Polypoidal choroidal vasculopathy with an appearance similar to classic choroidal neovascularisation on fluorescein angiography

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Differentiation between type 2 CNV and pure fibrinous tissue

Polypoidal choroidal vasculopathy (PCV) is a distinct clinical entity characterised by a branching choroidal vascular network and polypoidal vascular dilations at the border of the vascular network. PCV is associated with serous or haemorrhagic detachments of the retinal pigment epithelium (RPE) and neurosensory retina secondary to leakage and bleeding from the choroidal vascular lesions. PCV has been referred to by various names, including idiopathic PCV,¹ posterior uveal bleeding syndrome,² and multiple recurrent retinal pigment epithelial detachment in black women.^{3,4} Although the pathogenesis of PCV is not clearly understood, it is thought to originate in an abnormality of the inner choroidal vessels.^{1-5,7} Clinically, polypoidal vascular dilations may appear as orange-red lesions under the RPE on biomicroscopic examination. Indocyanine green angiography (IA) provides definitive diagnosis of PCV because it clearly shows a branching vascular network and polyp-like structures from the choroidal circulation.^{5,6} These vascular abnormalities seem to be a variant of choroidal neovascularisation (CNV), referred to as polypoidal CNV,⁸⁻¹⁰ and PCV is often referred to as polypoidal age-related macular degeneration (ARMD).¹¹

PCV previously was thought to be a rare disease. Recent reports have shown that PCV occurs extensively in patients of all races.^{6,9,10,12-18} It also seems to be more common in pigmented individuals, such as African-Americans and Asians. In Japanese patients, Shio *et al*¹⁰ reported that 100 (24%) of 418 patients with neovascular ARMD had PCV, Yoshimura¹⁷ reported 42% and Maruko *et al*¹⁸ reported 54.7%; the discrepancies might be due to the facts that PCV recently has become well recognised and the resolution of IA devices has improved. PCV was originally reported to be a peripapillary disease; however, it is located primarily in the macula.^{8-10,12-19}

In the natural history of PCV, half of the patients had persistent leakage or

repeated bleeding and a poor visual outcome.²⁰ Shio *et al*¹⁰ also reported severe visual loss in 35% of eyes. Photodynamic therapy (PDT) with verteporfin is an effective treatment for maintaining or improving visual acuity in patients with symptomatic PCV with macular lesions.²¹⁻²⁴ Stable or improved vision was achieved in 81% to 95% of eyes after PDT. Chan *et al*²³ reported that the use of PDT resulted in the complete absence of leakage on fluorescein angiography and total regression of polypoidal lesions on IA in 91% and 95% of patients, respectively. Besides the beneficial effects of visual outcome and angiographical responses, the number of PDT sessions necessary to treat PCV was lower than that of typical ARMD, such as predominantly classic, minimally classic or occult with no classic lesions.²¹⁻²⁶

PCV lesions usually look similar to occult CNV on fluorescein angiography.^{5,9} However, eyes with PCV sometimes appear similar to those with classic CNV on fluorescein angiography.^{1,5,6,9,10,18} Shio *et al*¹⁰ reported that 10 (9%) of 110 eyes with PCV had classic CNV on fluorescein angiography. Maruko *et al*¹⁸ also observed classic CNV lesions in 23 (11.3%) of 203 eyes with PCV. When they evaluated the 203 eyes with PCV using only fluorescein angiography, they classified predominantly classic CNV in three eyes (1.5%), minimally classic CNV in 20 eyes (9.8%) and occult with no classic CNV in 180 eyes (88.7%). Spaide and colleagues⁵ reported that classic CNV developed in 3 (15.8%) of 19 eyes (3 of 12 patients) with PCV during the follow-up period. Histopathological studies of surgical specimens of PCV showed a fibrovascular membrane within Bruch's membrane, whereas some eyes also showed CNV in the subretinal space (type 2 pattern).²⁷⁻³² Type 2 CNV, growth of new vessels between the sensory retina and RPE, often has the appearance of classic CNV on fluorescein angiography. Eyes with PCV that appear to have classic CNV

on fluorescein angiography and subretinal material have been thought to have type 2 CNV, which disrupts the RPE and invades the subretinal space from the sub-RPE PCV lesion.

