Optical Coherence Tomography of Idiopathic Polypoidal Choroidal Vasculopathy

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Purpose: To characterize cross-sectional images of idiopathic polypoidal choroidal vasculopathy (PCV).

Methods: A cross-sectional and retrospective study was performed involving 28 eyes with PCV and 112 eyes with exudative age-related macular degeneration. The frequency and dimensions of the retinal pigment epithelial detachment (RPED) with attenuation of internal reflectivity on optical coherence tomographic (OCT) examination were compared in both diseases.

Results: OCT showed the RPED with attenuation of internal reflectivity corresponding to the polypoidal structure in the indocyanine green angiogram in 75% of eyes with PCV, which was significantly more frequent than in eyes with exudative age-related macular degeneration (3.6%). In lesions suspicious of exudative age-related macular degeneration or PCV, the RPED with attenuation of internal reflectivity on OCT images strongly supported PCV diagnosis with a sensitivity of 84% and a specificity of 94%. The base diameter (p=0.010) and base diameter times height (p=0.028) of RPED were smaller in PCV than in exudative age-related macular degeneration.

Conclusions: An RPED with attenuation of internal reflectivity in OCT examination is a highly sensitive and specific finding which characterizes PCV. Recognition of this RPED appearance, as well as the evaluation of its size, aids in the diagnosis of PCV. Korean Journal of Ophthalmology 19(4):275-280, 2005

Key Words: Optical coherence tomogram, Polypoidal choroidal vasculopathy

Idiopathic polypoidal choroidal vasculopathy (PCV) is characterized by multiple recurrent serous, hemorrhagic detachments of the retinal pigment epithelium, and particular polypoidal vascular lesions at the termination of a choroidal network of vessels.1 Indocyanine green angiography shows distinctions in the choroidal vasculature: a branching network of vessels in the inner choroids, and vascular dilatations at the border of the network of vessels.1 Many previous reports revealed that patients with PCV, when compared to patients with age-related macular degeneration, have lesions at more distant sites from the fovea, show better visual prognosis, are not associated with soft drusen, lacquer cracks, or pathologic myopia, and do not show evidence of intraocular inflammation.1,3 Many authors insisted that the disorder should be regarded as a distinct clinical entity. However, the pathophysiology and clinical nature of PCV is still unclear, and it is controversial whether or not PCV is a variant of choroidal neovascularization.

Optical coherence tomography (OCT) is a relatively new technique that is useful for high depth resolution and for cross-sectional visualization of the retina and choroids. OCT is a powerful tool for detecting, monitoring, and quantitatively assessing macular diseases, including macular edema, macular holes and RPE detachments.4,6 OCT can provide much information about PCV because the majority of choroidal vascular lesions in PCV are located in the macular or peripapillary area, and hemorrhagic and/or serous retinal pigment epithelial detachments are associated with PCV. In our experience, many of the patients with PCV showed an RPED with attenuation of internal reflectivity on OCT examination. A small number of previous studies used OCT in eyes with PCV,3,8 but none mentioned how often the RPED with attenuation of internal reflectivity on OCT examination was detected in PCV compared with other macular diseases, nor whether this typical finding can be used to detect and characterize PCV.

The purpose of this study was to characterize the RPED with attenuation of internal reflectivity from the OCT examination of PCV.

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Materials and Methods

We reviewed 28 eyes with PCV in 26 patients who visited our clinic due to decreased vision, metamorphopsia or scotoma, from December 1999 to January 2003. Each of these patients underwent standardized, clinical, fluorescein and indocyanine green angiography and OCT examinations.

