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## Polypoidal choroidal vasculopathy in elderly Chinese patients

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**Abstract Purpose:** To investigate the frequency and clinical features of polypoidal choroidal vasculopathy (PCV) in a consecutive series of elderly Chinese patients. **Methods:** A retrospective analysis of 166 consecutive patients 50 years or older with diagnosis of exudative age-related macular degeneration (AMD) was conducted. Color fundus photographs were taken and fluorescein and indocyanine green (ICG) angiography were performed in all patients. **Results:** Of the 166 patients, 37 patients (22.3%) initially suspected of having exudative AMD were ultimately diagnosed as having PCV. Twenty-seven men (73.0%) were affected, 32 patients (86.5%) were unilaterally involved. Of 42 eyes with

PCV, 27 eyes (64.3%) demonstrated polypoidal dilations with branching vascular network, and the other 15 eyes (35.7%) showed scattered polypoidal dilations without identifiable continuous branching vascular network on ICG angiography. The predominant location for these lesions was at the macular region in 26 eyes (61.9%), the temporal vascular arcade in 9 eyes (21.4%), the peripapillary area in 6 eyes (14.3%), and the midperiphery in 1 eye (2.4%). **Conclusions:** PCV is a common disease in elderly Chinese patients. In our study group PCV mainly affected men and was mostly unilateral. Most of the lesions were located in the macular region and temporal vascular arcade.

### Introduction

Polypoidal choroidal vasculopathy (PCV) is characterized by branching choroidal vessels with polyp-like terminal aneurysmal dilations [5, 13, 15]. These vascular abnormalities seem to be a variant of choroidal neovascularization (CNV) and are termed polypoidal CNV [15]. PCV causes subretinal hemorrhage and serous or hemorrhagic pigment epithelial detachment (PED), and usually occurs in people over 50 years of age. These characteristics are shared with neovascularized age-related macular degeneration (AMD). However, the course, visual prognosis and potential response to treatment differ between PCV and AMD [5, 15]; hence, it is important to find the characteristics to distinguish PCV from AMD.

The definition of PCV has expanded over the past 10 years, and the diagnosis is no longer restricted to specific

demographic attributes or to a specific retinal location. Previous studies on white, black and Japanese populations suggested that PCV may be common in Asian people [3, 10, 11, 13, 15]. The purpose of our study was to establish the frequency and clinical characteristics of PCV in consecutive series of elderly Chinese patients.

### Patients and methods

A retrospective study was conducted on 166 consecutive elderly patients (211 eyes) with a presumed diagnosis of exudative AMD. All patients were Chinese. Every patient was seen in the macular service of Zhongshan Ophthalmic Center. Visual acuity testing, color retinography, fundus fluorescein angiography and indocyanine green (ICG) angiography were performed in both eyes of all patients. Fluorescein angiography was performed with a Topcon 50 VT fundus camera or a Zeiss FF450 fundus camera after injection

of 3 ml of 20% sodium fluorescein into the cubital vein. ICG angiography was performed with a Topcon TRC-50IA fundus camera or a Zeiss FF450 fundus camera

For the ICG procedure, 50 mg of ICG (Dianogreen, Daichi Pharmaceutical, Tokyo, Japan) dissolved in 5 ml of distilled solution was injected into the cubital vein within 5 s. The ICG angiograms were obtained over a period of at least 30 min. Informed consent was obtained from all patients before angiographies.

The diagnosis criteria of PCV were as follows: (1) age older than 50 years at onset; (2) subretinal hemorrhage, serous and/or hemorrhagic PED at the posterior pole; (3) branching choroidal vessels with polyp-like terminal aneurysmal dilations in ICG angiography; (4) scattered polypoidal dilations without identifiable continuous branching vascular network in ICG angiography. Pathological myopia, angioid streaks, central serous chorioretinopathy, presumed ocular histoplasmosis, and other retinal or choroidal diseases that could account for exudation and CNV were excluded, as were any patients with previous laser photocoagulation.

The polypoidal CNV was defined as "macular", "peripapillary", "arcade" or "midperipheral" polypoidal CNV when respectively the majority of the polypoidal dilations were located in the macula, within one disc diameter of the disc edge, under the temporal retinal vascular arcade (within one disc diameter of the temporal retinal vein outside the peripapillary area), or outside the posterior pole. In cases with polypoidal vascular complexes in multiple sites, the assigned location was determined by the largest complex.

