Cranial Motor Neuropathies

Diplopia – Pearls and Pitfalls

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Key Points

• Involved cranial nerves
  – Single, multiple, associated symptoms
• When to image
• What type of imaging
• Ancillary testing
• Confounding factors
Anatomy

• Eye Movements
  – 3 ocular motor nerves
    • CN III (Oculomotor)
    • CN IV (Trochlear)
    • CN VI (Abducens)
  – 6 extraocular muscles
    • Medial rectus, lateral rectus, superior rectus, inferior rectus, superior oblique and inferior oblique
Anatomy Cranial Nerve III (Oculomotor Nerve)

• Dorsal midbrain (midline)
  – Composed of several subnuclei
    • Superior rectus nuclei – contralateral innervation of the superior rectus muscle
    • Central caudate nucleus – single nucleus for both levator palpebrae superioris muscles
Anatomy and Function Cranial Nerve III

**Brainstem**
- Travels by the red nucleus, cerebral peduncles

**Subarachnoid space**
- Passes between the SCA and PCA

**Cavernous sinus**
- Lateral wall

**Superior orbital fissure**
- Superior and inferior division CN III
Function Cranial Nerve III

• **Superior Division**
  – Superior rectus – elevates the eye
  – Levator palpebrae superioris – elevates eyelid

• **Inferior Division**
  – Medial rectus – aDducts the eye
  – Inferior rectus - depresses the eye
  – Inferior oblique – elevates the eye
  – Pupillary sphincter
Cranial Nerve III Palsy

- Type of diplopia
  - Variable (vertical, horizontal, oblique)
Cranial Nerve III Palsy
Clinical Features to Help with Localization

• **Nuclear**
  – Ipsilateral mydriasis, ipsilateral weakness of medial rectus, inferior rectus, inferior oblique with bilateral ptosis and difficulty elevating both eyes

• **Fasicular**
  – Cerebral peduncle
    • contralateral hemiparesis (Weber’s Syndrome)
  – Red Nucleus
    • contralateral limb tremor (Benedikt Syndrome)
  – Brachium conjunctivum
    • contralateral cerebellar ataxia (Claude’s Syndrome)
Anatomy and Function of Cranial Nerve IV

Pontomesencephalic junction
  Ventral cerebral aqueduct

Inferior colliculus
  Exits dorsal
  Crosses prior to exit
Cavernous Sinus

Cavernous sinus
  Lateral wall
  Enters below CN III

Superior orbital fissure
  Innervate contralateral superior oblique
Anatomy of Cranial Nerve IV

• Superior oblique muscle
  – depresses and intorts the eye
Cranial Nerve IV Palsy

• Type of diplopia
  – Vertical diplopia often with torsional component
Cranial Nerve IV Palsy
Cranial Nerve IV Palsy
Cranial Nerve IV Palsy

• Misalignments as small as 200 um can cause diplopia
• No abnormalities on gross examination
  – Ask the patient to view a horizontal straight line
    • Edge of a door frame
  – Horizontal straight line with a second tilted line above or below
Anatomy and Function of Cranial Nerve VI

Dorsal pons
- Floor of 4th Ventricle
- Corticospinal tract

Subarachnoid space
- Clivus

Dorello’s Canal
- Petrus ridge
Anatomy and Function Cranial Nerve IV

**Cavernous sinus**
- Medial wall
- Lateral to carotid artery and medial to the 4th nerve

**Superior orbital fissure**
- Ipsilateral lateral rectus muscle
Function of Cranial Nerve VI

• Lateral rectus muscle
  – Abduct the eye
Cranial Nerve VI Palsy

• Type of diplopia
  – Horizontal
  – Better at near, worse at distance
Cranial VI Nerve Palsy - Left
Clinical Features to Help Localize Cranial Nerve VI Palsy

• Nuclear
  – Abduction deficit of ipsilateral eye with adduction deficit of the contralateral eye (nuclear conjugate gaze palsy)

• Fasicular
  – Isolated abduction deficit
  – 7th nerve nucleus (ipsilateral facial weakness)
  – Corticospinal tract – contralateral hemiparesis
Diagnostic Evaluation

• Three Features to Consider
  – Which cranial nerve
  – More than one cranial nerve involved
  – Age
    • Older or younger than age 50
Etiologies of Single Ocular Motor Nerve Palsy

- Ischemic
- Compression
- Trauma
- Inflammation
- Neuro – Muscular Junction Disorder
# Table 9-2  Etiology of Adult Isolated Cranial Nerve Palsy

