An Editorial on Optical Coherence Tomography Angiographic (OCTA) Findings in Polypoidal Choroidal Vasculopathy (PCV)

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Yanuzzi, et al. in 1990 described the orange pink polypoid lesions in the peripapillary and macular area with underlying abnormal choroidal vascular network as idiopathic polypoidal choroidal vasculopathy (IPCV) [1]. Later on, multiple authors performed studies to evaluate these abnormal choroidal branching vascular networks (BVN) and described the clinical features, imaging findings and treatment modalities of polypoidal choroidal vasculopathy (PCV) [2,3]. PCV is more common among the pigmented ethnicities such as Asians and Africans than the Caucasians and can present as exudative maculopathy, hemorrhagic serous macular detachment [2,4,5]. Various clinical manifestations of PCV are orange-red polypoidal lesions, pigment epithelial detachment (PED), serous subretinal fluid (SRF), hemorrhagic PED’s, massive subretinal hemorrhage, and/or exudative detachment [2,4,5]. PCV can be mistaken clinically for the occult choroidal neovascularisation (CNV) of age-related macular degeneration (ARMD) and chronic central serous chorioretinopathy (CSCR) [6,7]. There are recent advances in the imaging technology of retino-choroidal pathology that describe precisely the vascular changes of PCV from wet ARMD. PCV can be differentiated from CNV of ARMD and CSCR by indocyanine green angiography (ICGA). ICGA is the gold standard test to diagnose the choroidal polyps of PCV. PCV if not diagnosed and treated properly can lead to permanent visual deterioration due to the damage to the retinal pigment epithelium (RPE), hemorrhage, disciform scars through chronic progression [6,7]. The major study which was performed to describe these lesions was EVEREST study followed by many studies [2].

It commonly presents after 60 years of age and occurs both in females (Caucasians) and male (Asians) genders. The risk factors associated with PCV are cardiovascular diseases, systemic hypertension, diabetes mellitus, obstructive sleep apnoea and thrombocytopenia [3,5,8]. The pathogenesis involved in PCV is due to abnormality in the inner choroidal vessels that bulge and form polypoidal protrusions. PCV like ARMD is also considered to be due to abnormalities in the complement cascade pathway. The complement factors that have significant association with PCV are the complement factor-H (CFH) and complement component 2 (C-2) polymorphisms which are also seen in neovascular ARMD. The PCV can be unilateral or bilateral with single or multiple polypoid lesions in the macular and peripapillary area [5,8]. The visual acuity (VA) can be good in the initial stages, but with progression of PED, SRF, subretinal haemorrhages or exudates, the VA can be diminished. Apart from decreased VA, patients can also complain of metamorphopsia, central scotoma or floaters [5,8]. Various imaging modalities were performed to describe these lesions such as fluorescein angiography (FA), ICGA, and spectral domain

