Review of spinal oncology

Katie Krause MD, PhD

Virginia Mason Neurosurgery

Spine tumors are rare.

- Overall, spine tumors in the US are a rare entity
- Data from National Program of Cancer Registries and Surveillance Epidemiology, and End results between 2004-2007
 - Covers 99.2% of US population
 - Both malignant and non-malignant tumors included
 - 2576 malignant and 9236 non-malignant cases
 - Overall incidence of 0.97 per 100,000 persons/year
 - Overall incidence of brain tumor 14.8 per 100,000

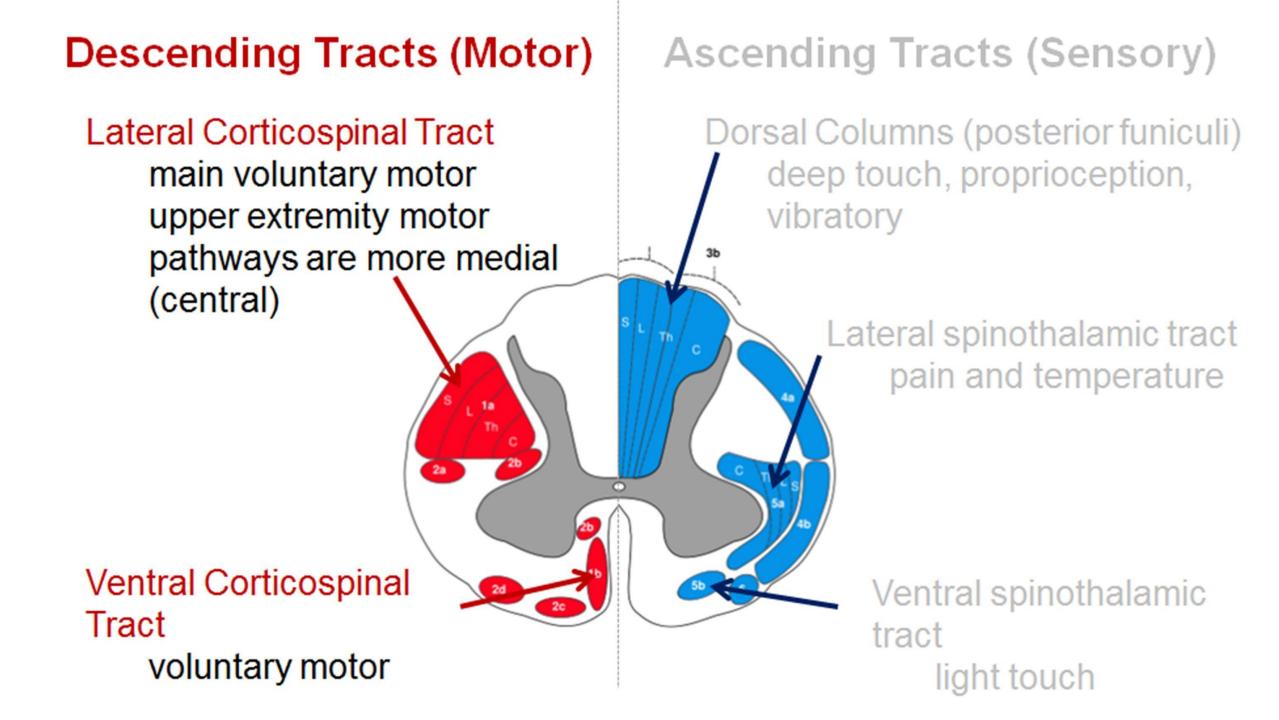


Type of spine tumors:

- Extradural 50%
- Intradural, extramedullary 40%
- Intradural, intramedullary 5-10%

Intramedullary vs Extramedullary

Intramedullary	Extramedullary
Poorly localized burning pain	Prominent radicular pain
"sacral sparing"	Early sacral sensory loss
Corticospinal tract signs appear later	Early spastic weakness in legs
Usually rapid progression (usually malignant lesion)	Usually <mark>slow progression</mark> (usually benign lesion)



