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ORIGINAL PAPER

Case series of coexistence of polypoidal choroidal vasculopathy with other rare fundus diseases

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Abstract
Purpose To record the coexistence of polypoidal choroidal vasculopathy (PCV) with other rare fundus diseases in a Chinese population.
Method In this retrospective hospital-based study, a chart review of 861 patients with newly diagnosed PCV was performed. The clinical features of rare fundus comorbidities of PCV were recorded.
Results Five eyes of 5 patients aged 63.4 ± 11.22 years (0.58%) had PCV coexisting with other fundus diseases in the same eye. Of the 5 PCV patients, 2 (0.23%) had myelinated nerve fiber, 2 (0.23%) had branch retinal vein occlusion, and 1 (0.12%) had retinal angiomatous proliferation.
Conclusion We reported rare fundus comorbidities of PCV in a large Chinese cohort. These comorbidities included myelinated nerve fiber, branch retinal vein occlusion and retinal angiomatous proliferation. The combination might constitute an accidental occurrence.

Keywords Polypoidal choroidal vasculopathy · Myelinated nerve fiber · Branch retinal vein occlusion · Retinal angiomatous proliferation

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Introduction

Polypoidal choroidal vasculopathy (PCV) is characterized by polyp-like terminal aneurysmal dilations with or without branching choroidal vessels [1]. Currently, PCV is considered a pachychoroid spectrum disease, receiving increasing attention over the past several years [2]. There have been various studies of the clinical manifestations and treatments for PCV [3]; however, there have been only limited reports regarding accompanying fundus diseases. To date, there remains a lack of systematic studies of the prevalence of coexisting fundus lesions. Because Chinese patients have a higher incidence of PCV [4], cases of PCV coexisting with other fundus diseases might be observed more often.

The purpose of this study was to record the coexistence of PCV with other rare fundus diseases in a Chinese population.

Methods

A retrospective study was conducted of 861 consecutive patients presenting to the Zhongshan Ophthalmic Center with PCV between September 2010 and September 2017. In this retrospective case series, patients were selected from case presentations. Medical and systematic histories were reviewed. Approval was obtained by the Zhongshan Ophthalmic Center

institutional review board at the Zhongshan Ophthalmic Center of Sun Yat-sen University. The study adhered to the tenets of the Declaration of Helsinki.

All patients underwent slit-lamp biomicroscopy, fundus examination, fluorescein angiography (FA) and indocyanine green angiography (ICGA) with a Zeiss FF450 fundus camera (Carl Zeiss, Oberkochen, Germany) or a Heidelberg scanning laser ophthalmoscope (HRA-2; Heidelberg Engineering, Heidelberg, Germany) to analyze the number of cases of PCV coexisting with other fundus diseases and the locations of PCV. The diagnostic criteria for PCV were branching choroidal vascular networks with polyp-like terminal aneurysmal dilations or scattered polypoidal dilations without identifiable continuous branching vascular networks on ICGA. Coexisting lesions were one or more kinds of fundus diseases presenting with PCV in the same eye.

Each angiogram was reviewed by at least two of the authors who were masked to the clinical findings. In a few cases (7 cases, 0.81%), there was disagreement in interpretation, and the determination was referred to Dr. Wen.

Results

PCV coexisting with other rare fundus diseases was identified in 5 patients (5 eyes) (0.58%) of 861 patients. The 5 patients with mixed lesions all had PCV coexisting with other fundus diseases in the same eye. The mean age was 63.40 years, with a range of 54–80 years, and the best-corrected visual acuity was 20/200 to hand motion. Most patients (80%) were male, and most of the PCV lesions were unilateral (60%). Two of the patients (0.23%) had myelinated nerve fiber (Case 1, Fig. 1), 2 (0.23%) had branch retinal vein occlusion (BRVO) (Case 3, Fig. 2), and 1 (0.12%) had retinal angiomatous proliferation (RAP). The characteristics of all 5 patients are summarized in Table 1.

On ICGA, polypoidal lesions were found in the macular region in 4 (80%) eyes and in the inferotemporal vascular arcade in 1 (20%) eye. The contralateral eyes were normal in 2 (40%) cases. PCV lesions were found in the contralateral eyes of 2 (40%) patients.

Discussion

PCV may result in recurrent serosanguinous retinal pigment epithelium detachment and neurosensory retinal detachment, which may obscure the observation of coexisting lesions. However, there have been only sporadic reported cases of PCV coexisting with other fundus diseases [5–8]. The current study investigated the prevalence and clinical features of PCV coexisting with other fundus diseases by observing a retrospective case series of Chinese PCV patients. Our study demonstrated that coexisting fundus diseases developed in 0.58% of PCV patients in the same eye. As far as we know, no other studies have systematically observed PCV coexisting with other fundus diseases.

