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Anatomical and Functional Outcome: A Case of Idiopathic Polypoidal Choroidal Vasculopathy

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Abstract

Idiopathic Polypoidal Choroidal vasculopathy (PCV) is characterized by subretinal vascular lesions leading to multiple, recurrent serosanguinous or hemorrhagic retinal pigment epithelial detachment. The classical PCV occurrence was initially reported in Asian descendent hypertensive middle aged women. Although anti-vascular endothelial growth factor (VEGF) is commonly used as first line treatment in Western countries, photodynamic therapy is the mainstay therapy for PCV in Asia. Here we describe a case of IPCV in a Malay lady who did not respond to initial anti-VEGF monotherapy and subsequently achieved stabilization of the disease with PDT. However, her functional vision was not able to be restored despite the anatomical improvement.

Keywords: Polypoidal choroidal vasculopathy; Ranibizumab; Photodynamic therapy

Case Report

Idiopathic Polypoidal choroidal vasculopathy (PCV) is characterized by subretinal vascular lesions leading to multiple serosanguinous or hemorrhagic retinal pigment epithelial detachments [1]. PCV which was initially known as 'Posterior uveal bleeding syndrome', recently been described as a variant of Type 1 Choroidal neovascularization. It is identified by branching vascular network and polypoidal aneurysmal vessels between the retinal pigment epithelium and Bruch's membrane [2]. Fifty percent of the patient inadvertently progress to have severe visual acuity loss secondary to recurrent episodes of exudation, hemorrhage, and fibrosis [3]. Clinical presentation of PCV varies among different ethnic populations. The classical hemorrhagic PCV occurrence was initially reported in Asian descendent hypertensive middle aged women. Caucasians tend to present with serosanguinous exudative detachment [4]. Here we describe a case of IPCV in a Malay lady who did not respond to initial anti-VEGF mono therapy and subsequently achieved stabilization of the disease with PDT. However, her functional vision was not able to be restored despite the anatomical improvement.

A 57-year-old Malay woman with no known medical illness, presented with right eye blurring of vision for 1 year, which was progressive and painless. She noted a grey spot in the central vision which was constant and affected her near vision. She did not complain of metamorphopsia. She had stable chronic floaters but no flashes of light, eye redness, or other ocular symptoms. She also denied any history of trauma to the eye.

There was no significant past ocular history. She has undergone caesarean surgery and a hernia repair surgery 20 years ago with no complications. She was allergic to Penicillin and seafood.

On examination, her Snellen visual acuities were 1/60 in the right and 6/6 in the left eye. Intraocular pressure was within the normal range in both eyes and anterior segment examination was unremarkable. Both lenses had nucleus sclerosis cataract. A subretinal hemorrhage, fibrin, hard exudate, and yellowish sub macular nodular lesion was recognized in the right macula. Optical Coherence Tomography showed distortion of retinal pigment epithelium with cystic intraretinal spaces and sub retinal fluid. Fluorescein angiography (FFA) showed leakage and hyper fluorescence at the lesion with the classic choroidal neovascularization (CNV) pattern (Figure 1). Indocyanine green angiography (ICG) showed two hypercyanescence spots in early frames with prominent choroidal vessels (Figure 2). She was diagnosed to have unilateral polypoidal choroidal vasculopathy from these findings.

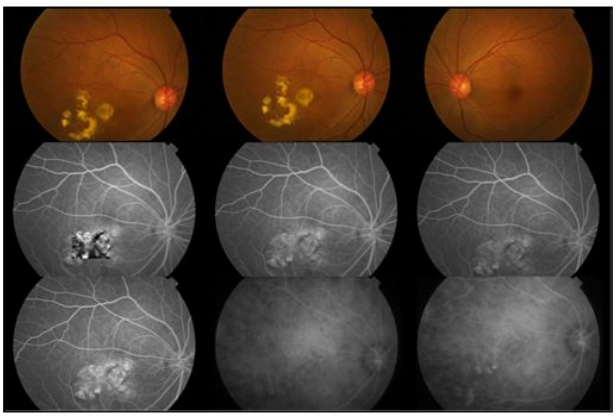

Figure 1: Colour fundus photo and serial FFA/ICG pictures showing exudative pattern PCV with extensive intraretinal lipid deposits adjacent to the macula. Late phase FFA shows leakage of undetermined origin (Occult neovascularization). ICG shows hypercyanescence spot in early frames.

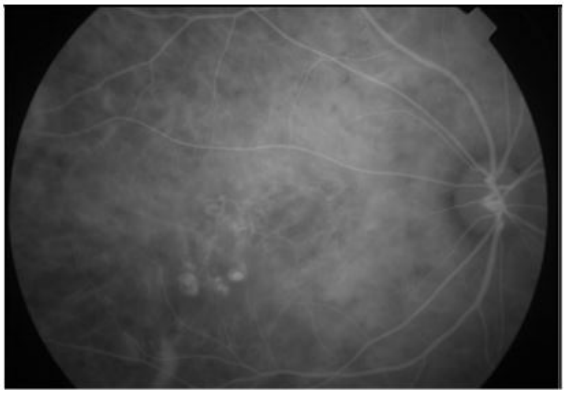

Figure 2: A closer look at ICG: Midphase indocyanine green angiography showing polypoidal lesions with branching vascular network.

