Idiopathic Polypoidal Choroidal Vasculopathy

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Idiopathic polypoidal choroidal vasculopathy (IPCV) has been recognized as a peculiar form of choroidal neovascularization. The clinical features of recurrent serous retinal leakage and retinal hemorrhage may vary from single lesion to multifocal, from self-limited to recurrent. Caucasian and Japanese patients with IPCV have been reported in the literature. However, research and case reports about IPCV in Chinese patients are relatively rare. We present 2 Chinese patients with IPCV in Taiwan with 2 different clinical pictures and visual outcomes during long-term follow-up. Further study for the etiology, clinical courses and treatments of the different subtypes of IPCV in Chinese is necessary. [*J Chin Med Assoc* 2007;70(2):84–88]

Key Words: choroidal neovascularization, idiopathic polypoidal choroidal vasculopathy, retinal pigment epithelial detachment

Introduction

In the past 2 decades, idiopathic polypoidal choroidal vasculopathy (IPCV) has been recognized by ophthalmologists as a peculiar form of choroidal neovascularization in the inner choroids. It was first described by Yannuzzi in 1982.[1] Clinically, it is characterized by serosanguineous retinal pigment epithelial detachments (RPED) in the posterior pole and can be single or multiple, persistent or regressed, recurrent or nonrecurrent. Previous studies have found that 7.8–23% of patients with neovascularized age-related macular degeneration (AMD) had PCV after evaluation by fluorescein angiography (FAG) and indocyanine green angiography (ICGA).[2,3] We followed 2 Chinese patients with IPCV for >18 months and herein present their different clinical pictures and visual outcomes.

Case Reports

Case 1

A 78-year-old man with medical history of arrhythmia, hypertension, diabetes mellitus, and chronic obstructive pulmonary disease had suffered from blurred vision in his right eye for a few days and came to our hospital. Best corrected visual acuity (BCVA) was 6/12 in the right eye and 6/5 in the left eye. After complete ophthalmic examination, the anterior segment showed only mild cataract of both eyes, and ophthalmoscopy revealed sensory elevation and hemorrhagic RPED (HRPED) at the temporal-lower juxtafoveal area in his right eye (Figure 1A). The retina of his left eye was unremarkable. FAG revealed fluorescence blockage (Figure 2A), and an underlying branching choroidal vascular network was noted with ICGA. Ultrasound B-scan and optical coherence tomography (OCT) (Figure 2B) findings were compatible with HRPED. The hemorrhage regressed spontaneously, leaving a pigmented area without any treatment in a few months.

Five months after the initial presentation, BCVA was 6/7.5. Some old lipid pigmentation in the temporal-lower juxtafoveal area and a new small HRPED in the temporal-upper juxtafoveal area were noted (Figure 1B). FAG revealed no fluorescence leakage, and the previous hemorrhagic area was flat.

Four months later, the patient complained of blurring sensation in the right eye again. BCVA was 6/8.6 in the right eye and 6/5 in the left eye, and new submacular HRPED and sub-RPE exudates in his right eye were noted (Figure 1C). ICGA was performed again and revealed a choroidal vascular network with 2 polypoid-like dilated hot spots nasal to the fovea