The mixed lesions in the 5 patients included myelinated nerve fiber, branch retinal vein occlusion and RAP. The combination of PCV with these fundus diseases has not been reported before. Our study found these new comorbidities, but some of the previously reported comorbidities were not screened out. The reason may be because the rate of PCV combined with other fundus diseases was very small; many of them were an accidental phenomenon, and other reports have to date reported only one case [9–11].

The present study found that, although combined with other fundus diseases, PCV in these patients remained male predominant, primarily unilateral, and located in the macular region and in the inferotemporal vascular arcade. The demographic features of PCV in the combined cases were similar with that of pure PCV. In the combined cases of PCV coexisting with BRVO and RAP, the combined diseases were at their common locations, while PCV was in the most common area, the macular region, in all cases. These results demonstrated that PCV and the fundus comorbidities do not significantly affect each other and continue to follow the gender and eye characteristics in the population.

The average prevalence rate of myelinated retinal nerve fiber in adult Chinese populations was 0.4% per eye [12]. The myelinated retinal nerve fibers were located most often in the temporal inferior fundus region and in the temporal superior fundus region. The prevalence rates of the almost completely encircled optic disk and nasal region were 0.09 and 0.11%, respectively [12]. In the present study, the myelinated retinal nerve fibers completely encircled the optic disk

Fig. 1 Fundus manifestation of Case 1. **a** Fundus photograph demonstrating myelinated retinal nerve fibers completely encircling the disk, a reddish-orange lesion and subretinal hemorrhages in the macular area. **b** FFA (venous phase) of the left eye showing weak fluorescence surrounding the disk and spotty hyperfluorescence in the macular area. **c** ICGA (early phase) of the left eye showing well-defined polypoidal dilations in the macular area

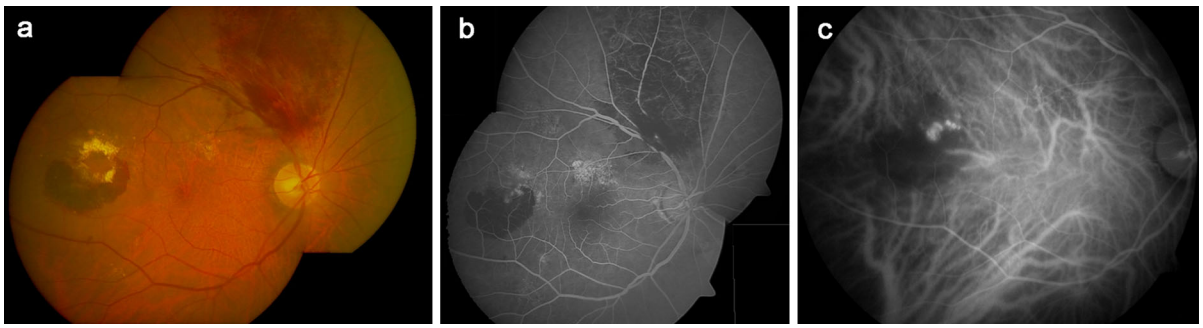

Fig. 2 Fundus manifestation of Case 3. **a** Fundus photograph showing branch retinal vein occlusion above the disk and subretinal hemorrhages and retinal exudation in the macular area. **b** FFA (venous phase) of the right eye demonstrating block fluorescence above the disk and spotty hyperfluorescence in the macular area. **c** ICGA (early phase) of the right eye showing weak fluorescence above the disk and well-defined polypoidal dilations in the macular area

Table 1 Angiographic and clinical characteristics of 5 patients with PCV coexisting with other fundus comorbidities

Case No.	Age (years)	Gender	Eye	Fundus comorbidities	Location of PCV	Visual acuity	Hyperpermeability of choroidal capillaries	Contralateral eye
1	54	M	OS	Myelinated retinal nerve fibers	Macular region	20/200	Yes	Normal
2	57	M	OS	Myelinated retinal nerve fibers	Macular region	6/100	Yes	Typical CNV
3	80	M	OD	BRVO	Macular region	6/100	Yes	PCV
4	70	F	OS	BRVO	Macular region	20/400	Yes	PCV
5	56	M	OS	RAP	Inferotemporal vascular arch area	HM/ 30 cm	Yes	Normal

PCV polypoidal choroidal vasculopathy, *BRVO* branch retinal vein occlusion, *RAP* retinal angiomatous proliferation

and nasal region, which are uncommon locations for this relatively rare disease. The coexistence of PCV with myelinated retinal nerve fibers in this study is therefore very rare.

