The Efferent Visual System: Disorders of Cranial Nerves III, IV & VI

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Course Goals

• To Become Familiar with Presentations of CN III, IV and VI Palsies
• To Understand the Relevant Neuro Anatomy
• To Understand the Neuro Imaging and Clinical Management
• To Obtain In Clinic Assessment Pearls

Course Format

• Case Presentation
  — Dr. Fanelli
• Relevant Neuro Anatomy
  — Dr. Lombardi
• Neuro Imaging and Management
  — Messner
• Clinical Take Home
  — Drs Fanelli, Lombardi and Messner

“THE FIRST 4 QUESTIONS”

1. WHO IS THE NEURO-OP ON CALL?
2. WHAT IS THEIR NUMBER?
3. HOW SOON CAN THE PATIENT BE SEEN?
4. WHAT IS THE DIAGNOSIS?

The Diplopic Patient

• Evaluation boils down to knowing the fields of action of the 6 EOM’s
• You know the actions, you can figure out the Palsy
  — Cerca Trova

“Do I see double?”
The Physiological “H”

- You are face to face with the patient
- Their EOM movements, as you view them, render the following “H” pattern:

4 Questions We Should Ask

1. Is Double Vision Present with one eye covered?
   - “Yes” eliminates neurologic etiologies
   - Usually a ‘windows’ problem
   - Media opacities

2. Does the Diplopia have a vertical component or a horizontal component
Questions We Should Ask

3-In which direction (R or L) does the diplopia worsen?

Questions We Should Ask

4-Is the diplopia greater at distance or near?

Clinical Assessment of Diplopia

- Begins with dissociating the presenting images before each eye
- Maddox Rod

“LANGUAGE OF THE LIGHT” (PATIENT’S VIEW)
Third Nerve Palsies

- CN III Innervates:
  - SR
  - IR
  - MR
  - IO
  - Levator
  - Parasympathetic Iris (constrictor)

So What is Presentation

- Go back to the Physiological H
- Assuming a RIGHT CN III Palsy:

EOM ACTIONS

- Hyper deviation which increases in upgaze and reverses in downgaze
- Exo which increases across from the vertically-limited eye

"The Signature" of CN III Paresis

Oculomotor nerve...

Its course and relationships

In the midbrain
Then CN III can run into trouble

Posterior Communicating artery

Aneurysms can rupture = pressure

Compresses Pupillary Fibers of III...
Dilated pupil
Extraocular Muscle Fibers of III

X-section III

Posterior communicating artery

Subarachnoid hemorrhage

Aneurysms can rupture

Meningeal irritation... pain

Sub-Arachnoid space

BRAIN

Compression (ie aneurysm)

vs

Vasculopathic (ie diabetes)

Lesion of CN III

CN III is peripheral nervous system

and it will regenerate

Sometimes to the wrong target organ...

Aberrant regeneration
Degeneration and regeneration of the axon

Aberrant Regeneration

“look down” Lid up

De generation and regeneration of the axon

The Clinical Picture

- CN III Palsy

CRANIAL NERVE PALSY STRATEGY

- IS THIS REALLY WHAT I THINK IT IS? (imposters)
- DOES IT COME WITH ANYTHING ELSE? (anatomically guided exam)
- IF IT IS ISOLATED, WHAT DO I DO? (management)
CN III palsy – pupil involved
52 y/o man with sudden onset/painful diplopia @ distance and near
CN III – pupil spared
65 y/o diabetic man with recent onset diplopia @ distance and near

“Rule of the Pupil”

Aberrant Regeneration of CN III

1. Pseudo-Graefe sign
2. Eyelid synkinesis
3. Light-gaze dissociated pupils

Neuroimaging for CNIII Palsy

- MRI
- MR Angiography
- Intra-arterial DSA
- CT Angiography

Clinical Kernels: CN III

- EOM pattern of hyper deviation that switches on up and down gaze and increases on gaze away from paretic eye
- Aberrant regeneration does NOT occur in cases of microvascular (diabetic) CN III
- Pupil sparing is NOT always indicative of microvascular etiology

Fourth Nerve Palsies

4th N Innervation & Motility

- Innervation is easy:
  - Superior Oblique
- Motility is more complex
  - Both a horizontal AND vertical component
  - AND……a TORSIONAL component
4th N Palsy

- The paretic eye is hyper in primary gaze
- The diplopia decreases on same gaze; increases on opposite gaze
- But........

4th N and SO Muscle

- The SO is primarily an INTORTER
  - Compensating for a faulty intorter, one would TILT your head in the opposite direction

Torsional Obliques

- Remember this:
  - SUPERIOR muscles INTORT
  - INFERIOR muscles EXTORT

Trochlear Nerve...