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optical coherence tomography (SD-OCT) [5,6,7,9-13]. The standardized diagnostic criteria of PCV on FA and ICGA are nodular appearance of the polyps on stereoscopic color fundus photographs, hypofluorescent halo around the nodule, abnormal vascular channels supplying the polyps, pulsatile filling of the polyps, orange subretinal nodules corresponding to the hyperfluorescent areas on ICGA, and massive subretinal hemorrhage (graded by Central Reading Center) [2,3]. The classic clinical finding of PCV is the reddish-orange subretinal nodules that can vary in size from small, medium to large depending on overlying RPE thinning caused by disease pathology. Based on the stereoscopic confocal scanning fundus photography, the polyps in PCV are classified as quiescent (presence of polyps with absence of SRF or subretinal haemorrhages), active, exudative (lipid exudates with intraretinal fluid, PED, serous macular detachment, absence of hemorrhage), hemorrhagic (subretinal or intraretinal hemorrhage and hemorrhagic PED) or mixed [5,8]. Upon fundus fluorescein angiography, PCV features can mimic CNV. Diffuse stippled hyperfluorescence is seen on FA in PCV. However, FA cannot definitely reveal the polypoidal lesions of PCV, but helpful in defining the greatest linear dimensions of the lesions area. ICGA is indicated when there is massive subretinal hemorrhage, serosanguinous macular detachment, notched PED’s, multiple or large orange-red nodules with no response to multiple injections of anti-vascular endothelial growth factor (VEGF) therapy [7,14]. ICG dye is 98% protein bound with longer wavelength, penetrates through the pigment, fluid, lipid or hemorrhage and is better imaging modality to study the choroidal vasculature. To confirm the diagnosis of PCV, ICGA is the investigation of the choice. Polyps appear as hyperfluorescent nodule in the initial phases after injections (approximately 2 - 5 minutes) surrounded by a halo of hypofluorescence. The polyps can be solitary or multiple on ICGA and can have ring (whorl pattern) or cluster (bunch of grapes) patterns [14]. Based on dynamic ICGA findings, the abnormal branching vessels of PCV can be classified into different patterns such as type 1 polyps with abnormal BVN (has both feeder and draining vessels), type 2 polyps without BVN (no feeder or interconnecting vessels). The characteristic feature of PCV on dynamic ICGA is the pulsatile filling of the polyps. Another advantage of dynamic ICGA is best visualization of the exact boundaries of the abnormal BVN in early phases of angiogram. ICGA is a two dimensional imaging modality that lacks the ability to localise the lesions in various layers of the retina and choroid as seen on OCT. ICGA is an invasive procedure with a need to inject intravenous dye which can have allergic reactions, and it will be difficult to inject dye at every follow-up visit after treatment, highly time consuming unlike OCT and ICGA may not be available in every eye clinic [7,14].

Optical coherence tomography is a non-invasive best imaging modality that provides high-resolution cross-sectional images of the retina and detects the morphological pathology of retina and choroid [9-13]. Both SD-OCT and swept source (SS) OCT provide better demarcation of outer retinal layers and morphological changes of the PCV. Spectral-Domain OCT is a useful tool to reveal the features of PCV such as sharp PED’s (thumb-like polyps), multiple and multilobulated PED’s, PED notching, double layer sign (DLS) at the RPE-Bruch’s membrane complex and round hyporeflective lesions corresponding to the polyp lumen. Underneath the RPE, attached are the hyperreflective lesions within which are these hyporeflective lesions [6,10,11,12]. The double layer sign, most important sign of PCV visualized on SD-OCT is described as separation of the irregular RPE (one layer, top) layer and inner layer of the intact Bruch’s membrane (second layer, bottom) [11]. The space within the DLS is occupied with fluid that accumulates between the basement membrane of the RPE and the inner layer of the Bruch’s membrane/choriocapillaris complex, which is secreted by the leaking vascular channels of the polyps. DLS on tomography is depicted as the BVN of PCV, which correlates to the early hypercyascence of the branching network on ICGA [11]. With SD-OCT, inner choroid cannot be visualized due to inability of signal to pass beneath the RPE-Bruch’s membrane complex. The main disadvantage of the basic examination via OCT is inability to reveal the pathological changes in the blood vessels of PCV. Swept source (SS)-OCT provides better details of choroidal pathology due to improved signal strength, better penetration into the choroid than SD-OCT. Swept source based Doppler OCT has higher penetration and provides 3D structural visualization of PCV including the branching vascular network and abnormal feeder vessels [9,13]. En face imaging by using SS-OCT is another step in the advancement of OCT to study the outer retinal layers, and choroid, choriocapillaris and choroidal vasculature pathology even upto the choroido-scleral junctio on [13]. En face SS-OCT clearly shows whether the abnormal BVN is beneath the RPE, above or beneath the Bruch’s membrane, within the choriocapillaris or large choroidal vascular layers. It also shows the focal or diffuse dilated choroidal vessels of the pathological choroidal vessels and thickened choroidal layer (pachychoroid), another characteristic of PCV but not of neovascular ARMD [13].

Another milestone in OCT technology is OCT angiography (OCTA) [14-20]. It is a non-invasive modality to visualize the pathology of retinal and choroidal vasculature. Also, OCTA measures the movement of red blood cells in the retinal and choroidal vessels over time and presents it as a B-scan of OCT [14-20]. OCTA is performed by RTvue XR Avanti (Optovue Inc., Fremont CA, USA) that has an A-scan.
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Conflicts of Interest

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