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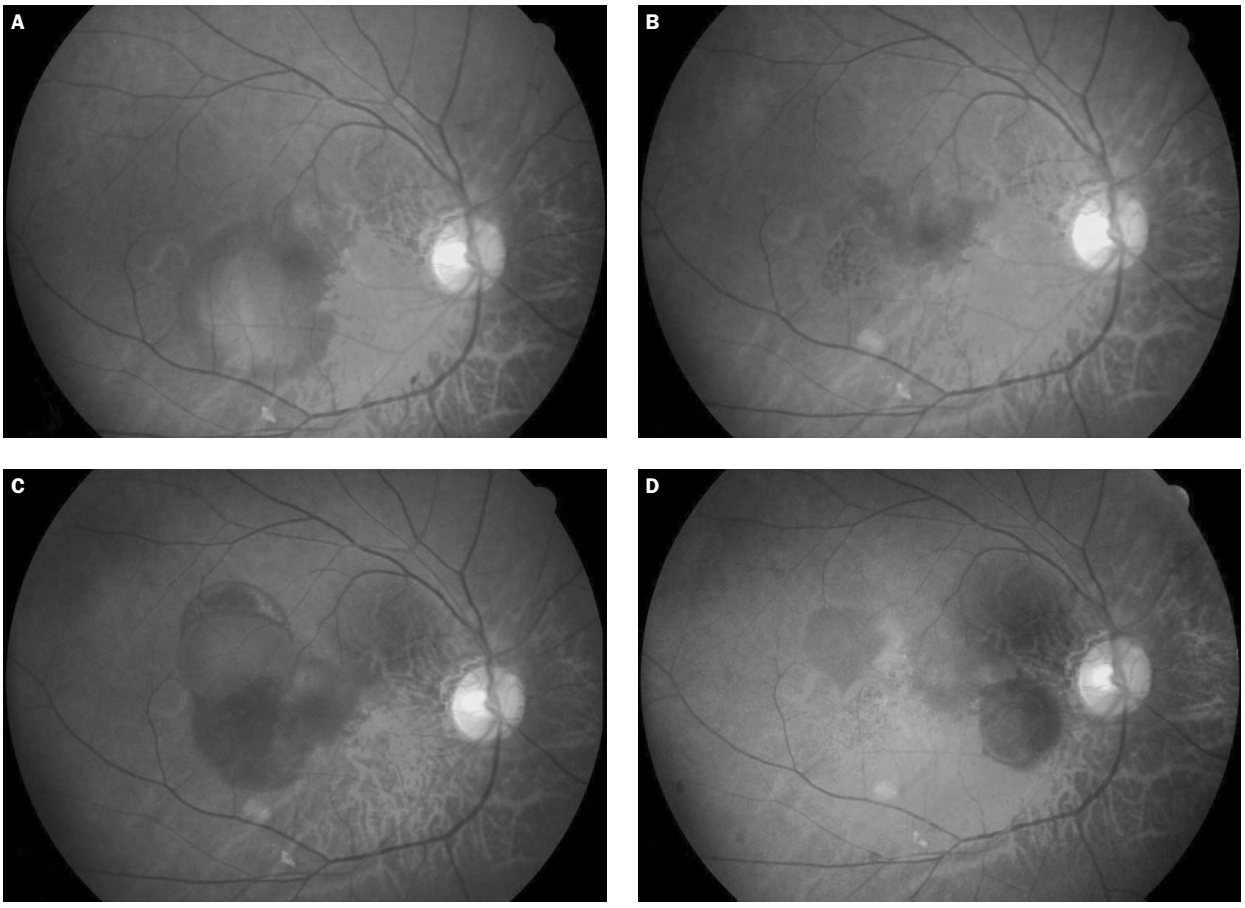

Figure 1. (A) At the first presentation, a sensory elevation and hemorrhagic retinal pigment epithelial detachment (HRPED) in the temporal-lower juxtafoveal area was noted in the patient's right eye. (B) Five months after the first presentation, the old hemorrhage was absorbed, leaving a pigmented area, and a new small HRPED was noted in his right eye. (C) Nine months after the initial presentation, recurrent HRPED in the temporal juxtafoveal area was noted. (D) Fifteen months after the initial presentation, the previous hemorrhage had been absorbed and a new HRPED area was noted in the nasal-lower juxtafoveal area.

(Figure 2C). FAG and OCT showed HRPED in the right eye.

Fifteen months after the first presentation, recurrent HRPED in the patient's right eye (Figure 1D) was noted, and ICGA revealed a new polypoidal dilated hot spot inferior to the previous polypoidal lesions developed from the same vessel network (Figure 2D). The hemorrhage was absorbed spontaneously 3 months later, and final BCVA was 6/8.6 in the right eye and 6/6 in the left eye.

Case 2

An 81-year-old man with medical history of hypertension and gouty arthritis had experienced sudden-onset blurred vision and central vision distortion of his left eye 10 years before coming to our hospital; his BCVA had recovered to 6/6 in his right eye and 6/7.5 in his left eye after laser therapy was performed at another hospital.

Eight years after the first episode, he suffered from sudden onset of vision decrease in his left eye, with visual acuity of 6/6.7 in his right eye and 1/60 in his left eye. Indirect ophthalmoscopy showed exudative RPED with lipid exudates and old laser scar in the patient's left eye (Figure 3A) and macular drusens in his right eye. FAG showed dye leakage that was like the appearance of occult subretinal neovascularization in his left eye. Transpupillary thermotherapy for his left eye was performed twice. The HRPED and exudates subsided, leaving a macular fibrotic scar 9 months after the second treatment.