The criteria for exudative AMD were as follows: (1) age greater than 50 years; (2) serous and/or hemorrhagic detachment of the macula noted on clinical slit-lamp biomicroscopy; (3) evidence of classic CNV or occult CNV on fluorescein angiography; (4) presence of "plaque" and "hot spot" CNV defined according to Guyer et al. on ICG angiography [2]; (5) no evidence of polypoidal dilations from choroidal circulation on ICG angiography.

## Results

### Patients

One hundred sixty-six consecutive elderly Chinese patients (211 eyes) with a presumed diagnosis of exudative AMD were studied retrospectively. There were 118 men (71.1%) and 48 women (28.9%). The subjects' ages ranged from 50 to 82 years with a mean of 68.3 years.

Of the 166 patients, 129 (77.7%) were diagnosed with exudative AMD. Among these cases, there were 16 of classic CNV (12.4%) and 113 of occult CNV (87.6%). The CNV were well defined by ICG angiography in 81 (71.7%) of the 113 occult CNV: 31 hot spots (38.3%) and 50 plaques (61.7%) were noted.

The remaining 37 (42 eyes) patients (22.3%) were diagnosed with PCV. Of these 37 patients with PCV, 27 (73.0%) were men and 10 (27.0%) were women (Table 1). Their ages ranged from 55 to 78 years (mean 66.5 years). One eye was affected in 32 patients (86.5%) and both eyes in 5 patients (13.5%). Their visual acuity of the affected eyes ranged from 0.02 to 0.7. The vision of 16 eyes (38.1%) was more than or equal to 0.3, 14 eyes (33.3%) had visual acuity of between 0.1 and 0.3 (23.3%), and 12 eyes (28.6) of less than 0.1.

**Table 1** Clinical characteristics of 37 patients (42 eyes) with PCV

Characteristics	Number	Proportion
Gender (patients)		
Male	27	73.0%
Female	10	27.0%
Laterality (patients)		
Unilateral lesions	32	86.5%
Bilateral lesions	5	13.5%
Type of polypoidal CNV (eyes)		
Macular	26	61.9%
Arcade	9	21.4%
Peripapillary	6	14.3%
Midperiphery	1	2.4%

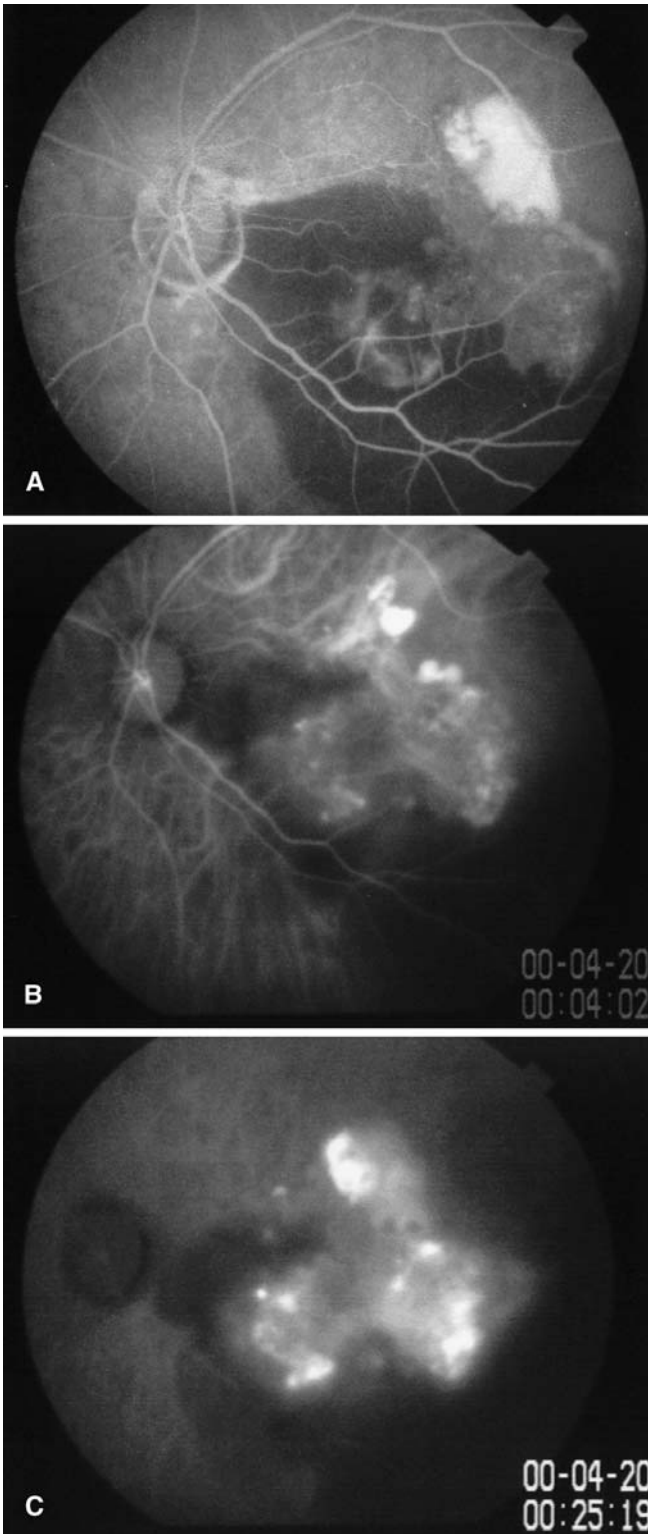
### Findings of fundus examination and fluorescein angiography

