Polypoidal Choroidal Vasculopathy
Category: Posterior Segment
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Abstract:
A 65-year-old African American male presents with mild ARMD. Examination reveals RPE changes and subretinal fluid near the optic nerve OD. IVFA demonstrates three foci of leakage near macula. The diagnosis is likely PCV.

Case History:
- Demographics:
  - 65-year-old African American male
- Chief Complaint:
  - Presents for comprehensive yearly eye exam
  - Slight distance and near blur with current spectacles
- Ocular History:
  - Mild dry age-related macular degeneration (AMD) in both eyes
    - Diagnosed one year ago at his last eye exam
- Medical History:
  - Diabetes Type II
  - Hypertension
  - Atrial fibrillation
  - Hypercholesterolemia
- No Ocular Medications

Pertinent Findings:
- Best corrected visual acuity:
  - 20/20-2 OD
  - 20/20 OS
- Dilated fundus examination:
  - Macula OD: cluster of RPE mottling and RPE drop out superior-nasal and inferior-temporal perimacularly
  - Macula OS: cluster of RPE mottling superior-temporal perimacularly
  - All other posterior pole findings were unremarkable
- Macular Optical Coherence Tomography (OCT):
  - OD: normal contour with subretinal fluid near the optic nerve head
  - OS: normal contour with no fluid
- Fundus Photos:
  - OD/OS: scattered areas of decreased lipofuscin on fundus autofluorescence
- Intravenous Fluorescein Angiography:
  - OD: three foci of leakage superior-nasal to macula and staining inferior-temporal to the macula
  - OS: leakage superior-temporal to the macula
**Differential Diagnosis:**
- Polypoidal Choroidal Vasculopathy (PCV)
- Wet Age Related Macular Degeneration (Wet ARMD)
- Central Serous (CSR)

**Discussion:**
PCV is more common in Asian and African American populations and is currently recognized as a phenotype of ARMD. The usual age of onset of PCV ranges between 50 and 65 years of age. However, the age of diagnosis ranges between the 20s and the 80s.

PCV can be clinically classified as quiescent, exudative, or hemorrhagic. In quiescent PCV, choroidal polyps exist without fluid or hemorrhage. Exudative PCV requires presence of subretinal or intraretinal fluid, but no hemorrhage. Lastly, hemorrhagic PCV includes all PCV with subretinal or sub-RPE hemorrhage.

The natural course of the disease often follows a relapsing-remitting course. Clinically, it is associated with chronic, multiple, recurrent serous or hemorrhagic detachments of the RPE and retina with long-term preservation of good vision.

The most common presenting symptom is decreased visual acuity due to subretinal fluid or subretinal hemorrhage, leading to common misdiagnosis and under-diagnosis due to its mimicry of wet AMD and CSR.

On fundus examination, the most suggestive findings of PCV are orange-red subretinal nodules, spontaneous subretinal hemorrhage, and serous or hemorrhagic pigment epithelial detachments (PED). However, relying solely on fundus appearance will not allow for a definitive diagnosis. It is important to utilize various modalities of testing including indocyanine green angiography (ICG), IVFA, OCT and fundus photography to determine the correct diagnosis. Fundus photography often shows only non-specific PED and leakage. However, utilizing the ICG will better highlight the dilated choroidal vascular changes that are pathognomonic for PCV, which appear as nodular polypoidal lesions. IVFA will show these choroidal vasculature abnormalities as non-specific hyperfluorescent spots. In some cases, OCT will show a double layer sign, with hyperreflectivity of the choroidal layers. The presence of one or more of these features suggests a diagnosis of PCV. If these features are not distinguishable initially, consider an ICG in a patient who is non-responsive to anti-VEGF therapy. An ICG is the preferred modality of imaging for PCV.
**Treatment and Management:**
An accurate diagnosis of PCV is essential for recommending appropriate treatment and management options. If a patient has retinal changes that do not threaten the macula or central vision, a conservative approach can be taken. Polypoidal lesions may undergo spontaneous regression in approximately 50% of patients with PCV have good visual prognosis with observation alone. Observation and regular follow-up is the treatment method we chose for our patient.

In cases in which persistent or progressive exudative changes threaten central vision, other treatment options are available. Although the initial acuity due to subretinal fluid is usually better in patients with PCV compared to patients with ARMD, PCV often more resistant to anti-VEGF therapy. Anti-VEGF is effective in stabilizing visual acuity and reducing exudation but has limited efficacy in causing regression of the polypoidal vascular abnormalities\(^2\). However, anti-VEGF treatment is still the primary method of treatment for PCV patients today.

Photodynamic therapy with verteporfin is an alternative to anti-VEGF injections and has shown promise in recent studies in the treatment of PCV. In a study with 22 eyes treated with ICGA-guided PDT using verteporfin, stable or improved vision was observed in 95% of eyes, absence of leakage in FA in 91%, and total regression of polyps in 95% with an average of 1.6 treatments.\(^{1,2}\)

**Conclusion:**
Polypoidal choroidal vasculopathy should not be overlooked as a potential diagnosis in a patient presenting with serous, exudative, and/or hemorrhagic retinopathy, with the absence of drusen. PCV has been more readily diagnosed due to the increased understanding of its presentation and clinical characteristics. PCV has unique features that differentiate it from other forms of choroidal vascular abnormalities. When assessing patients that are suspect for PCV, various clinical tests are important to determine a definitive diagnosis. Clinical presentation, IVFA, ICG, and OCT should be utilized in combination to determine a diagnosis of PCV.
References:

