Carotid-Cavernous Fistula Diagnosis and Management

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Patient Presentation

• 69 year old female presents to ophthalmology clinic with complaints of persistent **right conjunctival injection** and **swelling** over the past 6 months. Patient also suffered from intermittent **diplopia** and **blurry vision**. A course of steroids improved the swelling, but redness has persisted in spite of over the counter remedies.

• On exam, patients has right sided conjunctival injection, right sided proptosis, and isolated right sided elevated intraocular pressure. Pupils are reactive to light. On cranial nerve exam, patient is unable to abduct her right eye.
What Imaging Should We Order?
Select the applicable ACR Appropriateness Criteria

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>RRL</th>
</tr>
</thead>
<tbody>
<tr>
<td>MRI orbits without and with IV contrast</td>
<td>Usually Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>CT orbits with IV contrast</td>
<td>Usually Appropriate</td>
<td>★★★</td>
</tr>
<tr>
<td>CT orbits without IV contrast</td>
<td>May Be Appropriate</td>
<td>★★★</td>
</tr>
<tr>
<td>CTA head and neck with IV contrast</td>
<td>May Be Appropriate</td>
<td>★★★</td>
</tr>
<tr>
<td>MRA head and neck without and with IV contrast</td>
<td>May Be Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>MRI head without and with IV contrast</td>
<td>May Be Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>MRI orbits without IV contrast</td>
<td>May Be Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>MRA head and neck without IV contrast</td>
<td>May Be Appropriate (Disagreement)</td>
<td>O</td>
</tr>
<tr>
<td>MRI head without IV contrast</td>
<td>May Be Appropriate</td>
<td>O</td>
</tr>
<tr>
<td>Arteriography cervicocerebral</td>
<td>May Be Appropriate</td>
<td>★★★</td>
</tr>
<tr>
<td>CT head with IV contrast</td>
<td>May Be Appropriate</td>
<td>★★★</td>
</tr>
<tr>
<td>CT head without IV contrast</td>
<td>May Be Appropriate</td>
<td>★★★</td>
</tr>
<tr>
<td>CT head without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>★★★</td>
</tr>
<tr>
<td>CT orbits without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>★★★</td>
</tr>
<tr>
<td>X-ray orbit</td>
<td>Usually Not Appropriate</td>
<td>★</td>
</tr>
</tbody>
</table>

This imaging modality was ordered by the ophthalmologist.
Findings (unlabeled)
Findings (labeled)

Dilated but patent right superior ophthalmic vein

Mild right sided retrobulbar fat stranding with enlargement of the extraocular muscles

Medial right cavernous sinus postcontrast enhancement and right sided proptosis
Initial Dx:

Cavernous carotid fistula vs chronic cavernous sinus thrombosis

Cerebral angiogram was scheduled for confirmation of diagnosis, and potential treatment.
Follow up Imaging

- Filling of the right ICA
- Filling of the inferolateral trunk from the ICA
- Early leak into the cavernous sinus, likely from the inferolateral trunk
- Persisting contrast in the cavernous sinus
- Filling of the right ECA
- Filling of the right middle meningeal artery from the ECA
- Early leak into the cavernous sinus, likely from the middle meningeal artery
- Persisting contrast in the cavernous sinus
Case Discussion

• Carotid-cavernous fistulas (CCF) can be formed spontaneously or due to a traumatic insult. They allow shunting of arterial blood from the internal carotid artery (ICA) or external carotid artery (ECA) into the cavernous sinus.

• 75% are due to trauma
  • 0.2% of patients with craniocerebral trauma and 4% of patients with basilar skull fractures develop CCF

• Spontaneous CCF are less common and found in older, female patients.
  • Associated with cavernous carotid aneurysms
  • Can be due to chronic venous thrombosis with revascularization and thus are similar to dural arteriovenous fistulas..
Classification

- Hemodynamic status: high flow vs low flow
- Cause: spontaneous vs traumatic
- Anatomical: direct vs indirect (from branches of carotid)

<table>
<thead>
<tr>
<th>Type</th>
<th>Indirect vs Direct</th>
<th>High Flow vs Low Flow</th>
<th>Site of Fistula</th>
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</thead>
<tbody>
<tr>
<td>Type A</td>
<td>Direct</td>
<td>High</td>
<td>ICA</td>
</tr>
<tr>
<td>Type B</td>
<td>Indirect</td>
<td>Low</td>
<td>Branch of ICA</td>
</tr>
<tr>
<td>Type C</td>
<td>Indirect</td>
<td>Low</td>
<td>Branch of ECA</td>
</tr>
<tr>
<td>Type D</td>
<td>Indirect</td>
<td>Low</td>
<td>Branches of ICA and ECA</td>
</tr>
</tbody>
</table>

Our patient had a Type D CCF
Management

• External manual compression: numerous case series have found repeated manual compression to be effective at fistula closure with symptom relief.
  • Success rates vary between 17%-86% after 2 years.

• Endovascular intervention: trans-arterial or trans-venous embolization with the use of metal coils or other embolic agents.
  • More immediate results, but risks include cerebral infarction, hemorrhage, and transient worsening of symptoms

• Open surgical intervention: can be considered when less invasive options are not possible.
  • Invasive and associated with longer recovery times

• Radiosurgery: effective in treating indirect CCF with success rates reported as up to 90%. Less efficacious at treating direct CCF.
References:


• https://radiopaedia.org/articles/caroticocavernous-fistula-1?lang=us