Nontraumatic Myelopathies

Spinal Cord Injury Medicine Review Course

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DISCLOSURE OF CONFLICT OF INTEREST

- Dr. Kryger has no financial conflicts of interest relevant to this activity.

LEARNING OBJECTIVES

- At the conclusion of this activity, the participant will be able to:
  - Describe epidemiology, symptoms and treatment of multiple sclerosis
  - Discuss how someone might present with symptomatic spinal stenosis, and what the “red flags” may be.
  - Identify where different spinal cord tumors may be found.

IMMUNE MEDIATED CONDITIONS

1. Multiple Sclerosis
2. Transverse Myelitis
3. Neuromyelitis Optica
4. Neurosarcoidosis

MULTIPLE SCLEROSIS

- Affects CNS through demyelination and degeneration
- Initially symptoms often visual (50%), optic neuritis (20%), fatigue, weakness, parasthesias, gait
- Subtypes based on course:
  - Clinically isolated syndrome, Relapsing-Remitting, Primarily progressive, secondarily progressive
  - RR often becomes 2ndary progressive in 10-20 yrs

MULTIPLE SCLEROSIS

- Avg disease onset = 30 yrs
- Generally seen farther from equator
- 80% affected are women, but male disease is more severe
- Multiple Risk factors: Family hx, HHV-6, Vit D deficiency, smoking, high BMI
- Major Contributor: Those with Epstein-Barr Virus 32x more likely to develop MS, according to a study of 10 million adults in the military. (Through molecular mimicry)

MULTIPLE SCLEROSIS

- Thought to originate from microglial activation resulting in demyelination and neuron damage- axonal damage can follow
- MRI: T2 enhancements indicate active lesions
- Disability measured by Expanded Disability Status Scale (EDSS)
- Treatment with Diseases Modifying Therapies, several immune-modulating agents

EXPANDED DISABILITY STATUS SCALE

- Examines“functional systems”
  - Pyramidal, cerebellar, brainstem, sensory, bowel/bladder, visual, cognition, other
- Level 1-4.5 look at normal ambulators, but with issues with a functional system
- Level 5-9.5 categorize progressive ambulation difficulties

2017 MCDONALD CRITERIA

2 or more clinical attacks (over time)

PLUS

- ≥2 lesions with objective clinical evidence (OCE) or
- 1 lesion with OCE and clinical history of prior lesion or
- 1 lesion with OCE and ≥2 MRI lesions over time

1 clinical attack

PLUS

- ≥2 lesions with OCE AND (≥2 MRI lesions over time or Oligoclonal Bands) or
- 1 lesion with OCE AND ≥2 MRI lesions over space AND (≥2 MRI lesions over time or Oligoclonal Bands)

ACUTE TRANSVERSE MYELITIS

- Long lesion in spinal cord, tendency to affect thoracic spine
- Affects woman>men, 20s-40s
- Often presents with weakness and numbness
- Presenting incident for 5-10% of MS Cases
- 1/3 recover completely, 1/3 do not recover, 1/3 recovery partially
- May be caused by viral prodrome, rare association with vaccination
- Treated with steroids, IVIG, plasma exchange

NEUROMYELITIS OPTICA

- Variant of neurodegenerative disease that has predominant optic neuritis and transverse myelitis features
- Detect NMO Antibody against Aquaporin-4 channel in CNS
- Generally more aggressive than MS
- Uses similar treatments: IV Steroids, plasmapheresis, immunosuppression

