Neurologic Emergencies

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Localization

**Central nervous system**
- Brain
- Spinal cord

**Peripheral nervous system**
- Nerve root
- Plexus
- Peripheral nerve
- Neuromuscular junction
- Muscle

The most important step in neurologic localization is differentiating a *central nervous system* lesion from a *peripheral nervous system* lesion.
Localization

**Upper Motor Neuron**
- Pathway from pyramidal cells to spinal interneurons
  - Predominantly corticospinal and corticobulbar fiber tracts
  - Modulate action of cranial and spinal motor neurons

**Lower Motor Neuron**
- Pathway from fascicles of spinal motor neuron to effector organ
Localization

Upper Motor Neuron
- Mildly reduced bulk
- Increased tone (spasticity)
- Mild/moderate weakness
  - Flexors > extensors in UE
  - Extensors > flexors in LE
- Hyperreflexia; pathologic reflexes (Babinski, Hoffman)

Lower Motor Neuron
- Severely reduced bulk with fasciculations
- Reduced tone (flaccidity)
- Severe weakness
- Hyporeflexia

Grading muscle power
5: Full strength
4: Movement against some resistance
3: Movement against gravity only
2: Movement with gravity eliminated
1: Flicker or trace contraction
0: No contraction

Grading reflexes
4: Hyper with pathologic responses or clonus
3: Hyperactive/spread
2: Normal
1: Diminished
0: Absent
<table>
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<tr>
<th></th>
<th>Neuropathy</th>
<th>Myopathy</th>
<th>Myelopathy</th>
<th>NMJ disorder</th>
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<tr>
<td>Weakness</td>
<td>Distal &gt; proximal</td>
<td>Proximal &gt; distal</td>
<td>Below level of lesion</td>
<td>Fluctuating/ muscle fatigue</td>
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<tr>
<td>Deep tendon reflexes</td>
<td>Severe reduction/ early loss</td>
<td>Mild reduction/ late loss</td>
<td>Increased</td>
<td>Normal or mildly reduced</td>
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<tr>
<td>Sensory</td>
<td>Distal/ ascending</td>
<td>Preserved</td>
<td>Sensory level</td>
<td>Preserved</td>
</tr>
</tbody>
</table>
Localization

• Spinal levels

It's easy to remember... 1/2, 3/4, 5/6, 7/8

ALWAYS ABNORMAL:
• Asymmetric reflexes
• Pathologic responses (Babinski, clonus = CNS)
• Absent* reflexes (PNS)

*Hypoactive reflexes, especially if symmetric are not necessarily pathologic
Case 1 - TM

- Young right-handed patient who presents with weakness that started in the left foot and now involves the left side to the hand and difficulty voiding
- Patient had flu two weeks ago; otherwise no significant PMHx
- Neuro Exam:
  - EOMI, no facial asymmetry, tongue midline
  - Motor 5/5 right UE/LE c normal tone, 4/5 left UE & 3/5 LE (flexor > extensor; proximal > distal) c increased tone
  - Sensory diminished to LT/PP/temp in left LE and in distal UE; preserved sensation at shoulders and bilateral face
  - DTRs 3/4 left LE (spread of reflexes)
  - Plantar response flexor right and extensor left
What is the diagnosis:

a) Pontine infarct
b) Guillain-Barré syndrome
c) Myasthenia gravis
d) Partial cervical myelitis
e) Todd’s paralysis
f) Conversion disorder
Case 1 – Transverse Myelitis

• Synopsis
  – Unilateral ascending weakness of proximal > distal muscle with hypertonicity, sensory loss to modalities of anteriolateral fiber tract, hyperreflexia and extensor plantar response

• Localization
  – Lateral cervical spinal cord

• DDx:
  – Infection – WNV, HSV, VZV, borrelia/brucella (less likely)
  – Inflammatory – sarcoidosis, ADEM, NMO, multiple sclerosis (CIS)
  – Vascular – infarct (less likely), AV fistula
  – Mechanical – meningioma, ependymoma, dural hemorrhage, cord compression
Case 1 – Transverse Myelitis

• Diagnostic Tests
  – Spinal cord imaging with contrast
  – CSF analysis – include viral tests, IgG synthesis, IgG index, oligoclonal bands, if indicated vasculitis w/u

