Polypoidal Choroidal Vasculopathy

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Abstract

Polypoidal choroidal vasculopathy (PCV) is a disease of the choroidal vasculature that may result in sub-retinal hemorrhage and serous detachment of the retinal pigmented epithelium (RPE), leading to sub-retinal fibrosis and, sometimes, permanent vision loss. This report describes a case of PCV in an African-American female over the course of 1 year and demonstrates the progression of PCV, from being relatively asymptomatic to the development of a visually significant sub-retinal hemorrhage. She is currently being treated with Avastin intravitreal injections with some resolution of her symptoms and a reduction of sub-retinal bleeding.

INTRODUCTION

Polypoidal choroidal vasculopathy (PCV) is a disease of the choroidal vasculature that includes a sub-retinal branching vascular network with interconnected reddish-orange dilated vessels, which form this condition's characteristic polyps. Sub-retinal hemorrhage and serous detachment of the retinal pigmented epithelium (RPE) may occur with this condition, leading to sub-retinal fibrosis and possible permanent vision loss. Patients are often asymptomatic, but may present with an acute or gradual decrease in vision if the macula is affected. This report describes a case of PCV in an African-American female as she progressed from being relatively asymptomatic through the development of sub-retinal hemorrhage. Optical Coherence Tomography (OCT), intravenous fluorescein angiography (IVFA), and fundus photography were used to demonstrate the different presentations associated with this condition and highlight the differences between PCV and one of its primary differential diagnoses, age-related macular degeneration (AMD).

CASE REPORT

A 62-year-old African-American female presented at the eye clinic for an annual exam with no ocular complaints. She reported stable vision, but needed a new spectacle prescription for reading and night-driving. Her medical history consisted of asthma and gastroesophageal reflux disease that were being controlled with albuterol and omeprazole, respectively. Her ocular history consisted only of refractive error and she had no pertinent family ocular history.

Uncorrected entering VA's were OD: 20/30, PH 20/25 and OS: 20/30, PH 20/25. Her refraction was OD: -0.25 -0.25 x140 and OS: -0.25 -0.75 x005 with an add of +2.50, and she was corrected to 20/20 OD, OS, and OU. Her pupils were equal, round, and reactive to light, and there was no afferent pupillary defect (APD). Ocular motilities were full and smooth OD, OS and her confrontation visual fields were full with finger-counting in all quadrants in both eyes. Anterior slit lamp examination showed no remarkable findings. Goldmann applanation tonometry measured pressures of 17 mmHg in both eyes. Dilated fundus exam showed trace nuclear sclerosis of the lens in both eyes and a posterior vitreous detachment in both eyes. The optic nerves had a C/D ratio of 0.5 round in both eyes, and were pink and distinct. The a/v ratio was 2/3 with normal caliber and no crossing changes in both eyes. The maculae were flat with no sub-retinal fluid. The right eye showed multiple peripapillary yellow-

orange sub-retinal clustered lesions inferior temporal to the optic nerve (Figure 1). There was a small area of thickening about 1 disc diameter inferior temporal to the optic nerve head in the right eye. The posterior pole of the left eye was normal. The peripheral retina was flat and intact in both eyes with mild areas of reticular changes. OCT showed a normal foveal contour and no abnormal findings in the left eye. The foveal contour of the right eye was normal, but there were several peripapillary RPE elevations with overlying sub-retinal fluid (Figures 2 and 3). After a careful review of these findings, the patient was diagnosed with polypoidal choroidal vasculopathy. No treatment was indicated at the time of the initial visit as there was no macular involvement. It was recommended that the patient be followed by a retina specialist and she had follow-ups for the next 3 months with no change in the retina appearance, no presence of leakage as measured by IVFA and no increase in thickness of sub-retinal polyps as assessed by OCT.

