Life Threatening Weakness

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Objectives

• Recognize life threatening weakness that can be confusing to diagnose
• Differentiate between central and peripheral weakness
Disclosures

• I do not have any relevant financial relationships with any commercial interests
• I developed this lecture without influence (financial/intellectual) from outside parties
• My involvement as a committee member is without influence (financial/intellectual) from outside parties.

Case #1

• 32 y/o male
• Weakness in his arms and legs that has been progressing over last 2 days
• PMHx: recent hospitalization for pneumonia
• PE: breathing and speech are normal; CN intact; weakness more distal than proximal (UE normal); areflexia at lower extremities but decreased at upper extremities
• What test I will need to do?
Case #2

- 4 y/o female that her legs and arms have become progressively weaker over the past several hours
- Now presents with paresthesia and a unsteady gait
- Examination reveals ataxia and areflexia with more distal than proximal weakness
- Vital capacity is reduced
- What will be?

Case #3

- Female 30 y/o presents with intermittent diplopia and ptosis for two weeks
- Symptoms get worst after chewing food or reading but improve after sleeping
- Trouble smiling
- Weak cough
- No respiratory complaints
- Extremities and reflexes normal
- What is it?
Case #4

• A family comes in with presentations of diplopia, dysarthria, dysphagia. One has sore throat, another has difficulty lifting his head. Two have dilated, unreactive pupils. Gag reflexes are diminished or absent.
• CN’s show multiple deficits. One has neck weakness
• All have normal sensation and are awake

Case #5

• Female 24 y/o presents with weakness in her arms for 4 hours
• She reported similar episodes in the past due to “kidney problem”
• Old chart revealed h/o RTA
• K= 1.8
Types of Weakness

• Dr. Google is excellent to get some diagrams and pictures from…
Characteristics

- Rapidity of onset
- Symmetry
- Progression
- Pattern
  - Distal vs. proximal
  - Ascending vs. descending
Types of Weakness

• Localized
• General
• Fatigue
• Breathlessness

• Paralysis
• Distal vs. proximal
• Gait change
• Grip strength

Weakness: Are the symptoms bilateral or unilateral?

• Unilateral
  • Stroke
  • Intracranial mass
  • Brown-Sequard Syndrome
  • Infectious
  • Hypoglycemia
  • Postictal Todd paresis
  • Hemiplegic migraine
  • Radiculopathy or peripheral nerve entrapment syndrome
Weakness: Are the symptoms bilateral or unilateral?

• Bilateral
  • Multiple sclerosis
  • Dysfunction of the spinal cord (myelopathy)
    • Compression
    • Infarction
    • Infection
    • Transverse myelitis
  • Usually will have UMN findings, a sensory level, and bladder dysfunction

Immediate Life-Threatening Causes

• Stroke syndromes
• Aortic dissection
• CNS infections
• Spinal cord syndromes
• Seizure disorders
• Envenomning (e.g. snake)
• Acute toxicity e.g. organophosphates, heavy metals
• Electrolyte/ metabolic disorders
Weakness

• Is the weakness is central or peripheral?
• Cortex (usually unilateral)
  • Stroke
    • Ischemic
    • Hemorrhagic
  • Infection (abscess)
  • Space occupying lesion

• UMN (usually bilateral)
  • PML
    • Progressive multifocal leukoencephalopathy
  • Demyelinating disease
    • MS
    • Transverse myelitis
• UMN (usually bilateral)
  • Trauma
  • Spinal stenosis
  • Spinal epidural abscess

Central

• Upper motor neuron findings (hyperreflexia, Babinski, spasticity)
• Dysarthria, ataxia, cortical signs (aphasia, neglect), or hemiparesis
• Anterior horn axon (usually mixed)
  • Polio
  • ALS
  • West Nile virus

LMN:

DANG THE RAPIST B

• DM
• Alcohol
• Nutrition
  • B12 def
• GBS
• Trauma
  • Distal to injury
  • Radiculopathy
• Hereditary
• Environmental
  • Lead

• Remote CA
• Autoimmune
• Porphyria
• Inflammatory
• Syphilis
• TB
• Botulism
Peripheral

• Lower motor neuron findings (hyporeflexia, absent Babinski, decreased muscle tone)

Peripheral

• Patients with neuropathy:
  • Presenting with weakness and sensory loss will be greater distally than proximally
  • Presence of pain is variable, but usually is distal
  • Reflexes are either decreased or none
Neuropathies

- GBS
- MS
- Tick paralysis
- Diphtheria
- Critical illness polyneuropathy
- Acute intermittent porphyria
- Toxin (arsenic, fugu---pufferfish in Japan)
- Snake venom (eastern coral snake)
- Medication (cisplatin)

- Lambert-Eaton
  - Ca channel receptor Ab
- Myasthenia gravis
  - Acetylch receptor Ab
Peripheral

- Patients with neuromuscular junction disorders
  - Weakness fluctuates, while sensory loss and pain are absent
  - Reflexes are normal

Neuromuscular junction disease

- MG
- Botulism
- Black widow spider bite
- Organophosphate poisoning
- Lambert-Eaton myasthenic syndrome
Peripheral

