Peripheral Neuropathy for the Internist

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Objectives

- What is peripheral neuropathy?
- Approach to diagnosis
- Length dependent
- GBS/CIDP
- Mononeuropathy
- Hereditary Neuropathy
- Motor neuron disease
Peripheral Nerves

- Sensation
- Motor function
- Autonomic function
  - Heart rate
  - Breathing
  - GI functions
  - Sexual functions
Parasympathetic

- Stimulates flow of saliva
- Slows heartbeat
- Constricts bronchi
- Stimulates peristalsis and secretion
- Stimulates release of bile
- Contracts bladder

Sympathetic

- Dilates pupil
- Inhibits flow of saliva
- Accelerates heartbeat
- Dilates bronchi
- Inhibits peristalsis and secretion
- Conversion of glycogen to glucose
- Secretion of adrenaline and noradrenaline
- Inhibits bladder contraction
Peripheral Neuropathy

- Incidence: 20 million people in the US
- Major types
  - Focal vs. generalized
  - Motor vs. sensory
  - Painful vs. nonpainful
  - Acquired vs. hereditary
  - Axonal vs. demyelinating
Nerve conduction measured

Electromyogram

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Case 1

- A 60 year old male
- Hypertension and diabetes
- 1 year history tingling of toes
- Exam
  - Weakness of toe dorsiflexion
  - Decreased sensation
  - Abnormal reflexes
Length Dependent symmetric sensorimotor axonal neuropathy

- Most common type of polyneuropathy
- Length dependent- stocking/ glove distribution of sensory loss
- Pain is usually a prominent complaint
- Most common cause is diabetes
- Next is idiopathic
- Also associated with uremia, hepatic failure, alcohol abuse, drugs (chemo), vitamin deficiencies
- Laboratory studies
- Treatment
- Red flags
  - Motor > sensory
  - Rapidly progressive
  - Not length dependent
Case 2

- 34 year old female
- no significant past medical history
- 3 days of progressive weakness and back pain. 1 weeks ago she had a GI illness with diarrhea.

Exam
- 4 extremity weakness
- areflexia
- mild sensory loss at the toes
- Some shortness of breath
Gullain-Barre Syndrome (Acute Inflammatory Demyelinating Polyneuropathy)

- Is a demyelinating polyneuropathy
- Is an acute disease and by definition the nadir is by 6 weeks
- Progressive 4 extremity and bulbar weakness and little sensory complaints
- Can involve the respiratory muscles (can happen acutely)
- Also affects the autonomic nerves
- Pathophysiology
Gullain-Barre Syndrome

- Diagnosis
- EMG/NCS
- *campylobacter jejuni* and anti-GQ1B antibodies (need percentage)
- Treatment
- Recovery
Case 3

- 50 year old female
- 3-4 months of progressive arm and leg weakness and numbness
- Exam
  - weakness of the proximal and distal muscles
  - Areflexia
  - Sensation abnormal
Case 3

- EMG/NCS show multifocal demyelination of motor and sensory nerves
- Lumbar Puncture
  - No cells, increased protein
Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP)

- Usually presents over 8 weeks
- Usually symmetric and involves both motor and sensory fibers
- Motor > sensory and large fiber > small fiber involvement
- CSF protein is usually elevated with little or few WBCs
  - If CSF WBC is elevated should suspect Lyme, HIV, lymphoma, leukemia, or sarcoidosis
- Monoclonal gammopathy is associated with CIDP
CIDP

- Treatment
  - IVIG
  - Prednisone
  - Immunosuppressants
    - Azathioprine
    - Mycophenylate
Case 4

- 35 year old female
- 6 months of R>L hand numbness and some weakness when holding a pen in her right hand
- Exam
  - abnormal sensation on the palmar surface of digits 1-4 on both hands
  - weakness on thumb abduction on the right.
Carpal tunnel syndrome

- Risk factors
  - repetitive flexion of the hand (typing), use of vibrating tools, hypothyroidism, acromegaly, RA, amyloidosis
- Must be differentiated from a C8/T1 radiculopathy (hand weakness) or a C6 radiculopathy resulting in pain and numbness over D1 and 2.
- Diagnosis clinically and can be confirmed by EMG/NCS
- Treatment
  - Conservative measures include wrist splints
  - Can undergo carpal tunnel release
Median nerve is compressed at the wrist, resulting in numbness or pain.

- Compressed median nerve
- Carpal ligament
- Median nerve
- Thenar atrophy
- Normal thenar eminence
Mononeuropathy

- Other common mononeuropathies
  - Ulnar neuropathy at the elbow
  - Peroneal neuropathy at the fibular head
Case 5

- 25 year old female
- 10 year history of “not walking right.”
  - Diagnosed with hammartoes
  - She also noticed a gradual progression of a lack of sensation in her feet but denies pain.
- Exam
  - pes cavus, hammartoes
  - weakness of toe dorsi and plantar flexion and ankle dorsiflexion weakness
  - mild weakness of finger abduction.
  - decreased PP and temperature sensation up to her knees
  - catches her toes on the carpet
Hereditary Motor and Sensory Neuropathy (Charcot Marie Tooth)

- Inherited neuropathies
- Classified based on whether they are demyelinating, axonal, or mixed.
- Dominant, recessive, and X-linked have been described
- Genetic tests are available for most of the demelinating forms but only a few of the axonal neuropathies
- No treatment- just symptomatic with braces and PT/OT
Case 6

- 67 year old male
- 6 months of progressive left arm weakness and difficulty swallowing.
- Exam
  - spastic dysarthria
  - weakness and atrophy of the left hand
  - Fasciculations are noted in the left arm and left leg
  - Reflexes are brisk in the left arm and there is a positive jaw jerk
Amyotrophic Lateral Sclerosis

- Neurodegenerative disorder involving the upper and lower motor neurons
- Usually presents between age 30-60
- Most cases are sporadic but in about 15-20% of cases it is considered familial
- Cause is unknown
- The only drug that has been approved is riluzole
  - The initial study showed that it prolongs survival (life or time to tracheotomy) by about 3 months
  - Works best in patients with bulbar onset disease
ALS

- Presentation
  - painless focal weakness in a distal extremity
  - On exam there is a combination of upper and lower motor signs
  - The bulbar muscles are involved using impairing speech, swallowing, and breathing
  - Usual disease course is about 3-5 years with death secondary to respiratory failure
  - There is no sensory involvement but pain can be a feature especially
    - Spasticity
    - Frozen joints
    - Contractures
ALS

- Diagnosis
  - Clinical exam
    - Need to see evidence of upper and lower motor neuron involvement in at least 3 regions (bulbar, cervical, thoracic, and lumbar)
    - Can use EMG to look for evidence of lower motor neuron involvement if this is not evident on exam
    - Exclude other possible causes
ALS

• Management
  • Gold Standard- multidisciplinary clinics
  • Follow breathing and weight
  • Support for families
  • End of life care