Clinical Neuroanatomy
Part I: Brain & Eyes
Introduction

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Objectives

• Identify the anatomical structures of the central nervous system in the Cerebral Hemispheres and structures involved in CNS function

• Develop a rational, systematic approach to localization in clinical neurology

• Recognize those areas of the Brain and Eyes involved in the development of common symptoms of multiple sclerosis
Localization of Symptoms

Localization Tips

- The neurological approach to localization within the nervous system is:
  - Take note of all symptoms and signs
  - For each, consider what system(s) or pathway(s) is(are) implicated
  - For each, decide on the potential location a lesion could be & the side of the lesion; focal vs. multifocal
  - Adding up this analysis for each S/S, we then ask "Where in the nervous system can each of these localizations overlap?" The answer is the location of the current problem! One lesion vs. 2 or more lesions!

Localization in Neurology

• The principal of "garbage in-garbage out" does apply: if you fail to identify the clinical signs correctly, then you will be unable to identify where the problem is

• Clues frequently come from each of the various systems such as cranial nerves, motor and sensory, so that the localization of the lesion may be confirmed by examining each system

Localization Shortcuts

• Sensory loss findings - rule out muscle disease & disorders of the neuromuscular junction as the primary localization

• Visual acuity issues due to neurological ds can be localized to structures above the brainstem that transmit info from the retina to the occipital cortex areas dedicated to the production of visual imagery

• Seizures without other deficits localized to the cortex or to the related subcortical modulating structures in the thalamus

• Cognitive dysfunction without disturbances of consciousness or attention localize above the brainstem
Clinical Pearls

- **FACE involvement** Contralateral or Ipsilateral to weakness or numbness localize to BS or brain/cortex
- **Unilateral symptoms** – brain; **Bilateral symptoms** – spinal cord;
- **Signs of asymmetric UMN dysfunction**
  - if the lesion is above the decussation of the pyramids of the medulla-the UMN signs will be Contralateral to the lesion
  - if the lesion is below the decussation – the UMN signs will be Ipsilateral to the lesion

8 Levels of Localization in the CNS

- 1) The cortex - motor & sensory
- 2) The subcortical WM
- 3) The brain stem – midbrain, pons, medulla; the thalamus / Basal ganglia
- 4) The spinal cord
- 5) The spinal roots
- 6) The peripheral nerves
- 7) The neuromuscular junction
- 8) The muscle
Approach to Localization

• 30 year old female
• Sudden onset
• Left hand/arm weakness

Left hand weakness - Localization

• 1. Cortex
• 2. Corona Radiata
• 3. Internal capsule
• 4. Brainstem
• 5. Spinal Cord
Left sided facial weakness

- Facial weakness on one side & weakness on opposite side or both on the same side
- Localization: Brain
  - Cortex
  - Internal capsule
  - Corona radiata
  - Brain stem

Localization of both?

- Left sided facial weakness
- Left hand weakness
- Localization?
Brain Site Localization

- Think about left facial weakness & left hand weakness
- Weakness on the same side!
- Lesion localization
  - Right side cortical lesion
  - Motor tracts

Localization in the Upper Motor Neuron (Pyramidal) System

*Principle: Tone is increased, causing spasticity and hyperreflexia*

<table>
<thead>
<tr>
<th>Site/Cortex</th>
<th>Symptoms</th>
<th>Signs</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Differential weakness</td>
<td>Fractionated weakness</td>
</tr>
<tr>
<td></td>
<td>limbs and face</td>
<td>(arms &gt; face and leg)</td>
</tr>
<tr>
<td>Sensory symptoms</td>
<td>Aphasia, hemianopia, or hemineglect</td>
<td></td>
</tr>
<tr>
<td>Language, visual or attentional alterations</td>
<td>Cortical and primary sensory loss</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td>Cognitive dysfunction</td>
</tr>
</tbody>
</table>
The CNS as the World View

- Pathways and tracts pass through the entire length of the brain, brain stem and spinal cord
- ‘Meridians of longitude’ & ‘parallel of latitude’
- Establishing where these intersect, then you establish the site of the lesion