It would be of great help to determine whether eyes with PCV have type 2 CNV or not in order to predict the visual outcome.^{9,10} In most patients with type 2 CNV, fibrotic tissue persists in the subretinal space even after successful treatment with PDT, resulting in a poor visual outcome.³³ However, we have seen some cases of PCV with classic CNV on fluorescein angiography that had an excellent response to PDT, including complete resolution of the subretinal tissue and a good visual outcome.

Recently, Otsuji and colleagues³⁴ reported an important contribution to our understanding of PCV. The authors evaluated the clinical characteristics in eight eyes with PCV with the appearance of classic CNV and subretinal greyish tissue using IA and optical coherence tomography (OCT). The authors disclosed that eyes with PCV with the appearance of classic CNV with a subretinal moderately reflective mass over the polypoidal lesion apparent on OCT are sometimes diagnosed with pure fibrinous tissue without type 2 CNV. Leakage from active polypoidal lesions can result in subretinal fibrinous exudation, which may mimic classic CNV on fluorescein angiography. However, it is often difficult to discriminate pure fibrinous exudation from type 2 CNV.

In this issue of the *BJO*, Tamura and colleagues³⁵ report an interesting study of the visual prognosis and ocular characteristics in 38 eyes with PCV and ostensibly classic CNV (see page 1152). In all cases, optical coherence tomography showed subretinal material with moderate reflectivity that corresponded in location to the classic CNV. In response to the subretinal material to PDT, the authors classified two groups: the resolved group and persistent group. The visual acuity in the resolved group was significantly better than that in the persistent group. The subretinal material seen before treatment was more frequently seen in the subfovea in the persistent group. The subretinal material was adjacent to polypoidal lesions more often in the resolved group. The authors believed that the persistent group had type 2 CNV before PDT, whereas the resolved group had pure fibrinous tissue. The authors concluded that when subretinal material corresponding to classic CNV is seen in the subfoveal region or is separate from the polypoidal lesions, the eye may actually have type 2 CNV. This article is clinically relevant for patient management, because there is a difference in the visual prognosis

between PCV with type 2 CNV and pure fibrinous exudation. Further studies are needed to determine the accurate diagnosis and proper treatment for patients with type 2 CNV associated with PCV.