All 42 eyes with PCV showed disorders in the posterior pole. Hemorrhagic PEDs were seen in 21 eyes (50.0%). Large subretinal hemorrhage was observed in 14 eyes (33.3%) (Fig. 1); 5 eyes had exudates and no hemorrhage; and 2 eyes (4.8%) had serous PED. Of the 32 patients with unilateral PCV, only 3 (9.4%) had soft drusen in the fellow eye.

Spotty hyperfluorescence was detected in 40 eyes on fluorescein angiography, and all showed slight leakage from the lesions. In two eyes with large subretinal hemorrhage no hyperfluorescence was detected throughout the angiography period. The branching choroidal networks and polyp-like terminal aneurysmal dilations were seen on fluorescein angiography in 3 eyes.

### Findings of ICG angiography

ICG angiography revealed a typical branching vascular networks and polyp-like aneurysmal dilations at terminals of branches in 27 eyes (64.3%). The remaining 15 eyes (35.7%) had polypoidal choroidal vascular dilations without identifiable continuous vessels. The vascular network was clearly demonstrated in the early phase of ICG angiography. Polypoidal hyperfluorescence was well demonstrated in the middle phase of ICG angiography. Persistent staining of active polypoidal lesions can be seen in the late phase of ICG angiography (Fig. 2). The inactive polypoidal lesions presented a hypofluorescent structure surrounded by ill-defined hyperfluorescent ICG dye diffusion, which demonstrated ring-like staining. The number of polypoidal dilations ranged from two to eight. Polypoidal dilations were found in the macular area (macular polypoidal CNV) in 26 eyes (61.9%), the temporal vascular arcade (arcade polypoidal CNV) in 9 eyes (21.4%), the peripapillary area (peripapillary polypoidal CNV) in 6 eyes (14.3%), and the midperiphery (midperiphery polypoidal CNV) in 1 eye (2.4%) (Table 1).



**Fig. 1A–C** A 64-year-old Chinese man with a large subretinal hemorrhage. **A** The venous phase fluorescein angiogram shows blockage of fluorescence by the subretinal hemorrhage with an area of ill-defined hyperfluorescence at the macula, and an irregular pigment epithelial detachment superotemporal to the fovea. **B** The