• Management
  – Cord compression – dexamethasone 16 mg divided daily dose tapered over 10-14 days*
    • Acutely typically use 24 mg dexamethasone divided daily IV
    • Neurosurgery consultation for decompression
  – Inflammatory – dexamethasone 100-200 mg or methylprednisolone 1000 mg IV over 3-5 days
    • If CSF analysis shows intrathecal IgG synthesis then brain MRI

Case 2

- Young right-handed patient who presents with weakness that started in the left foot and over the past 1 hour involves the left side to the hand

- Neuro Exam:
  - EOMI, no facial asymmetry, tongue midline
  - Motor 5/5 right UE/LE c normal tone, 4/5 left UE & 3/5 LE (flexor > extensor; proximal > distal) c increased tone
  - Sensory intact to all modalities
  - DTRs 3/4 left LE (spread of reflexes)
  - Plantar response flexor right and extensor left
What is the diagnosis:

a) Infarct in posterior internal capsule
b) Cortical stroke
c) Myasthenia gravis
d) Guillain-Barré syndrome
e) Todd’s paralysis
f) Conversion disorder
Case 2 – Subcortical Infarct

• Synopsis
  – Unilateral weakness of proximal > distal muscle (pure motor) with hypertonicity, hyperreflexia and extensor plantar response

• Localization
  – Right posterior internal capsule

• DDx:
  – Vascular – infarct vs hemorrhage
  – Inflammatory/infectious – demyelinating disease, abscess
  – Mechanical – tumor
Case 2 – Subcortical Infarct

- Caused by occlusion of small penetrating branches of cerebral arteries
- Chronic HTN > DM > emboli

<table>
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<tr>
<th>Syndrome</th>
<th>Presentation</th>
<th>Localization</th>
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<tr>
<td>Pure Motor</td>
<td>Face, arm, leg</td>
<td>Internal capsule</td>
</tr>
<tr>
<td>Pure Sensory</td>
<td>Face, arm, leg</td>
<td>Thalamus</td>
</tr>
<tr>
<td>Sensorimotor</td>
<td>Face, arm, leg</td>
<td>Thalamocapsular</td>
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Case 3

• Young right-handed patient who presents with weakness that started in the right arm and “confusion”
  – Global aphasia (expressive and receptive component)

• Neuro Exam:
  – Gaze preference to left, right lower facial droop, tongue midline
  – Motor 1/5 right UE/LE (flexor > extensor; proximal > distal) c increased tone
  – Fails to react to painful stimulus right
  – DTRs 3/4 right UE/LE (spread of reflexes)
  – Plantar response extensor right and flexor left
Case 3 – Cortical Infarct

• Synopsis
  – Unilateral weakness of proximal > distal muscle and hemispheric sensory loss with hypertonicity, hyperreflexia, extensor plantar response and global aphasia

• Localization
  – Left frontal/parietal cortex

• DDx:
  – Vascular – infarct vs hemorrhage
  – Epileptic
  – Inflammatory/infectious – demyelinating disease, abscess
  – Mechanical – tumor
Case 3 - Would you give tPA?
Case 2/3 – Non-hemorrhagic Infarct

• Diagnostic Tests
  – Brain imaging (CT scan initially); if not hemorrhage or HT then consider tPA
    • MRI brain without contrast with DWI
    • If large vessel suspected then CTA and TTE or TEE to evaluate for proximal thrombus in atria or aorta

1. NINDS tPA Stroke Study, 2. European Cooperative Acute Stroke Study III


**tPA**

- **Management**
  - A-B-C’s
    - NPO, intubate for inadequate airway, ventilate if needed
    - Correct hypotension, rule out acute MI or arrhythmia (a-fib)
  - Rule out hypoglycemia - glucose < 50 mg/dl
  - BP maintained < 185/110
    - Maintain perfusion with target SBP 130’s – 140’s
  - IV tPA – 0.9 mg/kg (90 mg maximum dose)
    - 10% given as IV push over the first 1 minute with remainder administered over the next 60 minutes
    - Only if stroke ≤ 4.5 hours$^{1,2}$
    - Especially with large-vessel ischemia, must r/p CT brain within 24 hours to access for HT

- **Absolute contraindications to tPA**
  - Stroke within past 3 months
  - Major surgery or trauma within 14 days
  - GI hemorrhage within 21 days
  - Arterial puncture in non-compressible site within 7 days
  - History of ICH

- **Relative contraindications to tPA**
  - Neuro deficit (NIHSS score 5 to 22) must not be rapidly improving (TIA) or post-ictal (seizure at presentation)
  - MI in past 3 months
  - pregnancy

- Must have normal PTT, PT ≤ 15 sec, platelets ≥ 100,000
Hemorrhagic Transformation
Case 4

• Young right-handed patient who presents with left leg weakness that now involves the hand with associated sensory loss. His is afebrile, pulse 88, respirations shallow with rate of 18 bpm and BP on admission of 210/130.