Its course and relationships
The Clinical Picture

- CN IV Palsy
“The Signature” of CN IV Paresis

- A hypertropia that increases across from the vertically-limited eye and on ipsilateral head tilt

Evaluation of CN IV Palsy

- Which eye is higher in primary gaze?
- Hyper worse in right or left gaze?
- Which eye is higher on head tilt?
- Is there excyclotorsion?
Evaluation of CN IV Palsy

- Which eye is higher in primary gaze?
- Hyper worse in right or left gaze?
- Which eye is higher on head tilt?
- Is there excyclotorsion?
Traumatic CN IV palsy
28 y/o woman s/p closed head trauma
Right hyper greater in left gaze and right head tilt

Measuring Excyclotorsion

• Subjective
  – Maddox rod
  – Bagolini striated lenses

• Objective
  – Fundus photos
**Objective vs. Subjective Excyclotorsion**

- Objective = Subjective → Recent onset
- Objective > Subjective → Long-standing
- Objective without subjective → Infantile

**Etiology of Adult Superior Oblique Palsies**
(Mollan SP, et al. Eye 2009)

- N = 150
- 133 unilateral-isolated:
  - 38% congenital
  - 29% trauma
  - 23% vasculopathic
  - 7% undetermined
- 10 bilateral:
  - 50% trauma
  - 20% tumor
  - 20% undetermined
- 7 unilateral—complicated
  - 71% trauma
  - 14% tumor
  - 14% undetermined

**Isolate CN IV Palsy Management**

- Observation (improvement within several months for ischemic vascular)
- Prism (base-down over paretic eye/split between both eyes)
  - Rx vertical prism as single vision/NVO
- Surgery
  - Wait for spontaneous improvement (at least 6 months)
- Check for V-pattern eso in kids (indicator for bilateral CN IV palsy)
**Clinical Kernels: CN IV**

- ALL cranial nerves travel through the subarachnoid space, and as such they are susceptible to compression
  - Trauma and bleeds
  - CN 4: Long intracranial/subarachnoid course
- SO is primarily an intorter, therefore HEAD TILT will be an integral finding

**Clinical Kernels: CN IV**

- Assessment of excyclotortion is helpful in determining onset of problem:
  - Long standing, patients adapt, and objective measure > subjective complaints
  - Recent onset, patients haven’t ‘adapted’, noticeable findings, noticeable complaints

**Sixth Nerve Palsies**

**6th N Innervation and Motility**

- Innervation is easy:
  - Lateral Rectus
- Motility is easy:
  - No vertical component
  - Only horizontal component

**EOM ACTIONS**

![EOM ACTIONS Diagram]
**Motility Pattern**

- Inability to Abduct, therefore paretic eye has eso posture IN PRIMARY GAZE
- Eso increases on gaze TOWARD paretic eye

**Compensation for CN VI Palsy**

- Since the paretic eye cannot Abduct and is eso, the patient will TURN THEIR HEAD to the SAME side

**Abducens Nerve...**

Its course and relationships
The Clinical Picture

- CN VI Palsy

“The Signature” of CN VI Paresis

- Eso which increases in the action of the paretic eye

CNVI Palsy Motility Evaluation

- Duction > version
- “Glissades”
- Asymmetric OKN
- Negative forced duction

CN VI Motility Evaluation
27 y/o AA Woman

- c/o horizontal diplopia (right gaze > left)
- h/o recurrent headaches (am > pm)

BVA:
- 20/20 OD
- 20/20 OS
S/P Surgical Decompression

Etiology of CN VI Palsy
Mayo Clinic Study of Olmstead Co. MN USA from 1978-1992
(n = 137)

- Undetermined: 26%
- Hypertension: 19%
- HTN & diabetes: 12%
- Trauma: 12%
- MS: 7%
- Neoplasm: 5% (complicated)
- Diabetes (alone): 4%
- CVA: 4%
- s/p neurosurgery: 3%
- Aneurysm: 2% (complicated)
- Other: 8%

40 y/o woman
Acute horizontal diplopia greater at distance and on left gaze
Recent onset paresthesias R > L

**Clinical Kernels: CN VI**
- Sudden onset unilateral, think small vessel occlusive disease in vasculopathic population
  - But trauma is trauma
- Acquired bilateral: look at the optic nerves and think about increased ICP
  - Long climb up the clivus in sub arachnoid space
- BO prism can optically correct

**Poly Cranial Neuropathies**
- Involvement of CN III, IV and/or VI can be found simultaneously
- Investigation centers on locations in the head where III, IV and VI travel together
  - Orbital apex
    - Any associated proptosis??
  - Cavernous sinus
    - Can’t have a complete Neuro-op lecture without mentioning the Cavernous Sinus
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