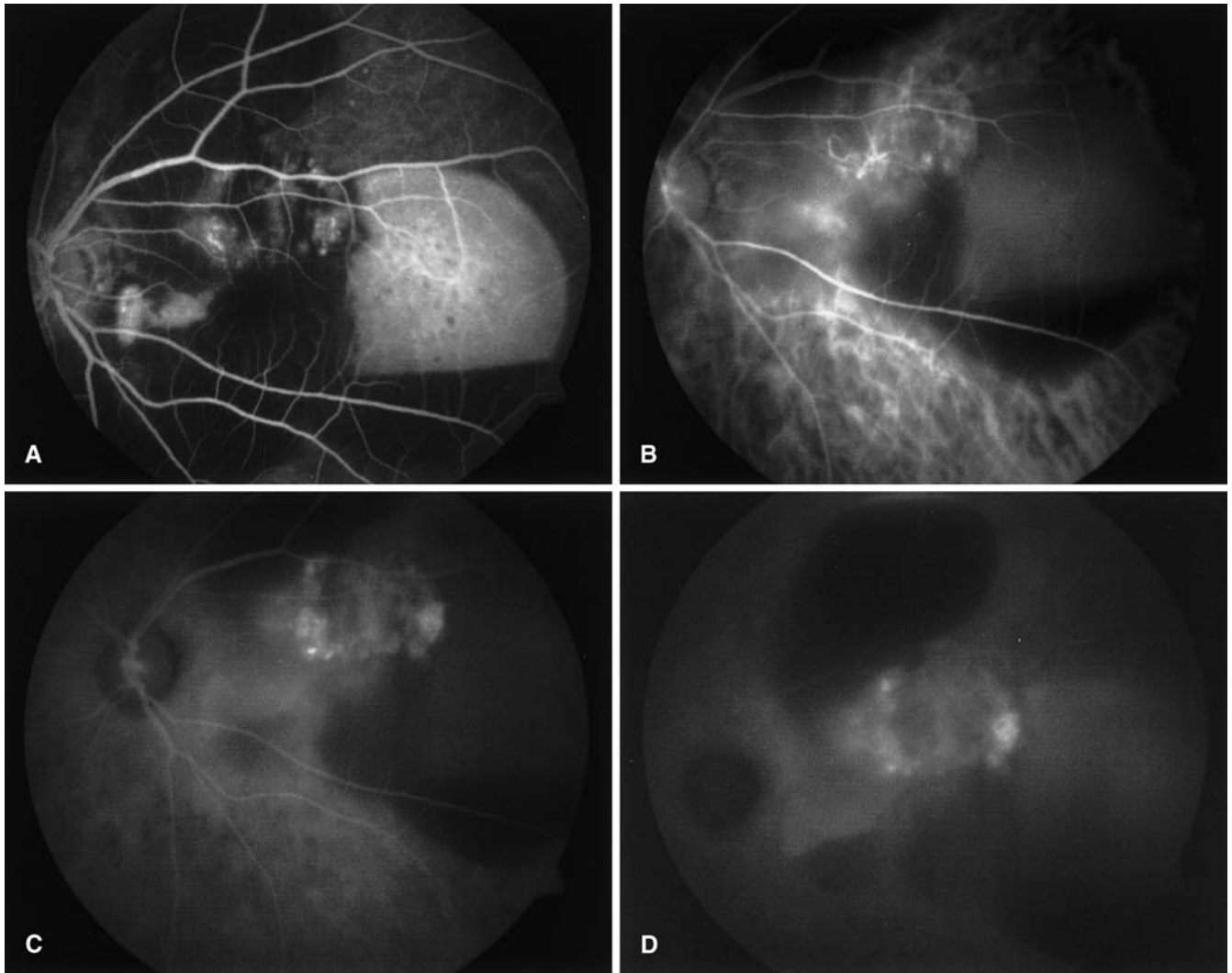
## Discussion

PCV as a cause of recurrent hemorrhagic and exudative PED and neurosensory retinal detachments was first described by Yannuzzi in 1982 [12]. A variety of terms such as “multiple recurrent serosanguinous retinal pigment epithelial detachment” and “posterior uveal bleeding syndrome” have been used to designate this disorder [3, 10]. Because the pathogenesis is unknown and the primary abnormality involves the choroidal vessels and has the character of choroidal vascular network with polyp-like terminal aneurysmal dilations, Yannuzzi et al. suggested the term idiopathic polypoidal choroidal vasculopathy (PCV) in 1990 [13]. At present, the term “PCV” is used to describe this distinct and new clinical entity [15].

PCV is known to have a predilection for more darkly pigmented individuals and Asians, but whites can develop the disorder as well. Yannuzzi et al. [13] and Spaide et al. [9] found that this disorder mainly affected female African Americans. More cases have been reported in Caucasians in Italy and other locations [5, 6, 7]. Uyama et al. [11] reported the characteristics and pathogenesis of PCV in Japanese patients. Our observations support previous reports that PCV may be common in Asian people and suggest that PCV may be frequent in Chinese people. Of the 166 patients in our study, 37 (22.3%) had PCV, a frequency much higher than the 9.3% seen in Chinese patients by Kwok et al [4]. The reason for the difference may be that Kwok et al. did not include the cases of PCV in which there were polypoidal dilations without identifiable continuous branching vascular network on ICG angiography. Our observed frequency was also much higher than that in Italy (9.8%) [7]. An investigator in Singapore has reported that his experience indicates a frequency of approximately 30% in Singaporeans (Dr. Koh Adrian, oral communication, June 2002). We excluded from our survey patients with evidence of AMD, although it is unlikely that drusen protect against PCV. Thus, the frequency may be much higher than this study suggests.