NEUROSARCOIDOSIS

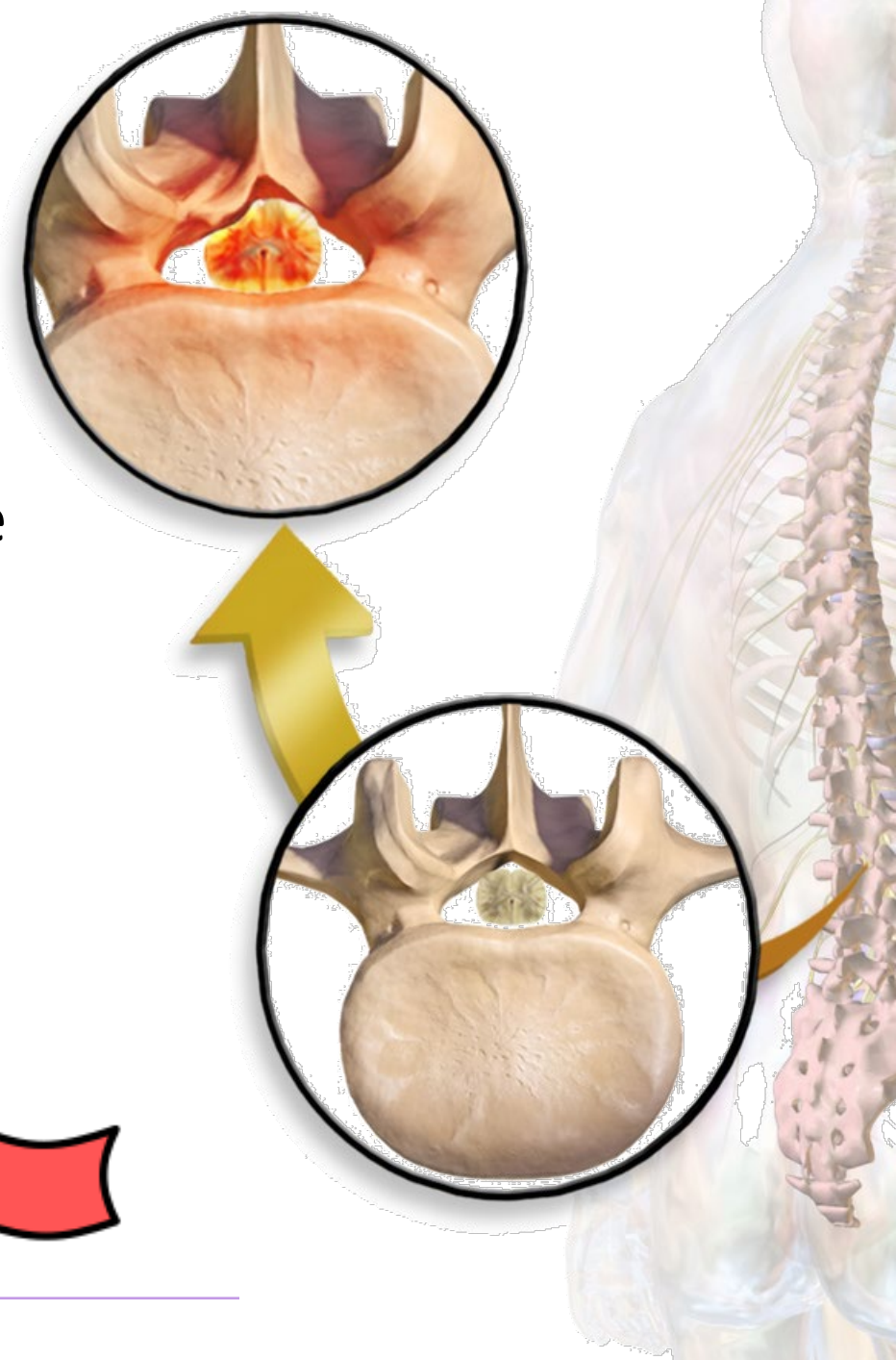
- Granulomatous disease
- 5% with sarcoidosis have neurologic complications
- Granulomas may be intramedullary or extramedullary
- Treated with steroids or immune suppression

DEGENERATIVE MYELOPATHIES

1. Spinal Stenosis
2. Intervertebral Disk Herniation

SPINAL STENOSIS

- Spinal Stenosis = ligamentum flavum hypertrophy and disk herniation resulting in central canal narrowing
- Very prevalent- 50% at age 45, 90% at age 60
- Most have neck pain
- Considered myelopathic when other neurologic symptoms present: most commonly UMN symptoms: spastic gait, hyperreflexia
- Weakness, sensory changes, bowel/bladder changes may be present, but less pronounced



