Atypical presentation of idiopathic increased episcleral venous pressure causing persistent bilateral hemorrhages in Schlemm’s canal

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I. Case History
• Patient demographics: 67 year old African American male
• Chief complaint: No visual/ocular complaints.
• Ocular history
  Mild cataracts OD, OS
  CMA with presbyopia OD, OS
  Open angle glaucoma – moderate stage OD, mild stage OS
  Persistent blood in Schlemm’s canal OD, OS
• Medical history
  Hypertension
  Obstructive sleep apnea
  Mixed hyperlipidemia
  Lumbar stenosis
  Cervical laminectomy
  Prostate hypertrophy
• Medications
  Amilodipine besylate
  Aspirin 81mg
  Atorvastatin
  Cholecalciferol 1,000unit
  Gabapentin
  Tamsulosin
• Ocular medications
  Latanoprost qbedtime OD – Monocular trial initiated 08/16

II. Pertinent findings
• Clinical
  Elevated IOP – OD: 18-28mmHg; OS: 18-22mmHg
  Persistent hemorrhages in Schlemm’s canal OD, OS
  Pertinent negatives: no evidence of tortuous/engorged conjunctival or episcleral vessels
  Optic nerve evaluation – OD: 0.65V/0.60H sloping inferior temporal and superior temporal; OS: 0.55V/0.50H thin superior rim
• Laboratory studies
  CBC with differential, syphilis panel (06/16) – WNL
  Sickle cell positive (06/16) – awaiting electrophoresis results
  TSH, Free T4, Free T3 (08/16) – WNL
• Radiology studies
  MRI (06/16) – no mass effect or acute infarcts seen
  MRV/MRA (06/16) – no signs of venous obstruction or arteriovenous abnormalities
Carotid duplex (04/16) – no evidence of hemodynamically significant internal carotid or vertebral disease bilaterally

- Others
  
  Gonioscopy: open grade III-IV 360
  
  03-06/16 – OD, OS: 1 clock hour inferior of blood in Schlemm’s canal
  07/16 – OD: no blood present in angle 360
  OS: 2 clock hours (3:00-5:00) of blood in Schlemm’s canal
  O8/16 – OD: 2 clock hours (9:00-11:00) of blood in Schlemm’s canal
  OS: 2 clock hours (3:00-5:00) of blood in Schlemm’s canal

  HVF 24-2 (07/16) – OD: superior arcuate defect; OS: essentially full

  Pachymetry (04/16) – OD: 535; OS: 533

  OCT RNFL (03/16) – no areas of thinning OD, OS

  Exophthalmometry – will perform at next visit to further rule out thyroid ophthalmopathy. However; MRI reveals no findings consistent with thyroid eye disease, and thyroid function is normal.

III. Differential diagnosis

- Primary/leading – Idiopathic increased episcleral venous pressure

- Others
  
  Venous thrombosis
  Thyroid Ophthalmopathy
  Retrobulbar tumor
  Sturge Weber Syndrome
  Carotid cavernous fistula
  Dural cavernous fistula
  Orbital arteriovenous fistula
  Superior vena caval obstruction
  Episcleral/orbital vein vasculitis
  Sickle cell hemoglobinopathy

IV. Diagnosis and discussion

- Aqueous outflow
  
  -Conventional outflow – pressure dependent
    
    From the anterior chamber to trabecular meshwork through Schlemm’s canal into episcleral venous plexus into aqueous veins of Asher, the long ciliary veins, vortex veins, ophthalmic veins, and finally to the cavernous sinus

  -Unconventional/uveoscleral outflow – pressure independent
    
    From the anterior chamber into the ciliary muscle through suprachoroidal and supraciliary spaces, then exiting the eye by uveal tract and sclera

- Episcleral venous pressure (EVP)
  
  -The pressure intraocular pressure (IOP) must flow against to exit the eye from Schlemm’s canal
  -Average EVP is thought to be between 6.3-9.9mm Hg measured non-invasively in humans by Zeimer’s episcleral venomanometer
  -According to one study, EVP is higher in patients with primary open angle
glaucoma than normal tension glaucoma, which is higher than patients without glaucoma.

• Causes for blood in Schlemm’s canal
  1) Compression during gonioscopy – Blood will retract as pressure is released perhaps leaving a temporary, slight pink stain to Schlemm’s canal
  2) Hypotony – As IOP decreases, episcleral venous pressure remains stable reversing the pressure gradient and allowing blood to flow into Schlemm’s canal.
  3) Increased episcleral venous pressure
    - Venous obstruction/arteriovenous abnormalities causing backflow throughout the orbital venous system resulting in blood within Schlemm’s canal. MRA/MRV can be utilized for possible etiologies.
    - Retrobulbar tumor – compressing distal portion of orbital venous system causing backflow and blood in Schlemm’s canal. MRI will reveal mass effect and acute infarcts.
    - Idiopathic
  4) Thyroid ophthalmopathy – Orbital tissue swells as a result of thyroid dysfunction, venous drainage can be altered and lead to increased episcleral venous pressure and increased IOP, thereby causing blood in Schlemm’s canal. Normal thyroid function, absence of exophthalmos, and no MRI findings consistent with thyroid eye disease will help rule out as possibility.
  5) Sickle cell – Two reported cases of sickle cell causing blood in Schlemm’s canal. Patients with sickle cell hemoglobinopathies are thought to have a higher number of sickled red blood cells in their anterior chambers, which can result in increased IOP due to decreased outflow. However, neither of the patients in the two cases mentioned had glaucomatous nerves or visual field defects.
  6) One case report showed spontaneous resolution of blood in Schlemm’s canal without pharmacologic or surgical intervention.

• Ocular presentation
  Most commonly unilateral, but can be bilateral
  IOP spikes usually greater than 30
  Episcleral and/or conjunctival venous engorgement/tortuosity
  Blood in Schlemm’s canal

V. Treatment, management

• Treatment and response to treatment
  - Our patient recently began latanoprost at bedtime OD as monocular trial to lower IOP by increasing uveoscleral outflow, bypassing the need for episcleral involvement in drainage. Our patient has not been evaluated since starting latanoprost, but has an appointment scheduled in October 2016 before American Academy of Optometry meeting in November.
  - The initiation of treatment with latanoprost is chosen for two reasons; one for the patient’s early glaucoma and two, for attempting to decrease episcleral venous pressure (EVP). One study on mice suggests latanoprost decreases episcleral venous pressure, as determined by blood reflux into Schlemm’s canal.
  - Upon return if the IOP is still elevated, we will need to consider additional topical medications to lower IOP. However, one article suggested decreasing IOP
only creates smaller gap between IOP and EVP, it does not actually lower EVP. Thus, not treating the underlying etiology of the increased IOP. Multiple articles suggested increased IOP secondary to increased EVP, is often resistant to topical medications, requiring surgical intervention. If glaucoma surgery is necessary, the patient is at greater risk for complications due to unopposed increased episcleral venous pressure.

VI. Conclusion
• Some experts in the field of glaucoma consider blood in Schlemm’s canal to be a pathognomonic sign for increased episcleral venous pressure. Foremost, hypotony and compression during gonioscopy should be eliminated as obvious etiologies. Before making any idiopathic diagnosis, other causes of the increased pressure need to be investigated including venous thrombosis, arteriovenous abnormalities, and systemic/autoimmune etiology. If found as cause, appropriate referrals need to be made.
• Elevated intraocular pressure secondary to increased episcleral pressure is often resistant to maximum medical therapy and can be referred for glaucoma procedure(s) with increased risk of complications.

VII. References