The diagnosis of PCV was made according to evidence of a branching network of inner choroidal vessels with polypoidal vascular dilatations at the border of the network of vessels via indocyanine green angiography, and the presence of a serous and/or hemorrhagic pigment epithelium detachment and retinal detachment in the posterior pole. Exclusion criteria were as follows: other diseases such as exudative age-related macular degeneration, high myopia, angioid streaks, and any previous ocular surgery. Indocyanine green angiography was performed with a fundus camera and the IMAGEnet system (IMAGEnet for Windows, version 1.55; Topcon, Tokyo, Japan) after the injection of 50 mg of indocyanine green dye in the antecubital vein. All patients underwent serial vertical and horizontal OCT scans (OCT 2000, software version A 7.0; Carl-Zeiss ophthalmic systems, San Leandro, California) through the lesion. An infrared light with a wavelength of 840 nm and energy of 750 µW was incident on the retina. The location and direction of the scan were determined to display the most verifiable image of the orange-red nodular elevation. All OCT images were evaluated to find the RPED with attenuation of internal reflectivity. It exhibits the dome-like focal protrusion of a highly reflective band corresponding to the detached RPE, and sometimes shows a cluster of multiple protrusions. There is moderate focal reflection just beneath the RPE protrusion and the characteristic attenuation of the reflectivity within the RPE detachment (Fig. 1A). When the RPED with attenuation of internal reflectivity was detected in the OCT images, we measured the base diameter and height of each lesion after

![Image](image_url)

Fig. 1. (A) Fundus photogram of the right eye shows pigment epithelial detachment, subretinal hemorrhages and orange-red nodular elevations of the retinal pigment epithelium (arrows). The optical coherence tomographic (OCT) scan line is indicated by a whitish horizontal line. Indocyanine green angiogram reveals polypoidal vascular dilatations (arrows) at the termini of a branching network of inner choroidal vessels (arrowhead) and confirms the diagnosis of polypoidal choroidal vasculopathy. OCT shows a dome-like elevation of a highly reflective band corresponding to the detached retinal pigment epithelium. There are moderate focal reflections just beneath the elevation and attenuation of optical reflectivity under these reflections. (B) OCT image of serous retinal pigment epithelial detachment in age-related macular degeneration demonstrates an elevation of the retinal pigment epithelium over an optically clear space corresponding to a serous fluid collection without shadowing of the choroids. (C) OCT image of hemorrhagic retinal pigment epithelial detachment developed in a case of exudative age-related macular degeneration shows a dense reflective band beneath the pigment epithelium and complete optical shadowing under the reflective band.
performing vertical and horizontal OCT scans through the center of the RPED, as described by Iijima et al.9 (Fig. 2). The base diameter was measured as the longest distance between the two points of the detached RPE's elevated upper margin. The height was measured as the longest distance between the extension line of the upper margin of the attached RPE and the upper margin of the detached RPE. The base diameter and height of each lesion were determined by averaging the two values from the vertical and horizontal scans.

To evaluate whether the RPED with attenuation of internal reflectivity is a typical OCT finding for PCV, we compared it with the OCT findings of RPEDs presented in exudative age-related macular degeneration. Criteria for exudative age-related macular degeneration included: (1) the fundus appearance of subretinal fibrovascular membranes, serous and/or hemorrhagic neurosensory detachment, and serous and/or hemorrhagic RPEs; (2) the presence of CNV revealed with fluorescein and indocyanine green angiography; and (3) no evidence of polypoidal dilatations at the border of a branching network of inner choroidal vessels via indocyanine green angiography.

The 112 eyes with exudative age-related macular degeneration examined in our clinic from April 2002 to March 2003 met the above inclusion criteria. The mean age of the patients was 67.3 years (range, 54-88) and included 65 men and 26 women. In the review of OCT images of exudative age-related macular degeneration, we focused on the prevalence and size of the RPED with attenuation of internal reflectivity.

The Mann-Whitney test was used for statistical analysis and conducted using SPSS statistical software (version 11.0; SPSS for Windows, 2001). A p value of less than 0.05 was considered significant.

**Results**

1. Clinical features

   The mean age of the 26 patients having PCV was 68.9 years (range, 54-87 years) (Table 1). The patient group included 18 men and 8 women. The affected eyes were unilateral in 24 patients (92.3%) and bilateral in two patients (7.7%), for a total of 28 eyes. In the patients with unilateral disease, the right eye was affected in 11 patients and the left eye in 12 patients. Six patients affected with diabetes and eight with hypertension were compared with the age matched prevalence of diabetes (6.63%) and hypertension (44.7%) acquired from a nation-wide survey (Korean National Health and Nutrition Examination Survey, 1998). The prevalence of a systemic disorder in the patients with PCV was not different from that of the general population (Z-test).