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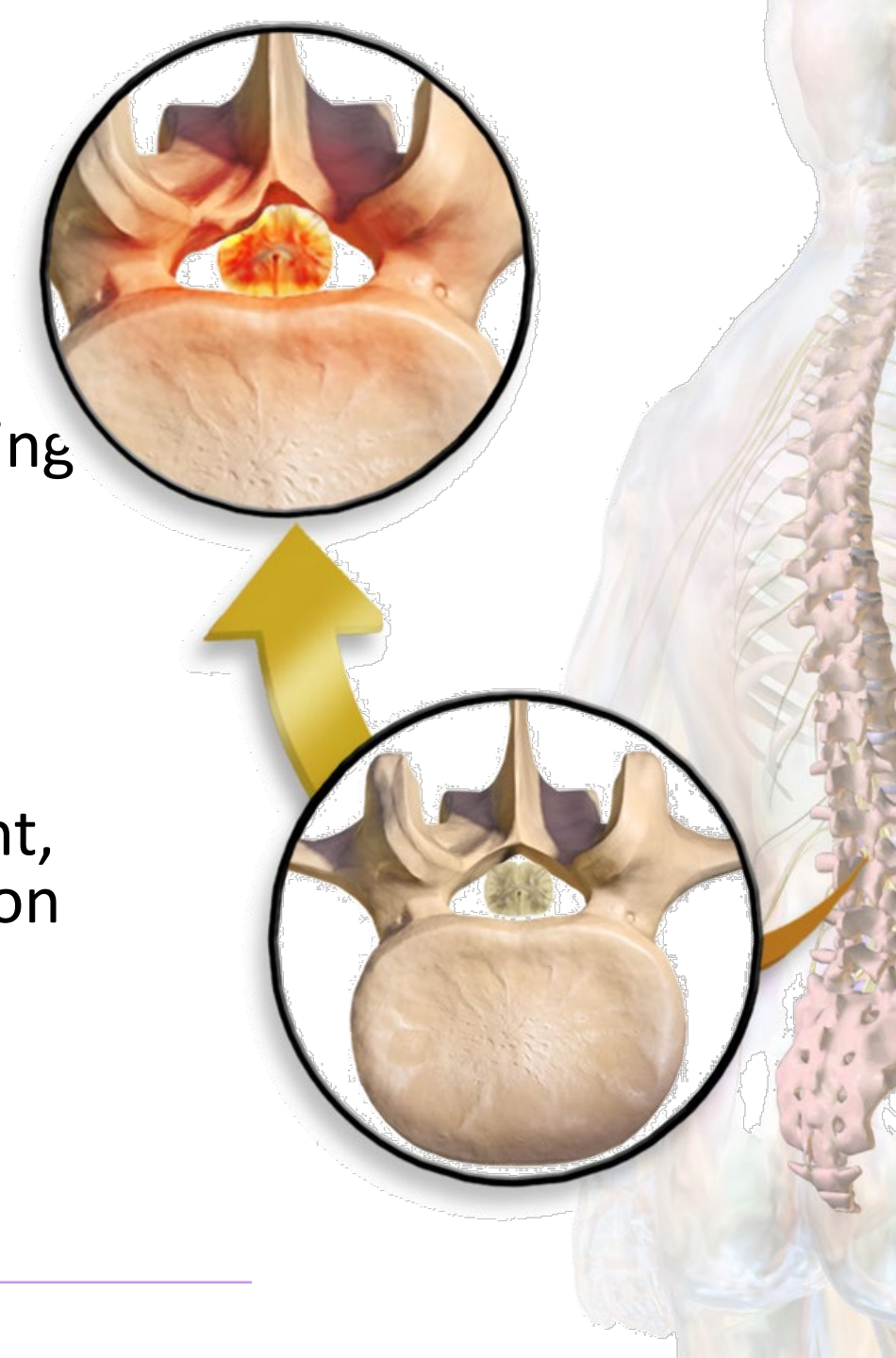
SPINAL STENOSIS

- If lower back/cauda equina is compressed, may have neurogenic claudication, hunched posture (“shopping cart sign”)
- Mild disease likely just requires conservative management
- Red Flag symptoms, or those not responsive to conservative management, may benefit from surgical decompression and fusion
- Cervical decompression has high complication rate (17%)
- Fall + Stenosis may result in severe SCI!