Our findings are similar to those of previous authors in many respects. Our results indicated that 73.0% of the PCV patients were male, which was similar to the reports of Uyama et al. [11] (69%) and Kwok et al. [4] (68.4%). Meanwhile, our study revealed that 86.5% of PCV cases were unilateral, close to the 91% observed by Uyama et al. [11] and the 84.2% seen by Kwok et al. [4]. However, the results differed from those of Yannuzzi et al [13] (80% bilateral). There may be differences between populations in the clinical characteristics of PCV. The epi-

early indocyanine green angiogram reveals a choroidal branching vascular net terminating in multiple polypoidal-like elements in the macular region and superotemporal to the macula. **C** The late indocyanine green angiogram displays persistent staining of the multiple polypoidal-like terminal aneurysmal dilations



**Fig. 2A–D** A 67-year-old Chinese woman with a large serosanguineous retinal pigment epithelium detachment. **A** The venous phase fluorescein angiogram shows subretinal hemorrhage with an area of ill-defined hyperfluorescence and a large serosanguineous retinal pigment epithelium detachment at the posterior pole and temporal vascular arcade. **B** The early indocyanine green angiogram reveals a choroidal vascular network terminating in multiple

polypoidal-like elements at the superotemporal vascular arcade. **C** The polypoidal-like hyperfluorescences were well demonstrated in the middle phase of indocyanine green angiography. **D** The late indocyanine green angiogram displays persistent staining of the multiple polypoidal-like lesions at the superotemporal vascular arcade

demiological features of PCV in Chinese patients are similar to those in Japanese patients.

There is great variation in terms of the location of polypoidal lesions. Early reports emphasized the peripapillary location [13]. Similar anomalies were reported in elderly Caucasians and Japanese, that the polypoidal lesions were often revealed in the macula and rarely in the fundus periphery [5, 11] Our study revealed that more than four fifths of eyes had the polypoidal lesions located predominantly in the macular and temporal vascular arcade areas, and less than one fifth in the peripapillary area, which differed from the observations of Yannuzzi et al [13], but resembled those of Uyama et al. [11] and

Kwok et al. [4]. In our study, the location of the polypoidal lesions fell into four categories: macula (61.9%), temporal vascular arcade (21.4%), peripapillary area (14.3%), and midperiphery (2.4%).

PCV was originally described as an inner choroidal vascular abnormality with two distinct components: (1) a network of branching vessels external to the choriocapillaris and (2) terminal aneurysmal dilations sometimes seen clinically as reddish-orange, spheroidal, polyp-like structures or polypoidal vascular lesions [13]. In the diagnosis of PCV, the presence of an associated network of branching vessels was not considered necessary [5, 7]. In our study, 64.3% of cases of PCV demonstrated poly-

oidal dilations with branching vascular network, and 35.7% showed scattered polypoidal dilations without identifiable continuous branching vascular network on ICG angiography. Sometimes small reddish-orange nodular elevations of retinal pigment epithelium (RPE) were found corresponding to these polyp-like dilation vessels on fundus examination.

PCV can easily masquerade as exudative AMD, but their therapy, natural history and visual prognosis are different. It is important to be aware of their characteristics. AMD is virtually always a bilateral disease, but most patients with PCV have no drusen in the fellow eye [11, 15]. Only in three patients (9.4%) were soft drusen found in the fellow eye in our study. The lesions of AMD occur predominantly in the foveal area, while PCV occurs in the macular, temporal vascular arcade, and peripapillary areas [5, 11, 13]. Especially choroidal vasculopathy in the peripapillary area should be considered PCV. With regard to the clinical characteristics of PCV that closely simulate AMD, ICG angiography is the best tool to image the polypoidal-like lesions and make the correct diagnosis.

Surgical intervention and photocoagulation may be of benefit in eyes with PCV, but the outcome is not certain [4, 8]. Laser photocoagulation should not be used as first-choice treatment for PCV. The natural history of PCV is not completely understood, but many patients have demonstrated chronic multiple recurrent serosanguineous detachments of the RPE and neurosensory retina with long-term preservation of good vision. Some eyes do develop chronic atrophy and cystic degeneration of the fovea with severe vision loss [14]. Our investigation suffers from the limitations of a retrospective study. We are not sure about the terminal visual acuity, natural history and effect of laser photocoagulation in Chinese patients. More cases and prospective, randomized clinical trials are necessary to assess the natural history and the role of conventional laser photocoagulation or photodynamic therapy in Chinese patients with PCV.

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