<table>
<thead>
<tr>
<th>Site</th>
<th>Symptoms</th>
<th>Signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Corona Radiata</td>
<td>Differential weakness of limbs and face</td>
<td>Fractionated weakness</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Primary sensory loss</td>
</tr>
<tr>
<td>Internal Capsule</td>
<td>Weakness only</td>
<td>Face, arm and leg affected equally and densely</td>
</tr>
<tr>
<td>Brainstem</td>
<td>Unilateral or bilateral weakness</td>
<td>Dense hemiparesis</td>
</tr>
<tr>
<td></td>
<td>Diplopia or vertigo</td>
<td>Ocular or oropharyngeal weakness</td>
</tr>
<tr>
<td></td>
<td>dysarthria or dysphagia</td>
<td></td>
</tr>
<tr>
<td>Spinal Cord</td>
<td>Difficulty with gait</td>
<td>No face involvement</td>
</tr>
<tr>
<td></td>
<td>Difficulty walking</td>
<td>Spastic quadriparesis (cervical) or paraparesis (thoracic)</td>
</tr>
<tr>
<td></td>
<td>Urinary Incontinence</td>
<td>Sensory level</td>
</tr>
</tbody>
</table>
Pathways & Tracts

Localization: Why bother?

• Offer a differential diagnosis
• Gives an indication of a potential cause
• Allows better selection of imaging & other diagnostic tests
• Directs for appropriate treatments
• Gives an indication of prognosis
Localization in MS

- Precise localization is crucial, in:
  - Distinguishing between a true exacerbation and a pseudoexacerbation (heat, fever, 'symptom reactivation')
  - Correlating clinical findings with MRI changes
  - Distinguishing between physiologic and psychologic symptoms/signs
  - Distinguishing between MS attacks and other problems in MS patients (strokes, disc disease, etc)

Brain Localization Summary

- This process of localization is important in neurology because it enables us to narrow the differential diagnosis of a problem, leading to more effective, and efficient management of patients with neurological diseases!
Overview

- Approach to localization in clinical neurology
- Cerebral signs
- “The Eyes Have it”
- Brainstem syndromes
- Spinal cord syndromes
  - and
- One bonus localizing sign
3-D, neuroanatomical thinking:
picturing the structures as you perform the neurological exam

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

The Nervous System: overview

- Brain
  - Brainstem/Cerebellum → nerves to the face
- Cervical Spinal Cord
- Brachial Plexus → nerves to the arm
- Thoracic Spinal Cord
- Lumbosacral Nerve Roots
  - Lumbosacral Plexus → nerves to the leg
Approach to Localization

- Within the CNS: Streets and Avenues: Identifying the "intersection"
  - Avenues: long tracts and pathways
    - Sensory: Pain/Temp vs. Proprioception/Vibration
    - Motor: Descending pyramidal tracts
    - Coordination: pathways to and from the Cerebellum
  - Streets: horizontal sections/exiting pathways
    - Cerebral: cortical functions; homunculus
    - Brainstem: exiting cranial nerves define level
    - Cord: exiting roots (dermatomes) define level

- Neighborhood Signs: looking for dysfunction in “what’s nearby”

Approach to Localization

Zooming in and identifying the “intersection” like a google map
Approach to Localization

- Localization principles: Dividing up the map
  - Front to Back:
    - Anterior → Motor;
    - Posterior → Sensory
  - Left to Right:
    - Crossed Sides (Left Hemisphere → Right body)
  - Bottom to Top
    - Lumbar → Thoracic Spine → Cervical Spine → Brain
      - Leg involvement
      - Arm/Hand involvement
      - Facial involvement

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

Planes of Section: terminology

(B) Coronal Sagittal Horizontal
Approach to Localization

- Anteriorly: Descending Motor
- Posterior: Ascending Sensory

Cerebral Signs

- Cerebral Cortex: the outermost surface

1) Corpus Callosum: crossing fibers

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Cerebral Signs

- Cortical Sensory and Motor homunculus

Sensory: postcentral gyrus      Motor: precentral gyrus

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

Cerebral Signs

- Looking for “Neighborhood Signs”: In Cortex, face & hand are together
Cerebral Signs

- Homuncular representation.

Primary cortices are made of cells dedicated to a particular basic function.
Cerebral Signs

- Focal dysfunction localizable to Cerebrum
  - If lesion is at cortical level, can look for signs of focal cortical dysfunction
    - Subtle errors in Mental Status testing
    - “Street level” = cortex

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Cerebral Signs
  o Focal dysfunction localizable to Cerebrum
    - Aphasias
      "Sensing"
    - Apraxias
    - Neglect/Extinction
    - Frontal Lobe Dysfunction:
      o 3 Behavioral syndromes
    - Affect
      o "Belle Indifference"
      o Pseudobulbar Affect