Intradural, intramedullary

TABLE 1. Intramedullary spinal cord tumors

Incidence	Location	Prognosis
Most common (50-60% of IMSCTs)	Cervical > thoracic > lumbar	Good
Rare	Filum terminale & conus medullaris	Excellent
Second most common	Cervical > thoracic > lumbar	Poor
Very rare; increased incidence in VHL disease patients	Cervical > thoracic > lumbar	Excellent
Very rare	Cervical > thoracic > lumbar	Good
Rare	Cervical > thoracic > lumbar	Good
Rare	Cervical > thoracic > lumbar	Poor
Very rare	Cervical > thoracic > lumbar	Poor
	Most common (50-60% of IMSCTs) Rare Second most common Very rare; increased incidence in VHL disease patients Very rare Rare Rare	Most common (50-60% of IMSCTs)Cervical > thoracic > lumbarRareFilum terminale & conus medullarisSecond most commonCervical > thoracic > lumbarVery rare; increased incidence in VHL disease patientsCervical > thoracic > lumbarVery rareCervical > thoracic > lumbarRareCervical > thoracic > lumbarRareCervical > thoracic > lumbarRareCervical > thoracic > lumbarRareCervical > thoracic > lumbar

https://doi.org/10.3171/2015.5.FOCUS15158



Ependymoma

- Most common, accounting for 50-60% of all intramedullary tumors
- Mid-adulthood, slightly more common in males
- Associated with NF
- Develop from ependymal cells
 - Epithelial-like cells lines the ventricles and central canal of spinal cord
- Most are slow growing and benign
- Four histologic subtypes:
 - WHO I: myxopapillary
 - WHO I: sub-ependymoma
 - WHO II: ependymoma
 - WHO III: anaplastic

Ependymoma

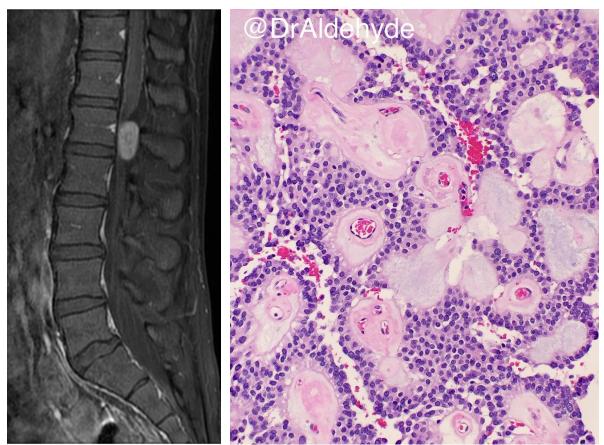
- Ependymoma (WHO II)
 - Arise from cervical >> thoracic cord
 - Radiographic features:
 - Widen the spinal centrally
 - Unecapsulated, although well-circumscribed
 - Associated with cysts and syringohydromyelia
 - Enhance strongly





Ependymoma (myxopapillary)

- Myxopapillary (WHO I) most common
 - Arises from conus/filum terminale
 - May present with sphincter dysfunction or hemorrhage
 - Prominent mucus component
 - Small, well circumscribed, ovoid



Astrocytoma

- Second most common type of intramedullary spine tumor
- Tends to be younger, with males slightly more affected
- Four histologic subtypes
 - I: pilocytic
 - II: diffuse
 - III: anaplastic
 - IV: glioblastoma
- In contrast to brain, spinal astrocytoma tend to be grade I or II

Astroctyoma

- Radiographic features:
 - Tend to look similar to ependymoma on MRI
 - Less enhancement
 - May be off-center or asymmetric, compared to ependymoma
 - Tend to span over 4-7 levels
 - Thoracic >> cervical
 - Unlike ependymoma, do not often have welldefined planes, and are more infiltrative



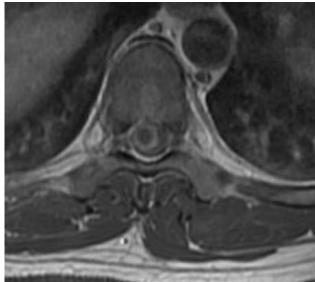


TABLE 2: Spinal cord ependymoma vs astrocytoma.