Ten years after first presentation, in the patient's recent follow-up visit, a new HRPED with extensive diffuse exudates in the superior juxtapapillary area of his left eye (Figure 3B) was noted. ICGA revealed a branching choroidal vascular network and a collection of small polypoidal dilatations of the vessels resembling a cluster of grapes (Figure 3C).

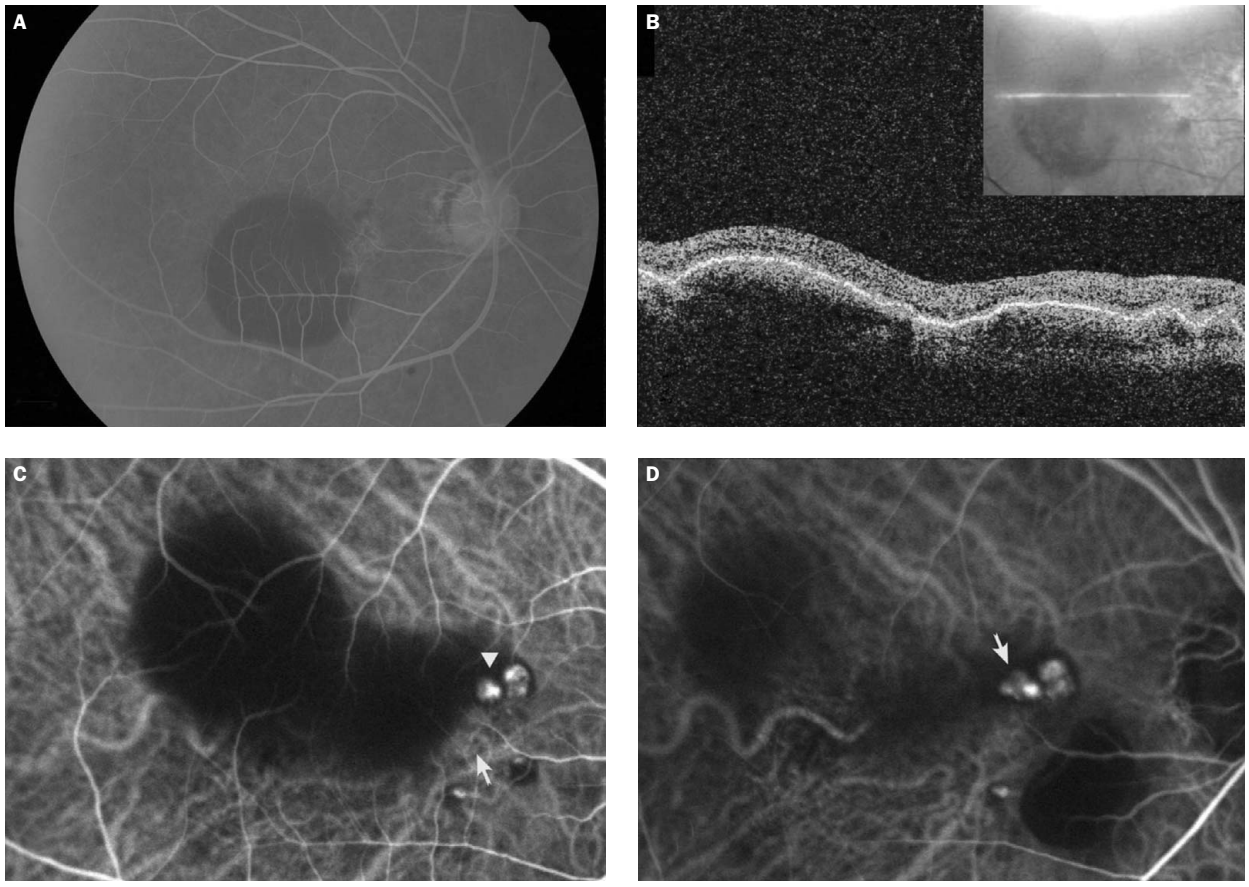

Figure 2. (A) At the first examination, fluorescein angiography revealed fluorescence blocked by hemorrhage; some leakage of undetermined source in the late phase was noted in his right eye. (B) Optical coherence tomography was compatible with retinal pigment epithelial detachment. (C) Indocyanine green angiography at the third episode revealed a choroidal vascular network (arrow) with 2 polypoid-like dilatations (arrowhead) nasal to the fovea. (D) The vascular network persisted, and a new polypoidal dilated hot spot (arrow) was noted 6 months later.