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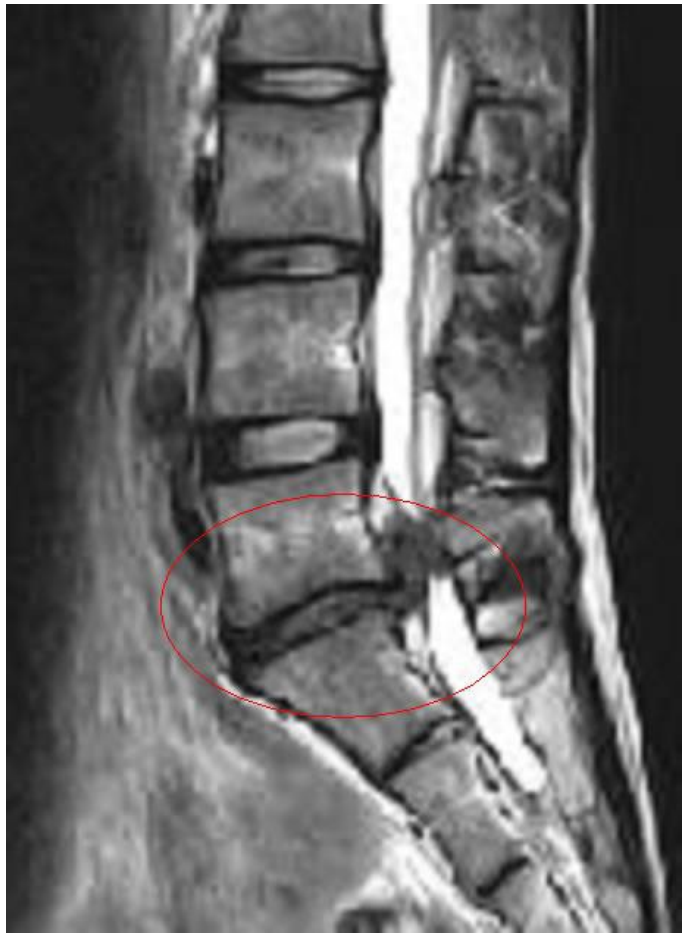
INTERVERTEBRAL DISK HERNIATION

- Most often occurs in dorsolateral direction, resulting in radiculopathy- pain without weakness or UMN signs
- Central herniation more rare. Occurs in lower cervical or lower lumbar spine
- May be sudden “pop” or slow progression
- Often starts as back pain, develops into neurologic symptoms later in course
- Herniation at L5-S1 can result in cauda equina, sciatica, los of ankle reflexes, foot drop.



REGARDING SPINE IMAGING

- Just because you have spondylosis does not mean you will have symptoms
- In 20s, 37% have disc degeneration, in 30s : 52%, in 40s, 68%

SPINAL CORD VASCULAR DISEASE

1. Arteriovenous Malformations
2. Infarctions
3. Hemorrhages

ARTERIOVENOUS MALFORMATIONS

- Fistulas between arteries and veins
- Type 1 – dural- Extramedullary, symptomatic at middle age (85%)
- Type 2 - Less common intramedullary, symptomatic in children- these often present suddenly due to hemorrhage/infarct
- Symptoms caused by compression from vascular dilatation or “steal” syndrome
- Progressive LE weakness/sensory loss, back pain, neurogenic claudication
- Diagnosis via angiography
- Treatment with embolization

SPINAL CORD INFARCTS

- Results from lack of blood flow to spinal cord
- May originate from aorta, segmental arteries or arteries within cord
- Dissecting aortic aneurysm, thrombosis, surgery with clamping of aorta, atherosclerotic plaques, severe hypotension
- Most occur in thoracic spine
- Infarct of anterior 2/3s called Anterior Spinal Artery Syndrome, though it is not usually the cause (dorsal column remains intact)
- MRI may be normal for first 24 hours, then hyperintense on T2, and hypointense on T1
- No effective management, but about 50% show decent motor recovery

SPINAL HEMORRHAGE

- May be epidural, subdural, subarachnoid, or intraspinal
- Pain, then rapid sensory loss/weakness
- 60% of cases have no cause. 10% had preceding trauma, 25% were anticoagulated, 5% from vascular malformation
- Often not visible for first 24 hours on CT or MRI
- Treat with surgical decompression/removal of hematoma

