Nervous System
10/29/14

Case one:

1. Weakness is assessed with muscle testing
2. Sensory modalities: Proprioception (flex/extend the big toe or the thumb while their eyes are closed), vibratory sense (tuning forks)
3. Sensory modalities in the spinal cord
   Primary Modalities:
   - ALS: pain, temperature, light touch
   - Dorsal column: light touch, pressure, vibration, and position

   Secondary modalities:
   - Two point discrimination: see how close you can get between two points of contact of a paper clip before the patient is unable to discriminate between the two points (varies by region of the body).
   - Stereognosis: ability to recognize common objects by holding it (closed eyes)
   - Graphesthesia: determine a number that is drawn by your finger or an instrument in the palm

4. Cranial nerve review:
   1. CN1: Olfactory nerve (smell-use familiar smells such as coffee or orange).
   2. CNII: optic nerve, eye exam
   3. CNIII: oculomotor nerve; H in space
   4. CNIV: Trochlear nerve; Superior oblique-down and in
   5. CNV: Trigeminal nerve; sensory and motor (mastication) to the face
   6. CNVI: Abducens nerve; H in space again - lateral rectus
   7. CNVII: facial nerve; facial expression, smile! Puff cheeks, raise eyebrows.
   8. CNVIII: Vestibulocochlear
   9. CNIX: glossopharyngeal, have patient say "ah", taste to posterior tongue (facial nerve for anterior tongue).
   10. CNX: vagus nerve, super important; also indicated in pharyngitis
   11. CNXI: accessory nerve; raise shoulders
   12. CNXII: Hypoglossal nerve; stick tongue out; if it deviates it will deviate to the side of the lesion.

Gaits:

- Diplegic/scissor gait: legs move but there is no movement in the arms
- Shuffling gait: Parkinson's disease; often with very small steps (march of small steps).
- Steppage gait: can't dorsiflex, stepping gait
- Cerebellar ataxia: usually associated with people under the influence of alcohol; wide based walking
- Hemiparetic gait: circumduction of foot
- Myopathic gait: can't stabilize the pelvis (hyperlordotic over weight bearing leg).

UMN lesion:
Muscles are affected in groups and there is muscle weakness (weakness with LMN too).
There can be ankle clonus: pronounced hyperreflexia with dorsiflexion

- Babinski's sign (plantar response)
- Hoffman's sign: flick the middle phalanx to see if there is flexion at IP of thumb
- Decerebrate posturing

LMN Lesion:
Individual muscles can be affected and there can be muscle weakness accompanied by atrophy. There is up to 70% loss of bulk.
Fasciculations: not always pathologic (can be due to caffeine)
Hyporeflexia
There are abnormal nerve conduction studies in this case

Categorize signs or symptoms of the case:

1. "Paresthesia in hands/feet for two years, extending to the knees." - primary sensory
   Secondary sensory: cortical interpretation occurs in brain (ie: graphesthesia, Stereognosis, interpreting what fabric is touching your skin).
1. "Leg weakness for one year, progressing to need for a walker a few months after onset."-Can't distinguish between UMN or LMN lesion.
2. "High arches and hammer toes since childhood-considered weaker than peers." - Could be UMN or LMN
3. "Scissoring on ambulation, due to spasticity." -Usually a sign of UMN lesion (ie: cerebral palsy)
4. "Stocking glove pattern of sensory loss." - primary sensory
5. "Marked loss of joint position and vibratory sensation with an upper level at the midthoracic spine." -primary sensory
6. "Hyperactive DTRs except for absent ankle jerks, extensor plantar response bilaterally." - UMN lesion.
7. Head enlargement-Can't determine if it is abnormal without looking at the height; also look at the family members as it could be a trend. If it is truly enlarged it will occur prior to 2 years old (when sutures close).

Dementia:
Higher increase rates outside of developed countries. The cause of this is unknown.
Remember that dementia is a symptom not a diagnosis (10% of those causes are treatable-ie: normal pressure hydrocephalus).
Two types of dementia: one with other neurological symptoms, one without.

