Idiopathic Polypoidal Choroidal Vasculopathy (IPCV)

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Background: Idiopathic polypoidal choroidal vasculopathy (IPCV) is a disease of choroidal vasculature resulting in idiopathic exudative and or haemorrhagic disorder of the macula. Its end point is subretinal fibrosis that is associated with severe ocular morbidity.[1-4] The aim of this article is to provide a basic and current information on IPCV.
Methods: The Internet was searched in English with Google Scholars, Hinari and Elsevier's Scopus for key words such as idiopathic polypoidal choroidal vasculopathy, update, latest discovery and new treatment options.
Results: Twelve recent and relevant peer reviewed articles were downloaded after an internet search.
Discussion: Typically bilateral, IPCV affects all races but commoner in heavily pigmented people with no gender bias. Cases thought to be wet age-related macular degeneration (AMD) have been reported to be IPCV.[4]

The etiology is not clearly understood. However, reviewed articles agreed that there is a disorder of inner choroidal vasculature in which there is a network of branching vessels deep to choriocapillaris in association with terminal aneurysmal dilatations. The assumption that IPCV is a subtype of choroidal neovascular membrane (CVNM) is not supported by its course and worse visual prognosis.[1,5,6]

Patients' complaints are decreased and/or distorted vision, central or paracentral scotoma from sub-foveal fluid accumulation.

On slit lamp bio-microscopy with 90/78D lens, lesions are orange-reddish bulb-like lesions budding from choroid into the subretinal space, with predilection for peripapillary area. Macula and even the periphery are not exempted. Subtotal or total haemorrhagic /exudative retinal detachment

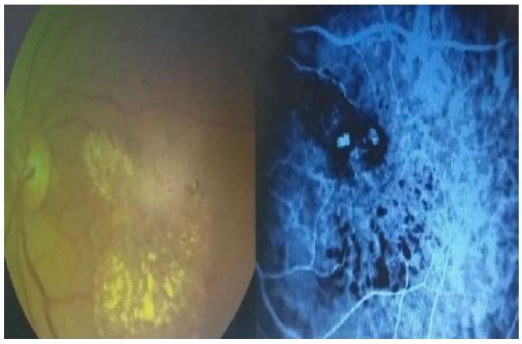

Figure 1: Fundus photographs, showing exudates and Indocyanide green angiography, showing polys and branching vascular network

and pigment epithelial detachment (PED), with or without breakthrough vitreous haemorrhage or hard exudates may also be seen (Figure 1).[3, 7,8]

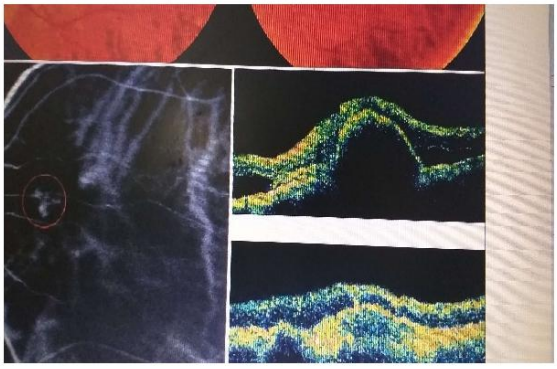

Figure 2: Optical coherence tomography showing tall peaked pigment epithelial detachment

Optical coherence tomography (OCT) shows tall peaked serous/haemorrhagic PED and sub-retinal fluid (SRF) (Figure 2). The polyps are seen as dome-like elevations of the retinal pigment epithelium with moderate internal reflectivity. A highly reflective line just below these lesions is consistent with location of the branching vascular network (BVN). The dual reflective layers are also called "double-layer sign," and are seen in 59% of eyes with IPCV.[2,10,11]

Indocyanide green angiography (ICGA) is the gold standard and shows the BVN better than fundus fluorescein angiography. Polyps are seen as focal hyperflourescent spots (Figure 1). Two types of polyps could be seen based on ICGA: type 1 (polypoidal CNV): polyp(s) with well-defined BVN (both feeder and draining vessels), and type 2

(typical PCV): polyp(s) with absent BVN (neither feeder nor draining vessels).[2,12]

Treatment is based on ablation of leaks seen on ICG with photodynamic therapy (PDT). Intravitreal injection of anti-vascular endothelial growth factors (anti-VEGF) have variable success. Combination of both treatments seem to yield greater therapeutic results.[1-5]

In conclusion, IPCV affects all races but tends to be commoner in heavily pigmented people. Visual outcome could be good especially with combined PDT and intravitreal anti-VEGF injections

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