

Clinical & Refractive Optometry is pleased to present this continuing education (CE) article by Dr. William J. McGill and Dr. Breyne Middleton, entitled **Polypoidal Choroidal Vasculopathy**. In order to obtain a 1-hour Council of Optometric Practitioner Education (COPE) approved CE credit, please refer to page 50 for complete instructions.

Polypoidal Choroidal Vasculopathy: A Case Report and Review

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ABSTRACT

Polypoidal choroidal vasculopathy (PCV) is a rare form of choroidal neovascularization. Once thought to occur only in middle aged darkly pigmented females, the prevalence of this disease is growing as it is identified in men, Caucasians, and elderly patients. The case of a 61-year-old African American female with juxtapapillary choroidal neovascular membrane and recurrent pigment epithelial detachments is presented. Photographs, fluorescein angiography and indocyanine green angiography were used to diagnose and document the disease's dramatic progression in this patient. PCV should be considered in cases of atypical wet macular degeneration (AMD) or recurrent idiopathic central serous chorioretinopathy (ICSC). Differentiating PCV from AMD-related choroidal neovascular membrane (CNVM) is important because of their different risk factors, clinical interpretation, natural course and outcome of treatment. PCV often has a better response to treatment than do other diseases involving CNVM; therefore, the importance of early detection and referral by optometrists cannot be understated.

CASE REPORT

A 61-year-old African American female veteran returned to the eye clinic for follow-up on a resolving branch retinal vein occlusion OS. The vein occlusion had been found incidentally at her annual diabetic eye exam five months prior. At the six-week follow-up, the vein occlusion had showed some signs of resolution.

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Her medical history was significant for diabetes mellitus type II (controlled with glyburide, metformin and insulin), hyperlipidemia (controlled with simvastin), and systemic hypertension (controlled with felodipine, clonidine patch, metoprolol, hydrochlorothiazide, and lisinopril). She also had a history of arthritis and sciatica for which she was taking tramadol, cyclobenzaprine and diclofenac.

At this visit her vision was stable at 6/6 (20/20) OS, but dilated fundus exam revealed a large area of retinal elevation nasal to the optic nerve that also spread inferior and superior to the macula with hemorrhages and hard exudates within the area of thickening (Fig. 1).

Fluorescein angiography (FA) performed two weeks later showed a parapapillary choroidal neovascular membrane (CNVM) with what appeared to be choroidal aneurysmal malformations (Figs. 2A, B). Polypoidal choroidal vasculopathy (PCV) was suspected and treatment options including focal laser photocoagulation and Macugen injections were discussed. The patient opted for continued observation.

Four months later, the patient returned complaining of vision loss OS over the previous two weeks. Visual acuity was count fingers at three feet. Dilated fundus exam at this visit revealed intraretinal and subretinal hemorrhages temporal to the optic nerve with a large orange-yellow lesion in the superior macula. The lesion



Fig. 1 Multiple parapapillary PED and hard exudate.