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

Cerebral Signs
  o **Aphasia**: Focal disturbance of language
    - Dominant Hemisphere (usually L) –
      o most L handed are still L hemisphere dominant.
    - 2 basic forms:
      o **Motor/Expressive Aphasia** (Broca’s): Frontal
      o **Sensory/Receptive Aphasia** (Wernicke’s): Posterior

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Cerebral Signs

- **Apraxia**: Focal disturbance of planning/"doing"
  - Non Dominant Hemisphere (usually R) –
  - Definition:
    - inability to perform an act despite intact pathways
    - "Dressing apraxia"
    - On exam: “brush your teeth”, “comb your hair”
  - Distinguish from Dementia

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Cerebral Signs

- **Sensory Neglect**: Focal disturbance of “sensory integration”
  - Non Dominant Hemisphere (usually R) –
  - Definition:
    - inability to perceive a stimulus despite intact sensory pathways
    - "Left hemineglect"
Cerebral Signs

- **Sensory Neglect:** Focal disturbance of “sensory integration”
  - Non Dominant Hemisphere (usually R)
  - Definition:
    - inability to perceive a stimulus despite intact sensory pathways
    - "Left hemineglect"
    - On exam: “double simultaneous stim”
      - visual, tactile extinction
      - Asterognosis, agraphesthesia
  - Distinguish from Dementia

Cerebral Signs

- **Frontal Lobe Dysfunction:**
  - 3 Behavioral Syndromes
    - **Orbitofrontal:** Disinhibited, lewd, loud
      - (See Mr. Gage, below)
    - **Dorsolateral Prefrontal:** poor planning, dysexecutive function (set shifting, Go/NoGo)
    - **Ventralmedial Frontal:** Flattened affect; avolitional; "lobotomized"
  - “Belle Indifference” – Classic for MS.
  - Distinguish from: Dementia, Personality d/o.
The Eyes Have It

- Two ways MS generally affects the eyes:
  - Optic neuritis
  - Extra-ocular movement abnormalities
    - (Very few other conditions do both.)

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

The Eyes Have It

- Optic Neuritis: blurry vision, grayed out, central scotoma
  - Afferent Pupilary Defect ("Marcus Gunn Pupil"): Input lesion

Visual Pathway

Pupilary Response Pathway

Note: Loss of Vision in both eyes: Bilateral ON (ADEM) or single lesion in optic chiasm (MS)...

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
The Eyes Have It

- Extra-ocular movements: CN3, 4, and 6:

  Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

The Eyes Have It

- Extra-ocular movement abnormalities I:
  - Unilateral: Single cranial nerve defects; Not usually within Brainstem.
    - CN6 lesion (Abducens): Ipsilat poor lateral gaze.

  Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
The Eyes Have It

- Extra-ocular movement abnormalities II:
- MS effects on lateral gaze:
  - Often *Bilateral*: Impairments of conjugate gaze
    - Localize to *within* the brainstem

**Midbrain (CN3)**
**Pons (CN6)**

Lateral Gaze Center; “Abducting Eye Leads”

The Eyes Have It

- Conjugate eye movement abnormalities caused by MS:
  - Really, only Three Brainstem Lateral Gaze Syndromes
    - 1) MLF lesion: **INO** (“tug of war”)
    - 2) B/L MLF lesions: **b/l INO**
      - midline – unlike strokes, demyelination doesn’t respect the midline
    - 3) MLF + PPRF: **1 ½ syndrome**
  - Tectum: Upgaze/convergence center
    - upbeat nystagmus (always pathological)

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
The Eyes Have It

• Single MLF lesion: **INO** (“tug of war”)

![Diagram](image)

The abducting eye’s nystagmus = “tugging” the opposite eye medially

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

The Eyes Have It

• Bilat (midline) MLF lesion: **B/L INO**

![Diagram](image)

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
The Eyes Have It

- Bilat (midline) MLF lesion: **B/L INO**

And...When a bilat INO results in lateral deviation (skew) of eyes at rest, The result is a Wall-Eyed Bilateral INO = **“WEBINO syndrome”**

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

VI. The Eyes Have It

- Single MLF + PPRF lesion

  **“1 ½ syndrome”**

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
VI. The Eyes Have It

- Single MLF + PPRF lesion

“1 ½ syndrome”

And... Nucleus of CN6 and the PPRF right near the nucleus of CN7 in pons:
When lesion produces a 1 ½ syndrome + a 7th palsy = “8 ½ syndrome”

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Clinical Neuroanatomy
Part 2: Brainstem & Spinal Cord
Introduction
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Objectives
• Identify the anatomical structures of the central nervous system in the brain stem and the spinal cord
• Develop a rational, systematic approach to localization in clinical neurology
• Recognize those areas of the Brain Stem and the spinal cord involved in the development of common symptoms of multiple sclerosis
The Brain Stem

