diopathic polypoidal choroidal vasculopathy (PCV) is a hemorrhagic disorder of the macula. The characteristic PCV lesion is a vascular network in the inner choroid ending in an aneurysmal bulge. This outward projection is visible clinically as a reddish-orange polyp-like structure.1

When this entity was first identified, PCV was thought to preferentially affect pigmented individuals, such as those of African or Asian descent.2 PCV has since been described in people of European descent as well,1 although prevalence is higher in Asian people than in whites.3

The disease follows a remitting-relapsing course and is associated clinically with chronic recurrent serosanguineous pigment epithelial detachment (PED).1 Although PCV shares some characteristics with choroidal neovascularization (CNV) secondary to age-related macular degeneration (AMD), they are recognized as separate entities. Successful treatment of PCV can be more difficult than treatment of CNV associated with more classic AMD. Investigators have reported some success with photodynamic therapy (PDT) and with vascular endothelial growth factor (VEGF) inhibitors such as bevacizumab (Avastin, Genentech) or ranibizumab (Lucentis, Genentech), either alone or in combination.3

Indocyanine green angiography (ICGA) has been the mainstay for diagnosis and follow-up of individuals with PCV.1,3 More recently, optical coherence tomography (OCT) has helped to establish the diagnosis and more fully elucidate the morphologic features of PCV and accompanying PED.4-7

In eyes with PCV examined using ICGA and OCT, Tsujikawa and colleagues4 found that polypoidal lesions are located at the margins of the PED. Fluid from the lesions can cause these lesions to detach from Bruch’s membrane and appear to be located inside the PED. In eyes with a PED, en face OCT shows protrusions of the retinal pigment epithelium (RPE) corresponding to polypoidal lesions seen on ICGA.4-6 In eyes without a PED, en face OCT shows distinctive rings of a highly reflective RPE line corresponding to polypoidal lesions seen on ICGA.6 Ojima and colleagues7 found that an enhanced spectral-domain OCT (SD-OCT) system clearly depicted Bruch’s membrane beneath areas of abnormal RPE in the locations where ICGA showed the vascular abnormalities associated with PCV. Clearly, OCT imaging has become an important diagnostic tool for use in eyes with PCV.

Case Report: Polypoidal Choroidal Vasculopathy Responds to Anti-VEGF Injections

Patient’s vision is stable 11 months after last injection.

BY SZILÁRD KISS, MD

Figure 1. Initial visit. Visual acuity 20/400. Fundus photography (A) shows sub-RPE and subretinal blood of varying ages. SD-OCT 3D (B) and slice (C) images show the appearance of the multiple PEDs, the morphology of which is suggestive of PCV.
CASE REPORT

A 50-year-old woman from Africa presented with visual acuity in the right eye of 20/400. She reported a history of 2 years of progressively worsening vision in that eye. Color fundus photograph revealed areas of subretinal and sub-RPE hemorrhages of variable duration (Figure 1A). On SD-OCT (3D OCT-1000; Topcon Medical Systems, Paramus, NJ), the appearance of the PEDs was suggestive of PCV (Figures 1B and 1C). Intravenous fluorescein angiography (FA) and ICGA confirmed the presence of PCV.

The patient specifically requested intravitreal injections of ranibizumab (Lucentis, Genentech). She declined any combination therapy. After three consecutive monthly intravitreal injections, her visual acuity in the right eye improved to 20/200. Clinically, the polypoidal lesions were regressed, with decreased hemorrhage (Figure 2A) and flattening of the PEDs (Figures 2B and 2C).

After a total of five monthly ranibizumab injections, visual acuity in the right eye improved to 20/150 and the sub-RPE blood had resolved (Figures 3A-3C). At her most recent follow-up, 11 months after the last injection, the visual acuity in the right eye was stable and the patient was delighted with the improvement from her baseline.

DISCUSSION AND CONCLUSIONS

As described recently in the literature, the peaked appearance of the PED in this patient on SD-OCT was suggestive of PCV. Subsequent ICGA demonstrated the polyps and confirmed our diagnosis.

Some authors have reported that VEGF inhibition is less effective for treatment of idiopathic PCV than for CNV secondary to AMD. This well educated patient specifically requested (and paid for) treatment with ranibizumab. A course of five treatments resolved the sub-RPE blood, flattened the PEDs, and stabilized the vision in the affected eye.

The ultimate visual acuity improvement, from 20/400 to 20/150, was limited by the chronic nature of the subretinal blood and some resultant subfoveal fibrosis. Nonetheless, the patient was extremely happy with her improved vision because she had been living with 20/400 vision for 2 years.

Spectral-domain OCT is a useful imaging modality for diagnosing and following PCV.

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