Idiopathic Polypoidal Choroidal Vasculopathy

Outline

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Abstract
Polypoidal choroidal vasculopathy is a disorder characterized by recurrent sub-retinal hemorrhaging due to abnormalities in the choroidal vasculature. This case demonstrates the diagnosis and management a 47 year old male afflicted with the condition.

I. Case History
- Patient demographics
C.W., a 47 year-old African American male

- Chief complaint
Walk-in with complaints of sudden decrease in the central vision of his left eye over the past two days with gradual worsening since. His vision was stable in the right eye. He denied any ocular pain, flashes, or new floaters.

- Ocular, medical history
A review of his ocular history revealed previous subretinal hemorrhage in the left eye, treated with intravitreal Lucentis injection. There was no history of glaucoma or trauma. He had no history of ocular disease in his family. A detailed review of medical records revealed a previous diagnosis of essential hypertension, morbid obesity, hyperlipidemia and type II diabetes mellitus.

- Medications
Omepradole
Metoprolol
Tramadol
Lisinopril
Spironolactone
Diltiazem
Metformin
Simvastatin
Pravastatin
Eplenone

II. Pertinent findings
- Clinical
Entering Acuitities measured 20/20 in the right eye and 20/30 PHNI in the left. His spectacle prescription measured OD -2.50 +4.00 x 177 and OS -1.00 +0.75 x 137 with an add of +2.00. There was no improvement in acuity with that correction. Pupils were equal, round and reactive to direct and consensual illumination and there was no afferent papillary defect in either eye. Confrontational visual fields were full to finger counting in both eyes and extraocular motility was full in both eyes. Slit lamp examination revealed clear corneas, clean lids and lashes, white and quiet conjunctivas, flat irides and clear lenses OD, OS. Intraocular pressures by Goldmann applanation tonometry measured 15 mm Hg OU at 10:31 am using one drop 0.5% proparacaine hydrochloride ophthalmic solution. Upon dilated fundus examination (using one drop 1% tropicamide and 2.5% Phenylephrine OU), the cup to disc ratios were found to be 0.3 OD, OS. The neuroretinal rims were pink and had distinct margins in both eyes. The macula in the right eye was clear and flat, with two 0.5 mm areas of mottling of the retinal pigment epithelium (RPE) superior to it in the posterior pole. There were no hemorrhages, drusen or fibrosis. The left eye however, had severe RPE changes surrounding the fovea with active subretinal hemorrhage and pigment epithelial detachment (PED) 1 disc diameter superior temporal to the optic nerve head and another active subretinal hemorrhage in the temporal macula. Retinal vasculature was of normal caliber without emboli and the vitreous was clear and quiet OD, OS. There were no breaks in the retinal periphery, no angioid streaks, laquer cracks, staphylomas, punched out lesions, peripapillary atrophy or choroidal ruptures OD, OS.
Optical coherence tomography (OCT) Optical revealed a new pigment epithelial detachment (PED) with subretinal fluid (SRF) located in the posterior pole 1 disc diameter superior temporal to the optic nerve head in the left eye. Also present was a residual central PED with SRF from two years prior. There was an elevation of the RPE temporal to the fovea, a large empty space representing SRF and a zone of hyper reflectivity in Henle’s layer. A fluorescein angiogram (FA) was ordered when the patient first presented two years ago. The early phase demonstrated a large area of hypofluorescence extending throughout the entire macula caused by subretinal blood. The overlying retinal vessels were clearly visible. There were a few hyperfluorescent spots in the peripapillary area caused by retinal pigment epithelium defects. In the late phase, the center of the hemorrhage had a well-delineated hyperfluorescent nodule.

III. Differential diagnosis
Leading differential diagnoses for this case point to various causes of subretinal or juxtrapapillary subretinal neovascularization. These include but are not limited to Idiopathic choroidal neovascular membrane, Polypoidal Choroidal Vasculopathy (PCV), Exudative Age-related macular degeneration (ARMD) and Idiopathic Central Serous Chorioretinopathy (ICSC).