• Neuro Exam:
  – Poorly responsive, pupils small but reactive bilaterally, lower left face droop, tongue midline
  – Motor 3/5 left LE > UE (flexor > extensor; proximal > distal) c increased tone
  – Sensory loss to all modalities left side
  – DTRs 3/4 left UE/LE (spread of reflexes)
  – Plantar response extensor left and flexor right

• To what structure does the hemisensory loss localize?

• What would you like to do next?
Case 4 – Thalamic ICH

- Over the next hour, he is obtunded with respiratory rate at 16 bpm and his right pupil is dilated and poorly responsive
Intracranial hemorrhage

- Approximately 10% of all strokes
- Most common cause hypertension
  - Small penetrating vessels
  - Thalamus, basal ganglia, cerebellum
- Lobar hemorrhage: consider anticoagulation, amyloid angiopathy, AVMs, cavernous malformations, metastatic or primary tumors
- Neurologic deterioration primarily due to hematoma expansion and worsening cerebral edema
- Management:
  - Gradually reduce BP to maintain MAP <150 mmHg
  - Maintain airway/treat hypoxia, cardiac monitoring, avoid hypotonic solutions
  - Treat glucose disorders, treat seizure
Increased intracranial pressure

• General medical treatment of increased ICP:
  – Intubate
  – ICP monitoring
  – Hyperventilation ($pCO_2 < 33$ mm) – vasoconstriction with reduction of blood volume; aggressive hyperventilation may cause worse outcome
  – Hyperosmolar therapy: mannitol 20% (0.25 gm/kg q6 hrs if $S_{osm} < 310$)
    • Temporizing measure only

• Specific treatment of increased ICP:
  – CSF drainage
  – Surgical evacuation of hematoma
  – Craniotomy
  – Tumor, encephalitis, abscess (vasogenic edema): dexamethasone 4 mg IV q6 hrs
Case 5

- Young right-handed patient who presents with worsening weakness that started in the feet and now involves the hands with associated sensory loss
- Neuro Exam:
  - EOMI, no facial asymmetry, tongue midline
  - Motor 5/5 bilateral proximal UEs, 4/5 bilateral distal UEs (flexor = extensor) c normal tone; 2/5 bilateral proximal/distal LEs (flexor = extensor) c diminished tone
  - Diminished sensation of all modalities in legs but intact in arms
  - DTRs 0/4 patella and ankles bilaterally, 1/4 in the UEs
  - Plantar response flexor bilaterally
What is the diagnosis:

a) Pontine infarct
b) Guillain-Barré syndrome
c) Myasthenia gravis
d) Partial cervical myelitis
e) Todd’s paralysis
f) Conversion disorder
Guillain-Barre Syndrome

• CSF: cytoalbuminological dissociation (elevated protein with few or no mononuclear cells)
  • May be normal in the first week
  • If WBC count >10 consider infectious/inflammatory

• Electromyography/nerve conduction study
  – May be normal early
    • Reduced nerve conduction velocities
    • Conduction block
    • Prolonged F-waves

• Antiganglioside antibodies
  • GM1 Abs (correlate with C. jejuni infection)
  • GQ1b associated with C. Miller Fisher variant (ataxia, areflexia & ophthalmoparesis)
Guillain-Barre Syndrome

**Acute Inflammatory Demyelinating Polyneuropathy (AIDP)**
- Typically follows an infectious process (2/3)
  - C. jejuni, CMV, EBV, M. pneumoniae
- Presents with ascending numbness/tingling, can be painful
- Weakness typically follows sensory disturbances
- *Areflexia – this might be delayed*
- Autonomic dysfunction
  - Labile BP, arrhythmia
  - Bowel and bladder function typically spared
- Symptoms should not proceed >8 weeks
  - 98% achieve “plateau phase” by 4 weeks
  - Duration of “plateau” 12 days

**Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)**
- Typically has insidiously slow process (>60%) though some will have relapsing presentation (33%) with partial or complete recovery between events
- Presents with proximal & distal weakness (proximal = distal at times)
  - Motor predominant
- Autonomic dysfunction
  - Labile BP, arrhythmia
  - Bowel/bladder can be involved
- Minimum 8 weeks duration to consider CIDP
**Treatment - AIDP**

- Telemetry, respiratory parameters, ICU monitoring for dysautonomia and respiratory compromise
- IVIG (0.4g/kg/day for 5 days) versus plasma exchange
  - Ease of administration, fewer complications, preferred in hemodynamically unstable patients
  - Should be started within 2 weeks
  - No difference b/w IVIG or TPE*; should continue for additional course if no response c poor prognosis if no response
- Corticosteroids have not been shown to be beneficial
- Intubation criteria:
  - VC <15 mL/kg or NIF worse than -25 cm water
  - PO$_2$<70 mmHg
  - Oropharyngeal weakness, weak cough, suspected aspiration

*Hughes, RA, Pritchard J, Hadden RD, Cochrane Database Syst Rev, 2013*
Case 6 - MG

- Young right-handed patient who presents with generalized weakness, ptosis and SOB. One week ago patient had URI and treated with 3 days of azithromycin.

- Neuro Exam:
  - EOMI, bilateral ptosis (worsens with sustained upgaze), no facial asymmetry, tongue midline but increased salivation noted, dysarthric
  - Motor 3-/5 bilateral UE > LE (flexor = extensor; distal > proximal) c normal tone; weakness worsens with repetition
  - Sensory intact to all modalities
  - DTRs 1/4 throughout
  - Plantar response flexor bilaterally
What is the diagnosis:

a) Pontine infarct
b) Guillain-Barré syndrome
c) Myasthenia gravis
d) Partial cervical myelitis
e) Todd’s paralysis
f) Conversion disorder
MG – Acute Therapy

• Respiratory parameters
  – 30% of pts develop respiratory muscle weakness and crisis occurs in 15-20%

• Telemetry: 14% of pts in myasthenic crisis have some degree of arrhythmias

• Intubation criteria
  – VC < 15 mL/kg
  – Stop anti-cholinesterase medication (causes excessive bronchial secretions and diarrhea)

• PE (improvement in 75%), IVIG
  – Steroids may worsen weakness in acute setting
Myasthenia gravis

- Ice pack test: improves ptosis in MG > 80%
- Repetitive stimulation or single fiber EMG
- Autoimmune work up
- AchR Ab
  - 85% c global MG
  - 50-60% c ocular MG
  - Others: anti-MuSK, anti-striated muscle, anti-titin/Ryanodine receptor (RyR)
- Chest CT – evaluate for thymoma
  - Thymectomy may improve chances of remission
  - May be less effective in ocular myasthenia and those >60
  - Thymoma associated with anti-titin/RyR
- Trigger or worsen exacerbations
  - Surgery
  - Immunization
  - Emotional stress
  - Menstruation
  - Intercurrent illness (eg, viral infection)
  - Medication (eg, aminoglycosides, clindamycin, macrolides, quinolones, chloroquine, lithium, phenytoin, beta-blockers, procainamide, statins)
Case 7

• Young patient who was found poorly responsive with only minimal and occasional spontaneous limb movements. The last time patient was seen normal was one day prior to presentation. The patient has infrequent generalized myoclonic movements of the limbs that last about 30 seconds and known h/o epilepsy

• Neuro Exam:
  – Obtunded, no facial asymmetry, tongue midline, Doll’s present
  – No spontaneous motor activity but patient localizes to pain throughout c normal tone throughout
  – DTRs 3/4 throughout with diffuse spread of reflexes ?
  – Plantar response flexor bilaterally
Encephalopathy/Status epilepticus

• Benzodiazepine as temporizing measure
  – 0.1 mg/kg lorazepam (max dose 4 mg)
  – Generalized GABA activity to inhibit excess glutaminergic activity

• Load with AED (ideally use home medication)
  – Fosphenytoin (20 mg/kg), valproate (40 mg/kg), levetiracetam (2000 -4000 mg), lacosamide (400 mg)

• Emergent EEG
  – Evaluate for non-convulsive status epilepticus
    • In single prospective study, between 14-34% patients had either non-convulsive status or intermittent ictal discharges after convulsive status episode
      – Add second AED if necessary

• CSF evaluation if no known h/o epilepsy or no cause for breakthrough seizure in person with known history
  – Evaluation for virus/bacteria
  – Evaluation for inflammatory process