Discussion

IPCV is a disease entity characterized by recurrent serous retinal leakage and hemorrhage in the elderly population, caused by vascular abnormalities in the inner choroids. This disease entity was first described by Yannuzzi in 1982.[1] Kleiner and Brucker[4] subsequently described the term "posterior uveal bleeding syndrome" as with multiple recurrent hemorrhage or serous fluid beneath the RPE and neurosensory retina.

In 1 previous study of PCV, Yannuzzi et al originally demonstrated a predominance of blacks, women, bilateral involvement, and peripapillary location.[2] However, in Italy, Sforzolini et al found a preponderance of unilateral involvement and extramacular location of abnormal vessels in PCV patients.[5] Uyama described the preponderance of men, unilateral involvement, and macular location of PCV in Japanese patients.[6] In 2002, Kwok et al reviewed 204 Chinese patients with a provisional diagnosis of AMD and found that the diagnosis of 22 eyes (9.3%) could be revised to IPCV.[7] The predominance of males (68.4%), unilaterality (84.2%) and macular location of polyps (63.6%) were the same as in Japanese. To our knowledge, the first case of IPCV in Taiwan was reported in 2001.[8] Beyond that, case reports and the study of IPCV patients in Taiwan has been rare. We have presented here 2 Chinese males with IPCV with unilateral involvement, the first 1 presented with recurrent juxtafoveal HRPED and the second presented with peripapillary massive serous and exudative RPED involving the fovea area.

Uyama et al observed the natural history of 14 eyes with PCV for at least 2 years without any treatment.[9] Fifty percent of the patients had a favorable course, and the disease persisted and resulted in a poor visual outcome in the remaining half of patients. They indicated that there are 2 patterns of clinical manifestation of PCV, exudative and hemorrhagic. Theoretically, polypoidal vascular dilations may consist of aneurysmal dilation of vessel wall; these structures can disappear