Figure 1: *Fundus photograph of initial presentation, OD*

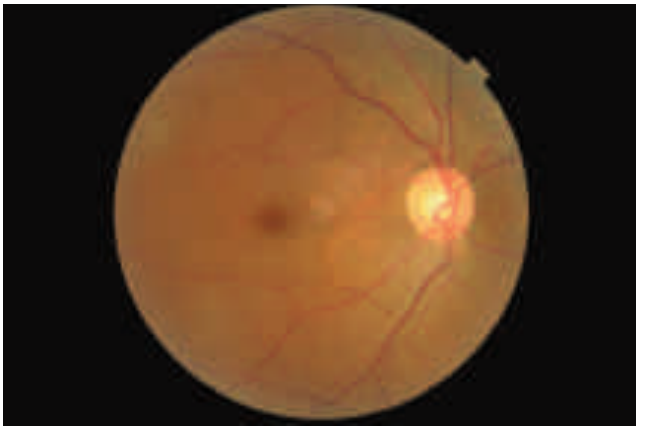


Figure 2: *Spectralis OCT of initial presentation, foveal cross-section*

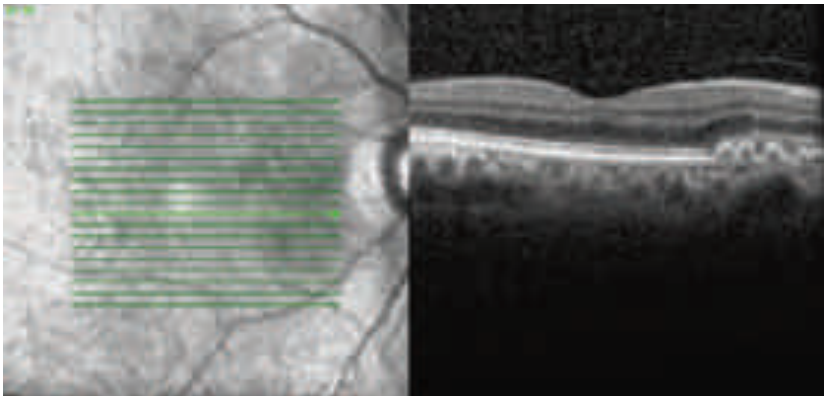
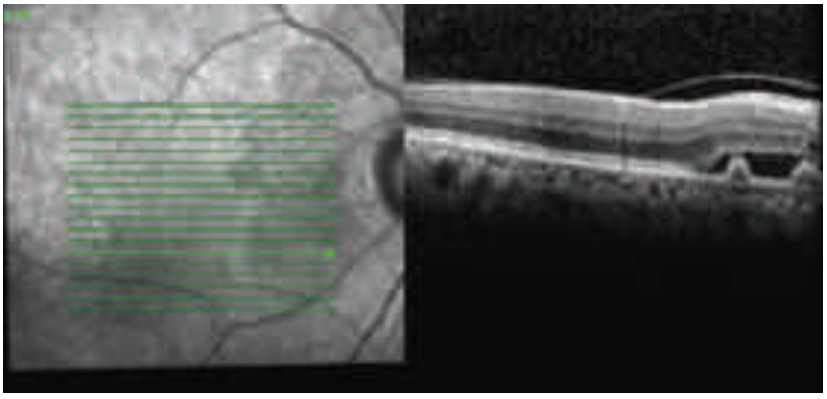


Figure 3: *Spectralis OCT of initial presentation, inferior macular cross-section*



Approximately 4 months after the diagnosis, the patient presented at the clinic for an emergency visit with complaints of blurred vision and new onset of flashes and floaters in the right eye for two days. Her vision with glasses was 20/40-1 with eccentric viewing OD and 20/20 with the left eye. There was no improvement with refraction. Her pupils were equal, round, and reactive to light, and there was no APD. Ocular motilities were full and smooth OD, OS and her confrontation visual fields were full with finger-counting in all quadrants in both eyes. The right eye had central metamorphopsia and an incomplete central scotoma as measured on an Amsler grid. Anterior slit lamp examination showed no remarkable findings. Goldmann applanation tonometry measured pressures of 17mmHg in the right eye and 19mmHg in the left eye. Dilated fundus exam showed trace nuclear sclerosis of the lens in both eyes and a posterior vitreous detachment in both eyes. The optic nerves had a C/D ratio of 0.5 round in both eyes, and were pink and distinct. The a/v ratio was 2/3 with normal caliber and no crossing changes were noted in either eye. The macula and posterior pole of the left eye were flat without fluid. In the right eye, a large sub-retinal hemorrhage measuring 5 disc diameters (DD) horizontal by 4DD vertical was present throughout the macula and inferior to the optic nerve with overlying sub-retinal fluid extending into the inferior arcade as viewed on clinical exam and confirmed by OCT (Figure 4). The peripheries of both the right and left eyes were flat and intact without breaks, holes or tears. OCT showed a large area of significant sub-retinal fluid and large elevations in the RPE inferior to the optic nerve extending to the macular region in the right eye (Figures 5 and 6). An appointment was scheduled for evaluation by a retina specialist within a week. The retina specialist initiated treatment with Avastin intravitreal injections. The patient returned for a follow-up appointment in our clinic four months later, after receiving four Avastin injections in the right eye. Her vision was corrected to 20/30 OD and 20/20 OS. All exam findings were stable, but she showed a significant reduction in sub-retinal fluid in the right eye (Figure 7). There was no sub-retinal fluid in the macular region and the sub-retinal hemorrhage had decreased substantially to a small area approximately 1DD in size (Figures 8 and 9). She has continued to be followed by a retina specialist.