- Midbrain: Vision, hearing, eye movement, and body movement
- Medulla: maintaining vital body functions, such as breathing and heart rate
- Pons: involved in motor control and sensory analysis

The Spinal Cord

- 3 kinds of neurons connect CNS to the body
  - Ascending sensory
  - Descending motor
  - Interneurons
- Motor - CNS to muscles and organs
- Sensory - sensory receptors to CNS
- Interneurons: Connections Within CNS
Spinal Cord Syndromes

- Partial transverse cord lesions
- Hemi section of the spinal cord
- Central spinal cord lesion
- Posterior column syndrome
- Anterior spinal syndrome
- Disseminated / Multiple Sclerosis Acute & Subacute: Degeneration of the spinal cord

Clinical Shortcuts

- **FACE** involvement Contralateral or Ipsilateral to weakness or numbness localize to BS or brain/cortex
- **Unilateral symptoms** – brain; **Bilateral symptoms** – spinal cord;
- **Signs of asymmetric UMN dysfunction**
  - if the lesion is above the decussation of the pyramids of the medulla-the UMN signs will be contralateral to the lesion
  - if the lesion is below the decussation – the UMN signs will be Ipsilateral to the lesion
Clinical Pearl: Cervical Spinal Cord

- The higher the spine lesion leading to below the level signs and symptoms
- C2-3 lesions innervate below and above the level of lesion / injury
- Ex. Lesion at C2-3 leads to numbness in the back of the head

Localization of S/S

- **At what level(s)?**
- cerebral hemispheric cortex/subcortical
- Brain stem: midbrain /pons /medulla
- cerebellum
- spinal cord
- nerve root
- peripheral nerve
- neuromuscular junction / muscle
Localization of S/S

- **What longitudinal system(s) is (are) involved?**
  - consciousness
  - language
  - vision
  - motor function
  - coordination
  - pain, temperature
  - proprioception, vibration

- **What is the lateralization?**
  - right?
  - left?
  - bilateral?

- **What is the course?**
  - acute?
  - subacute?
  - chronic?

- **Is the process . . .**
  - focal?
  - multifocal?
  - diffuse?
Types of Localization

• Single lesion theory: can all symptoms/signs be explained by one lesion?

• Types of Localization
  Focal, Multifocal, or Diffuse
  MS, at different points, can be all three. Exacerbations themselves can be focal or multifocal

Types of Localization

• Focal, Multifocal, or Diffuse
  Focal: Glioma
  Multifocal: Metastases
  Diffuse: HIV dementia
Approach to Localization

- 30 year old female
- Sudden onset
- Left sided weakness

Left sided weakness - Localization

- 1. Cortex
- 2. Internal capsule
- 3. Corona radiata
- 4. Brainstem
- 5. Spinal Cord
Right Eye deviates medially

- Right-eye deviates medially
- Lateral-Rectus muscle weakness
- Localization
  - Brain stem
    - EOM
    - Cranial Nerves
      - CN III
      - CN IV
      - CN VI

Localization of both?

- Right eye deviates medially
- Right Lateral Rectus muscle weakness
- Left sided weakness
- Localization
Brain Stem Localization

Ipsilateral CN VI

Contralateral Weakness

Brain stem

PONS

Localization Within the Brain Stem

Principle: Specific cranial nerve involvement guides localization

<table>
<thead>
<tr>
<th>Site</th>
<th>Signs and Symptoms</th>
</tr>
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<tbody>
<tr>
<td>Midbrain</td>
<td>Impaired vertical gaze</td>
</tr>
<tr>
<td></td>
<td>&gt;CN 3 palsy (plus contralateral ophthalmoplegia (INO))</td>
</tr>
<tr>
<td></td>
<td>&gt;CN 4 palsy</td>
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<tr>
<td></td>
<td>Contralateral motor signs (hemiparesis suggests Weber's syndrome, ataxia suggests Claude's syndrome)</td>
</tr>
<tr>
<td></td>
<td>Tremor or chorea suggests Benedikt's syndrome</td>
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<tr>
<td></td>
<td>Alterations in consciousness, perception or behavior (peduncular hallucinosis)</td>
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<tr>
<td>Site</td>
<td>Signs and Symptoms</td>
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<td>--------------</td>
<td>------------------------------------------------------------------------------------</td>
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<tr>
<td>Pons</td>
<td>Dysarthria and dysphagia</td>
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<tr>
<td></td>
<td>Contralateral hemiparesis or hemisensory loss</td>
</tr>
<tr>
<td></td>
<td>Ipsilateral facial nerve loss (CN5)</td>
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<tr>
<td></td>
<td>Ipsilateral gaze palsy (paramedian pontine reticular formation PPRF) or one and a half syndrome (PPRF and MLF)</td>
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<tr>
<td></td>
<td>Locked in syndrome (bilateral basis pontis with ocular bobbing)</td>
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<tr>
<td></td>
<td>Horizontal nystagmus (brachium pontis)</td>
</tr>
<tr>
<td></td>
<td>Ataxia</td>
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</table>

