MKSAP Neurology Review

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Approach to neurology

- LOCALIZE the LESION!
  - "What means where"
- ELIMINATE EMERGENCIES FIRST!
  - VET those emergencies
    - Vascular
    - Electrical
    - Traumatic
- Short differential diagnosis
- Treat the underlying pathology

2 overriding concepts

- Ockham's Razor for lesions
  - Put the lesion in ***one site*** if possible OR
  - Localize to a particular tissue such as:
    - Gray matter
    - White matter
    - Muscle
- "WIRE CROSSING IS BAD"
  - Tracts are PROPERLY laminated
  - KNOW where the tracts cross

4 Tracts for lesion localization

- MOTOR system [most important for localization]
  - Corticospinal
    - Cortical
    - Subcortical
  - Anterior horn cell starts the peripheral nervous system
- 2 sensory systems
  - DORSAL columns
  - Spinothalamic
- Cerebellar [least important for localization]

Motor system first! [Strength, tone, atrophy, ±reflex]

- CNS lesion?
  - Hyper-reflexia (after initial shock)
  - Increased tone
  - NO ATROPHY (caveat: anterior horn cells)
- Peripheral lesion?
  - Decreased reflexes
  - Decreased tone
  - Atrophy (caveat: demyelinating neuropathies)

Reflex levels-count to 8

REFLEX	ROOT LEVELS [1st of pair is predominant]
Ankle	S1 – S2
Knee	L3 – L4
Biceps brachii	C5 – C6
Brachioradialis	C5 – C6
Triceps brachii	C7 – C8

Central nervous system

- Cortical?
  - Aphasia?
  - Visual field cut?
  - Abulia? Apraxia?
  - Contralateral neglect?
- Subcortical?
  - Hemiparesis
- Brainstem
  - Cranial nerve lesion IPSILATERAL
  - Hemiparesis CONTRALATERAL
- Spinal cord?
  - Both sides of body usually affected
  - SENSORY LEVEL LOCALIZES TO SPINAL CORD

Peripheral nervous system

- Anterior horn cell/motor nerve roots
  - Invariant weakness with atrophy
  - NO SENSORY LOSS
- Peripheral nerve
  - Distal weakness WITH sensory loss
- NMJ
  - Waxing/Waning weakness
  - NO SENSORY LOSS
- Muscle
  - Invariant proximal>distal weakness
  - NO SENSORY LOSS

SENSORY SYSTEM (2 types)

- LARGE FIBER/DORSAL COLUMN
  - Vibration/Position sensation
  - Afferent portion of reflex arc
  - HIGH CROSS
- SMALL FIBER/SPINOTHALAMIC
  - Light touch/Pin/Temperature sensation
  - LOW CROSS

Sensory tracts just below the cervicomedullary junction

- Dorsal columns: [CROSS at C1]
  - Sacral fibers are most MEDIAL
  - Cervical fibers are most LATERAL
- Spinothalamic: [IMMEDIATE CROSS]
  - Sacral fibers most LATERAL
  - Cervical fibers most MEDIAL

2nd Step: Sensory system

- If peripheral: NO DISSOCIATION
  - Sensory modalities lost together
  - No “splitting”
- If central: DISSOCIATION or LEVEL
  - Sensory level = SPINAL CORD
  - Dissociation
    - Pain and temperature affected on one side
    - Vibration and position affected on the other

Cerebellum

- ABOVE cervicomedullary junction
  - Usually contralateral to affected side
- BELOW cervicomedullary junction
  - Tracts cross twice
  - Lesion is ipsilateral to affected side

Major Neurology Topics

- Headaches/Brain Tumors
- Delirium/Dementia/Stupor and Coma
- Dizziness/Vertigo/Syncope/Hearing Loss
- Movement disorders
- MS/Spinal cord diseases
- Neuromuscular disease
- Epilepsy
- Stroke

Headaches

- RED FLAGS:
  - "Worst headache of life"
  - Headache with ANY neurological deficits
  - Headache with papilledema
  - Headache with fever
  - Headache with jaw claudication
- ALL other headaches are usually migraine
  - Cluster headaches are extremely rare
  - Tension headaches are rarer still

Headaches

- Any “RED FLAG” headache requires:
  - Head CT WITHOUT contrast (exclude bleed)
  - LP if CT is negative
  - ESR if temporal arteritis is suspected
- Non red-flag headaches
  - Migraine
  - Cluster
  - Chronic parxoysmal hemicrania