Figure 4: *Fundus photograph at four-month follow up*

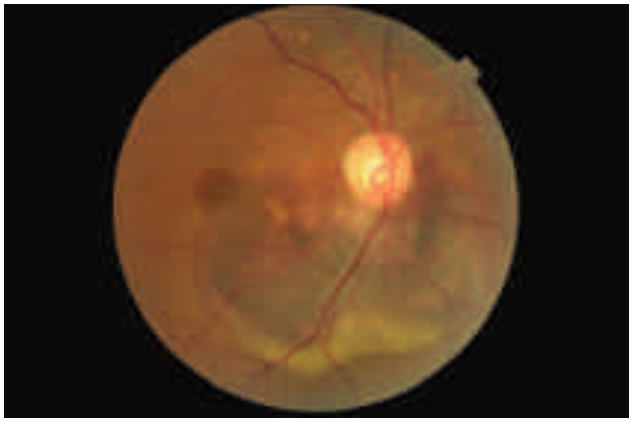


Figure 5: *Spectralis OCT at four-month follow-up, foveal cross-section*

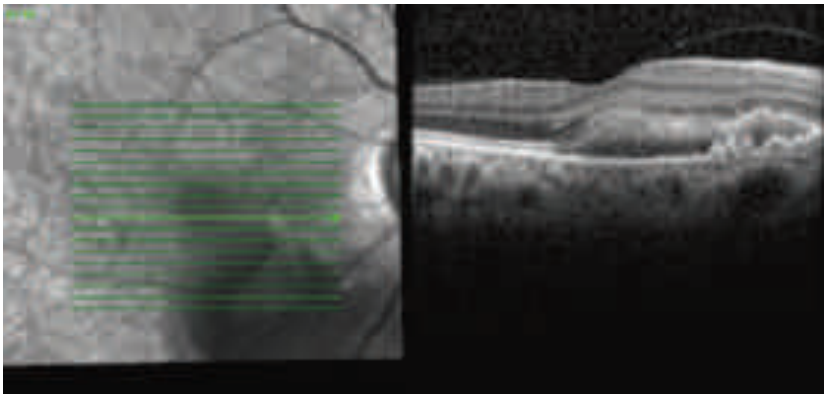


Figure 6: *Spectralis OCT at three-month follow up, inferior macular cross-section*