According to previous studies, a few combined diseases were thought to be related to PCV with similar pathogenic mechanisms [13–15]. For example, PCV in systemic lupus erythematosus and associated antiphospholipid syndrome might be caused by

chorioretinal vasculitis and ischemia with increased vascular endothelial growth factor levels [13]. However, according to the present and other studies, the majority of combined diseases might be coincidental, including myelinated nerve fiber, BRVO, RAP and retinitis pigmentosa [6, 11]. These entities are independent of each other; whether they were related to one another necessitates further investigation.

Conclusion

We reported rare fundus comorbidities of PCV in a large Chinese cohort. These comorbidities included myelinated nerve fiber, branch retinal vein occlusion and retinal angiomatous proliferation. The combinations might be accidental occurrences.

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Compliance with ethical standards

Conflicts of interest The authors declare that there are no conflicts of interest regarding the publication of this paper.

Ethical approval All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional ethics committee and with the 1964 Declaration of Helsinki and its later amendments.

References

1. Yannuzzi LA, Ciardella A, Spaide RF, Rabb M, Freund KB, Orlock DA (1997) The expanding clinical spectrum of idiopathic polypoidal choroidal vasculopathy. Arch Ophthalmol 115(4):478–485
2. Lee JH, Park HY, Baek J, Lee WK (2016) Alterations of the lamina cribrosa are associated with peripapillary retinoschisis in glaucoma and pachychoroid spectrum disease. Ophthalmology 123(10):2066–2076
3. Tomiyasu T, Nozaki M, Yoshida M, Ogura Y (2016) Characteristics of polypoidal choroidal vasculopathy evaluated by optical coherence tomography angiography. Investig Ophthalmol Vis Sci 57(9):OCT324–OCT330
4. Liu Y, Wen F, Huang S, Luo G, Yan H, Sun Z et al (2007) Subtype lesions of neovascular age-related macular degeneration in Chinese patients. Graefes Arch Clin Exp Ophthalmol 245(10):1441–1445
5. Nakagawa S, Yamashiro K, Tsujikawa A, Otani A, Tamura H, Ooto S et al (2013) The time course changes of choroidal neovascularization in angioid streaks. Retina 33(4):825–833
6. Ishida T, Moriyama M, Morohoshi K, Furuse Y, Fukuda T, Ohno-Matsui K (2013) Polypoidal choroidal vasculopathy in a case with retinitis pigmentosa. Int Ophthalmol 33(3):305–308
7. Peiretti E, Iranmanesh R, Yannuzzi LA, Fossarello M (2008) Polypoidal choroidal vasculopathy associated with tilted disk syndrome. Retin Cases Brief Rep 2(1):31–33
8. Mauget-Faysse M, Cornut PL, Quaranta El-Maftouhi M, Leys A (2006) Polypoidal choroidal vasculopathy in tilted disk syndrome and high myopia with staphyloma. Am J Ophthalmol 142(6):970–975
9. Baillif-Gostoli S, Quaranta-El Maftouhi M, Mauget-Faysse M (2010) Polypoidal choroidal vasculopathy in a patient with angioid streaks secondary to pseudoxanthoma elasticum. Graefes Arch Clin Exp Ophthalmol 248(12):1845–1848
10. Bartlett HM, Willoughby B, Mandava N (2001) Polypoidal choroidal vasculopathy in a patient with melanocytoma of the optic nerve. Retina 21(4):396–399
11. Li H, Wen F, Wu D (2004) Polypoidal choroidal vasculopathy in a patient with circumscribed choroidal hemangioma. Retina 24(4):629–631
12. You Q, Xu L, Jonas JB (2007) Prevalence of myelinated retinal nerve fibres in urban and rural adult Chinese populations: the Beijing Eye Study. Acta Ophthalmol Scand 85(6):631–632
13. Chin YC, Bhargava M, Khor CC, Cheung CM, Wong TY (2014) Polypoidal choroidal vasculopathy and systemic lupus erythematosus. Lupus 23(3):319–322
14. Toyama T, Ohtomo K, Noda Y, Ueta T (2014) Polypoidal choroidal vasculopathy and history of central serous chorioretinopathy. Eye (Lond) 28(8):992–997
15. Nicolas El-Haddad M, Clement Fernandez F, Perez-Salvador E, Martin Reyes C (2002) Polypoidal choroidal vasculopathy in a patient with a melanocytoma of the optic nerve. Arch Soc Esp Oftalmol 77(8):455–458