Monthly Intravitreal Ranibizumab 0.5 mg was given for the next 3 months. There was no improvement observed in the visual acuity. However, the repeated OCT showed minimal resolution in the subretinal fluid and reduced central subfield thickness. The intraretinal cysts became more numerous and larger (Figure 3).

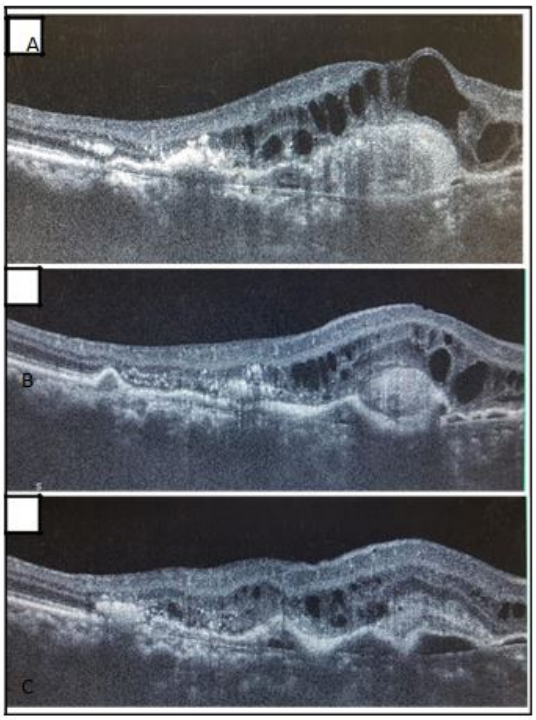

Figure 3: Serial OCT images showing structural improvement of the lesion post monthly Ranibizumab. (A) Initial OCT showing intraretinal cysts and exudates. Dome shaped elevation of RPE-lipid filled (B) cystic spaces and exdates improving (C) macula thickness improved with residual cystic spaces and RPE irregularities.

A repeat FFA and ICG was performed to ascertain the nature of the underlying lesion. FFA showed leakage of undetermined origin in the arteriovenous phase. The hypercyanescence spots are persistent with evident of polyps in ICG (Figures 4 and 5). The area of leakage was more than one-disc diameter (1DD).

Patient was scheduled for photodynamic therapy with verterporfin. Her body surface area (BSA) was 1.94 m^2 (weight: 100.7 kg, height 151.4 cm). The 5.7 ml verteporfin diluted with 24.3 ml dextrose 5% solution and was infused over 15 minutes. The largest diameter of the lesion was 5375 μm. Thus, PDT was performed with a 6400 μm spot size, 83 sec durations. There were no post procedural complications.

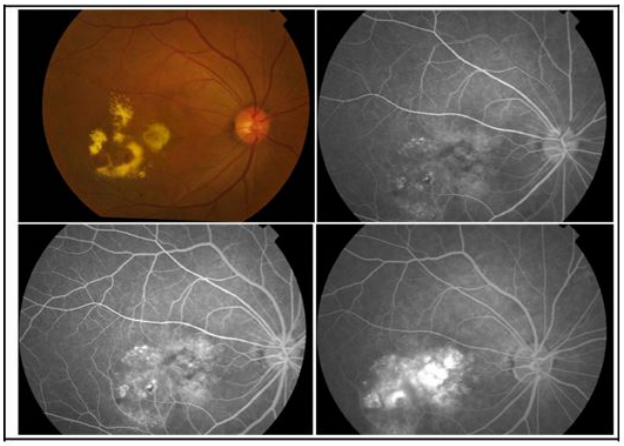

Figure 4: Colour fundus photo and serial FFA imaging prior to PDT: note the presence of polypoidal lesions and subretinal scarring at the macula (staining effect in late FFA).

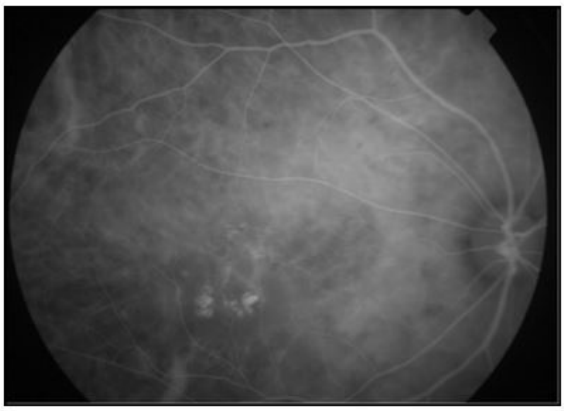

Figure 5: Mid-phase ICG showing multiple polypoidal lesions with branching vascular network.