Diagnostic guide	Ependymoma	Astrocytoma	
Percentage of total intramedullary glial neoplasms (%)	60	33	
Mean age of presentation (years)	39	29	
Most common cord segment affected	Cervical	Thoracic	
Location in the spinal cord	Central	Eccentric	
Haemmorhage ('cap sign')	Common	Uncommon	
Morphology	Well circumscribed	Infiltrative	
Enhancement	Focal, intense homogenous	Patchy, irregular	
Average extent of lesion (vertebral body segments)	3.6	7	
Diffusor tensor imaging (fibre tractography)	Displaces white matter fibres	Disrupts white matter fibres	
Associated syndromes	Neurofibromatosis 2	Neurofibromatosis 1	

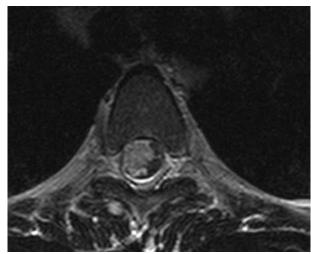
Hemangioblastoma

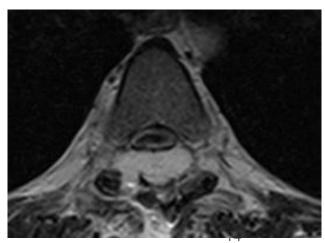
- By far, much more rare accounting for 3-4%
- Do not develop from intrinsic cells of spinal cord, but has close association which vasculature
- Strong association with VHL
 - 30% with spinal hemangioblastoma will have VHL
- Tend to develop in dorsal (posterior) cord
 - Sensory and proprioception symptoms

Hemangioma

- 35 y/o F with 3-month hx of LLE paresthesias
 - 5/5 throughout UE and LE
 - Lumbar MRI unremarkable
 - EMG normal
 - Neuro-axis imaging ordered for further workup



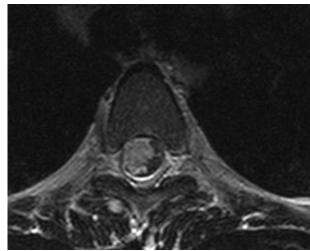


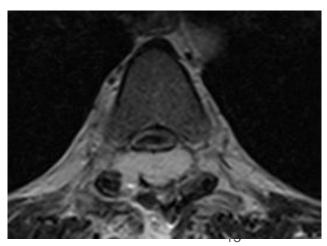


Hemangioma

- Underwent a T3-5 laminoplasty for resection
 - Remained motor intact, with some decreased sensation at T5 level
 - Discharged home POD 2
 - Pathology with cavernous hemangioma

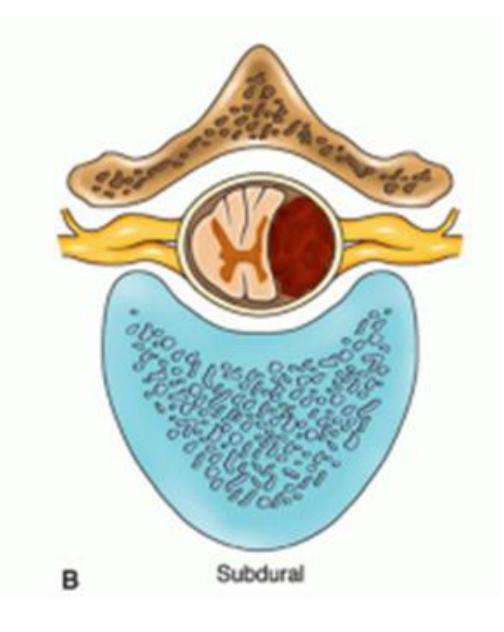






Intradural, Extramedullay

Neoplasm	Relative Frequency	Clinical Correlates*	Typical Location	Imaging Features
Meningioma	Common	Strong female predilection (great- er than for intracranial tumors) Ionizing radiation a documented risk factor Other risk factors: hormonal vari- ation, association with NF2 Symptoms related to compression of spinal cord or nerve roots Most tumors WHO grade I	Thoracic spine much more com- mon than cervical spine; lumbar spine rare Usually lateral to spinal cord (ante- rior location more common when tumor in cervical spine)	T1 iso-/hypointense and slightly T2 hyperintense to spinal cord Intense enhancement with common dural tail Calcification possible at CT
Schwannoma	Common	Association with NF2 Sensory symptoms and pain com- mon Schwannomatosis in young adults without NF1 or NF2; severe pain	Dorsal spinal nerve root Foraminal extension common Spinal cord rare (1%)	Well-circumscribed T1 hypointense/T2 hyper- intense mass; intense enhancement but may be heterogeneous (cystic change) in larger lesions Heterogeneity at MRI correlates with Antoni B tissue Slight hyperattenuation at CT correlates with Antoni A tissue
Melanotic schwannoma	Rare	Association with Carney complex Guarded prognosis		Melanin: T1 hyperintensity
Neurofibroma	Common	Usually asymptomatic (pain rare) Association with NF1 Noonan syndrome with multiple lentigines (NSML)	common "Kissing," paraspi-	Tumors encase rather than displace nerve roots Marked T2 hyperintensity Plexiform tumors: "bag of worms" appearance
Malignant peripheral nerve sheath tumor (MPNST)	Rare	Adults (30–60 years old) most common 50% occur in NF1 (28–36 years old) Prior radiation therapy: 10% Usually radicular pain Poor prognosis, especially in children	Paraspinal most common	May mimic appearance of benign nerve sheath tumor Heterogeneous appearance typical

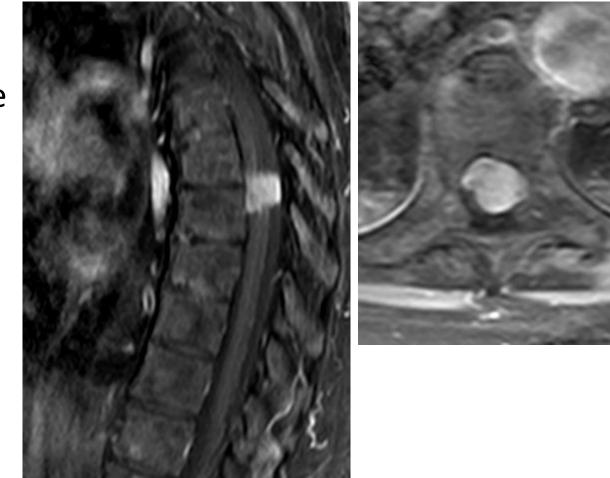


Meningioma

- Second most common extramedullary lesion
- Generally benign, slow growing
 - Arise from arachnoid cap cells of the meninges
 - 70-90% WHO I
- Middle aged females
 - Associated with NF 2
- Thoracic (80%) >> cervical (15%)
- Well circumscribed, associated with a "dural tail"
 - May be associated with Ca++
 - Often lateral to spinal cord

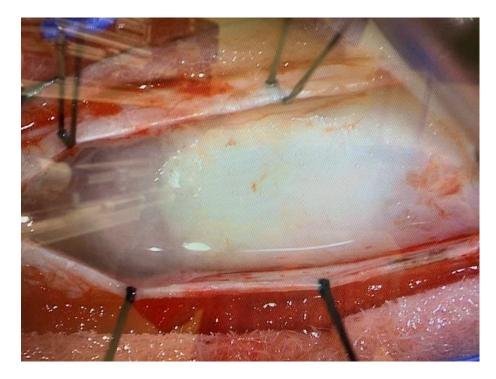
Meningioma

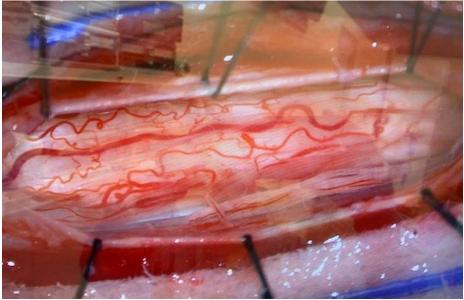
- 78 y/o F with 1 yr hx of progressive motor and sensory decline
 - Went from active retired RN to unable to ambulate and wheelchair bound
 - Bilateral LE with 1/5 throughout
 - T8 sensory level
 - Loss of proprioception