Migraine headaches

- Unilateral pulsatile HA with photophonophobia
- Pathophysiology-Low pontine serotonin
- Females > Males
- Prophylaxis
  - Tricyclic antidepressants
  - Beta-blockers
  - Anticonvulsants (Depakote, Topamax)
- Abortive
    - Triptans
    - Sedatives
    - Ergotamines

Cluster headaches

- Similar pathophysiology
- Males > females
- Other differences
  - Wakes people from sleep in early AM
  - Relieved by 10-12 liters of oxygen by mask
  - Extreme agitation (migraineurs are reclusive)
  - Rhinorrhea and lacrimation

Brain Tumors

- Infiltrators
  - Primary CNS neoplasms
  - Neural crest derived tumors [melanoma]
- Displacers
  - Originate elsewhere
  - Supporting elements
    - Meningioma
    - Ependymoma

Tumor Comparison

	Infiltrators	Displacers
Size at detection	Large	Small
Usual presentation	Seizures Minimal deficits	Seizures/HA Obvious deficit
Location	Diffuse	Gray-White junction

Delirium, Dementia, Stupor, Coma

- RED FLAGS:
  - Acute onset
  - Focal neurological deficits
  - Metabolic derangements
- DISEASES you should NEVER MISS:
  - Wernicke's (always give Thiamine)
  - B12 or Folate deficiency
  - Thyroid disease induced encephalopathy

Delirium, Dementia, Stupor, Coma

	Sensorium	Interaction with environment
Delirium	Agitated	Hostile but purposeful
Dementia	Clear	Normal on the surface
Stupor	Clouded	Minimal but purposeful
Coma	Obtunded	Reflex actions only

Approach-
Focal neurological deficit?

- Yes: The brain is likely at fault
  - Head CT WITHOUT CONTRAST-STAT
  - LP if head CT is negative
  - EEG if LP is negative
- No: The brain is secondarily affected
  - This is a TOXIC-METABOLIC process
  - Medication/drug abuse history
  - CBC, CMP, TSH, B12, Folate
  - RPR, HIV with high index of suspicion

Dementia

- Do not diagnose until you have excluded:
  - Pseudodementia (e.g. depression)
  - Treatable dementias (B12, Folate, TSH)
- Otherwise, it is probably Alzheimer's
  - Procholinergics slow the rate of decline
  - They do NOT reverse the disease process

Delirium

- 99.9% is toxic-metabolic in origin
- Exception: Non-dominant hemisphere lesions
  - Acute stroke
  - Herpes simplex encephalitis
- Treatments
  - Find the underlying cause and address it
  - Tincture of time

Stupor and Coma

- FOCAL DEFICIT = Brain is at fault
- LOCALIZE the lesion
  - Examine ALL cranial nerve reflexes:
    - Pupillary
    - Blink
    - Oculocephalic [NOT Doll's eyes]
    - Pharyngeal [NOT gag]
  - Posturing?
    - Decorticate or arm flexion = above red nucleus
    - Decerebrate or arm extension = below red nucleus

Dizziness, Vertigo, Hearing Loss

- Define vertigo
  - Illusion of motion
  - “Spinning” is not necessary
- Occurs from mismatch of sensory inputs
  - Vision
  - Posterior columns
  - Vestibular system
- MUST differentiate “central “ from “peripheral”

Central versus peripheral

	Central	Peripheral
Long tract signs	Presence confirms central	NEVER
Hearing loss	EXTREMELY RARE	Virtually diagnostic
Fatigues	No	Yes

Common causes of vertigo

- Benign Positional
  - Provoked in 1 position ONLY
  - Fatigues with each provoking maneuver
- Meniere's disease is TRIAD of
  - Hearing loss [Localizes to periphery]
  - Tinnitus
  - Vertigo
- Stroke
  - Long tract signs
  - Cerebellar deficits

Movement disorders

- BALANCE of Ach and DA
  - Increased Ach = Decreased DA
  - Increased DA = Decreased Ach
- Excess DA leads to:
  - Psychosis
  - Chorea
- Reduced DA leads to
  - Parkinsonism
  - Dystonia/Rigidity

Movement disorders

- Parkinson's Disease [Idiopathic]
  - Too little DA in Substantia Nigra
  - FOUR cardinal features
    - Tremor
    - Rigidity
    - Bradykinesia
    - Postural instability
  - Should respond to DA agonists
- Parkinson's Plus syndromes do NOT respond to DA therapy

Tremors

- Essential
  - Action tremor
  - Disappears at rest
- Parkinson's tremor
  - Tremor at REST
  - Disappears wit movement
- Cerebellar Tremor [RARE]
  - Worsens as the target is approached
  - Clinically obvious