Her vision improved to 6/60, (BCVA 6/36) over the next 4 weeks. Her OCT showed reduced central subfield thickness with subretinal fluid. There was subretinal fibrosis with distortion retinal pigment epithelium as well. Patient was not keen for anymore intervention. Patient was given gutt nevanac tds to improve the subretinal fluid.

At 6 month follow up, patient's vison remained at 6/60. On fundus examination, the sub retinal lesion and exudates appear to be stable with no evidence of new lesion. OCT showed intraretinal cystoid edema with subfoveal scarring.

Discussion

Idiopathic polypoidal choroidal vasculopathy (IPCV) is a variant of Type 1 choroidal neovascularisation, characterized by the presence of a network of polyps with associated feeder vessels. They are visualized as red-orange nodules in the fundus. The areas of RPE detachments are multifocal and often seen at the peripapillary and macular region [4].

Interestingly, there are well documented ethnic variations in the clinical presentation of PCV. Epidemiological studies have shown that the incidence of PCV is high in middle aged African American or Asian descendent women, relatively low in Caucasian population as compared to the age-related macular degeneration. Few studies from the Asian population have shown unilateral presentation in males with predominantly macular involvement [1].

Our patient, an elderly Malay lady falls within the typical age group for neovascular age related macular degeneration (ARMD). However, unilateral presentation and lack of drusens in both eyes alerted us regarding an alternative diagnosis. On fundus examination, PCV can present with subretinal fluid and subretinal hemorrhage or hemorrhagic pigment epithelial detachment (PED) which is similar to neovascular AMD. The classical orange-red nodules are often missed due to the surrounding exudation. In this patient, there was a yellowish submacular nodular elevation mimicking a pigment epithelial detachment (PED).

PCV patients may have no visual symptoms when there is no leakage from the polypoidal lesions. Acute visual loss is commonly due to spontaneous rupture of the polypoidal lesions, leading to submacular hemorrhage causing scotoma or breakthrough vitreous hemorrhage. Patients may also present with progressive visual loss with metamorphopsia due to accumulation of subretinal fluid and exudates around the polypoidal lesion. The natural course of PCV can vary among patients and it has been stipulated that in 50% of patients spontaneous regression of polyps can occur without treatment [5].

Spectral domain optical coherence tomography is sensitive and specific in distinguishing PCV from neovascular AMD. The presence of at least two out three signs in OCT (pigment epithelium detachment, double-layer sign, and thumb-like polyps) indicates a positive test with the sensitivity of 87.5% [6]. Indocyanine green angiography (ICGA) is useful in identifying branching choroidal vascular network and polyps projection from the internal choroid. These structures manifest as early vascular hypercyanescence in ICGA. It is difficult to identify the polyps on the fundus fluorescein angiography (FFA), as they are located beneath the RPE. However on ICGA, the polyps are clearly visible as single or multiple vascular aneurysms with a diameter of 100-500 mm. They will fill after a short delay and remain hyperfluorescent until the late phases. Recent guidelines for the clinical diagnosis and treatment of IPCV identified ICGA as the gold standard for the diagnosis. It is agreed that ICGA should be performed when routine ophthalmoscopic examination indicates a serosanguineous maculopathy with one of the following features: Red-orange subretinal nodules visible with the ophthalmoscope; Spontaneous massive subretinal hemorrhage; Notched or hemorrhagic PED; Occlusion of polyps with low response rate to anti-VEGF therapy [6].

PDT and anti-VEGF are the treatment options for PCV. PDT has been proven to stabilize the vision of PCV patients while anti-VEGF has been found to be effective in reducing the fluid from PCV lesions. The aim of PCV treatment is to achieve complete regression of polyps as the presence of polyps significantly increases the possibility of recurrent bleeding and collection of subretinal fluid [6]. Hence, PDT is desirable treatment in these patients although its potential side effects can include retinal pigment epithelial (RPE) rips, sub retinal haemorrhage, scarring or atrophy which can lead to long term visual impairment [7]. In the above case, the final visual outcome was not optimal due to the presence of subfoveal scarring.

In the EVEREST study, PDT alone or combined with ranibizumab was proven to be superior to ranibizumab monotherapy in achieving complete regression of polyp [8]. Anti VEGF injection can be useful in treating the eyes with complete regression of polyps in indocyanine green angiography, but with persistent leakage on fluorescein angiography.

Globally, the incidence of PCV is increasing as the awareness is raised about the diagnosis and treatment of this disease. It is less frequently identified in centers without ICGA and where there is lack of access to PDT. The treatment of PCV is complex and differs from that of neovascular AMD algorithms. Superior anatomic and visual outcomes can be achieved with PDT alone or in combination with anti-VEGF agents. Treatment plan need to be tailored individually according to the location of polyps, degree of leakage and the regression of the polyp.

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