| Pontomedullary Junction | Vertigo (CN 8)                                                                 |
| Lateral Medulla (Wallenberg) | Dysarthria                                                                  |
|                          | Horizontal or vertical nystagmus                                               |
|                          | Contralateral hemisensory loss and hemiparesis                                 |
| Medulla                  | Ipsilateral Horner’s syndrome                                                   |
| Medial medulla           | Ipsilateral limb ataxia                                                        |
|                          | Ipsilateral face and contralateral body numbness                               |
|                          | Gait ataxia                                                                     |
|                          | Vertigo, dizziness (CN 8)                                                       |
|                          | Dysphagia (CN 9, 10, 12 palsy)                                                  |
|                          | Hemiplegia                                                                      |
|                          | Contralateral posterior column sensory loss                                     |
|                          | Ipsilateral tongue weakness (12)                                                |
Localization in the Spinal Cord

<table>
<thead>
<tr>
<th>Site</th>
<th>Signs and symptoms</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemicord</td>
<td>Ipsilateral hemiparesis</td>
<td>Penetrating trauma</td>
</tr>
<tr>
<td>(Brown</td>
<td>Contralateral spinothalamic sensory loss</td>
<td>Extrinsic cord compression</td>
</tr>
<tr>
<td>Sequard)</td>
<td>Ipsilateral dorsal column sensory loss; spinchter dysfunction</td>
<td></td>
</tr>
<tr>
<td>Anterior cord</td>
<td>Upper and lower motor paralysis</td>
<td>Anterior spinal artery</td>
</tr>
<tr>
<td></td>
<td>Spinothalamic sensory loss</td>
<td>infarction</td>
</tr>
<tr>
<td></td>
<td>Spinchter dysfunction</td>
<td>(often T4-T8)</td>
</tr>
<tr>
<td></td>
<td>Sparing of posterior column</td>
<td></td>
</tr>
<tr>
<td>Central cord</td>
<td>Paraparesis</td>
<td>Syringomyelia</td>
</tr>
<tr>
<td></td>
<td>Lower motor paralysis; wasting</td>
<td></td>
</tr>
<tr>
<td></td>
<td>paralisis; wasting</td>
<td></td>
</tr>
<tr>
<td></td>
<td>and fasciculations in arms</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Sensory loss in “shawl” distribution (cervical)</td>
<td></td>
</tr>
</tbody>
</table>

Posterior cord

|               | Proprioceptive and vibratory       | Vit B12 def                  |
|               | Segmental tingling and numbness    | Demyelination (ms)           |
|               | Sensation of constricting “bands”  | Extrinsic compression        |

Foramen magnum

|             | Spastic quadripareisis             | Tumor                        |
|             | Neck and pain stiffness            | (meningioma, chordoma)       |
|             | C2 to C4 and upper facial numbness | atlantoaxial                 |
|             | Ipsilateral Horner’s syndrome      | subluxation                  |
|             | Ipsilateral tongue and trapezius muscle |                          |
Conus Medullaris
- Lower sacral saddle
- Sensory loss (S2-S5)
- Sphincter dysfunction
- Impotence
- Back or rectal pain
- L5 & S1 motor deficits
  (ankle & foot weakness)

Cauda Equina
- Sphincter dysfunction
- Weakness or paraparesis
- Sensory loss in multiple bilateral dermatomes

- Intrinsic tumor
- Longitudinal spine lesion
- Extrinsic cord compression

Extrinsic tumor
- Spinal stenosis

Localization: Why bother?
- Offer a differential diagnosis
- Gives an indication of a potential cause
- Allows better selection of imaging & other diagnostic tests
- Directs for appropriate treatments
- Gives an indication of prognosis for example a brain stem episode in a male patient
Localization in MS

- Precise localization is crucial in MS
- Distinguishing between a true exacerbation and a pseudo-exacerbation (heat, fever, ‘symptom reactivation’)
- Distinguishing between physiologic and psychologic symptoms/signs
- Distinguishing between MS attacks and other problems in MS patients (strokes, disc disease, etc.)