- Other causes: chronic drug intoxication, pernicious anemia, hyper/hypothyroidism, pseudodementias (depression, hypomania, schizophrenia).

Most common causes: Alzheimer's disease (risk increases with age), frontotemporal dementia with parkinsonism linked to chromosome 17, vascular dementia.
Important to inform patients and families of the risks with driving.

Neurodegenerative cousins: damaging processes that occur from genes and the environment that lead to early symptoms of disease entities.

- Alzheimer's disease: Without an autopsy Alzheimer's is not absolute. Most patients are in their 60s or older. The prevalence rate is 3x higher in women (but women do live loner).
    - Course: symptomatic course extends over about 5 years.
    - Microscopic changes: neurofibrillary tangles in the cytoplasm. The tau fragments are from disintegrating microtubules related to beta amyloid plaque deposition (unsure what starts that deposition). There is degeneration especially in the hippocampus (memory).
        - Presenilin 1 and 2: with even one mutation in these enzymes there is commitment to early onset Alzheimer's disease.
    - Research: two weeks ago researchers were able to simulate beta amyloid generation. Showed that beta amyloid deposition happens before the tau fragments.
    - Mildly impaired visuospatial and construction skills early, executive functioning is impaired later, fluctuating mental status is less pronounced, psychotic features are not typical early on, delusions are related to memory loss, depression and anxiety are common and parkinsonism is later in the disease course (these are in comparison to lewy body dementia)
    - 3rd leading cause of death in the US.
    - Can have mild cognitive impairment (not severe enough to disrupt the person's day to day life).

- Circumscribed cerebral atrophy: Pick's disease/frontotemporal dementia (many names): Progressive dementia with cerebral atrophy in frontal and temporal lobes.
    - Onset: typically earlier than Alzheimer's disease at 45-60.
    - There is a familial history present in about 50% of cases.
    - Men and women are equally affected (women more affected in Alzheimer's).
    - Course: Duration is 8 years (longer than 5 year course of Alzheimer's).
    - Clinical presentation: Often younger person that presents with behavioral changes (apathy, repetitive/compulsive behaviors like picking at carpet over and over again).

- Huntington Disease:
    - Very late onset (oral facial movements), older onset (more associated with choreiform movements).

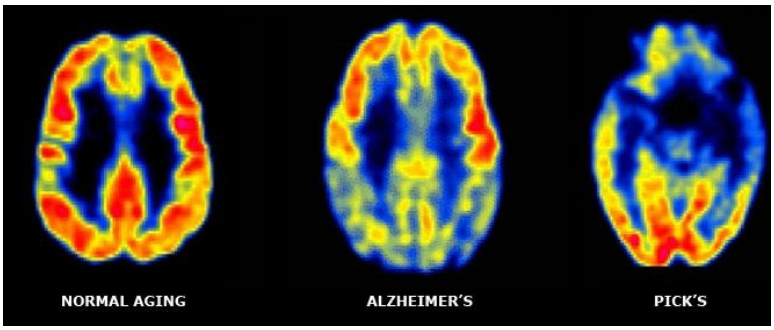
- Lewy body disease:
    - Dementia with lewy bodies, parkinsonism features.
    - Early on memory dysfunction is less pronounced than Alzheimer's
    - Executive functioning impaired early
    - Pronounced fluctuating mental status
    - Can have prominent psychotic features early with bizarre delusions

    - Can have depression and anxiety
    - Parkinsonism within 1-2 years of dementia

- <u>Mild cognitive impairment:</u> can stay here and not progress to Alzheimer's. This is a clinical diagnosis.

<u>PET scans:</u>
Positron emission tomography
Red: more active brain function

- Alzheimer's disease: more red present in the anterior part of the brain
- Pick's disease: huge reduction in activity in the anterior part of the brain



<u>Frontal lobe signs:</u>

- Grasp reflex (babies)-disappears in second year of life. If it re-emerges in adulthood that is a sign.
- Snout reflex: touching the lip will create pursed lips. Again if it re-emerges in adulthood that is a sign.
- Sucking reflex
- Palmomental reflex: ipsilateral jaw movement with touch of the thenar imminence.