A Rare Case of Bilateral Peripheral Exudative Hemorrhagic Chorioretinopathy (PEHCR)
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An 83 year old male with bilateral idiopathic peripheral subretinal neovascularization experiences massive vitreous hemorrhaging and subsequent loss of vision while on therapeutic levels of Coumadin. Receiving off-label Avastin injections in seeing eye.

- Case History
  - Patient Demographics
    - 83 year old white male living near Boston, MA and spending his summer in FL
  - Chief Complaint
    - Sudden reduction in vision OD after diagnosis of peripheral choroidal neovascular membranes OU.
  - Ocular History
    - Bilateral peripheral choroidal neovascular membranes OD>>OS with associated pigment epithelial detachments OD
      - Diagnosed in April 15, 2009 at Bay Pines VA in Florida
      - Fundus photos and OCT images showed choroidal neovascular membranes, subretinal hemorrhaging and PEDs OD; chorioretinal scarring with sub- and intraretinal hemorrhages present OS
      - Subsequent photos and IVFA revealed subretinal fluid extending ~1DD inferior to fovea. Early and late hypofluorescence secondary to subretinal hemorrhage OD.
    - OD eventually suffered breakthrough vitreous hemorrhage leading to light perception vision
    - Monocular, OS seeing. S/p off-label Avastin injections x 3 OS only to prevent potential hemorrhaging and to preserve vision OS.
    - Extensive chorioretinal degeneration OU
      - Consistent with prior episodes of peripheral choroidal neovascularization, though none had been noted since 2003 when he first entered the VA system.
    - Peripheral hemorrhages OU first noted in 2004 with gradual increase in the following years
    - Primary open angle glaucoma OS with inferior notching and corresponding visual field loss
    - Multiple areas of bullous and flat retinoschisis superior/temporal and temporal OU
    - Atrophic retinal hole inferior OD well-sealed with pigment
    - Epiretinal membrane OD
    - Mild cataracts OU-not visually significant
    - Asteroid hyalosis inferior OD
    - Hyperopia, astigmatism, and presbyopia OU
Medical History
- Atrial Fibrillation
- Normyocytic anemia of chronic disease
- Hypothyroidism
- Hypertension
- Diverticulosis
- Chronic airway obstruction secondary to pulmonary coin lesion
- Malignant neoplasm of laryngeal cartilages
- Actinic keratosis
- Xerosis
- Constipation

Medications
- Alphagan 0.2% TID OS
- Trusopt 2% TID OS
- Celecoxib 200mg capsule po BID
- Diltiazem 240mg capsule po QD
- Formoterol fumerate 12mcg capsule by inhalation QD
- Furosemide 20mg tablet po QD
- Levothyroxine NA 0.05mg tablet po QD
- Metoprolol tartrate 1/2 25mg tablet po TID
- Mometasone 220mcg/inhaler, 1 puff QHS
- Omeprazole 20mg po QD
- Tiotropium 18mcg capsule by inhalation QD
- Warfarin NA 1/2 5mg tablet QHS; 1 tablet Sunday and Wednesday

Other Salient Findings
- International Normalized Ratio (INR) profile: Goal 2-3
  - Between 0.9 to 5.7
  - INR at time of choroidal neovascular membrane diagnosis 1.7
  - INR at time of large sub-and intra-retinal and vitreous hemorrhages 3

Pertinent Findings
- Clinical
  - OD
    - Light perception in inferior quadrants only
    - Positive 3+ APD
    - Exo position with Hirschberg
    - Mild cataract-not attributing to decrease in vision
    - 3+ red blood cells in vitreous restricting view to retina
    - B Scan: PVD with hemorrhagic retinal detachment vs dense vitreous hemorrhage temporally and extending to posterior pole and inferior/nasally
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- Choroidal melanoma ruled-out previously with non-hyperfluorescent IVFA and with subsequent B scans which revealed a medium to high intensity mass not consistent with choroidal tumor.

  - **OS**
    - BCVA 20/20
    - Kinetic visual field constricted to ~35 degrees nasally secondary to temporal chorioretinal scarring
    - Mild cataract-not visually significant
    - Large band of chorioretinal scarring with associated RPE changes extending from 8DD temporal to optic nerve to equator between 12-5:30 o'clock
    - Patches of intraretinal hemorrhaging noted at 2 and 5 o'clock
    - Superficial hemorrhaging inferior to macula along inferior arcade

  - **Physical**
    - Atrial fibrillation requiring Coumadin
    - Hypertension controlled with medication

  - **Laboratory Studies**
    - INR at 08/12/10 exam 1.9

- **Differential Diagnosis**
  - **Primary**
    - Bilateral idiopathic peripheral choroidal neovascularization with secondary peripheral exudative hemorrhagic chorioretinopathy

  - **Others**
    - Choroidal melanoma
      - Ruled out with IVFA; medium to high intensity mass on B-scan rather than smooth dome with hollow center and choroidal excavation consistent with choroidal melanoma
    - Hemorrhagic retinal detachment
      - Retina intact on B-scan
    - Macroaneurysm
      - Ruled out with IVFA
    - Polypoidal choroidal neovascularization
      - No polypoidal lesions noted
    - Breaks in Bruch's membrane with subsequent choroidal neovascular membrane formation-negative patient history
      - Pathologic myopia
      - Collagen disease
      - Trauma
      - Ocular histoplasmosis
      - Multifocal choroiditis, etc.
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- Retinal neovascularization-negative patient history
  - Diabetes
  - Sickle cell anemia
  - Ocular ischemic syndrome
  - Retinal vein occlusions
  - Retinopathy of prematurity, etc.
- Hemorrhagic posterior vitreous detachment
- Chorioretinal scarring secondary to trauma-negative history of trauma
- Choroidal osteoma-ruled out on B-scan

