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Multifocal Polypoidal Choroidal Vasculopathy in Caribbean and African-Americans

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Editorial

Polypoidal Choroidal Vasculopathy (PCV) is a relatively uncommon form of exudative Age-Related Macular Degeneration (ARMD) first described in 1990 by Yannuzzi et al. [1]. It presents as multiple, branching networks of small, polyp-like clusters, or larger identifiable characteristic polyp-like out-pouchings. Manifestations include lipid exudation, serosanguinous pigment epithelial detachments and sub-retinal hemorrhage. Indocyanine-Green (ICG) angiography has been said to be the necessary for delineating the polypoidal vascular abnormalities, but recently hi-resolution spectral domain Ocular Coherence Tomography (OCT) has been demonstrated to perhaps be diagnostic as well [2]. Recently it has been associated with a pachychoroid [3].

The vast majority of cases noted and reported on have involved Asians (in whom it is a very common form of exudative AMD) and Caucasians [4-8]. We have examined, imaged and treated a different ethnic and underserved population of Caribbean and African-Americans who display a different form of PCV. It is characterized by its multifocal nature, prominent hypertrophic pigmentary scarring, abundant circinate lipid exuation, and large polyps. Just two such cases have been reported in the literature in 2016 [9], Both of which were males. Furthermore, a thickened, pachychoroid is usually associated with PCV. We reported a unique case of PCV ARMD in the setting of drusen, and bilateral, chronic hypertrophically pigmented retino-choroidal folds [10].

PCV genetic studies have been conducted in populations of Asians and have found some putative linkage [11]. Since our population is of a different ethnic background, and phenotype is so different from that seen in Asians and Caucasians, I speculate that the incident genotype may be diverse. It is unfortunate that this ethnic group has been underrepresented in prior observational studies and clinical trials, perhaps because of it rarity.

We are actively collating our cases (several hundred) that we have seen in Brooklyn over the course of the last 15 years in hope of highlighting the unique variant of PCV seen in male and female Caribbean and African-Americans. They are distinctly different that cases seen in Asians, which have been reported in abundance. The polyps are often quite large and almost always multifocal over the course of time. The late appearance is reminiscent of a white-dot syndrome, Serpiginous Retinochoroidopathy. Some cases look like a scarified version of Serpiginous, sometimes known pejoratively as "Black Serpiginous" by some, due to the great abundance of hypertrophic-black scarring of the retina. I do not think our PCV cases represent "Black" Serpiginous retinochoidopathy, as there is no prior or coincident ocular inflammation. Our cases respond well to anti-VEGF intravitreal injection, owing to their pathological vasculogenic nature. It may come to pass that our observations will lead to a better understanding of ethno-racial differences that exist in PCV. We may ascertain new genetic markers. Further, its distinct patterns may lead to a change in classification of PCV, and addition of a category that may come to be known as Multifocal PCV. There are some late-cases that are so striking that they may be aptly considered as "Black"Polypoidal Choroidal Vasculopathy.

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