An atypical benign case of subretinal neovascularization: Peripapillary subretinal neovascularization

A 66 year old asymptomatic male presents with a recurrent subretinal hemorrhage adjacent to the left eye optic nerve, signifying the presence of subretinal or choroidal neovascularization in an otherwise unremarkable posterior segment exam.

- Case History: 66 year old male without visual complaints presenting for an updated glasses prescription
  - Ocular History
    - Pseudophakia OU
    - Low risk glaucoma suspect due to moderate cupping
    - Subretinal hemorrhage secondary to peripapillary subretinal neovascularization membrane (PSRNVM) nasal to optic nerve OS 7/2008, OD 10/2009
  - Med History
    - Obstructive sleep apnea
    - Hypertension and Hyperlipidemia
    - Chronic Obstructive Pulmonary Disease
    - Obesity
  - Medications
    - Hydrochlorothiazide/Lisinopril
    - Albuterol
    - Budesonide
  - Findings
    - Cataract surgery OU
    - Pseudophakia with moderate myopic refractive error
    - Best corrected visual acuity: 20/15 OD/OS
    - Unremarkable anterior segment exam
    - Intraocular pressure (IOP): 15/15
    - Ocular fundus exam
      - Posterior pole, macula, peripheral exam unremarkable
      - Low risk glaucoma suspect based on optic nerve cupping: previous glaucoma workup unremarkable for ocular coherence tomography (OCT) measured retinal nerve fiber layer thinning or visual field defects
      - OD: Recurrent subretinal peripapillary hemorrhage associated with subretinal neovascular membrane
        - Photos taken 2009 and 2014
        - Line scan OCT: no subretinal edema, no exudates
  - DDx
    - Primary: Idiopathic PSRNVM
    - Secondary: Age-Related Macular Degeneration associated PSRNVM
    - Others including Polypoidal Choroidal Vasculopathy (PCV)
• Diagnosis and discussion
  o Diagnosis: Bilateral recurrent idiopathic PSRNVM, not vision threatening due to nasal location and small size
  o Pathophysiology
    ▪ 57% of PSRNVM develop through breaks in Bruch’s membrane near the optic disc
    ▪ 43% wrap around the termination of Bruch’s adjacent to the disc
  o 14% of elderly eyes have non vision-threatening PSRNVMs
    ▪ Usually small, nasal in location, and self-involuting
    ▪ Mostly asymptomatic, but can cause enlarged blind spots or paracentral/cecocentral scotomas
  o Vision-threatening or symptomatic PSRNVMs typically temporal to disc
    ▪ 45.5% PSRNVMs are associated with ARMD
      • Can be visually symptomatic even if PSRNVM is not directly extending under macula or fovea
      • No specific features of the optic disc or peripapillary region have been identified that increase the risk of development of PSRNVM
    ▪ A variant of PSRNVM: Polypoidal Choroidal Vasculopathy
      • Classic predilection for patients of pigmented races, however, PCV is known to occur around optic disc in white patients
      • Thought to be a form of neovascular ARMD, but interestingly not dependent on VEGF related pathways
      • 25-47% of membranes presumed due to ARMD may have a PCV etiology instead

• Treatment options
  o Monitor closely
    ▪ Treatment utilized in the reported case
    ▪ Reported case is bilateral, recurrent, idiopathic, and non vision-threatening similar to those described by Sarks 1973
  o Laser photocoagulation and Photodynamic Therapy (PDT)
    ▪ Possible damage can occur to optic nerve or the papillomacular bundle with conventional laser
    ▪ Unclear whether laser or PDT is effective at stabilizing/improving visual acuity or limiting recurrence
    ▪ Laser treatment guidelines by Macular Photocoagulation Study limit laser to small membranes without adjacent submacular hemorrhaging and limited temporal peripapillary retinal involvement
    ▪ PDT viable alternative for patients not qualifying for laser
  o Anti-VEGF agents
    ▪ Landmark studies of anti-VEGF agent efficacy are focused on macular membranes
    ▪ Studies demonstrate variation in improvement in BCVA in peripapillary membranes after anti-VEGF injections
    ▪ Current studies directed specifically at PSRNVM and anti-VEGF agents are limited by small populations and short follow up
• Conclusion
  o In an otherwise unremarkable clinical exam, it is difficult to detect presence of PSRNVM unless subretinal hemorrhages or edema develop
  o Clinicians should recognize the risks associated with PSRNVM, especially in patients with other retinal conditions such as Age-Related Macular Degeneration or Polypoidal Choroidal Vasculopathy
  o Suggested ancillary testing
    ▪ Fundus photos
    ▪ Optical Coherence Tomography: clinically accessible means of identifying subretinal fluid
    ▪ Fluorescein Angiography if indicated
      • Categorize neovascularization as occult or classic and visualize extent of membrane
      • Some evidence that Indocyanine Green Choroidal Angiography is better at detecting the edges of CNV