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REFERENCES

- 1 Yannuzzi LA, Sorenson J, Spaide RF, *et al.* Idiopathic polypoidal choroidal vasculopathy (PCV). *Retina* 1990;**10**:1–8.
- 2 Kleiner RC, Brucker AJ, Johnston RL. The posterior uveal bleeding syndrome. *Retina* 1990;**10**:9–17.
- 3 Stern RM, Zegarra H, Kleiner RC, *et al.* Multiple recurrent serosanguineous retinal pigment epithelial detachments in black women. *Am J Ophthalmol* 1985;**100**:560–69.
- 4 Perkovich BT, Zakov ZN, Berlin LA, *et al.* An update on multiple recurrent serosanguineous retinal pigment epithelial detachments in black women. *Retina* 1990;**10**:18–26.
- 5 Spaide RF, Yannuzzi LA, Slakter JS, *et al.* Indocyanine green videoangiography of idiopathic polypoidal choroidal vasculopathy. *Retina* 1995;**15**:100–10.
- 6 Yannuzzi LA, Ciardella A, Spaide RF, *et al.* The expanding clinical spectrum of idiopathic polypoidal choroidal vasculopathy. *Arch Ophthalmol* 1997;**115**:478–85.
- 7 Yuzawa M, Mori R, Kawamura A. The origins of polypoidal choroidal vasculopathy. *Br J Ophthalmol* 2005;**89**:602–7.
- 8 Uyama M, Matsubara T, Fukushima I, *et al.* Idiopathic polypoidal choroidal vasculopathy in Japanese patients. *Arch Ophthalmol* 1999;**117**:1035–42.
- 9 Yannuzzi LA, Wong DWK, Sforzolini BS, *et al.* Polypoidal choroidal vasculopathy and neovascularized age-related macular degeneration. *Arch Ophthalmol* 1999;**117**:1503–10.
- 10 Sho K, Takahashi K, Yamada H, *et al.* Polypoidal choroidal vasculopathy. Incidence, demographic features, and clinical characteristics. *Arch Ophthalmol* 2003;**121**:1392–6.
- 11 Singerman LJ, Brucker AJ, Jampol LM, *et al.* Neovascular age-related macular degeneration: roundtable. *Retina* 2005;**25**:S1–22.
- 12 Ahuja RM, Stanga PE, Vingerling JR, *et al.* Polypoidal choroidal vasculopathy in exudative and haemorrhagic pigment epithelial detachments. *Br J Ophthalmol* 2000;**84**:479–84.
- 13 Lafaut BA, Leys AM, Snyers B, *et al.* Polypoidal choroidal vasculopathy in Caucasians. *Graefes Arch Clin Exp Ophthalmol* 2000;**238**:752–9.
- 14 Scassellati-Sforzolini B, Mariotti C, Bryan R, *et al.* Polypoidal choroidal vasculopathy in Italy. *Retina* 2001;**21**:121–5.
- 15 Kwok AKH, Lai TYY, Chan CWN, *et al.* Polypoidal choroidal vasculopathy in Chinese patients. *Br J Ophthalmol* 2002;**86**:892–7.
- 16 Wen F, Chen C, Wu D, *et al.* Polypoidal choroidal vasculopathy in elderly Chinese patients. *Graefes Arch Clin Exp Ophthalmol* 2004;**242**:625–9.
- 17 Yoshimura N. Polypoidal choroidal vasculopathy. [in Japanese] *Folia Ophthalmol Jpn* 2004;**55**:155–72.
- 18 Maruko I, Iida T, Saito M, *et al.* Clinical characteristics of exudative age-related macular degeneration in Japanese patients. *Am J Ophthalmol*. In press.
- 19 Moorthy RS, Lyon AT, Rabb MF, *et al.* Idiopathic polypoidal choroidal vasculopathy of the macula. *Ophthalmology* 1998;**105**:1380–5.
- 20 Uyama M, Wada M, Nagai Y, *et al.* Polypoidal choroidal vasculopathy: natural history. *Am J Ophthalmol* 2002;**133**:639–48.
- 21 Spaidey RF, Donsoff I, Lam DL, *et al.* Treatment of polypoidal choroidal vasculopathy with photodynamic therapy. *Retina* 2002;**22**:529–35.
- 22 Lee SC, Seong YS, Kim SS, *et al.* Photodynamic therapy with verteporfin for polypoidal choroidal vasculopathy of the macula. *Ophthalmologica* 2004;**218**:193–201.
- 23 Chan WM, Lam DSC, Lai TYY, *et al.* Photodynamic therapy with verteporfin for symptomatic polypoidal choroidal vasculopathy. One-year results of a prospective case series. *Ophthalmology* 2004;**111**:1576–84.
- 24 Silva RM, Figueira J, Cachulo ML, *et al.* Polypoidal choroidal vasculopathy and photodynamic therapy with verteporfin. *Graefes Arch Clin Exp Ophthalmol* 2005;**243**:973–9.
- 25 Treatment of Age-related Macular Degeneration With Photodynamic Therapy (TAP) Study Group. Photodynamic therapy of subfoveal choroidal neovascularization in age-related macular degeneration with verteporfin: two-year results of 2 randomized clinical trials - TAP report 2. *Arch Ophthalmol* 2001;**119**:198–207.
- 26 Verteporfin in Photodynamic Therapy Study Group. Verteporfin therapy of subfoveal choroidal neovascularization in age-related macular degeneration: two-year results of a randomized clinical trial including lesions with occult with no classic choroidal neovascularization - verteporfin in photodynamic therapy report 2. *Am J Ophthalmol* 2001;**131**:541–60.
- 27 MacCumber MW, Dastgheib K, Bressler NM, *et al.* Clinicopathologic correlation of the multiple recurrent serosanguineous retinal pigment epithelial detachments syndrome. *Retina* 1994;**14**:143–52.
- 28 Kuroiwa S, Tateiwa H, Hisatomi T, *et al.* Pathological features of surgically excised polypoidal choroidal vasculopathy membranes. *Clin Experiment Ophthalmol* 2004;**32**:297–302.
- 29 Shiraga F, Matsuo T, Yokoe S, *et al.* Surgical treatment of submacular hemorrhage associated with idiopathic polypoidal choroidal vasculopathy. *Am J Ophthalmol* 1999;**128**:147–54.
- 30 Terasaki H, Miyake Y, Suzuki T, *et al.* Polypoidal choroidal vasculopathy treated with macular translocation: clinical pathological correlation. *Br J Ophthalmol* 2002;**86**:321–7.
- 31 Nakajima M, Yuzawa M, Shimada H, *et al.* Correlation between indocyanine green angiographic findings and histopathology of polypoidal choroidal vasculopathy. *Jpn J Ophthalmol* 2004;**48**:249–55.
- 32 Gass JDM. Biomicroscopic and histopathologic considerations regarding the feasibility of surgical excision of subfoveal neovascular membranes. *Am J Ophthalmol* 1994;**118**:285–98.
- 33 Rogers AH, Martidis A, Greenberg PB, *et al.* Optical coherence tomography findings following photodynamic therapy of choroidal neovascularization. *Am J Ophthalmol* 2002;**134**:566–76.
- 34 Otsuji T, Tsumura A, Takahashi K, *et al.* Evaluation of cases of polypoidal choroidal vasculopathy showing classic choroidal neovascularization in their natural course. [in Japanese] *J Jpn Ophthalmol Soc*, 2006;**110**:454–61.
- 35 Tamura H, Tsujikawa A, Otani A, *et al.* Polypoidal choroidal vasculopathy appearing as classic choroidal neovascularisation on fluorescein angiography. *Br J Ophthalmol* 2007;**91**:1152–9.

Polypoidal choroidal vasculopathy

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From the viewpoint of an Asian ophthalmologist

Polypoidal choroidal vasculopathy (PCV) has been recognised for almost two decades. In 1990, Yannuzzi and colleagues first described idiopathic polypoidal choroidal vasculopathy (PCV) macular disorder—a choroidal vasculopathy that causes haemorrhagic and exudative macular degeneration—in 11 patients.¹ The patients had peculiar polypoidal subretinal

vascular lesions associated with serous and haemorrhagic detachments of the retinal pigment epithelium (RPE). In the late 1990s, Yannuzzi and colleagues expanded the clinical spectrum of PCV and established the concept of PCV.^{2,3}

In 1995, Spaide and colleagues identified the precise choroidal abnormalities associated with PCV and examined them using indocyanine green angiography

(IA).⁴ IA showed two basic choroidal vascular changes: a branching network of vessels in the inner choroid and polypoidal vascular dilations at the border of the network of vessels. IA confirmed the definitive diagnosis of PCV. In 1999, using optical coherence tomography (OCT), Iijima and colleagues showed that on images of the inner choroid in eyes with PCV orange-red lesions protruded anteriorly.⁵ These lesions had a sharper peak than serous RPE detachments in the subretina, suggesting that polypoidal vascular lesions in eyes with PCV lie beneath Bruch's membrane and are covered anteriorly by the RPE and Bruch's membrane.⁶ IA and OCT, relatively new diagnostic devices, are useful for understanding the pathophysiology and for diagnosing PCV. However, the cause and the pathogenesis of PCV are not clearly known.