SPINAL CORD INFECTIONS

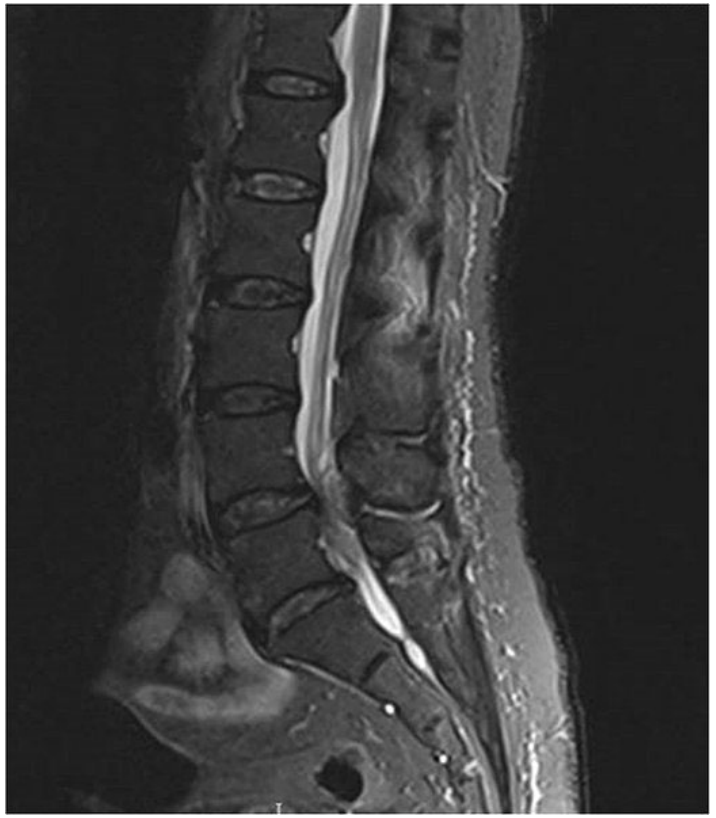
1. Epidural Abscesses
2. Potts Disease
3. Neurosyphilis
4. West Nile Virus
5. Vacuolar Myelopathy
6. HTLV-1
7. Post-polio Syndrome

EPIDURAL ABSCESS

- Purulent fluid collection within spine outside the dura
- Causes:
  - One third extend from local infection (osteomyelitis/discitis)
  - One third hematogenous spread (IV drug use, GU, endocarditis)
  - One third idiopathic
- Initially presents as local pain, majority with fevers, radicular pain, leg weakness
- Usually gradual evolution

EPIDURAL ABSCESS

- Diagnose with MRI (ring-enhancing mass)
- Treat with surgical drainage/excision
- IV Abx for 6-8 weeks
- Recovery often completely if caught before LE paraplegia