Movement disorder therapy

- “Move too much…give them sedatives”
  - Beta-blockers
  - Barbiturates
  - Benzodiazepines
- “Move too little…try some Sinemet”
  - Use DA agonists below age 60
  - Start Sinemet at age 60 or greater

Some exceptions

- Acute dystonic reaction from DA blocker
  - Block Ach to restore balance
  - Use diphenhydramine IV
- Parkinson's tremor does NOT respond to DA therapy
  - Non-anticholinergic sedatives required
- Focal dystonia responds best to Botulinum toxin injectins

Multiple Sclerosis

- A WHITE MATTER CNS disease
- Two distinct clinical episodes separated in SPACE and TIME
  - Space: Must affect DIFFERENT white matter locations
  - Attacks must be more than 30 days apart
- MRI criteria can be used instead of an actual SECOND clinical attack

1 lesion ≠ Multiple Sclerosis

- Diagnosis based on location
  - Optic tract = Optic Neuritis
  - Spinal cord = Transverse myelitis
  - Cerebellum = Acute cerebellitis
- Risk of MS increased after first attack
- IV steroids hasten recovery but have NO EFFECT on probability of second attack
- IV steroids are superior to oral steroids which are = placebo

MS therapy

- Acute attack- IV methylprednisolone
  - Given as 500 mg IV bid for 3-5 days
- Reduce relapse frequency/progression
  - Interferons
  - Copaxone
- When above agents fail:
  - Cyclophosphamide
  - Mitoxantrone

Spinal cord disorders

- ALWAYS eliminate cord compression:
  - Plain X-rays to exclude bony pathology
  - MRI
- CSF exam if previous studies are normal:
  - Transverse myelitis
  - Meningomyelitides
- Other odd causes
  - Spinal cord strokes
  - B12 deficiency

Neuromuscular

- LOCALIZE the lesion
  - Motor, Sensory or both?
  - Trunk, shoulder or hip girdle involved?
  - Face involved?
- If motor affected:
  - Constant or variable weakness?
  - Atrophy?
  - Tone increased or decreased?

Nerve structure

- Myelin sheath
  - For rapid conduction of impulses
  - Vibration and position sense
  - Motor function
- Vasa nervorem
  - Blood vessels end on nerve surface
  - Loss of perfusion affects small fibers in nerve center
- Axons
  - Most metabolically active
  - Atrophy when the motor axons are injured

Neuropathy vs. myopathy vs. NMJ

- Neuropathy vs. myopathies
  - Most neuropathies cause distal before proximal dysfunction
  - Muscle diseases affect proximal before distal
- NMJ disorders usually involve the CNs
  - NO sensory loss
  - NO atrophy
  - Extraocular muscles affected early

Myelinopathy versus axonopathy

- Reflexes lost early
- Large fiber sensory loss
- Weakness with minimal atrophy
- CSF protein may be increased
- Patterns other than distal to proximal

- Reflexes retained until late
- Small > large fiber sensory loss
- CSF protein normal
- Distal to proximal dysfunction

Myopathies

- Inflammatory usually have increased CPK
  - Polymyositis: Cellular infiltrate
  - Dermatomyositis: A vasculitis
- Non-inflammatory have normal to minimally elevated CPK
- Diagnosis can only be established by biopsy

Epilepsy

- A disorder of the cortical gray matter
- Definition:
  - 2 UNPROVOKED seizures
  - A CLINICAL diagnosis
- Provoked seizure examples:
  - Syncope
  - Sleep deprivation
  - Alcohol withdrawal

Epilepsy diagnosis

- Clinical to classify the epilepsy
  - Reliable witness
  - Tongue laceration
  - Prolonged LOC
- EEGs do NOT diagnose epilepsy
  - Many patients have normal EEGs
  - Spikes on an EEG do NOT prove epilepsy

Evaluation

- Elicit complete history to exclude a provoked seizure
- CBC, CMP, UDS at a minimum
- Imaging
  - Generalized only requires head CT
  - Focal onset REQUIRES MRI
- EEG
  - Highest positives within 24 hrs
  - Anticonvulsants do NOT suppress abnormalities

Treatment

- Treatment of a first unprovoked seizure is controversial but most will do so if any of the following are noted:
  - Focal onset
  - EEG abnormalities noted
  - MRI abnormality in cortical gray noted
- All anticonvulsants have the same efficacy for most seizure types-they differ in side effects

Treatment exceptions

- Juvenile myoclonic epilepsy
  - Valproic acid
  - Lamotrigine
- Absence seizures
  - Valproic acid
  - Ethosuximide