to OCT showing intrachoroidal cavitation underlying the area of yellow-orange discoloration.*

## Conclusions:

Peripapillary Choroidal Cavitation (PCC) classically presents as a yellow-orange lesion typically inferior to the myopic conus in patients with high or moderate myopia. OCT imaging definitively identifies this condition as thickening, cavitation, and schisis of the peripapillary choroid with intact overlying retinal pigment epithelium and retina. In PCC eyes, 71% exhibit glaucomatous visual field defects (despite normal IOP) which may not always correspond to the location of the PCC as compared to 23% of similarly highly myopic eyes. Most PCC patients carry a diagnosis of glaucoma or are glaucoma suspects.

**References:**


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5. Shimada N, Ohno-Matsui K, Yoshida T, et al."Characteristics of peripapillary detachment in pathologic myopia." Arch Ophthalmol. 2006; 124(1): 46-52.