Idiopathic Polypoidal Choroidal Vasculopathy in a Young Man

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Abstract

Purpose: To report a case of idiopathic polypoidal choroidal vasculopathy and its angiographic characteristics in a young man.

Method: Clinical data including visual acuity, color fundus photography, fluorescein angiography and indocyanine green results of this case were reviewed.

Case: A 21-year-old Saudi healthy man presented with sudden loss of vision in the left eye for 3 weeks duration. Visual acuity in the right eye was 20/20 and in the left eye was 1/200. Fundus examination of right eye was normal; however, in the left eye showed vitreous hemorrhage with massive sub retinal yellowish old blood in the posterior pole with epiretinal fresh blood on the fovea. Intra venous fluorescein angiography and indocyanine green tests showed features of idiopathic polypoidal choroidal vasculopathy. Patient received photodynamic therapy twice without significant visual improvement.

Conclusion: Idiopathic polypoidal choroidal vasculopathy can occur in a young age group and can lead to severe visual impairment.

Key words: Idiopathic polypoidal choroidal vasculopathy, young age, severe visual loss, photodynamic therapy

Discussion

Idiopathic polypoidal choroidal vasculopathy is a disease entity characterized by recurrent serous retinal leakage and hemorrhage in the elderly population, caused by vascular abnormalities in the inner choroid. The definition of IPCV has expanded over the past 10 years and the diagnosis is no longer restricted to specific demographic attributes or to specific retinal location. One large study done by Yannuzzi et al showed that IPCV may occur in all races but with a predilection for heavily pigmented races. Women tend to be affected more often than men; the average age of onset of IPCV is 60 years. However, one patient was noted to have clinical manifestations as early as the age of 20 years. Idiopathic polypoidal choroidal vasculopathy is more commonly bilateral than unilateral and has a preferratory location. However, Stiltsen et al found a predominance of unilateral involvement and extra macular location of IPCV in Italian patients. Uyama and Kwok et al described the preponderance of men; unilateral involvement and macular location of IPCV in Japanese and Chinese patients.

Indocyanine green test angiography is the best tool for diagnosis of IPCV. It usually shows a branching vascular network from the choroidal circulation and characteristic polypoidal, and aneurismal dilatation at the terminals of branching vessels. The branching vascular networks may last for a long period of time; however, the polypoidal dilations at terminals of the network may change configuration, new dilations grow, while other regress.

Uyama et al described two patterns of polypoidal dilations in ICGA 1) a large solitary round aneurysmal dilation in which usually the hemorrhage and sub macular fluid subsided leaving a macular fibroid scar after a follow-up period of 9 months from the treatment (fig 4). Final vision was 2/200.

Idiopathic polypoidal choroidal vasculopathy (IPCV) as cause of recurrent hemorrhagic and oxidative retinal pigment epithelium (RPE) and neurosensory retina detachment was first described by Yannuzzi.

In the past, a variety of terms such as “posterior uveal bleeding syndrome” and “multiple recurrent serous retinal pigment epithelial detachments in black woman” were used to designate this disorder. The primary abnormality involved the choroidal circulation and the characteristic lesions were in the inner choroidal vascular network of vessels ending in an aneurysmal bulge or outward projection visible clinically as a reddish orange spheroidal polyplike structure. It is associated with multiple, recurrent, seroussecesssive detachment of RPE and neurosensory retina secondary to leakage and bleeding from the peculiar choroidal vascular lesion. Vitreous hemorrhage, relatively minimal scarring and absence of druse, retinal vascular disease and signs of intracranial inflammation were also seen.

The diagnosis of IPCV is best made by using indocyanine green test (ICG) because it permits visualization of the choroids vasculature and will show the typical aneurismatic and spheroidal dilatation of the choroidal vessels.

The disease onset was typically seen among the elderly population. The reported average age of onset of IPCV was 60 years which is significantly younger in age than related macular degeneration. We are aware of one report by Yannuzzi et al in which the clinical manifestation was seen in a 28-year-old patient, and to the best of our knowledge this will be the second reported case. Previous reports of IPCV were also reviewed.

Idiopathic polypoidal choroidal vasculopathy can occur in a young age group and can lead to severe visual impairment.

Case Report

A 21-year-old Saudi healthy man suffered from sudden loss of vision in the left eye for duration of 3 weeks, he denied history of trauma. Best corrected visual acuity in the right eye was 20/20 and in the left eye was 1/200. Fundus examination in right eye was normal; however, in left eye showed a significant vitreous hemorrhage obscuring the view with massive sub retinal yellowish old blood in the posterior pole with epiretinal fresh blood on the fovea (fig 1). Intra venous fluorescein angiography (IVFA) revealed fluorescence blockage by blood and dye leakage at a late frame that resembled the appearance of occult sub retinal neovascularizations (fig 2). Optic coherant tomography test showed neurosensory retinal detachment. Indocyanine green test (fig 3) revealed a branching choroidal vascular network and collection of small polypoidal dilatation of the vessels. Based on that, the diagnosis of IPCV was made and the patient received photodynamic therapy (PDT) twice in which the hemorrhage and sub macular fluid subsided leaving a macular fibroid scar after a follow-up period of 9 months from the treatment (fig 4). Final vision was 2/200.

References