IV. Diagnosis and discussion
Two years prior, the patient presented with an orange-red spheroid, polyp like subretinal hemorrhage in the left eye which may have been consistent with polypoidal choroidal vasculopathy (PCV). FA, clinical presentation and OCT findings correlated well with this condition. On initial presentation to the author, the patient presented with a recurrent orange-red lesion very similar to the previous macular lesion. The recurrence of the bleeding is classic of PCV; this gave further support to the diagnosis.¹

Exudative ARMD was not likely given the patient’s young age, race and the absence of drusen. The average age of onset of PCV is typically younger than that of ARMD. The range of ages is also wider as it has been reported in individuals from 25 to 85 years of age.¹ The presence of hemorrhage rules out ICSC. This condition is reported to have serous retinal detachment but not subretinal blood. When PCV is predominantly serous, the disease may be easily confounded with CSRS but subretinal hemorrhage was evident in this case.¹ Fundus and angiographic findings differed from that of idiopathic CNVM, the latter showing a well defined area of hyperfluorescence in the early phase.¹ The presence of recurrence in another location in the posterior pole was an additional finding indicative of PCV. Indocyanine Green Angiography (ICGA) for choroidal vascular evaluation was not available.

-Elaborate on the condition

Polypoidal Choroidal Vasculopathy (PCV) was first described by Yannuzzi et al in 1982 at a macula society as a “peculiar hemorrhagic disorder of the macula caused by a primary abnormality of the choroidal circulation causing serosanguineous detachments of the retinal pigment epithelium and neurosensory retina”.² Since then, the condition has been named posterior uveal bleeding syndrome³, multiple recurrent serosanguineous retinal pigment epithelial detachment⁴ and Idiopathic Polypoidal Choroidal Vasculopathy⁵.

The clinical hallmark lesion is an inner choroidal vascular network ending in an aneurismal bulge or outward projection visible clinically as an orange-red, spheroid, polyp-like structure. These are believed to come from branches of the short posterior ciliary arteries through defects in Bruch’s membrane.¹ The protrusions in PCV have been suggested to cause long-term compression on structures overlying the RPE and Bruch’s membrane. This may also result in thinning and defects in the RPE and Bruch’s membrane.
Age-related changes such as calcium deposition may also contribute to Bruch’s membrane damage. This in turn may allow the proliferation of choroidal neovascularization, which has been often reported in PCV. Patients may present with subretinal and vascular lesions associated with chronic, multiple, recurrent serous and hemorrhagic detachments of the neurosensory retina or the retinal pigment epithelium. Presence of lipid exudate is not infrequent.6

The exact pathogenesis remains unknown7 but it has been recently suggested that the retinal changes in PCV might be the results of vascular endothelial growth factor (VEGF), which extend their angiogenic effects to the overlying neurosensory retina. The initial stages manifest as subretinal hemorrhage, retinal pigment epithelial atrophy and in later stages, subretinal fibrosis. It often has a unilateral presentation but it is considered a bilateral disease.2,6,7,8,9,10 It is thought to be a variant of choroidal neovascularization without any systemic involvement.1

Diagnosis is typically made with clinical examination and use of ancillary testing such as fluorescein angiography, ICGA angiography, and Optical coherence tomography (OCT). Polypoidal lesions appear as hyperfluorescent nodules on fluorescein angiography. This type of imaging can be used to identify retinal vascular problems and gross choroidal lesions.1 However, it cannot properly detect subtle changes in the choroid and therefore cannot be used to differentiate between PCV and exudative AMD caused by choroidal neovascularization.10 The reason for this is that major choroidal vessels are impermeable to fluorescein molecules. In addition, PEDs, exudation and hemorrhages mask the fluorescein and the choroidal plexus cannot reliably evaluated. The fluorescein dye travels so quickly through the choriocapillaris that it does not allow enough contrast to identify and locate choroidal lesions.1

ICGA however, clearly delineates the polypoidal lesions and aneurismal vascular abnormalities in the choroidal circulation better than fluorescein angiography.7 ICGA is highly protein-bound and fluoresces at longer wavelengths than fluorescein dye. This allows subtle choroidal alterations to be better visualized.1 These lesions are hyperfluorescent in the early phases with a surrounding hypofluorescent area behind the lesion.6 Indocyanine green angiogram is therefore the gold standard for the diagnosis of PCV.10

OCT is an imaging technique that is often being used to identify the choroidal lesions of PCV. The images reveal elevation of the RPE with low reflectance underneath it. This is indicative of a serous retinal detachment. Because of greater amount of protein in the serous fluid, it appears to be more reflective than subretinal fluid found in idiopathic serous chorioretinopathy.1

The retinopathy caused by PCV resembles exudative ARMD. However, this disease does not fall into any of the well-defined categories of ARMD; there is absence of drusen and it has a better prognosis. Whereas, exudative ARMD usually becomes progressively worse, those who get PCV usually retain a good visual acuity and the retinal manifestations may even spontaneously regress.2,6 This seems to occur more frequently than in all other forms of choroidal neovascularization.1

