Guest Editorial

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Pachychoroid Spectrum - An Emerging Clinical Entity

The Pachychoroid clinical spectrum is a group of macular disorders that manifest similar choroidal findings. Optical coherence tomography (OCT) shows increased choroidal thickness (>300 µm) and dilated outer choroidal vessels are seen on OCT/Indocyanine green (ICG) angiography. These dilated vessels of the Haller's layer are referred to as 'pachyvessels', which compress Sattler layer and choriocapillaris causing attenuation at the inner aspect of the choroid. Central serous choroidopathy (CSC), pachychoroid pigment epitheliopathy (PPE), pachychoroid neovasculopathy (PNV) and polypoidal choroidal vasculopathy (PCV) are entities of pachychoroid clinical spectrum. This spectrum of disorders is more commonly seen in Asians.

Chronic CSC presents as non-resolving longstanding CSC with widespread retinal pigment epithelium (RPE) decompensation or focal leaks. Fundus autofluorescence (FAF) shows areas of RPE atrophy as gravitational tracks in these cases. Mineralocorticoid antagonists, laser photocoagulation, photodynamic therapy (PDT) and subthreshold micropulse laser have got a potential role in management of such cases.

Pachychoroid pigment epitheliopathy was first described in the fellow eyes of patients with CSC. It is considered as forme fruste CSC with pigmentary changes like RPE mottling and no subretinal fluid. It is often misdiagnosed as atypical age-related macular degeneration (AMD) or pattern dystrophy.

Pachychoroid neovasculopathy is considered as a precursor lesion of PCV where long standing CSC or PPE causes microbreaks in Bruch's membrane causing in growths of sub-RPE endothelial cells.

PCV is considered as a variant of neovascular AMD. PCV is a slow growing, complex form of neovascularisation that has a branching vascular network (BVN) with aneurysmal dilations at the outer border of the network. Hemorrhagic detachments are more common in PCV. Clinical course of PCV is more stable and visual outcomes are more favourable from those of AMD. ICG Angiography has gained an important role in diagnosis and monitoring of pachychoroid disorders and non-responding patients of AMD. It has become the gold standard to detect polyps in PCV. Video ICG angiography shows focal hypercyanescent lesions appearing before 6 minutes which are pulsatile and associated with hypocyanescent halo. BVN may also be present. PCV is characterised on OCT by thumb like pigment epithelial detachments (PED), "double layer sign" due to presence of branched vascular network (BVN), subretinal fluid and thickened choroid as seen on OCT. Enface imaging using OCT Angiography (OCTA) has an immense role in providing details about retinal and choroidal microcirculation like branching vascular network. Slow flowing lesions like polyps of PCV however are easily missed on OCTA. PLANET Study showed that intravitreal Aflibercept monotherapy improves visual acuity in PCV patients, without the need for PDT and became the current first-line treatment option for PCV. Direct thermal photocoagulation may be considered for extrafoveal symptomatic PCV lesions. For symptomatic juxtafoveal or subfoveal polyps, full fluence or reduced fluence PDT with or without intravitreal anti-vascular endothelial growth factor (anti-VEGF) injection should be considered as second line management. PDT alone has some limitations like subretinal hemorrhage and up regulation of VEGF which causes secondary neovascular membrane formation and PCV recurrence. Anti-VEGF agents used alone have low polyp regression rates and negligible effect on BVN. However, if used along with PDT these can probably suppress the pro angiogenic activity and result in lesser complications than PDT monotherapy alone. For PCV with subretinal hemorrhage larger than 4 disc area and presenting within 10-14 days of onset, pneumatic displacement with intravitreal gas and tissue plasminogen

activator can be performed. Vitrectomy with subretinal injection of tissue plasminogen activator may be considered as alternative in cases with massive subretinal hemorrhage or breakthrough vitreous hemorrhage. A long term follow-up is must in these disorders to look for complications or recurrences.

With the advent of multimodal imaging modalities, new insights have been obtained in the pathogenesis, clinical features and management of pachychoroid spectrum of disorders. One must be aware of this clinical entity for better diagnosis and management of these cases as they are distinct from AMD and other hereditary macular degenerations.

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