2. Indocyanine Green Angiographic Findings

   The abnormalities characteristic of PCV were demonstrated in all cases; a large choroidal vessel in the PCV lesion became hyperfluorescent in the early phases before the retinal vessels were filled with dye, after which the vascular network began to fill, followed by the polypoidal structure becoming increasingly hyperfluorescent. In the late phases, the polypoidal structure became hypofluorescent in the majority of cases. The late indocyanine green staining characteristic of CNV was seen in 16 eyes, and in some cases, minimal leakage revealed the RPED overlying the polyp.

   The mean number (±standard deviation) of polyps found in one eye with PCV was 4.0±2.1 (range, 1-10). The interconnecting vessels among the polyps were visible in 22 eyes (78.6%). Subretinal hemorrhage was evident in 21 eyes (75.0%). RPEDs found by indocyanine green angiography were observed in 8 eyes (28.6%). CNV concurred with PCV in 16 eyes (57.1%).

3. Optical Coherence Tomographic Findings

   Several OCT images scanning through the orange-red subretinal masses on clinical examination were obtained. The RPED with attenuation of internal reflectivity in OCT examination was recognized in 21 eyes (75.0%, 21/28) with PCV and the mean number (±standard deviation) of RPEDs with attenuation of internal reflectivity for one eye was 1.5±0.85 (range, 1-4). In contrast, only 4 eyes (3.6%, 4/112) with exudative age-related macular degeneration revealed RPED with attenuation of internal reflectivity in the OCT examination, and each one showed singularity and unilaterality of the lesion. Thus, the presence of RPED with attenuation of internal reflectivity on OCT images strongly supported PCV diagnosis, with a sensitivity of 84% and specificity of 94% in the lesions suspected of either exudative age-related macular degeneration or PCV.

   The RPED with attenuation of internal reflectivity in eyes with PCV showed a steeper protrusion and a smaller size...
than those of exudative age-related macular degeneration.

The mean base diameter of RPED with attenuation of internal reflectivity in eyes with PCV was 1097±574 µm, which is smaller than that of exudative age-related macular degeneration (1860±677 µm, p=0.010). The mean height was 219±129 µm in eyes with PCV and 362±158 µm in exudative age-related macular degeneration, showing no statistical difference (p=.057). The mean value of height times the base diameter of lesions was smaller in eyes with PCV (252493±244514 µm²) than that of exudative AMD (741777±531672 µm², p=0.028) (Fig 3). The mean ratio of the height to the base diameter of the RPED in eyes with PCV was 0.21±0.11, which was not statistically different from that of exudative AMD (0.19±0.05, p>0.05).

Table 1. IPCV study group

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Little data exists regarding the sensitivity and specificity of the OCT findings and the difference between the RPED in PCV and RPED in exudative age-related macular degeneration. Since the serous and/or hemorrhagic RPED typically presents in the exudative age-related macular degeneration and PCV, our study compared the OCT images of patients with PCV and those with exudative age-related macular degeneration.

The RPED detected in PCV typically showed attenuation of internal reflectivity on OCT examination, and was distinguishable from serous or hemorrhagic RPED in eyes with exudative age-related macular degeneration (Fig. 1B, 1C). The optical reflectivity of the sub-RPED fluid in PCV is presumed to be between that of pure blood and serosanguineous fluid, which causes the RPED attenuation of internal reflectivity on OCT typical in PCV. Figure 1B shows an example of a serous RPED containing an optically empty space due to low-reflective serous fluid. An OCT image of hemorrhagic RPED (Fig. 1C) can be distinguished from a serous RPED by the presence of optical backscattering arising from blood beneath the detached RPE and the shadowing of deeper structures. The sub-RPED fluid in PCV may have the optical reflectivity anywhere between serous RPED and hemorrhagic RPED, and show moderate reflectivity directly beneath the detached RPE, but increasing attenuation of reflectivity in deeper space. The characteristic results from OCT examination appear to be correlated with those from ICGA, in which small polyps become increasingly hyperfluorescent in the middle phase rather than early phase. The polyps in PCV are considered to have a slow blood flow and are clinically visible as a pinkish rather than dark red structure. The above findings may be correlated with the typical RPED of PCV having attenuated internal reflectivity on OCT.