• Patients with myopathy
  • Weakness is usually worse in the proximal muscles and is accompanied by pain.
  • Sensation is unaffected and reflexes are normal
Myopathy

- Periodic paralysis
- Critical illness myopathy
- Electrolyte disorders (Ca$^{+2}$, Mg$^{+2}$, K$^{+}$)
- Trichinosis
- Polymyositis
- Dermatomyositis

Peripheral vs Central

- Weakness that involves the muscles of respiration is usually due to disorders of the peripheral nerves affecting the nerve, neuromuscular junction, or muscle
Case #1

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- Weakness in his arms and legs that has been progressing over last 2 days
- PMHx: recent hospitalization for pneumonia
- PE: breathing and speech are normal; CN intact; weakness more distal than proximal (UE normal); areflexia at lower extremities but decreased at upper extremities
- What test I will need to do?
- LP to r/o
- GBS

GBS

- Autoimmune (most of the cases) attack on peripheral myelin (acute inflammatory demyelinating polyradiculoneuropathy)
- Most common cause of non-traumatic acute paralysis
- 2/3 of patients have had a viral illness (CMV, EBV, HSV, HIV) or bacterial infection (Mycoplasma, Campylobacter jejuni-most common cause)
GBS

• Influenza vaccine risk: 1.7/million (1/6th overall GBS rate)

• Possible OPV or Menactra meningococcal vaccine link

• Symptoms peak in 4 weeks or earlier after URI or GI illness

• Ascending paralysis, symmetrical weakness of the lower legs first, then the thighs, and finally the arms; no reflexes

vs Miller-Fisher syndrome

• Triad:
  • Partial or complete ophthalmoplegia
  • Severe ataxia
  • Areflexia is a less common variant

• Acute onset

• Descending
Case #2

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• Now presents with paresthesia and a unsteady gait
• Examination reveals ataxia and areflexia with more distal than proximal weakness
• Vital capacity is reduced
• What will be
  • GBS?
Tick Paralysis

- Clinical findings are similar to GBS
- Female children are most common (less weight?) and longer hair
- Treatment:
  - Remove tick (Daa!!)
  - Symptoms will improve within hours

Case #3

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Myasthenia Gravis

• Autoimmune disorder in which antibodies are directed at acetylcholine receptors in the neuromuscular junction
  • The reduced receptors mean decreased transmission

• Is the most common disease of the neuromuscular junction

• Women are affected as twice as men

Myasthenia Gravis

• Ocular symptoms
  • Ptosis and extraocular muscle weakness are present in 50% of cases
  • Half of those will develop general weakness

• 25% will have bulbar symptoms
  • Difficulty chewing or swallowing or slurred speech

• Improves with rest
vs Lambert-Eaton myasthenic syndrome

• Rare condition

• Weakness results from an abnormality of acetylcholine (ACh) release at the neuromuscular junction

• Results from an autoimmune attack against voltage-gated calcium channels (VGCC) on the presynaptic motor nerve terminal

vs Lambert-Eaton myasthenic syndrome

• Weakness increased at proximal muscles

• Symptoms improve with increased use due to increased released of calcium (over-coming the antibodies) stimulating the pre-synaptic release of acetylcholine

• Associated with cancer
Case #4

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Botulism

• Caused by an exotoxin of the anaerobic bacillus, *Clostridium botulinum*, which results in irreversible blockade of synaptic vesicle release

• Infantile botulism is the most common form
  • Due to intestinal colonization of ingested bacteria
  • Honey is one of the source; canned foods; wound botulism (S/Q skin poppers)
Botulism

• Pathology
  • Toxins binds to presynaptic terminals of peripheral cholinergic synapses
  • Within the cell, it lyses proteins responsible for Ach release
  • Symptoms only resolve when the axon makes another terminal

Botulism

• Classic presentations
  • Symmetric
  • Descending paralysis
  • Involves cranial, spinal, and peripheral nerves
  • Alert
  • No sensory changes
Case #5

- Female 24 y/o presents with weakness in her arms for 4 hours
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Hypokalemic Paralysis

- Localized or generalized weakness
- Reduction or loss of deep tendon reflexes
- Short duration of symptoms followed by complete recovery
- Attack begins proximally and spread distally (bulbar and diaphragm are spared)
Periodic Paralysis

- It is important to distinguish between periodic paralysis and hypokalemic paralysis due to K wasting diseases because treatment depends on the etiology
- K can shorten acute attacks of both but, it is more important in patients with K wasting
- Rebound hyperkalemia occurs in approximately half of the patient with HPP as K abruptly returns to the extracellular compartment

To recap

- Do a complete history and physical
- It may tell you if:
  - Central vs peripheral
  - Symmetrical or asymmetrical
    - Symmetrical usually peripheral
    - Asymmetrical usually central
  - Rapid or gradual onset
    - Both can be life threatening
  - Ascending or descending
- Sometimes, you can treat and “fix” immediately
YES!!

YOU'RE STILL AWAKE! ANY QUESTIONS?