- Diagnosis and Discussion
  - Elaborate on the condition
    - Peripheral subretinal neovascularization with secondary peripheral exudative hemorrhagic chorioretinopathy (PEHCR) is most often found in the temporal quadrants and occupy several clock hours. The lesions are often found between the equator and ora serrata.
    - Lesions are typically characterized by hemorrhage, lipid exudation, and subretinal fluid.
    - This condition is often misdiagnosed as choroidal melanoma.
    - Often, subretinal membranes involute spontaneously and hemorrhages may resorb leaving residual RPE atrophy and chorioretinal disciform scarring in the periphery. Residual subretinal hemorrhages and/or fibrotic subretinal membranes may also occur.
    - Extramacular subretinal membranes infrequently affect central vision but have been implicated as a common cause of hemorrhagic retinal detachments; blood can dissect the retina to cause vitreous hemorrhaging which can be detrimental to vision.
    - Vicrectomy is indicated if no spontaneous resorption occurs after 6 months, the hemorrhage occurs in the only good eye, a need to binocular vision is present, or resultant hemolytic glaucoma develops.
    - Peripheral neovascularization differs from macular neovascularization because of the physiological differences in choroidal, Bruch's membrane, and the RPE in the posterior pole and the peripheral retina.
    - Systemic hypertension and arteriosclerotic heart disease were associated with PEHCR. Anticoagulation therapy increases the risk of intraocular hemorrhage in these eyes despite therapeutic levels.
    - Idiopathic peripheral choroidal neovascular membrane occur most often in the presence of breaks in Bruch's membrane and are a fairly common subclinical finding in extramacular regions in senile eyes. The patient's history was negative for typical causes for breaks in Bruch's membrane leading to the diagnosis of idiopathic peripheral neovascularization.
  - Expound upon unique features
    - PEHCR occurs most often in elderly women between the ages of 60 and 90 years.
The bilaterality of this patient's condition is a fairly unique feature as it only occurs bilaterally in approximately 25-31% of patients.

- Patient was being treated with therapeutic doses of Coumadin.
- Subretinal fluid and hemorrhaging extended to the posterior pole and began to affect the macula. Breakthrough retinal hemorrhage obscured vision completely rather than the typical course of involution and spontaneous resorption.
- This patient deferred vitrectomy with the hope that the hemorrhage would resorb. The vitreous hemorrhage occurred between 08/28/09 and 09/01/09 and has not cleared. His visual prognosis is poor as the vitreous hemorrhage is very dense.
- Because the patient is left with one seeing eye, off-label Avastin injections are being administered to protect remaining vision in that eye.

- **Treatment, Management**
  - **Treatment and response to treatment**
    - After diagnosis in April 2009, pt was observed closely at both the Bay Pines and the West Roxbury VAs. Once subretinal hemorrhaging and fluid approached 1 disc diameter from the fovea, a referral to Jamaica Plain ophthalmology was made for further evaluation and IVFA.
    - Close monitoring was prescribed and a consult placed with hematology to switch the patient from Coumadin to levonox on August 28, 2009. The patient was maintained on Coumadin as his INR levels were therapeutic and not exceeding his goal INR at that time.
    - By 09/01/09, patient had a vitreous hemorrhage and finger counting vision. The patient obtained a second opinion by a retinal specialist outside the VA system. The retinal specialist educated the patient that the hemorrhaging could potentially resorb or vitrectomy could be performed to remove the blood. The patient chose to defer vitrectomy with the hopes that the blood would resorb.
    - There is a very dense vitreous hemorrhage remaining in the right eye with no apparent resorption having occurred. Both the patient and the retinal specialist, by whom he is now followed, do not think that vitrectomy at this time would benefit the patient.
    - The fellow eye is undergoing Avastin injections because it is his only seeing eye. To date, he has had 3 injections in the left eye with good success. His best corrected vision remains 20/20.
  - **Refer to research where appropriate**
    - Typical course for extramacular subretinal neovascularization is involution. If vitreous hemorrhaging does occur and the blood has not resorbed after 6 months, vitrectomy is recommended. If the affected eye is the patient's only seeing eye or if binocularity is required by the patient, earlier intervention is recommended (Orth, Flood).
Patients undergoing vitreoretinal surgery while on Coumadin have had no intraoperative complications reported. Postoperative hemorrhagic complications have resolved spontaneously (Dayani, Grand).

Subretinal hemorrhagic complications have been reported in patients receiving long-term anticoagulation treatment with concomitant age related macular especially in conjunction with systemic hypertension (Hattenbach, et al).

Bibliography, literature review encouraged

- Carter, J. Anticoagulant, antiplatelet, and fibrinolytic (thrombocytic) therapy in patients at high risk for ocular hemorrhage.
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**Conclusion**
- Clinical pearls, take away points
  - PEHCR generally spontaneously involutes but can rarely be associated with subretinal, and occasionally vitreous, hemorrhaging which can disrupt vision.
  - Treatment options should include:
    - Close observation.
    - Vitrectomy is encouraged after 6 months observation with no resorption, binocularity is required, or if the hemorrhaging occurred in the only seeing eye.
    - Alternative treatment, such as anti-VEGF, should be considered.
  - Very careful observation of patients who are at risk for subretinal or vitreous hemorrhaging should be implemented especially when they are on concomitant anticoagulation or antiplatelet therapy. Systemic hypertension can cause further complications.