Brain & Spinal Cord Localization Summary

- This process of localization in the Brain & the Spinal Cord is important in neurology because it enables us to narrow the differential diagnosis of a problem, leading to more effective, and efficient management of patients with neurological diseases!
Clinical Neuroanatomy
Overview for the MS clinician

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CMSC 2015

AN EXAMPLE OF LOCALIZATION IN NEUROLOGY

A preview of clinical localization...

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
AN EXAMPLE OF LOCALIZATION

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AN EXAMPLE OF LOCALIZATION IN NEUROLOGY
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  - Avenues: long tracts and pathways
    - Sensory: Pain/Temp vs. Proprioception/Vibration
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  - Streets: horizontal sections/exiting pathways
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    - Brainstem: exiting cranial nerves define level
    - Cord: exiting roots (dermatomes) define level
  - Neighborhood Signs: looking for dysfunction in “what’s nearby”

Approach to Localization

- Localization principles: Dividing up the map
  - Front to Back:
    - Anterior ➔ Motor;
    - Posterior ➔ Sensory
  - Left to Right:
    - Crossed Sides (Left Hemisphere ➔ Right body)
  - Bottom to Top
    - Lumbar ➔ Thoracic Spine ➔ Cervical Spine ➔ Brain
      - ?Leg involvement
      - ?Arm/Hand involvement
      - ?Facial involvement

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
The Funnel

- Below the cortex: descending/ascending tracts from the cortex on out
- The Funnel: why lesions in Brainstem/Spinal Cord cause more symptoms than in cerebrum

Why the Brainstem Matters

- In addition to being densely packed with avenues to and from the body...
- Brainstem Nuclei and Cranial Nerves are responsible for the most vital functions of the organism:
  - Vision
  - Respiration
  - Articulation
  - Speaking
  - Swallowing
Brainstem Syndromes

- The Brainstem = The base of The Funnel.
- Intermediary between Brain and Spinal Cord organization
  - Same Rules of organization apply
    - ie, Anterior = Motor; Posterior = Sensory
- Cranial Nerves define the level (the “streets”)

Brainstem Syndromes:

- “Crossed Signs”
- Three Main Brainstem Divisions, and Twelve Cranial Nerves.
  - nerves exit ipsilateral to their symptoms
- Top to Bottom, divided up:
  - First 4; Middle 4; Inferior 4:
    - Midbrain: CN3
    - Pons: CN5, CN6, CN7
    - Medulla: CN9, CN11, CN12

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Classic Brainstem Syndrome #1

- Man presents with double vision and left sided weakness.
  - On exam, his right eye is deviated down and out
  - His right eye will not move medially and ptosis is present
  - He is weak on the left side, arm and leg
  - He cannot walk or lift his left arm

R eye down & out with ptosis
(All planes of gaze)

The Avenue:

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ANTERIOR: Descending Motor
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The Street:

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CN III: Oculomotor Nerve
- Medial Gaze
- Ptosis
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Classic Brainstem Syndrome #1

- Man presents with double vision and left sided weakness.

"Problems Doing"

MIDBRAIN
Classic Brainstem Syndrome #1

- Man presents with double vision and left sided weakness.
  - Motor signs: Anterior
  - Cranial Nerve III: The Midbrain
  - Which side? The Right side.
    - Crossed Signs
    - Nerve is ipsilateral (right eye medial gaze), weakness is contralateral (left)

Classic Brainstem Syndrome #2

- Man presents with right facial weakness and left sided weakness.
  - On exam, his right nasolabial fold is flat
  - He cannot move the corner of his right mouth he is drooling from the right side of this mouth
  - His speech is slurred
  - He is weak on the left side, arm and leg
  - He cannot walk or lift his left arm
Man presents with right face and left body weakness.

- Motor signs: Anterior
- Cranial Nerve VII: The Pons
- Which side? The Right side.
  - Crossed Signs
  - Nerve is ipsilateral (right), weakness is contralateral (left body)

Left

Millard-Gubler Syndrome
Classic Brainstem Syndrome #3

- Woman presents with slurred speech, imbalance, and numbness

  - What do you suppose the ER would think of a stumbling, drooling patient with slurred speech and no focal weakness?