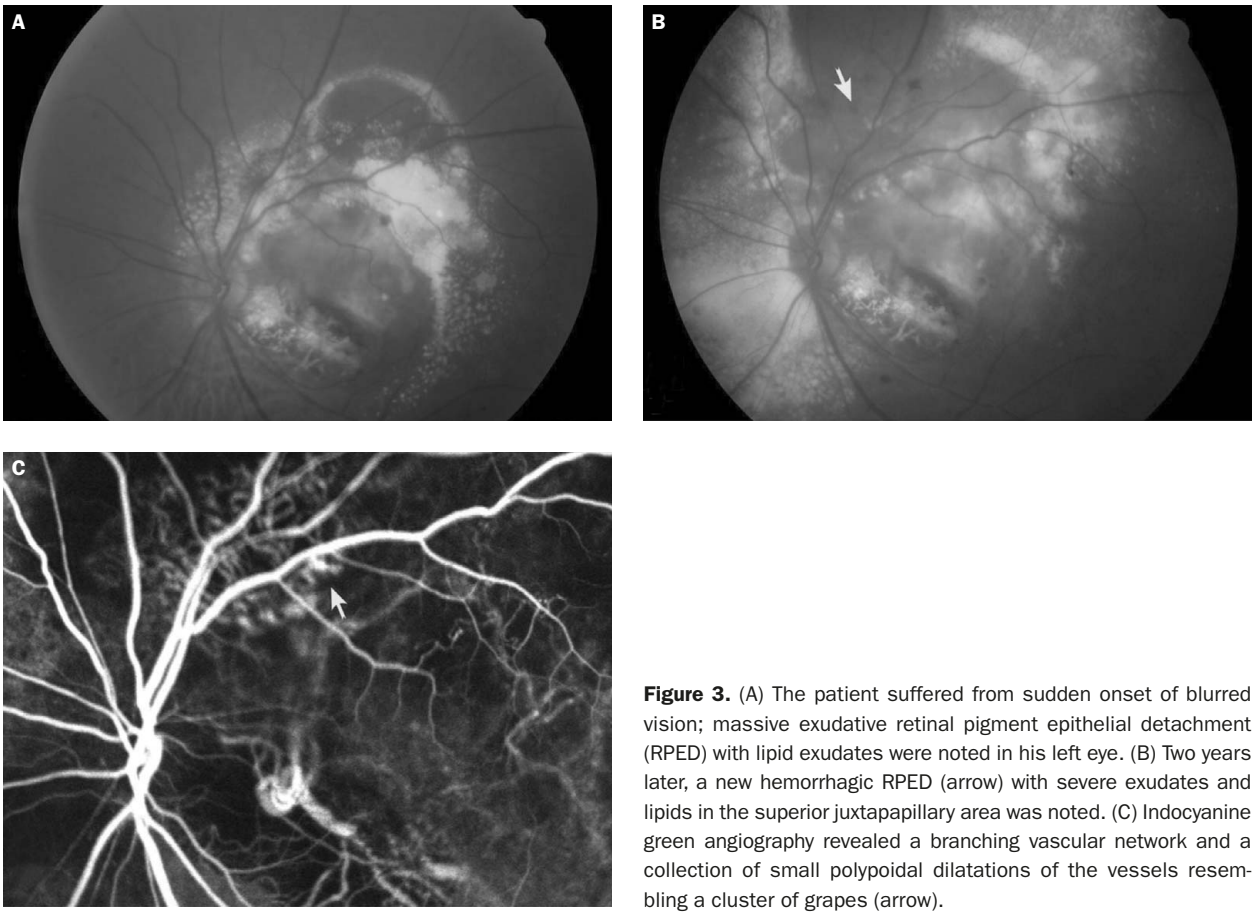

Figure 3. (A) The patient suffered from sudden onset of blurred vision; massive exudative retinal pigment epithelial detachment (RPED) with lipid exudates were noted in his left eye. (B) Two years later, a new hemorrhagic RPED (arrow) with severe exudates and lipids in the superior juxtapapillary area was noted. (C) Indocyanine green angiography revealed a branching vascular network and a collection of small polypoidal dilatations of the vessels resembling a cluster of grapes (arrow).

due to occlusion of the vessels by thrombus formation within the vessels in a given area, and can grow due to dilation of the vessel walls in other areas. Different clinical manifestations may develop due to either leakage or hemorrhage. Leakage in the vessel wall at aneurysmal dilations leads to the exudative pattern. Hemorrhage may derive from rupture of aneurysmal dilations, and massive hemorrhage may derive from rupture of venules and sometimes of arteries. In our report, case 1 showed recurrent hemorrhage in the parafoveal area, with visual acuity of 6/8.6 in his right eye during 18 months of follow-up. In contrast, case 2, who had a clinical picture of massive lipid exudates in the peripapillary area, presented a poor visual outcome, with visual acuity of 1/60 in his left eye at 10 years after the first presentation.

ICG angiography is the best tool available today for the visualization of PCV. It usually shows a branching vascular network from the choroidal circulation and the characteristic polypoidal and aneurysmal dilations at the terminals of branching vessels.[10,11] The branching vascular networks may last for a long period of time, however, the polypoidal dilations at the terminals of the network may change configuration; new dilations grow, while others regress. Uyama et al pointed out that there were 2 patterns of polypoidal dilations in ICGA: (1) a large solitary round aneurysmal dilation; and (2) a collection of small aneurysmal dilations resembling a cluster of grapes.[9] They also demonstrated that patients with lesions of the first pattern had relatively favorable clinical course and better visual outcome. Lesions of the second pattern, in contrast, were usually active and tended to bleed or leak and cause severe visual loss. The ICGA pictures and outcomes in our cases were similar to Uyama et al's observation. Case 1 had the first pattern, with relatively good visual outcome, case 2, presenting a group of dilations resembling a cluster of grapes in ICGA, had poor visual outcome.

Clinicopathologically, Robert et al described the dilated polypoidal lesions as large thin-walled cavernous vascular channels without muscular layer that originated from branches of the short posterior ciliary arteries in a patient with IPCV undergoing enucleation.[12] Both of our 2 cases had medical history of hypertension. Although the relationship between hypertension and IPCV has not been well demonstrated, 1 theory is that increased perfusion pressure secondary to hypertension

within the arteriolar branches of the short posterior ciliary arteries stimulates the prolapse and protrusion of these vessels through defects or gaps in Bruch's membrane in the peripapillary region.

In summary, we presented 2 typical cases of IPCV with different clinical presentations and visual outcomes during long-term follow-up. Case 1, which can be classified as a hemorrhagic type of IPCV, with several recurrent large round aneurysmal dilations, had the relatively better visual outcome. Case 2, with a collection of small aneurysmal dilations resembling a cluster of grapes, can be classified as an exudative type of IPCV and had a poor visual outcome. Further study of the etiology, clinical courses, and treatments of the different subtypes of IPCV is necessary.

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