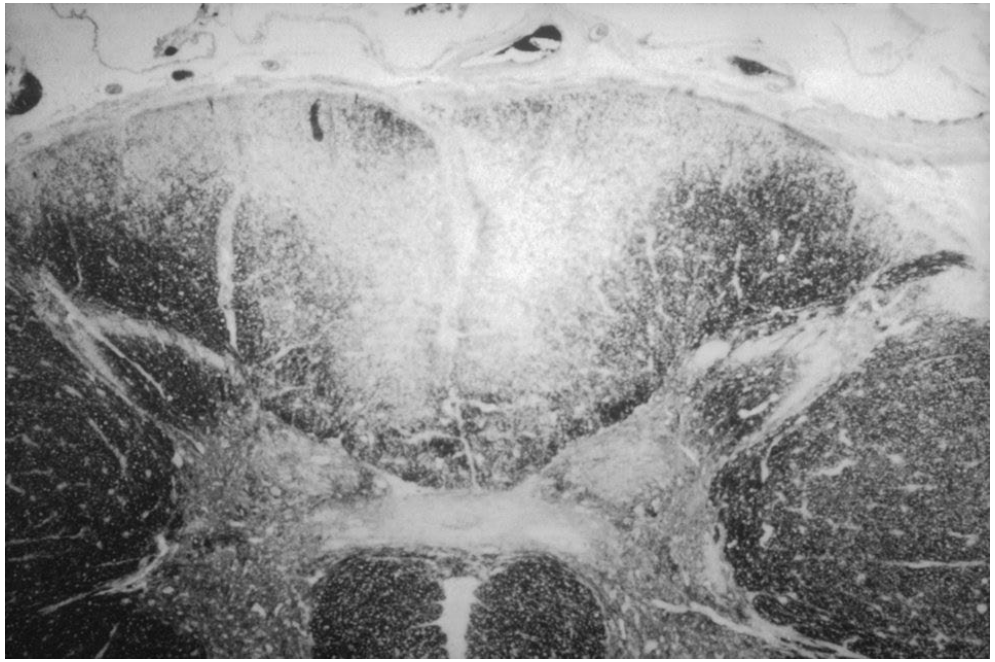


NONPYOGENIC OSTEOMYELITIS

- Aka Potts Disease. Arises from Tuberculosis.
- <5% have skeletal involvement, half of those have spine involvement
- Back pain, night sweats, weight loss
- Diagnosis: TB Test, MRI Spine
- Treatment: Multi-drug chemotherapy

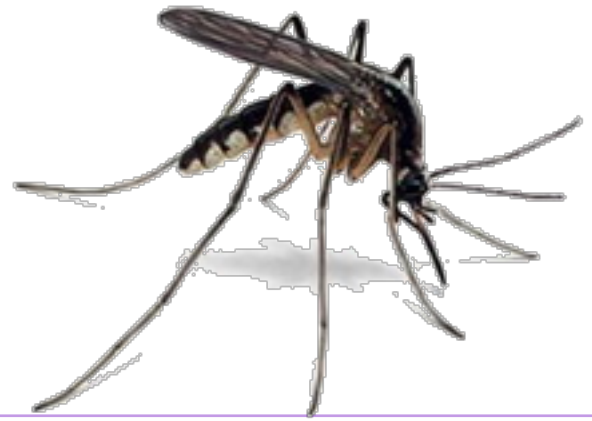
NEUROSYPHILIS

- From Treponema pallidum bacteria
- Tertiary Syphilis can have tabes dorsalis (dorsal column affected)- paresthesia, loss proprioception, eventually weakness and death
- Treated with Penicillin G



WEST NILE VIRUS

- From an RNA virus, presents similarly to poliomyelitis, but in adults
- Acute asymmetric weakness after febrile illness with CSF pleocytosis
- Diagnosis: Seropositive for virus, EMG/NCS (asymmetric impact on anterior horn cells and axons)
- Treatment: Supportive



HIV ASSOCIATED MYELOPATHY (VACUOLAR)

- Develops during late-stage AIDS
- Progressive LE weakness, sensory loss, loss of bowel/bladder
- Pathology shows intramyelinic and peri-axonal vacuolation and demyelination.
- Prevented, with some improvement with HAART