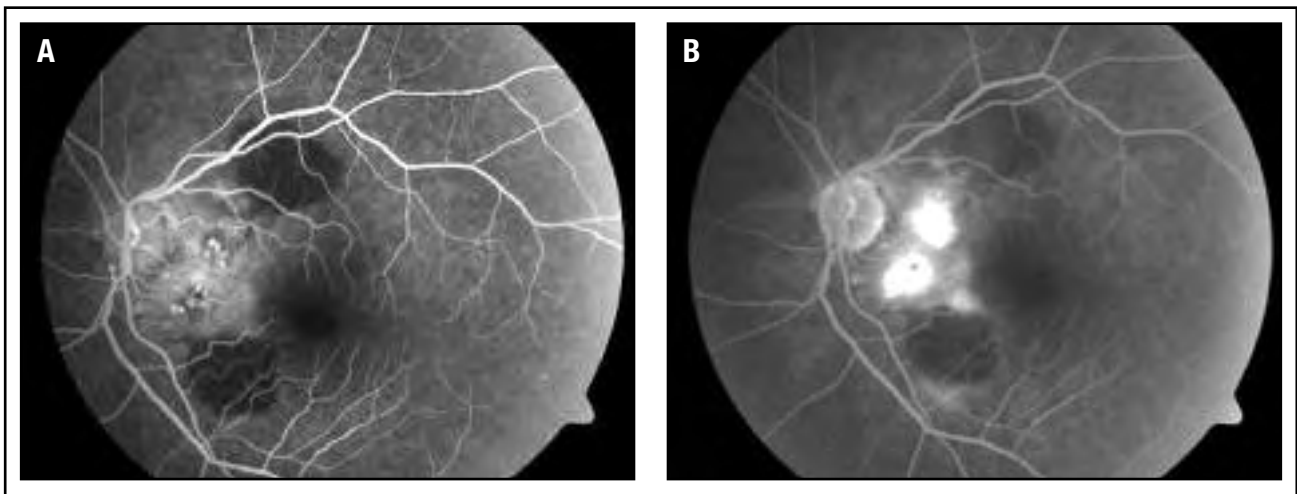


Fig. 2 (A) Note hyperfluorescence **(B)** and late staining.



Fig. 3 Large PED with retinal hemorrhages and large lipid exudate.

involved the superior fovea and was approximately 4-5 disc diameters in size. Retinal pigment epithelial elevation was also noted temporal to the optic nerve (Fig. 3).

Due to the elevation and appearance of the lesion, neoplastic disease needed to be ruled out. An MRI, a mammogram, and additional laboratory testing were ordered, all of which returned as normal. FA was repeated and revealed no intrinsic circulation in the area of the lesion, only blockage. B-scan ultrasonography was performed and showed serous fluid with a hypochoic mass and no choroidal excavation.

Indocyanine green angiography (ICGA) was ordered to confirm the diagnosis of PCV. The ICGA showed choroidal blockage superior to the macula. Typical of PCV, the peripapillary CNVM had a “cluster of grapes” appearance which hyperfluoresced (Fig. 4). The patient

was followed closely over the next year while the exudates and hemorrhages resolved and the serous detachment improved. The patient’s vision fluctuated between count fingers and 6/9 (20/30-). It seems to have stabilized at 6/15 (20/50-) at the last two visits (Fig. 5).

DISCUSSION

Since the initial description by Stern and colleagues in 1985,¹ the clinical entity now known as polypoidal choroidal vasculopathy (PCV) has been increasingly recognized.^{2,4} PCV was initially referred to as the “posterior uveal bleeding syndrome”⁵ and “multiple recurrent serous sanguineous retinal pigment epithelial detachments in black women”.¹ In 1990, Yanuzzi et al² coined the term idiopathic polypoidal choroidal vasculopathy, referring to peculiar nodular protrusions and their connected vascular channels emanating from the normal surface layer of the choroid.

Although PCV is most commonly diagnosed in patients between the ages of 50 and 65 years, the age of diagnosis can range from 20s to 80s.⁶ Sex predilection points toward women, who are affected over men by a ratio of 4.7:1.³ African-Americans and Asians are more at risk in developing this distinct choroidal disorder.^{1,6} However, PCV has been found to be present in about 8-13% of white patients, and has also been reported in Irish, French, German, and Italian patients.⁶ While PCV tends to be bilateral,^{2,3} the majority of patients will initially present unilaterally; eventually, similar lesions will likely develop in the fellow eye.⁶

PCV is characterized by the presence of recurrent serosanguineous pigment epithelial detachments (PED) and neurosensory retinal detachments.^{2,3, 6-7} To date, the pathogenesis is unknown; however, recent findings have determined that increased concentrations of vascular