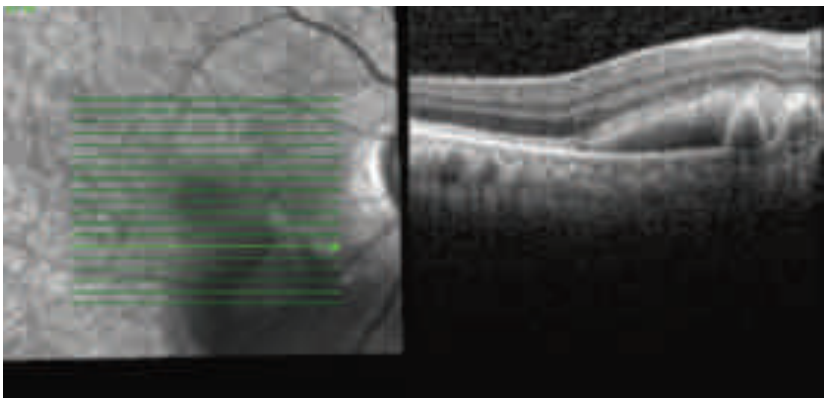


Figure 7: *Fundus photograph at seven-month follow-up*

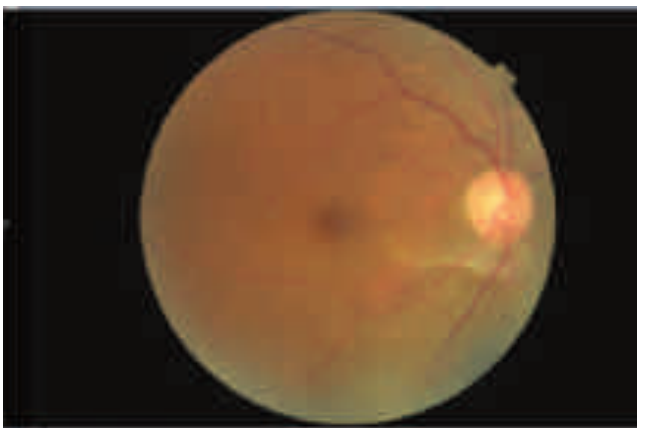


Figure 8: *Spectralis OCT at eight-month follow-up, foveal cross-section*

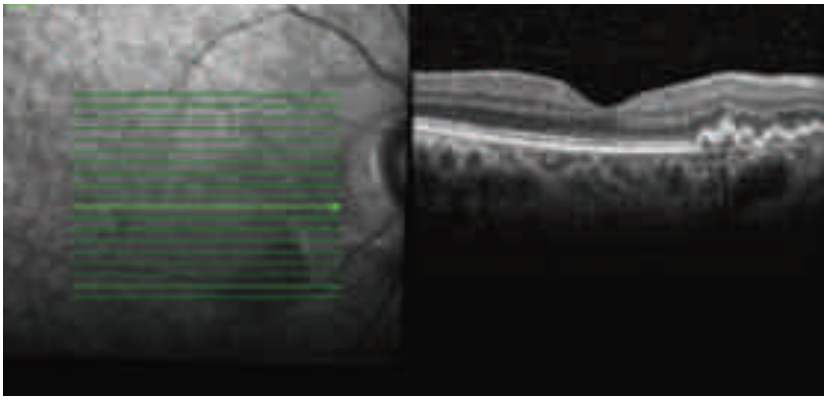
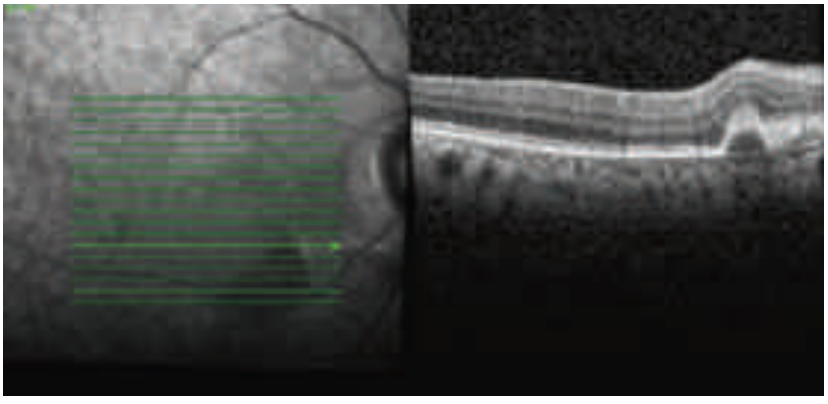


Figure 9: *Spectralis OCT at seven month follow- up, inferior macular cross-section*



DISCUSSION

While PCV is currently considered a variation of exudative age-related macular degeneration (AMD), some reports suggest that PCV may be a distinct vascular abnormality.[2] Although PCV is phenotypically similar to AMD, it tends to have a very different natural progression, target demographic and response to treatment.[2,6] PCV is more common in Asian and African-American populations, while AMD is more common in Caucasian patients.[2,6] Different races tend to display different genetic markers for PCV and AMD, but risk alleles tend to be the same in both conditions, suggesting that the clinical appearance and efficacy of treatment for PCV may be affected by other additional genes or modulating factors amongst different ethnicities.[1,2,6] PCV is more common among men in Asian populations (22-37% female), but more prevalent in women in Caucasian populations (52-65% female).[2] While PCV affects patients aged 21-93 years (mean 68.4 years), AMD is most often seen at around 80 years of age.[2,6,7] Several studies have determined the systemic and ocular risk factors for PCV, which include systemic hypertension, elevated C-reactive proteins, history of central serous chorioretinopathy and cigarette smoking.[2]