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Cranial Nerve III</th>
<th>Cranial Nerve IV</th>
<th>Cranial Nerve VI</th>
</tr>
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<tbody>
<tr>
<td>Ischemic</td>
<td>49%</td>
<td>23%</td>
<td>45%</td>
</tr>
<tr>
<td>Aneurysmal</td>
<td>9%</td>
<td>0%</td>
<td>0%</td>
</tr>
<tr>
<td>Tumor</td>
<td>7%</td>
<td>1%</td>
<td>2%</td>
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<tr>
<td>Congenital</td>
<td>0%</td>
<td>38%</td>
<td>0%</td>
</tr>
<tr>
<td>Trauma</td>
<td>6%</td>
<td>29%</td>
<td>15%</td>
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<tr>
<td>Myasthenia gravis</td>
<td>0%</td>
<td>0%</td>
<td>1%</td>
</tr>
<tr>
<td>Multiple sclerosis</td>
<td>3%</td>
<td>0%</td>
<td>7.5%</td>
</tr>
<tr>
<td>Other</td>
<td>16%</td>
<td>1%</td>
<td>22%</td>
</tr>
<tr>
<td>Undetermined</td>
<td>10%</td>
<td>8%</td>
<td>7.5%</td>
</tr>
</tbody>
</table>

Single Nerve Palsy > 50

- **Imaging**
  - Disagreements exist on urgency
- **ESR/CRP**
  - Headache, jaw claudication
  - 12% of single cranial nerve palsies may be related to GCA
- **Acetylcholine Receptor Antibodies, SF EMG**
  - Variability
  - No improvement at 4 weeks
- **Lumbar Puncture**
Single Nerve Palsy > 50

• Imaging (no formal guidelines)
  – Original recommendations wait 12 weeks, if no improvement/resolution then image
  – 4 to 6 weeks if no improvement
  – In today’s medical – legal environment consider imaging earlier then later
What Type of MRI

• MRI ORBITS with and without Contrast
  – Brain stem and skull base
  – Orbits
  – Cavernous sinus
  – Axial FLAIR of the entire brain
  – Diffusion gradient ECHO
Communication with Radiology is Key

• Protocols vary by institution
  – Seattle Radiology
    • TEC
    • Swedish First Hill – Fiesta Coronal: high resolution sequence that allows visualization of CN III and CN VI as they exit the brainstem

• Protocols vary within an institution
  – Seattle Radiology: Swedish First Hill
  – RADIA: Swedish Cherry Hill/Issaquah/PRMC

• The more you can put on the requisition in terms of symptoms or possible etiologies, the higher the yield
MRI ORBITAL APEX CASE
Cranial Nerve III Palsy – URGENT Imaging

- Incomplete or pupil involving CN III palsy
- URGENT
- Need to r/o aneurysm
- Originally cerebral angiogram was gold standard
- MRA Head, CTA Head
  – Ideal imaging varies with institution
Noninvasive Vascular Imaging – CTA and MRA

- Highly sensitive in detecting even small aneurysms
- Several studies have shown that when imaging originally read as normal, on retrospective review aneurysm was present
Single Nerve Palsy < 50

- Directed MRI Imaging
- Acetylcholine Receptor Antibodies/SF EMG
- Lumbar puncture
- Anti – GQ1b – areflexia or ataxia
Multiple Cranial Nerve Palsies

- Cavernous Sinus
- CSF based process affecting nerves in subarachnoid space
- Neuro-muscular junction disorder
  - May mimic multiple cranial nerve palsies
- Inflammatory
Multiple Cranial Nerve Palsies

- Directed MRI imaging
  - Cavernous sinus, skull base
- Acetylcholine Receptor Antibodies/SF EMG
- Lumbar puncture
- Anti – GQ1b
  - areflexia or ataxia
Confounding Factors

• **History of strabismus/prior strabismus surgery**
  – Esotropia or exotropia
  – Neuro – Ophthalmology can be helpful in determining what is “new” vs. “old” based on ocular alignment measurements

• **Internuclear Ophthalmoloplegia (INO)**
  – May be confused with a third nerve palsy

• **Congenital Syndromes**
  – Duane syndrome
Summary

- Identifying involved ocular motor nerve(s) will allow you to determine urgency of imaging and where to direct your imaging
- A pupil involving or partial CN III palsy requires STAT imaging to rule out an aneurysm
- Noninvasive imaging is sensitive in detecting even small aneurysms
Summary

• Communication with your radiologist is vital
  – Appropriate studies
  – Increase sensitivity in detecting subtle changes

• No consensus on the timing of imaging in patients > 50 with an isolated ocular motor nerve palsy

• Patients < 50 or with multiple nerve palsies require immediate imaging