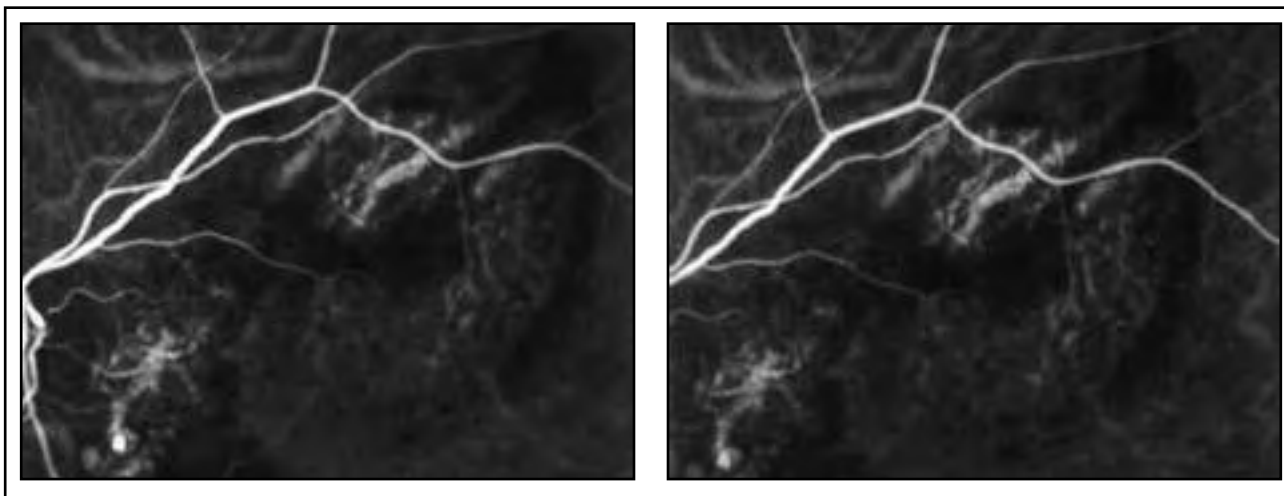


Fig. 4 ICGA indicates "cluster of grapes" hyperfluorescence near optic nerve.

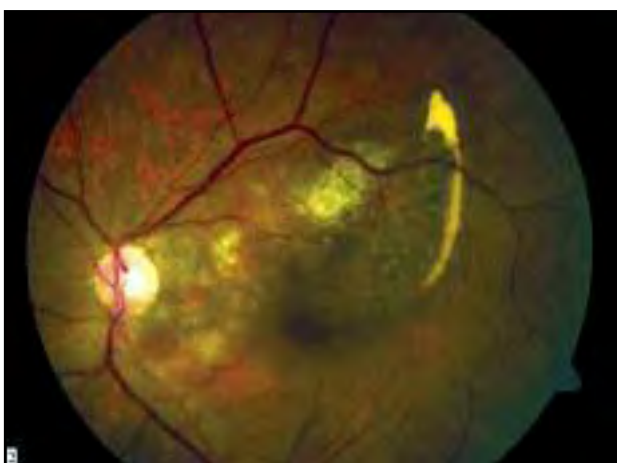


Fig. 5 Note residual lipid and RPE atrophy at site of former PED.

endothelial growth factor (VEGF) have been found in the aqueous humor and in histologic examination of active PCV eyes.⁸

Vascular abnormalities typically occur at the level of the inner choroid below the choriocapillaris, and present with two characteristic findings: branching choroidal vessels usually located peripapillary, but also found subfoveal and (less commonly) mid-peripheral, and reddish-orange aneurysmal projections (hence the term polypoidal) emanating from the branching vessels.^{2,3,6-7} The aneurysmal lesions account for the transient retinal PED, exudates and hemorrhage characteristic of PCV.⁶ Large, reddish-orange subretinal masses resembling an ocular tumor (such as that which occurred in our patient) can develop from a cluster of polypoidal components beneath a PED. These vascular elements are best visualized via indocyanine green angiography (ICGA),

and will be found to project anteriorly from the inner choroid toward the outer retina. This large PED frequently resolves, leaving retinal pigment epithelial (RPE) atrophy in place of the original lesion.³

Vision is usually not affected during this early phase as PED are located away from the macular center. Some patients with multiple recurrences of PCV maintain long-term good visual acuity, unless the vascular anomalies progress and/or enlarge to encroach and encompass the macula.^{2,3} Larger scale progression results in RPE atrophy or rarely disciform scarring.^{2,3} As detachments, hemorrhages, and exudates effecting the macula resolve, visual acuity may improve. However, the chance of recurrence and perhaps repeat visual demise is high and should be expected.^{2,3} Some patients may also experience bullous retinal detachment and vitreous hemorrhage.^{1,2,5,7}