HTLV-1 MYELOPATHY

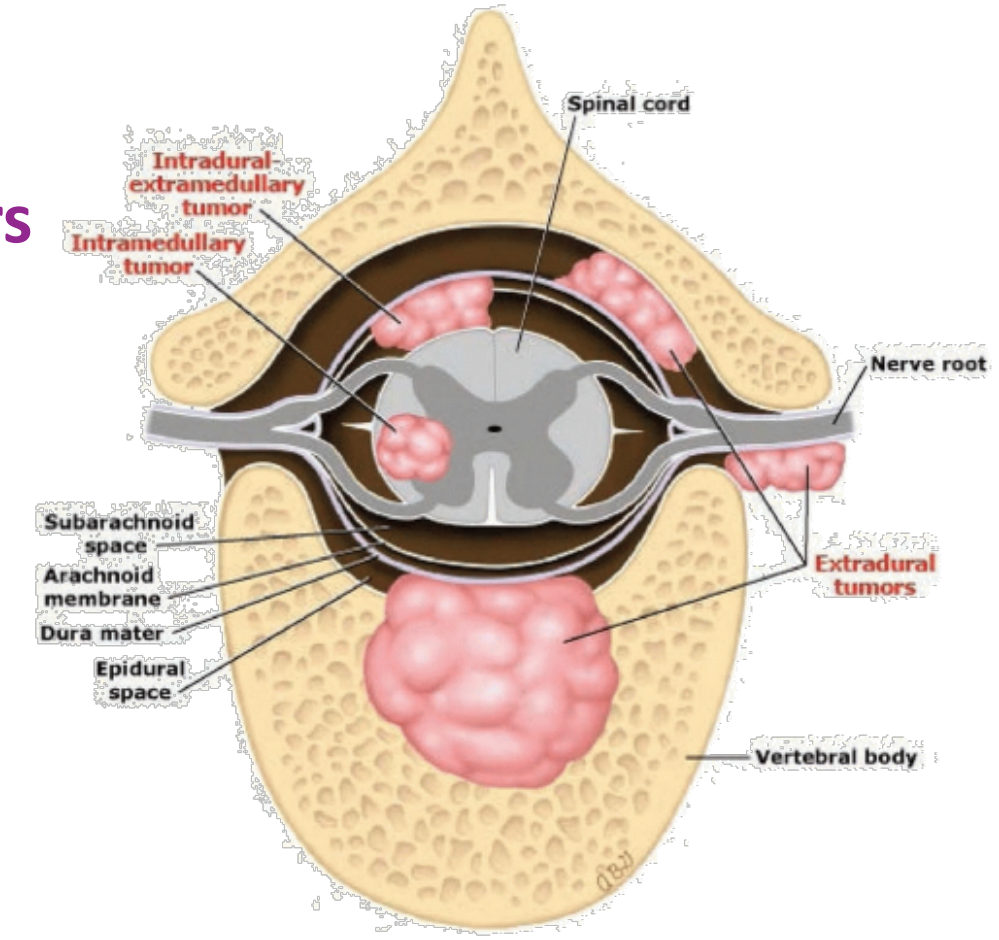
- T lymphotropic virus type I
- AKA Tropical Spastic Paraplegia
- Transmitted via blood/sexually
- Found in Caribbean, Africa, Japan
- Leg weakness/ataxia with spasticity
- Wheelchair use within 5-10 yrs
- Diagnosis: HTLV-I antibodies, CSF pleocytosis. MRI normal
- No definitive treatment. Treat symptomatically

POST-POLIO SYNDROME

- New progressive weakness/pain/fatigue years after initial event (>15 yrs usually)
- Persists for > 1 year
- Incidence 22-68% in survivors of Polio
- Requires multi-disciplinary rehabilitation, symptom management

SPINAL TUMORS

1. Epidural tumors
2. Intradural/Extramedullary tumors
3. Intramedullary tumors



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EPIDURAL TUMORS

- Most common
- Usually malignant, originate from metastatic region
- Lung/breast cancers 50%, with compression (rarely hematogenous spread). Prostate also common
- Treated with steroids, radiation, excision of masses

INTRADURAL/EXTRAMEDULLARY TUMORS

- In subarachnoid space
- Generally benign, slow growing
- Schwannoma>Meningioma>ependymoma
- Treatment: Surgery, often curable

INTRAMEDULLARY TUMORS

- Inside of cord
- Ependymomas 1/3- usually in cauda equina
- Astrocytomas 1/3- usually in upper cord
- Astrocytomas can be malignant
- Not usually operable

NUTRITIONAL DEFICIENCIES

- Subacute Combined Degeneration (SCD)- From Vit B12 deficiency usually due to pernicious anemia, 42% with neuropathy/myelopathy, 12% with just myelopathy
- Vit E deficiency- Spinocerebellar degeneration with dorsal column affected
- Copper Deficiency- clinically similar to SCD. Presents with gait and parasthesias, ataxia, UMN signs

MANY OTHER NONTRAUMATIC DISEASES AFFECTING SPINAL CORD…

- Amyotrophic Lateral Sclerosis
- Primary Lateral Sclerosis
- Spinal Muscular Atrophy
- Hereditary Spastic Paraplegia
- Friedreich Ataxia
- Radiation myelopathy
- Toxic myelopathy
- Decompression myelopathy

THANK YOU!

Q & A

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QUESTION

- A patient with progressive lower extremity weakness and sensory loss had a spinal cord biopsy that appeared to show vacuoles. What test results could confirm your diagnosis?
  1) Oligoclonal bands from spinal fluid
  2) Low CD4 count in blood test
  3) Large compressive disc herniation on MRI
  4) Vitamin B12 deficient in blood test