As a result of our study, this typical appearance of RPED in OCT examination proved to be highly sensitive (84%) and specific (94%) for PCV when compared with exudative age-related macular degeneration. This OCT manifestation therefore strongly supports the diagnosis of PCV. The result was compatible with one report insisting that 85% of the cases presenting large hemorrhagic and exudative pigment epithelial detachments in the absence of drusen were PCV. Iijima et al. reported that OCT images in eyes with PCV show a highly reflective band of remarkable anterior protrusion that is thicker and steeper than that of the serous RPED. Our study results indicate that the steepness of the RPED, which was calculated by the ratio of its height to base diameter, cannot differentiate the RPED in PCV from the RPED in exudative age-related macular degeneration. Instead, the dimensions of the typical appearance of RPED on OCT examination were different between the two disorders. We found that the base diameter and the product of the base diameter and height of the characteristic RPED on OCT examination was statistically smaller in eyes with PCV than in eyes with age-related macular degeneration. Although the value calculated by multiplying the base diameter by the height provides the area of a rectangle, it can be interpreted as the area of RPED because of its low height-to-base ratio (0.2).

RPED with attenuation of internal reflectivity on OCT examination represents a dome-like focal protrusion of a highly reflective band corresponding to the detached RPE with moderate reflections below the protrusion and shadowing underneath. The detachment of RPE appears to be accompanied by the detachment of Bruch’s membrane. The moderate reflections and shadowing within the focal protrusion may correspond to the fibrovascular proliferation, dilated vessels and voluminous accumulation of extravasated blood and exudate per diapedesis in the sub-RPE space. Marked attenuation of incident light through the RPE produces moderate reflections under the detached RPE. The shadowing under moderate reflections is derived not only from the increased attenuation of the incident light due to a high light scattering coefficient of blood, but also from the enhanced reflectivity of the overlying layer.

Although indocyanine green angiography is usually indispensable for the diagnosis of PCV, it is not always diagnostic of PCV. In a recent report on the differential diagnosis of hot spots detectable by indocyanine green angiography, the majority of the lesions were categorized into PCV, retinal angiomatous proliferation and pure occult CNV. When only one hot spot is detected on indocyanine green angiography, or when an interconnecting vessel is not detected in apparent PCV case, the diagnosis of PCV imposes challenges even with the indocyanine green angiography. Notably in our case studies, only one polyp was detected in 2 eyes (7.1%) and an interconnecting vessel was not detected in 6 eyes (21.4%). The detection of the characteristic RPEDs on OCT examination may provide a firm basis for the diagnosis of PCV in these cases.

Fig. 3. Scattergram demonstrating the relationship between the base diameter and height of the retinal pigment epithelial detachments with optical shadowing in eyes with PCV (triangle) and exudative age-related macular degeneration (dot). The RPEDs with optical shadowing in eyes with PCV tend to be smaller than those of exudative age-related macular degeneration.

**Discussion**

Previous reports suggested that elevated RPE and moderate reflectivity of the overlying layer are characteristic OCT findings of PCV. Little data exists regarding the sensitivity and specificity of the OCT findings and the difference between the RPED in PCV and RPED in exudative age-related macular degeneration. The above findings may be correlated with the OCT examination results from ICGA, in which small polyps become increasingly hyperfluorescent in the middle phase rather than early phase. The steepness of the RPED, which was calculated by the ratio of its height to base diameter, cannot differentiate the RPED in PCV from the RPED in exudative age-related macular degeneration. Instead, the dimensions of the typical appearance of RPED on OCT examination were different between the two disorders. We found that the base diameter and the product of the base diameter and height of the characteristic RPED on OCT examination was statistically smaller in eyes with PCV than in eyes with age-related macular degeneration. Although the value calculated by multiplying the base diameter by the height provides the area of a rectangle, it can be interpreted as the area of RPED because of its low height-to-base ratio (0.2).