This disease was originally described as a recurrent subretinal hemorrhage in middle-aged to elderly African-American women.2 However, further research has demonstrated its prevalence in all races and both sexes.7,8 The incidence varies with different ethnic groups. It seems to be highest in those of African American descent, considerably high in Asians and lowest in Caucasians. This is exactly opposite to the incidence of ARMD, which is highest in Caucasians.9 In Asians; PCV is more common in males with more macular involvement. In African American patients, it occurs more commonly in female with
V. Treatment, management
-Treatment and response to treatment

Many different treatment regimens have been suggested for macula involved PCV, however there is still no standard of care. Among these treatment strategies, photodynamic therapy (PDT) and anti-VEGF injections have the highest safety and efficacy. Several studies have also demonstrated good results with the combination of these two therapies. PDT has been shown to maintain good visual outcome in most PCV patients. Reported side effects include large post treatment subretinal hemorrhage with consequent vitreous hemorrhage. PDT itself can also cause complications including RPE tearing, retinal atrophy and scarring. Lastly, there can be recurrence of the lesions, which can decrease visual acuity.  

Intravitreal injection of anti-VEGF such as Avastin or Lucentis is another treatment option as it has been shown to reduce the subretinal fluid from PCV lesions. The choroidal polyp lesions, however, remained unaffected by this treatment alone. The effect of this treatment is temporary due to the drug being evacuated from the system with time. Complications with intravitreal injections include endophthalmitis and retinal detachment.

The 2012 EVEREST study shows that PDT in conjunction with intravitreal anti-VEGF therapy or PDT alone was demonstrated to be statistically superior to anti-VEGF monotherapy in achieving complete regression of the polyps in patients with PCV. For cases of PCV without foveal hemorrhage or exudative changes, observation has been recommended.

Management:

When the initial work-up was completed two years ago, the case was referred to retinal specialist for treatment with Anti-VEGF therapy, Lucentis injection, to improve visual acuity and prevent further damage of the macula.

One month after initial injection, the patient was re-examined and the visual acuity remained at 20/50 but the patient reported improved functional acuity. The OCT at that time showed increased subfoveal pigment epithelial detachment (PED) of the left eye. Subretinal blood and fluid were present with a stable size. Another injection of Lucentis was carried out to further reduce the subretinal fluid.

At the one-month post injection follow-up, the acuity improved further to 20/25. On fundus examination the macula appeared to have a resolving dark subretinal hemorrhage surrounding a lighter area of dehemoglobinized blood and RPE changes. There were no drusen seen, further confirming the absence of ARMD. The OCT showed a PED in the left eye and subretinal hemorrhage with some SRF. The subretinal hemorrhage was therefore resolving since the previous exam. The patient had a great response to two treatments of Lucentis. A third injection was given that day to reduce the residual SRF.

Six weeks later the acuity was 20/20. OCT of the left eye showed PEDs without SRF. An additional injection was not required at this time and monitoring the patient was recommended. If recurrent subretinal hemorrhage was to occur, ICGA was to be considered to further explore PCV.
Two months later the vision remained 20/20. Photos of the left eye showed a flat macula with a superior scar and a mostly resolved subretinal hemorrhage. The OCT continued to show a PED without SRF and a resolving hemorrhage with only a trace hemorrhage remaining. The patient continued to be monitored at 4-6 week intervals without intervention.

At the initial presentation to the author, the retina specialist was consulted on the same day and an injection of Lucentis was given. 1 month later, the vision improved to 20/20 and there was no SRF. Another injection was given for maintenance. The patient continues to be observed.

VI. Conclusion
-Clinical pearls, take away points if indicated

In conclusion, PCV seems to be a distinct condition that should be distinguished from other types of choroidal neovascularization. This disease does not only differ clinically but also demographically. It is important to correctly identify this clinical entity as treatment considerations, as well as response, natural courses and risk factors differ from ARMD. Indocyanine green videoangiography has been reported to be the most useful ancillary test for imaging PCV choroidal lesions. The condition has a good prognosis; most cases result in minimal or temporary vision loss. PDT alone or in conjunction with anti-VEGF therapy is the preferred treatment for this condition. Although subretinal fibrosis and RPE atrophy can cause permanent vision loss, the visual prognosis of PCV has been reported to be superior to that of exudative ARMD.


