Giant non-pulsatile polypoidal choroidal vasculopathy

Elton Lik Tong Tay, Augustinus Laude

National Healthcare Group Eye Institute, Tan Tock Seng Hospital, 308433, Singapore

Correspondence to: Elton Lik Tong Tay. Department of Ophthalmology, Tan Tock Seng Hospital, 11 Jalan Tan Tock Seng, 308433, Singapore. Elton_tay@ttsh.com.sg

Received: 2016-02-04 Accepted: 2016-04-12

DOI:10.18240/ijo.2016.12.26


Dear Editor,

Polypoidal choroidal vasculopathy (PCV) was first described by Yannuzzi et al. (1) in the 1980s as a distinct choroidal abnormality. Clinically, PCV is seen as orange-red subretinal nodules (2) frequently presenting with submacular haemorrhage or serosanguinous pigment epithelial detachments (PED) (3). Indocyanine green angiography (ICGA) provides the means to confirm the diagnosis of PCV, with angiographic features of early focal hyperfluorescence (within 6 min of angiography), with or without associated branching vascular networks.

Formal PCV definitions were first proposed by the Japanese Study Group of Polypoidal Choroidal Vasculopathy, which suggested a diagnosis of “definite” PCV in the presence of protruding elevated orange-red lesions on fundus examination and/or characteristic polypoidal lesions on ICGA. A diagnosis of “probable” PCV was made if only the abnormal branching vascular network was seen on ICGA and/or presence of recurrent hemorrhagic and/or serous detachment of the retinal pigment epithelium (RPE) (4).

More recently, the EVEREST trial defined PCV as presence of early (within 6 min of dye injection) subretinal focal ICGA hyperfluorescence, plus at least one of the following angiographic or clinical criteria: association with a branching vascular network, presence of pulsatile polyp, nodular appearance when viewed stereoscopically, presence of hypofluorescent halo (in the first 6 min of angiography), orange subretinal nodule on colour fundus photograph or association with massive submacular hemorrhage (at least 4 disc areas) (5).

In this report, we describe three cases that met the EVEREST trial definition of PCV, as well as the Japanese Study Group’s definition of “definite” PCV. However, they appear to be a unique subset of PCV as they presented with very large non-pulsatile polyps with an apparent internal architecture. The cases were also more resistant to treatment. To our knowledge, this distinctive phenotypic subset of PCV has not been described before.

Report of Cases A 64 year-old Chinese male presented with right poor vision of uncertain duration. He was a smoker and had hypertension, hyperlipidemia, and a history of a previous cerebrovascular accident.

His right best corrected visual acuity (BCVA) on presentation was counting fingers at half metre. In the right fundus, there was an orange-red nodule associated with a half disc diameter haemorrhagic PED at the macula with surrounding subretinal fluid (SRF) and hard exudates (Figure 1).

Right eye ICGA revealed a large polyp measuring 1381 μm (in the largest diameter). It was nodular stereoscopically, non-pulsatile, with no associated branching vascular networks (BVN). There was presence of a hypofluorescent halo around the polyp, and the polyp appeared to have an intraluminal architecture. Optical coherence tomography (OCT) showed a peaked sub-RPE lesion with SRF in the right eye. The patient declined anti-vascular endothelial growth factor (VEGF) injections in view of his vascular risk factors and opted for photodynamic therapy (PDT). He underwent 2 PDT treatments with no sustained improvement. His most recent BCVA is counting fingers closely in the right eye.

Case 2 A 55 year-old Chinese gentleman presented with right poor vision for a month. He had hypertension and was a smoker. His right BCVA on presentation was 20/80. In the right fundus, there was an orange-red nodule seen with subretinal haemorrhage and surrounding SRF (Figure 2).

Right ICGA revealed a large polyp measuring 1388 μm (in the largest diameter). It was nodular stereoscopically, non-pulsatile, with no associated BVN. There was presence of a hypofluorescent halo around the polyp, and the polyp appeared to have an intraluminal architecture. OCT showed a peaked sub-RPE lesion with SRF. The patient underwent 11 intravitreal ranibizumab injections. However, the SRF persisted and his most recent BCVA is 20/50.

Case 3 A 60 year-old Chinese gentleman presented with right blurred vision for 3mo. He was a smoker with hypertension, hyperlipidemia and ischaemic heart disease. His right BCVA on presentation was 20/40. In the right fundus, there were two orange-red nodules seen with PEDs at the macula (Figure 3).
ICGA revealed two large polyps measuring 1105 and 1448 μm (in their largest diameters). They were nodular stereoscopically, were non-pulsatile, and with no associated BVN. There were presence of a hypofluorescent halos around the polyps, and the polyps appeared to have intraluminal architectures. OCT of the right eye showed two peaked sub-RPE lesions with SRF. The patient underwent 3 intravitreal ranibizumab, 3 intravitreal aflibercept injections as well as PDT with no sustained improvement. He subsequently developed a macular scar and still has minimal macula thickening. His latest BCVA is 20/40.

**DISCUSSION**

PCV lesions often vary histopathologically. Lafaut *et al.* (1) described PCV as sub-RPE fibrovascular membranes containing dilated thin-walled vessels supporting the postulate that PCV could be a neovascular lesion. Another study showed histological similarities between branch retinal vein occlusion and PCV lesions, with vessel walls hyalinization (7). Nakashizuka *et al.* (8) reported histological findings of hyalinization of choroidal vessels similar to retinal macroneurysms, and proposed that arterosclerotic processes contributed to PCV lesions. One study compared larger and smaller-sized PCVs and proposed that in addition to histopathological differences, large-sized PCVs could reflect later stage of disease (9).

We present a case series of three patients with arteriosclerotic risk factors who presented with giant PCVs. In addition to similar patient profiles, the three giant PCV cases also shared clinical and angiographic similarities of very large non-pulsatile polyps with an internal architecture. We hypothesise that the internal filling pattern within the giant polyp represented angiographic dye filling up spaces around areas of partial thrombosis of stagnant blood within the lumen of the giant PCV. The intraluminal thrombosis hypothesis also supports the finding of absence of pulsatility on angiography. All cases of giant PCVs appeared more resistant to treatment, with poor visual acuity after treatment. In conclusion, we present a case series of a unique phenotype that we term giant PCV. We note that patients with giant PCV have similar risk factors of smoking, hypertension and vascular risk factors. Giant PCV have angiographic features of large non-pulsatile polyps with internal luminal architectures. Giant PCVs are also resistant to treatment with poorer resultant visual acuity. Histological studies will
benefit greater understanding of giant PCVs, and as this is a small case series, a review of more giant PCVs will elucidate more clearly its natural history and response to treatment.

ACKNOWLEDGEMENTS

Conflicts of Interest: Tay ELT, None; Laude A, None.

REFERENCES