Angioid Streaks in a Patient with Sickle Cell Disease

In a patient with sickle cell disease without proliferative retinopathy, the presence of angioid streaks spoking from the optic disc margin is detected during a routine dilated fundus examination.

I. Case History
- Demographics: 60-year-old African American male
- Chief Complaint: Patient lost glasses on the bus a few weeks ago and wants an updated prescription. No visual complaints.
- Ocular History: Last eye examination 6 months ago, ocular hypertension OU, mild cataracts OU, refractive error c presbyopia OU, herpes zoster virus x 2011
- Medical History: Sickle cell disease (HbSS), sickle cell anemia, depression, drug and alcohol dependence, hypertension
- Medications: Acetaminophen 750mg, Tramadol 50mg, Folic Acid 1mg, Thiamin 100mg, Diphenhydramine 25mg, Mirtazapine 45mg, Venlafaxine 225mg, Omeprazole 20mg, Hydroxyurea 1000mg, Lorazepam 2mg

II. Pertinent Findings
- Clinical:
  - BCVA OD: 20/20, OS: 20/20-2
  - EOMs: full/no restrictions OD/OS
  - Pupils: 3mm OD/3 mm OS ERRL, (-) APD OD/OS
  - Confrontations: FTFC OD/OS
  - IOP: OD- 19, OS- 18
  - Slit Lamp Examination: Mild EBMD OD/OS
  - Dilated Fundus Examination:
    - C/D = 0.20; healthy rim tissue; (-) edema/pallor OD/OS
    - Posterior pole OD: 1 nasal angioid streak spoking from the ONH
    - Posterior pole OS: 1 superior nasal angioid streak spoking from the ONH
    - Macula: flat and intact; (-) CNVM OD/OS
    - Periphery: flat and intact; (-) holes/tears/RDs/hemes/CNVM/sea fan retinopathy OD/OS
- Ancillary Testing
  - Spectralis ONH OCT: (+) RPE disruption, thickened choroid nasal to ONH; OS: (+) RPE disruption, thickened choroid superior nasal to the ONH
  - Spectralis macula OCT: normal foveal contour, (-) edema/CNVM OD/OS
  - Spectralis autofluorescence: hypoauflorescence of the angioid streaks OD/OS
  - Fundus photos: DFE serves as interpretation OD/OS
  - Cirrus RNFL OCT: WNL all quadrants OD/OS
  - Blood work: The patient has abnormally high levels of fetal hemoglobin, likely providing protecting against more severe sickle cell crises and allowing him to outlive the average life span of a sickle cell patient.

III. Differential Diagnosis
- (PEPSI): Pseudoxanthoma Elasticum, Ehlers-Danlos Syndrome, Paget’s Disease, Sickle Cell Disease, Idiopathic
- If not angioid streaks, what else could they be?
  - Lacquer cracks, myopic degeneration, choroidal folds, choroidal rupture
IV. Diagnosis and Discussion

- The patient, positive for sickle cell anemia and sick cell disease, presented with asymptomatic, non-proliferative angio streaks.
- The patient is asymptomatic, BCVA 20/20 OD/OS, and there is no choroidal neovascularization; therefore, this patient is monitored with DFE every 6 months. He was instructed to return to the clinic stat with any sudden visual changes.
- There are 5 different types of hemoglobinopathies. It is crucial to know which one a patient has, as certain forms of the disease affect the eyes more severely.
  - Sickle cell disease, sickle cell trait, sickle cell C disease, sickle cell thalassemia, hemoglobin C trait

V. Treatment and Management

- While there is no cure for sickle cell disease, treatments are designed to relieve symptoms and treat complications. Goals for treating sickle cell anemia include: pain relief and prevention of infections, organ damage, and stroke.
- Treatments include hydroxyurea, pain medications, lifestyle changes, antibiotics, blood transfusions, oxygen therapy, gene therapy, bone marrow and stem cell transplant.
- Due to the risk of developing choroidal neovascularization, patients should be dilated every 6 months, dispensed an Amsler grid for home monitoring and prescribed polycarbonate lenses for additional protection.
- If neovascularization is present, treat with cryotherapy, photocoagulation or anti-VEGF.
- Treat any retinal detachments if they should occur.

VI. Bibliography


VII. Conclusion

- Sickle cell disease often leads to sickle cell anemia, which predominately affects the African American population.
- Carefully look through a patient’s systemic history to know what type of hemoglobinopathy they have, as the severity of ocular findings is dependent upon the type of hemoglobinopathy.
- Perform a close examination of the retina to look for non-proliferative signs (angioid streaks, salmon patches, black sunbursts), proliferative signs (sea-fan retinopathy, fibrovascular changes, retinal detachment), and choroidal neovascularization.