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Short Communication

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

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Abbreviations

ARMD: Age Related Macular Degeneration; BVN: Branching Vascular Network; CNV: Choroidal Neovascularisation; CSCR: Central Serous Chorioretinopathy; FA: Fluorescein Angiography; ICGA: Indocyanine Green Angiography; OCTA: Optical Coherence Tomography Angiography; PCV: Polypoidal Choroidal Vasculopathy; PDT: Photodynamic Therapy; PED: Pigment Epithelial Detachment; RPE: Retinal Pigment Epithelium; SRF: Subretinal Fluid; VA: Visual Acuity; VEGF: Vascular Endothelial Growth Factor

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 compliment cascade pathway. The compliment 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

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 to 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 junction [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

rate of 70,000 scans per second using light source entering at 840 nm and bandwidth of 540 nm; its and resolution is 5 mm axially with beam width of 15 mm. Each B-scan contains approximately 316 A-scans and usually 2 consecutive B-scans are performed in a fixed position at each location [12]. OCTA helps in distinguishing the flow within the vessels by utilizing several protocols such as split-spectrum amplitude-decorrelation (SSADA), phase variance and spectral variance. Unlike FFA and ICGA, OCTA guides in better axial localization and three-dimensional reconstruction of the perfused microvasculature within the retino-choroidal layers, provides volumetric and depth-resolved images of the retinal and choroidal vessels and capillary networks, and doesn't need an invasive dye and works by using red blood cells as the motion contrast. It allows accurate visualization of branching vascular networks of the PCV. Without using an intravenous dye, OCTA provides clear visualization of the neovascular complex either above or beneath the RPE. OCTA is being performed by many authors to describe the abnormal choroidal vasculature in PCV, changes in the choroidal blood flow in abnormal branching vascular network of PCV and to observe the progression of PCV during the follow-up after treatment [13-20]. It is particularly helpful in cases of PCV with large polyps, multiple extramacular polyps, and polyps without the BVN's. The patterns of polyps on OCTA reported recently by authors from Korea are halo type (high-flow density surrounding the inner dark regular circular cavity), rosette type (high-flow density surrounding the inner irregular dark cavity) and vascular network type [14]. The polyps on OCTA appear as hypoflow (low signal) round structures (few reported as hyperflow lesions with surrounding hyporeflective halo) and BVN as hyperflow vascular networks. The PCV polyps recorded on OCT can be confirmed by ICGA. Sometimes the polyps in PCV may appear smaller on OCTA than ICGA [7,14]. On OCTA, BVN are identified as extremely bright vascular structures distinguishable from the surrounding normal blood vessels with a clear boundary unlike on ICGA. Huang CH et al reported three different types of BVN based on the distinct features on the OCTA such as trunk, glomeruli and stick patterns [15]. Type 1 is the "trunk" pattern with one or more main trunk of neovascular vessels with radiating branches pointing toward the periphery of the vascular network. Type 2 is the "glomerular" pattern, without a major vascular trunk and has vascular network with multiple interconnecting anastomosis and resembles the glomeruli of the nephrons in the kidney. Type 3 is "stick" pattern with localised fine vascular network without identifiable feeder vessels [15]. The relative blood flow speed in the choriocapillaris in PCV lesions can be studied by another novel OCT-based algorithm, called a variable interscan time analysis (VISTA) [18]. Some polyps can have faster flow in the periphery and slower flow in the center which may indicate turbulence within the polyps that may be related to the self-obliteration of the lumen walls as a natural course of the disease. Studies also reported blood flow speed in the BVN and noted that there is relatively faster flow in larger trunk vessels and slower flow in the smaller vessels [18]. After treatment with anti-VEGF agents or photodynamic therapy, changes in the polyps were observed (decrease in size and number) on OCTA but not much of the morphological changes of BVN could be identified on OCTA unlike ICGA [14]. The disadvantage of OCTA is it cannot reveal the pathology in cases of large submacular hemorrhage, hemorrhagic PED's, and SRF whereas ICGA can demonstrate the choroidal pathology under the same circumstances. OCTA is also limited to detect some polyps due to severe motion artefact, high PED and small polyps which could be detected only by ICGA in suspected PCV cases [15].

Treatment modalities of PCV are anti-vascular endothelial growth factors (VEGF) such as bevacizumab, ranibizumab, aflibercept, photodynamic therapy (PDT) and thermal laser photocoagulation [5,18,21]. Initial stages of PCV are followed-up by observation if there are small peripheral polyps with no bleeding, good visual acuity and asymptomatic patients. Prognosis for vision is poor if not treated. Permanent deterioration of VA in PCV could be due to damage to the RPC, large polyps with recurrent bleeding and disciform scars through chronic progression.

Polypoidal choroidal vasculopathy continues to be clinically challenging condition as its clinical features and presentation mimics the neovascular ARMD. Multimodality imaging is helpful in the diagnosis of PCV. Optical coherence tomography angiography, a novel advanced technology in OCT is a non-invasive imaging tool to detect the vascular changes of PCV without using any intravenous dye. It provides better visualization of the branching vascular networks and various patterns of polypoidal lesions of PCV. OCTA guides in understanding the anatomy, pathophysiology of PCV and also in monitoring the treatment changes during follow-up visits.

Conflicts of Interest

The author has no conflict of financial interest in the machines or medicines used in the manuscript.

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