Patient

HPI: 36 yo African American female seen as follow up from the ER with red OS x 6 months treated by multiple doctors for conjunctivitis that has not resolved. She was also being treated for elevated intraocular pressure with Brimonidine in her left eye. Also states that she has had a headache with occasional diplopia on and off for the past 5 months. Denies pain, trauma.
History

- POH: none
- Gtts: Brimonidine 0/1
- PMH: DM (diagnosed 2010), HTN
- Meds: Metformin
- All: nkda
- FH: no glaucoma/blindness
EXAM

- dVa sc: 20/20, 20/25
- Pupils: 3 → 2 ou, no apd
- EOMs: full ou, no diplopia
- CVFs: full ou
SLE

- K: clear ou
- A/C: d and q ou
- P/I: r and r ou
- L: clear ou
DFE

- V: clear ou
- C/D: 0.3/0.3, s and p ou
- M: flat ou, +flr ou
- V: wnl ou
- P: no holes/tears/heme seen ou
Differential Diagnosis
Differential Diagnosis

- Thyroid Eye Disease
- Idiopathic Orbital Inflammation
- Orbital tumors (benign or malignant)
- Arteriovenous Fistula
Goldman Applanation
Tapp: 15/27 @ 11:45am
Upon Specific questioning

- Patient states that she has been hearing a wooshing sound for the past month as well
  - Ocular auscultation WNL
Differential Diagnosis

- Thyroid Eye Disease
- Idiopathic Orbital Inflammation
- Orbital tumors (benign or malignant)
- Arteriovenous Fistula
What would you do next?
CT Angiogram
CT Angiogram

- Findings highly suggestive of an Arteriovenous fistula
Arteriovenous Malformation

- Developmental anomaly
- Presence of shunting through a nidus of coiled and tortuous vascular connections that join feeding arteries to draining veins without an intervening capillary bed

Arteriovenous Fistula

- Acquired lesion
- Caused by abnormal direct communication between an artery and a vein
<table>
<thead>
<tr>
<th>Symptom</th>
<th>Cavernous Sinus (%)</th>
<th>Transverse-Sigmoid Sinus (%)</th>
<th>Tentorium (%)</th>
<th>Superior Sagittal Sinus (%)</th>
<th>Anterior Fossa (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ocular symptoms</td>
<td>80–97</td>
<td>. . .</td>
<td>. . .</td>
<td>. . .</td>
<td>. . .</td>
</tr>
<tr>
<td>Cranial nerve deficits</td>
<td>44–77</td>
<td>7–12</td>
<td>14–17</td>
<td>. . .</td>
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</tr>
<tr>
<td>Bruit, tinnitus</td>
<td>40–50</td>
<td>40–42</td>
<td>70–88</td>
<td>. . .</td>
<td>. . .</td>
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<tr>
<td>Headache</td>
<td>. . .</td>
<td>46–76</td>
<td>8–24</td>
<td>50</td>
<td>12–15</td>
</tr>
<tr>
<td>Central nerve deficits</td>
<td>3</td>
<td>10–20</td>
<td>23–42</td>
<td>29</td>
<td>5–33</td>
</tr>
<tr>
<td>Intracranial hemorrhage</td>
<td>Rare</td>
<td>15–28</td>
<td>60–74</td>
<td>23</td>
<td>44–84</td>
</tr>
<tr>
<td>Dementia</td>
<td>. . .</td>
<td>Rare</td>
<td>. . .</td>
<td>5</td>
<td>. . .</td>
</tr>
</tbody>
</table>
Barrow Classification of Carotid-Cavernous fistulas

Type A

Type B

Type C

Type D
Etiology of Each Type

Type A (Direct)

- Trauma, Spontaneous (Ehlers-Danlos), secondary to aneurysms

Type B-D (Indirect)

- Atherosclerosis, hypertension, collagen vascular disease, seen in elderly most often
**Direct Carotid Cavernous Fistula**

- High blood-flow rate
- Classic triad: pulsatile proptosis, conjunctival injection/chemosis, and an ocular bruit
- Other findings
  - Tortuous epibulbar vessels
  - Elevated IOP
  - Choroidal effusions
  - Blood in Schlemm’s canal
  - Cranial nerve dysfunction of III, IV, and most commonly VI
  - Enlargement of EOM’s, & dilated SOV
**Dural Sinus Fistula**

- Low blood-flow
- Small meningeal arterial branches communicating with venous drainage
- Insidious onset

Findings (same as CC fistula but not as severe/acute)
- Arterialization of episcleral veins
- Elevated Intraocular Pressure
- Enlargement of EOM’s & dil SOV
Pathophysiology

Retrograde Venous Drainage → Venous Hypertension

- Enlarged EOMs → Restriction → Diplopia, Proptosis → Exposure
- Chemosis and Injection → Red Eye
- Increased Episceral pressure → Increased IOP → Secondary Glaucoma
Pathophysiology

Venous and Arterial Stasis

- Decreased Ocular/Retinal Perfusion
- Decreased perfusion of Cavernous Sinus Cranial Nerves

Decreased visual acuity
Anterior Segment Ischemia

Diplopia
Ophthalmoplegia
What imaging to order?