The clinical appearances of PCV and AMD are quite similar in some regards, but they do display some distinct variations. Both diseases show abnormal neovascularization of retinal tissues and sub-retinal fluid accumulation that result in sequelae such as sub-retinal hemorrhage and pigment epithelial detachment (PED).[1] PCV has a clinical appearance that differ from that of AMD. PCV is often found in the peripapillary and extramacular area whereas AMD exists exclusively within the macula.[2] PCV may also be located outside the vascular arcades of the posterior pole.[2] PCV presents with signs of a branching vascular network with interconnected orange-red dilated protrusions, or polypoids, below the RPE instead of the dru-

sen, pigment clumping and geographic atrophy seen most commonly in AMD.[1] Subretinal polypoidal vascular structures can be associated with PED and can induce exudative changes such as serous retinal detachment (SRD), sub-retinal hemorrhage, and sub-retinal fibrosis.[1,2] It has been reported that polyps are commonly present at the margins and inside the PED.[2] Micro-rips, hyperplasia and atrophy of the RPE are often found overlying or surrounding the vascular nodules.[2] Because of the variation in the locations of polypoidal lesions compared to AMD, the baseline mean visual acuity in PCV is, on average, better than that in AMD.[2,4,7] The presence of serous PED for longer than 12 months is a suspected risk factor for a reduction in visual acuity in PCV patients due to increased infiltration of the polyps into the subretinal pigment epithelial space.[2]

PCV is best diagnosed with indocyanine green angiography (ICGA) rather than IVFA due to its ability to image the choroidal circulation below the RPE.[2,4] With IVFA, one cannot reliably differentiate PCV from exudative AMD secondary to choroidal neovascularization.[4] Despite being the standard for diagnosis, ICGA is not widely used in countries that do not have a high prevalence of PCV.[1] SD-OCT can also aid in the early detection of the disease when other imaging studies are not readily available.[8]

Treatment strategies also differ between AMD and PCV. Research has demonstrated that the use of anti-VEGF therapy provides some improvement in vision in patients with PCV.[1] Unlike AMD, patients with PCV have been shown to respond more favorably to verteporfin photodynamic therapy (PDT) after failing to respond to anti-VEGF therapy.[1,2,6] However, despite a better initial reaction to PDT, research showed that the improvement in VA was not significant after 12-36 months of initial treatment with PDT alone.[2-4] Recent studies suggest that the use of PDT in conjunction with anti-VEGF therapy provides an added benefit in the treatment of PCV.[1] The EVEREST study reported that PDT monotherapy and combination therapy (PDT + anti-VEGF) achieved higher proportions of patients(77.8 % and 71.4%) with complete polyp regression than anti-VEGF injection therapy alone (28.6%) at 6 months, but did not conclude whether monotherapy or combination therapy was better for visual acuity outcomes.[1-4] Intravitreal injection of an anti-VEGF agent (ranibizumab or bevacizumab) remains a contentious treatment choice for PCV.[2] More recently, the "LAPTOP" study revealed that PCV patients who received a series of three monthly injections followed by as-needed injections of ranibizumab had a better improvement of visual acuity (30.4%) than those who received PDT monotherapy (17%) at 12 and 24 months, though the injections did not result in complete polyp regression.[2,3,9] In fact, treatment with anti-VEGF therapy exclusively in patients who respond favorably may avoid complications of PDT such as sub-retinal hemorrhaging, exudative retinal detachments and choroidal vessel occlusion.[1,3] In addition, recent studies have shown the efficacy of argon laser for PCV located outside of the foveal region and may be a potential treatment option for certain cases of PCV.[6]

This case demonstrates the importance of the appropriate diagnosis of a retinal condition that, at first glance, may be mistaken for exudative AMD. A correct diagnosis is critical for ensuring that the patient receives proper, timely care while avoiding unnecessary, costly and potentially harmful procedures. Readily available technology, such as OCT, can assist in making a correct diagnosis and help with monitoring for progression in an attempt to ensure a positive visual outcome for the patient. ●

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