RPED with attenuation of internal reflectivity on OCT examination represents a dome-like focal protrusion of a highly reflective band corresponding to the detached RPE with moderate reflections below the protrusion and shadowing underneath. The detachment of RPE appears to be accompanied by the detachment of Bruch’s membrane. The moderate reflections and shadowing within the focal protrusion may correspond to the fibrovascular proliferation, dilated vessels and voluminous accumulation of extravasated blood and exudate per diapedesis in the sub-RPE space. Marked attenuation of incident light through the RPE produces moderate reflections under the detached RPE. The shadowing under moderate reflections is derived not only from the increased attenuation of the incident light due to a high light scattering coefficient of blood, but also from the enhanced reflectivity of the overlying layer.

Although indocyanine green angiography is usually indispensable for the diagnosis of PCV, it is not always diagnostic of PCV. In a recent report on the differential diagnosis of hot spots detectable by indocyanine green angiography, the majority of the lesions were categorized into PCV, retinal angiomatous proliferation and pure occult CNV. When only one hot spot is detected on indocyanine green angiography, or when an interconnecting vessel is not detected in apparent PCV case, the diagnosis of PCV imposes challenges even with the indocyanine green angiography. Notably in our case studies, only one polyp was detected in 2 eyes (7.1%) and an interconnecting vessel was not detected in 6 eyes (21.4%). The detection of the characteristic RPEDs on OCT examination may provide a firm basis for the diagnosis of PCV in these cases.
Little is known of the histopathology of PCV, especially as to the spatial relation between polyp and the interconnected vessel, and as to whether PCV is a peculiar type of CNV or just a vascular dilation. One study disclosed extensive fibrovascular proliferation in the subretinal space and within the Bruch's membrane, and the presence of many choroidal vessels that transversed defects in the Bruch's membrane. The subsequent histopathological studies of PCVs noted sub-RPE and intra-Bruch's fibrovascular membrane. PCV has been regarded as a peculiar type of choroidal neovascularization in all of the above reports. Whether CNV was secondary to the RPED in PCV or not, it is remarkable that the coincidence of the RPED and CNV was identified in 12 of 28 eyes with PCV in our study. In contrast, a recent histopathological study on eyes with PCV suggested the lesion as a degenerated RPE-Bruch's membrane-choriocapillaris complex and inner choroids rather than an intra-Bruch's fibrovascular membrane. The study proposed that PCV might be caused by the stasis of the venule and arteriole at the arteriovenous crossing site associated with arteriolar sclerosis. Although the systemic condition of patients with PCV is generally healthy, the correlation between systemic hypertension and PCV has been suggested in a few reports. Some authors insist that hypertensive insult to the choroidal vascular beds involves the pathogenesis of PCV. Histopathological data from one study suggested that the arteriosclerotic change might also be involved in the pathogenesis. Since the correlation between PCV and systemic disorders including hypertension and diabetes was not noted in our study, we believe further data is necessary to clarify the role of systemic disorders in PCV.

Twenty-four (92%) of 26 patients in our study showed unilateral involvement of PCV, although the rate of bilaterality was about 70% in original reports on PCV. Recent large case studies indicate that the rate of bilateral involvement is around 10%. The dominance of unilaterality in our study may be attributed to the fact that the data from each case was mainly obtained from the patient's first visit to our clinic, implying that our data was collected from the acute symptomatic phases of PCV. Since the involvement in the contralateral eye may develop after a dozen years, the proportion of bilateral cases in our studies would increase with follow-up for an extended period. Other possibilities may include racial differences.

In conclusion, an RPED with attenuation of internal reflectivity on OCT examination is a sensitive and specific finding for PCV. Detection of this finding along with a small cross sectional area on OCT examination strongly supports the diagnosis of PCV.

References