  - On exam, her speech is dysarthric and she’s drooling
  - Her gag reflex is diminished on the right
  - Her right face is numb to pain and temperature
  - Her left body is numb to pain and temperature
  - She is imbalanced and clumsy

The Avenue:

The Street:

"Problems Sensing"

POSTERIOR: Ascending Sensory

CNs IX, X, XI

-Dysarthria
Dysphagia
Gag reflex

MEDULLA
Classic Brainstem Syndrome #3

- Woman presents with slurred speech, imbalance, and numbness
  - Sensory signs: Posterior (no Motor findings!)
  - Cranial Nerves IX, X, XI: The Medulla
  - Which side? The Right side.
    - Crossed Signs: right facial numbness and depressed right gag
    - Nerves are ipsilateral (right), body numbness is contralateral (left)
    - Limb clumsiness is ataxia from the cerebellar connections

![Wallenberg Syndrome]

Classic Brainstem Syndrome #4

- Man collapses in the grocery store, unresponsive, gets intubated, brought to Emergency Dept.
  - On exam, he makes no attempt to speak or move
  - He follows no commands and doesn't track left or right with his eyes
  - His face is motionless
  - He does not withdraw any limb to painful stimuli
  - He does not improve.

  - Weeks later he is transferred to a long-term care facility, where a therapist notes that he seems to blink his left eye to command.
Classic Brainstem Syndrome #4

- Man is quadriplegic, aphonic, no horizontal eye movement; blinking intact.

  - Motor signs: Anterior
  - Cranial Nerve III still functions; everything below does not: The Pons
  - Which side? BOTH SIDES – Bilateral transection at Pons

  - “This neurological injury paralyzes all expression – by word or movement – yet leaves the patient in possession of full sentient consciousness.”
    - Plum and Posner, 1972

Locked-In Syndrome
Locked-In Syndrome

Brain
- Brainstem/Cerebellum \( \rightarrow \) nerves to the face
- Cervical Spinal Cord
- Brachial Plexus \( \rightarrow \) nerves to the arm
- Thoracic Spinal Cord
- Lumbosacral Nerve Roots
- Lumbosacral Plexus \( \rightarrow \) nerves to the leg

Paraplegia

Quadriplegia (Christopher Reeve)

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Locked-In Syndrome

Brain

- Brainstem/Cerebellum ➔ nerves to the face
- Cervical Spinal Cord
- Brachial Plexus ➔ nerves to the arm
- Thoracic Spinal Cord
- Lumbosacral Nerve Roots
- Lumbosacral Plexus ➔ nerves to the leg

Quadriplegia And Facial Paralysis With Aphonia

Locked-In Syndrome

Brain still fully conscious.

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Brainstem Syndromes

- Cranial Nerves NOT usually involved in MS: (red flags for other etiologies)
  - CN1: olfactory (Dementias, degenerative dz)
  - CN4: downgaze (Meningeal processes)
  - CN8: hearing usually spared
    - Mitochondrial d/o’s, Vasculitis, Susac’s
    - Vestibular system, however, is often affected (Middle Cerebellar Peduncle)

- Ataxia is the overall “posterior fossa” localizer
  - Intention tremor; Dysmetria; Rebound; Titubation
  - Cerebellum and its connections
  - The Cerebellum as shock absorber
  - Cerebellar homunculus: midline = trunk; lateral = limbs
  - Cerebellar hemispheres are “doublecrossed” so Ipsilateral to body (unlike everything else in head)
Brainstem Syndromes

An example of Localization:

- **Case:** 34 year old man with headache, vertigo, clumsiness, and unsteady vision
- **Eye findings:** Extraocular motility was abnormal. The patient had conjugate eye movements. There was a jerk nystagmus with the fast phase down-going. The nystagmus was more prominent in down and lateral gaze, and dampened with up-gaze. Downward pursuit was abnormal.
- **Fundus examination:** Normal disc, macula, vessels OU
- **Visual fields:** normal
- **Neurologic Exam:** The neurologic examination was notable for restricted range of motion of the neck, a diffuse increase in muscle tone, decreased proprioception of hands and feet bilaterally, dysmetria of the left arm, positive Romberg sign, ataxic gait, abnormal tandem gait, hyperreflexia with bilateral upgoing toes and ankle clonus.

Focal, Multifocal, or Diffuse?
- Can one lesion explain all the symptoms/signs?
- Where?
Brainstem Syndromes

An example of Localization:

The Cervico-Medullary Junction: A Great Pretender.
“Bottom of the Funnel” + Myriad Cranial Nerves + Cerebellar tracts
Mimics multifocal disease, including MS.