- CT/MR Angiogram
- Orbital color Doppler U/S
- Intra-arterial Catheter Angiogram
  “Gold Standard”

- Zipfel GJ, Shah MN, Refai D, Dacey RG Jr, Derdeyn CP.
- Department of Neurological Surgery, Washington University School of Medicine, St. Louis, Missouri 63110, USA. zipfelg@nsurg.wustl.edu

- This article presents a modification to the existing classification scales of intracranial dural arteriovenous fistulas based on newly published research regarding the relationship of clinical symptoms and outcome.

- The most critical anatomical feature is the identification of cortical venous reflux.

- Based on recent data the annual rate of intracerebral hemorrhage is 7.4-7.6% for patients with symptomatic CVD compared with 1.4-1.5% for those with asymptomatic CVD. The addition of asymptomatic CVD or symptomatic CVD as modifiers to the Borden-Shucart and Cognard systems improves their accuracy for risk stratification of patients with high-grade dural arteriovenous fistulas.
When to Observe

- In lesions without CVR (Cortical Venous Reflux)
- Asymptomatic or tolerable symptoms
- May close spontaneously (10-60%)

FOLLOW-UP

- Serial MRI, MRA and DSA after 3 yr
- Any change in symptoms needs evaluation

CONCERNS

- 2-3% chance of developing CVR
- Intra-cerebral hemorrhage
  - Overall risk of hemorrhage – 1.6% / yr (Brown et al.)
- Exacerbation & remission
Indication for Treatment

- Lack of spontaneous closure
- Risk to eye/vision
- Intolerable symptoms
- “High-risk” for stroke
- Venous thrombosis
- Mental status changes
What is the treatment?

- Low grade
  - Observation
  - Compression Therapy

- High grade
  - Observation
  - Compression Therapy (unlikely to succeed)
  - Embolization (trans-arterial or venous)
  - Surgical Excision
Successful treatment of dural AV fistulas by manual compression—a matter of perseverance

Martin Schumacher • Norbert Szczepnik

Abstract A noninvasive treatment of an occipital dural AV fistula by external compression of the carotid artery and jugular vein is documented. The late successful occlusion of the malformation after almost 19 months and a total of 13,216 single manual compressions shows that fistula occlusion can be achieved late and is dependent on careful performance of the maneuver.

Keywords Dural arteriovenous malformation • Long-term treatment • Carotid artery compression

Cognard at al. [18] regard arterial compression as useful only in the treatment of type I or IIa asymptomatic fistulas. They recommend performing the compression with the contralateral hand in a sitting or lying position to avoid possible ischemia due to embolic complications or vagal reaction. The maneuver should be increased in duration from 10 to 30 s and in frequency from four to six times an hour.
Intra-arterial Angiogram

Indirect Meningeal feeders
A-V shunting – cavernous pouch and inferior petrosal sinus (IPS)

Courtesy of:
Sundeep Mangla, M.D.
Director of Interventional Neuroradiology
SUNY Downstate
NeuroInterventional Therapy

Interim coiling
Patient Care, Medical Knowledge

Final Post Coiling
(AV shunting eliminated)
Presentation

1 month post-embolization
Presentation

1 month post-embolization
1 month post-embolization
1 month post embolization

- Subjective
  - “My red eye went away in 2 days”
  - “The swooshing sound instantly disappeared”

- Changes on exam
  - dVa sc 20/20 od/os
  - Tapp: 16/16 @ 10:00am
  - Hertels: 16/17.5
Reflective Practice

This case demonstrated the importance of listening to all of the patient complaints and having the background knowledge to put all the clues together in order to properly diagnose and treat the proper disease etiology.

This patient had an expedited and excellent result and was very happy with the resolution of all her symptoms after treatment.
Core Competencies

- **Patient Care**: The case involved thorough patient care and attention to patient’s complaints. Once diagnosed, the patient received proper management and care.

- **Medical Knowledge**: This presentation allowed us to review the presentations, proper evaluation/work up, and different treatments.

- **Practice-Based Learning and Improvement**: This presentation included a current literature search of developing associations and current treatment modalities.

- **Interpersonal and Communication Skills**: The patient was treated with respect and every effort was made to communicate with the patient in a timely manner for the proper follow-up.

- **Professionalism**: The patient was treated in the proper manner. She was also referred to the proper specialist to treat her condition.

- **Systems-Based Practice**: The patient was discussed in detail with neuroradiologic colleagues in regard to follow up and treatment.
Thank you

- Dr. Shinder
- Dr. Mangla
- KCHC staff
References

- Sundeep Mangla, M.D.
  Director of Interventional Neuroradiology
- Dural AVF Classification and Management. Rakesh K. Singh
- Basic Clinical and Sciences Series. Orbit. Pages 64-70
- Interventional treatment of carotid cavernous fistula
- Richard C. Barry a, Mark Wilkinson b, Rebekah M. Ahmed c, Charmaine S.M. Lim a, Geoffrey D. Parker b,
  a Department of Ophthalmology, Royal Prince Alfred Hospital, Camperdown, New South Wales, Austra