Myelopathies and central demyelinating diseases

Didactic Session 6-Summary
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Myelopathy = Spinal cord

- Signs of spinal cord involvement
  - Symmetric muscle weakness
    - Paraparesis
    - Quadriplegia
  - Sensory Level
    - Sharp demarcation on the trunk
    - May be to one or more than one modality
  - Sphincter dysfunction
- Myelopathies are emergencies until proven otherwise
Beware of spinal shock!

- Upper motor neurons do not die at once
  - They use their reserve of inhibitory neurotransmitters
  - Spasticity and increased reflexes may not occur until days to up to 3 weeks later
- Initial phase of spinal shock
  - Decreased tone
  - Decreased reflexes
  - Urinary/Bowel content retention
Approach to the patient with myelopathy

- Localize the lesion
- Identify the underlying cause
  - X-ray should be done when bone pathology suspected
    - Examples: Fractures, metastatic lesions
    - Confirm with CT
  - MRI when intrinsic neural pathology is suspected
- Treatment is directed at underlying cause
Demyelinating diseases

- Acute demyelinating diseases – severe autoimmunity
  - Acute disseminated encephalomyelitis
- Chronic demyelinating diseases with acute events
  - Multiple sclerosis (2 phases)
    - Immune mediated
    - Degenerative
  - Neuromyelitis optica (antibody mediated)
- Inherited demyelinating disease
  - Adrenoleukodystrophy
  - Metachromatic leukodystrophy
Monophasic Demyelination - Named by site involved

- Optic neuritis
- Acute transverse myelitis
- Acute cerebellitis
Optic Neuritis Treatment Trial

- Randomized into 3 groups
  - IV Solumedrol for 3 days then oral prednisone for 11 days
  - Oral prednisone (1mg/kg/day) for 14 days
  - Placebo

- Results
  - Oral prednisone group did worse than placebo
  - IV treatment had better outcome
  - IV treatment delayed time to diagnosis of MS
Multiple sclerosis - central nervous system demyelination

- Two requirements
  - TWO different locations of CNS demyelination
    - One episode of demyelination ≠ MS
  - Separated in time by at least 30 days

- Can prove by
  - Two clinical events (no ancillary tests needed)
  - One clinical event plus ancillary test
    - MRI findings
    - Lumbar puncture (oligoclonal bands)
    - Visual evoked potentials

Types of MS

1. Benign Multiple Sclerosis
2. Relapsing Remitting Multiple Sclerosis
3. Secondary Chronic Progressive
4. Primary Progressive (10-20% of patients)
Treatment

- Acute exacerbation = IV Solumedrol
- Chronic suppression with injectable disease modifying agents
  - Avonex
  - Beta-Seron
  - Copaxone
- Oral agents
  - Fingolimod
- Tysabri when these fail
Neuromyelitis Optica (Devic’s)

- Affects two main locations
  - Optic nerves
  - Spinal cord (especially thoracic)
- Autoimmune disease mediated by IgG-NMO
  - This antibody is directed against aquaporin channels
  - Edema tends to be very significant
- Differs from MS in:
  - Pathophysiology
  - Location
  - Tendency to involve contiguous cord segments