The differential diagnosis for PCV includes the various causes of subretinal and juxtapapillary subretinal neovascularization.² Wet AMD is the most common diagnosis associated with PCV. The two are somewhat related, as the pathogenesis of the vascular abnormality in PCV is thought to be a variant of choroidal neovascularization (CNV) seen in wet AMD.⁷ PCV does not typically involve the central macula as AMD, although in Asians macular CNV usually will appear as a PCV.⁹ Interestingly, after ICGA was recognized as the diagnostic procedure of choice in the diagnosis of PCV, a significant proportion of patients thought to have wet AMD were actually found instead to be diagnosed with PCV.^{3,7,10}

Patients with PCV will tend to be slightly younger and more darkly pigmented than those with AMD. Furthermore, macular findings usually seen with AMD, such as drusen, RPE changes, and the classic grayish green appearance that points to CNV, are not apparent

with PCV.⁷ The natural course of wet AMD lacks multiple exacerbations and remissions seen with PCV and the prognosis is not as favorable.¹⁻³ Untreated and unsuccessfully treated wet AMD results in permanent vision loss due to disciform scarring—notably absent in patients diagnosed with PCV.^{7,11}

PCV has also been known to masquerade as idiopathic central serous choroidopathy (ICSC). ICGA is the best diagnostic tool for delineating the two. A PED caused by PCV will typically appear to hyperfluoresce, whereas one caused by ICSC will present as hypofluorescence.⁶

Non-polypoidal juxtapapillary subretinal neovascularization can also occur in association with papillitis, sarcoidosis, angioid streaks, drusen of the optic nerve head and presumed ocular histoplasmosis syndrome.² PCV has also been found to occur in association with tilted disc syndrome and high myopia with staphyloma.¹²

Although ICGA is the gold standard test used in diagnosing PCV, in many cases the classic spheroidal end bulbs are also visualized using fluorescein angiography. The infrared fluorescent characteristics of ICGA, however, afford penetration of retinal blood and exudates to provide clear choroidal images of PCV vascular abnormalities.³ Yannuzzi and Ciardella et al³ studied the role of ICGA in diagnosing PCV and discovered that the choroidal abnormalities of PCV were distinct entities from lesions noted in other choroidal vascular diseases.

Given the tendency for PCV to self-resolve and to be situated primarily peripapillary, conservative management is usually recommended.³ Uyama et al⁹ document that fifty percent of untreated eyes have a favorable outcome. However, if there is persistent or progressive exudative change that is threatening the central macula, treatment options typically include laser photocoagulation, photodynamic therapy (PDT) and/or injection of anti-VEGF agents.^{3,8} With its favorable outcome, PDT has recently been proposed as a standard treatment modality; however, its application has been found to be limited due to the difficulty in treating multiple polyps with a single PDT beam, the lack of ability to also treat the entire network of branching anomalous vessels, the recurrent nature of PCV, and the possibility of secondary massive submacular hemorrhage.^{6,12} Because of recent evidence that VEGF may have a similar role in PCV as it does in other choroidal neovascular networks, the anti-VEGF drug, bevacizumab, (Avastin®, Roche Canada) has been tried with favorable results.^{8,13} The combination of PDT and anti-VEGF agents have also shown improvement in best corrected visual acuity and reduced angiographic leakage in most patients.⁸ Other various treatment proposals for PCV include tissue plasminogen activator injection with gas displacement, submacular surgery, and macular translocation surgery.⁸ The beneficial effects from these latter modalities are in doubt owing to recurrence or poor long-term results.^{8,9}

CONCLUSION

Polypoidal choroidal vasculopathy is a distinct clinical entity that must be differentiated from other forms of choroidal neovascularization. Differentiating PCV from the classic or occult CNV of wet AMD is important because of the significant differences in the demographic risk profile, natural course, visual prognosis and management of affected patients.⁷ In patients with serosanguinous detachments of the RPE — particularly blacks and Asians — ICG angiography should be performed to evaluate the choroidal vasculature in an attempt to establish a more definitive diagnosis. Delineating PCV from other forms of CNV will enable the practitioner to render the proper management plan for patients with this unique and potentially sight threatening vascular abnormality.

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