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

Spinal Cord Localization

Peripheral Sensory Distributions: Roots/Dermatomes and Nerves

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Cord Localization

- Tightly packed bundle of sensory and motor fibers: “No silent cord”

- Pattern recognition
  - Hallmark features of myelopathy:
    - Bilateral weakness/sensory loss
    - Bilateral UMN signs (Babinski, etc) and UMN weakness
      - Finger extensors, knee flexors, foot dorsiflexors
    - Bowel and bladder symptoms

- The “Street” is the sensory level at which the cord is affected

- Pattern recognition of 4 key cord syndromes…(to follow)

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

Cord Localization

Anterior/Posterior distinction (Motor vs Sensory) is largely preserved
- Spinal roots: Sensory enters posteriorly, Motor exits anteriorly
- Anterior Horn cells = Ipsilateral Motor
- Posterior Column = Ipsilateral Touch, vibration, proprioceptive
- Spino-thalamic = Contralateral pain & temp (& anterior)
  - “the rebel”

That rebellious spino-thalamic tract: Sensory in the front, and On the wrong side.

The posterior columns: Ipsilateral, ascending vibration and proprioception.

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Cord Localization

• Classic Cord Syndromes:
  • 1. Anterior Cord Syndrome
  • 2. Posterior Cord Syndrome
  • 3. Lateral Cord Syndrome (Brown-Sequard)
  • 4. Central Cord Syndrome

  • Loss of ascending pain/temp.
  • Loss of descending motor.
  • Preserved vibration and proprioception.

  • Classic etiology:
    • Anterior Spinal Artery infarction ("cord stroke")...also, post traumatic (whiplash, chiropractor, etc)

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Cord Localization

• **Classic Cord Syndromes:**
  • 2. Posterior Cord Syndrome
    • Preserved ascending pain/temperature.
    • Preserved descending motor.
    • Loss of vibration and proprioception.

• Classic etiologies:
  • Low B12, Syphilis, HIV myelopathy, Demyelinating dz
  Heavily myelinated pathways – JPS is fast; pain is slow

(One needs rapid streams of proprioceptive information in order to do anything.)

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

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Cord Localization

• **Classic Cord Syndromes:**
  • 2. Posterior Cord Syndrome

Thoracic cord (1 posterior column)  Cervical cord (2 posterior columns)

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Cord Localization

• Classic Cord Syndromes:
  • 3. Lateral Cord Syndrome
    (Brown-Sequard…one guy)
    • Loss of ipsilateral
      vibration and
      proprioception
    • Loss of ipsilateral motor
    • Loss of contralateral
      pain/temp
    • One leg is weak, the
      other one is numb (to
      pinprick)
  • Classic etiologies:
    • Trauma, tumor,
      compression from
dural space

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.

Cord Localization

• Classic Cord Syndromes:
  • 4. Central Cord Syndrome
    • Loss of bilateral
      crossing pain and
      temperature fibers
    • Loss of bilateral motor
    • “Cape-like” distribution
      of numbness
  • Classic etiologies:
    • Syringomyelia,
      intrinsic cord tumor,
demyelinating dz

Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.
Cord Localization

- **Classic Cord Syndromes:**
  - 4. Central Cord Syndrome

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And one new localization case...

- **The Food Critic Who Couldn’t Taste (and Six Other Cases of Dysgeusia in Multiple Sclerosis)**

  - 40 y/o M, a prominent food and wine critic, developed imbalance and left-sided incoordination walking through a food market. He became aware that he had lost taste sensation and developed numbness of the left tongue.
  
  - When eating, he would move food to the right side of his mouth “to get all of the taste information.”
  
  - He was ultimately diagnosed with a first attack of Multiple Sclerosis.
  
  - Over a few months his hemiageusia 90% resolved. He had a sensory myelopathy a year later and was formally diagnosed with MS.
The Food Critic Who Couldn’t Taste and Six Other Cases of Dysgeusia in Multiple Sclerosis
The Food Critic Who Couldn’t Taste and Six Other Cases of Dysgeusia in Multiple Sclerosis

• These cases support that taste information runs medially in the ipsilateral dorsolateral pontine tegmentum.
  • The lesions cluster in one small region of the SNT, a tract with a long course throughout the brainstem.
  • This location may be more likely to produce dysgeusia of which the patient is aware.

• Dysgeusia may be the initial presenting symptom of MS.
  • Its presence likely portends a brainstem attack, which is a known predictor of poor clinical outcome.
Thank you!

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Slides: Stephen Krieger, Mount Sinai School of Medicine. Images: Sourced from Google.