TITLE:
When a red eye is a red herring: an evolving case of Carotid Cavernous Fistula

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ABSTRACT:
A 60 year-old Hispanic Male presents with unilateral redness and increased intraocular pressure progressing to proptosis and strabismus. CT angiography confirms diagnosis of Carotid Cavernous Fistula, followed by improvement of symptoms despite unsuccessful endovascular intervention.

I. Case History

- Patient demographics: 60 year old Hispanic Male
- Chief complaint: Bulging of the right eye with double vision and increasing pain, redness and tearing.
- Ocular history:
  - History of Glaucoma suspect both eyes.
  - Pingueculitis and Ocular Hypertension x 3 weeks:
    - Started on Lotemax QID right eye, Alphagan BID both eyes, and Latanoprost qHS both eyes.
    - Pressure was effectively lowered in the left eye but remained elevated in the right eye.
  - Abduction deficit of the right eye was noted at second visit
- Medical history:
  - Diabetes Mellitus type II
  - Hypertension
  - Heart attack (x2)
  - Hypercholesteremia
  - Stroke
- Medications:
• Brimonidine 1%
• Latanoprost 0.005%
• Carvedilol
• Clopidogrel
• Furosemide
• Isosorbide dinitrate
• Simvastatin
• Metformin

• Other salient information: severe motor vehicle accident in 2013 with cardiac arrest and acute myocardial infarction, followed by cerebral vascular accident.

II. Pertinent findings

• Clinical
  o Visual Acuity
    ▪ OD: 20/50 PH 20/40
    ▪ OS: 20/25 PH NI
  o Pupils
    ▪ trace (+) APD OD
  o Slit Lamp Exam
    ▪ OD
      • Proptosis
      • 2+ conjunctival injection and conjunctival vascular tortuosity
      • Optic nerve: vertical elongation of the cup, but within normal appearance.
      • No retinal hemorrhages or vessel tortuosity
    ▪ OS: no pertinent findings
  o Tonometry (GAT): OD 35 mmHg, OS 18 mmHg, at 1:56 PM
  o Gonioscopy
    ▪ OD: open, blood in Schlemm’s canal 360
    ▪ OS: open 360
  o Hertel Exophthalmometry: OD 28mm, OS 23 mm
  o Strabismus Exam: Right abduction deficit and right hypotropia
  o Red cap desaturation: 50% desaturation OD
  o Color Vision, Ishihara: OD reduced; OS normal

• Humphrey Visual Field 30-2: poor reliability, cloverleaf pattern both eyes; non-diagnostic

• Radiology studies
  o Of note: patient is MRI non-compatible due to pacemaker
  o CT angiogram cerebral with 3D reconstruction: Early venous enhancement of the right cavernous sinus in the earlier arterial phase images with prominent linear appearing venous structure in the delayed venous phase. Early venous enhancement on the right
superior ophthalmic vein. Subtle proptosis on the right, with engorged extraocular muscles.

III. Differential diagnosis

- Primary/leading: Dural Carotid Cavernous Fistula
- Others: Thyroid Eye Disease, Idiopathic Orbital Inflammatory Disease, Neoplasm, Cavernous Sinus Thrombosis

IV. Diagnosis and discussion

- Carotid Cavernous Fistula (CCF)
  - An abnormal communication between the carotid artery and the cavernous sinus. Can be classified by pathogenesis (traumatic or spontaneous); by hemodynamics (high or low flow); or by anatomy (direct or dural) (1).
  - Barrow’s classification: Type A fistulas are direct shunts between the internal carotid artery and cavernous sinus; Types B, C, and D are dural shunts (typically low flow) (1).
  - Arterialization of veins leads to elevation of the intravenous pressure as well as a change in the rate and direction of blood flow, which then causes venous and arterial stasis of the eye and orbit, increased episcleral venous pressure, and decreased blood flow to the cranial nerves (2).

- Dural CCF
  - Literature suggests that these low flow fistulas may open spontaneously in older women, as well as with hypertension, diabetes, atherosclerotic disease, childbirth, or collagen vascular disease (1, 2).
  - Typically have a more insidious onset with less severe symptoms which can lead to early misdiagnoses (gradual red eye, ocular hypertension) (2).

- Ocular manifestations
  - Proptosis (pulsatile with direct), epibulbar arterialized vessels, increased intraocular pressure and secondary glaucoma (as well as blood in Schlemm’s canal), motility restrictions (6th nerve likely). Direct CCF more likely to cause retinal ischemia and hemorrhages, optic disc swelling, and orbital bruit (2, 5).
  - Usually unilateral and ipsilateral to the fistula (2).

V. Treatment, management

- Our patient’s intervention: Catheter Angiogram under general anesthesia for endovascular repair.
Revealed a small fistula without connection to the SOV. The fistula drains superiorly into what is possibly a cortical vein although it is difficult to ascertain. Attempted a transvenous approach but could not access the small pouch and arterial feeders could not be catheterized. Procedure was then terminated.

Plan: observe the possible cortical venous drainage as treating it surgically “would be a major endeavor for someone on ASA and Plavix with a cardiac history”. Repeat angiogram in 6 months.

- At 1 month neuro-ophthalmology follow-up:
  - Improved proptosis (from a difference of 5 mm to 1.5 mm), improved eye movements (mild restriction of abduction and elevation of the right eye with ability to fuse in primary gaze), normalized intraocular pressure (with topical medications), improved arterIALIZATION in conjunctiva, and no evidence of residual optic neuropathy.

- CT angiography (and to a lesser extent MR angiography) has been shown to be as effective as the more invasive digital subtraction angiography (DSA) in visualizing fistulas (4).

- Direct CCF’s typically require urgent treatment. Dural CCF’s may spontaneously resolve (20-50% in various literature). However, treatment may still be indicated, especially if there is significant vision loss, diplopia, a bruit or headache that is intolerable to the patient, or severe proptosis causing corneal exposure. Symptoms must be weighed against the anatomy of the fistula and the possible treatment modality (1, 2).

- With direct CCF’s the goal is to occlude the site of communication between the ICA and the cavernous sinus. With indirect CCF’s the goal is to interrupt the communication and lower the pressure. Options include a transarterial or transvenous approach with a balloon, stent, coil or liquid (such as n-butyl cyanoacrylate) to embolize. Other less frequent techniques include surgery and radiotherapy (5, 6).

- The success rate for closing direct fistulas is 85%-99% with risks of occluding the ICA completely, and worsening the cranial nerve palsy. The reported cure rate for indirect CCFs is 70%-78% with a complication rate of 5%. It has been reported that 20%-30% of patients without complete angiographic success will have still experienced improved symptoms (6).

**VI. Conclusion**

- Patients with CCF will often present to an eye care professional initially due to ocular symptoms such as a progressive red eye, ocular motility restriction and/or proptosis.

- It is important for the optometrist/ophthalmologist to make a prompt referral to a neurosurgeon and neuro-ophthalmologist for imaging and subsequent intervention as needed (3).

- Subsequent monitoring of the patient by an optometrist/ophthalmologist (especially if intervention was not indicated or was not successful) is required to monitor vision, pupillary reaction, intraocular pressure, visual fields, exophthalmometry, gonioscopy, strabismus examination and fundoscopy (1).
Bibliography