Meningioma

- Taken from T6-8 laminoplasty for resection
- Immediate post op with improvement of LE strength
- 6 months post op, walking with a walker!

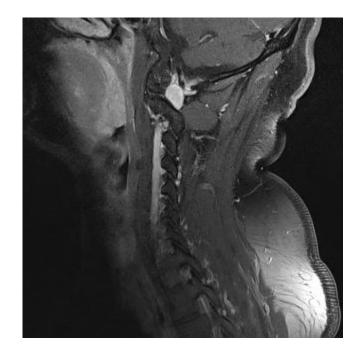


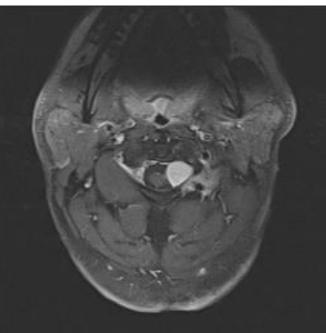


Nerve sheath tumors

• Schwannoma

- Most common nerve sheath tumor
- Peak 5-7th decade, equal sex
- Generally sporadic (95%)
- Arise of the dorsal root
 - Generally does not involve the nerve fibers themselves, but abuts them
 - Well circumscribed
 - Strong enhancement



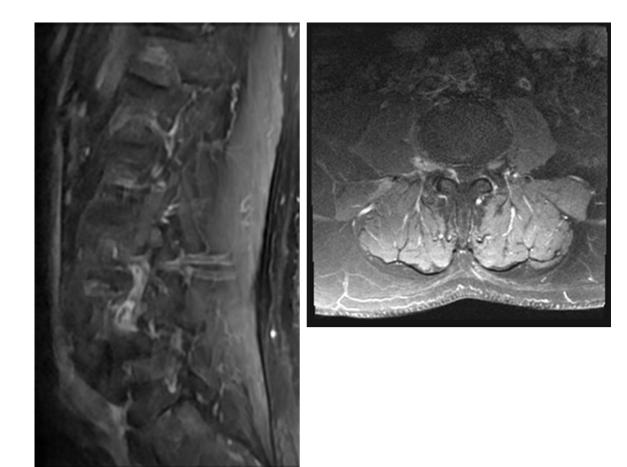


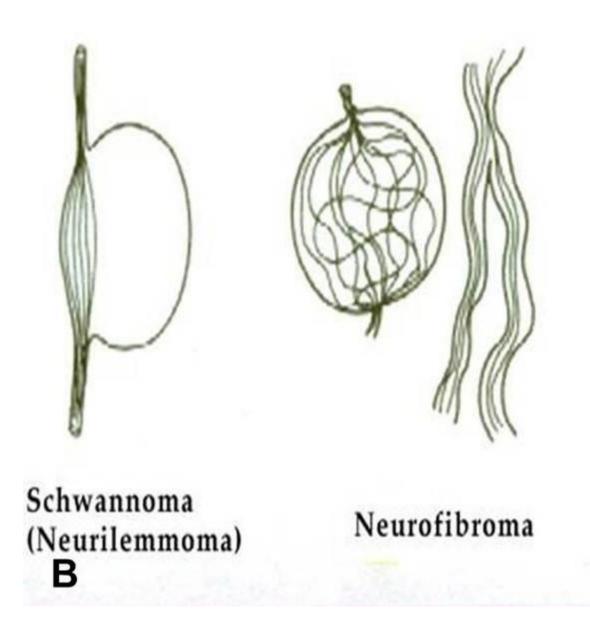
Neurofibroma

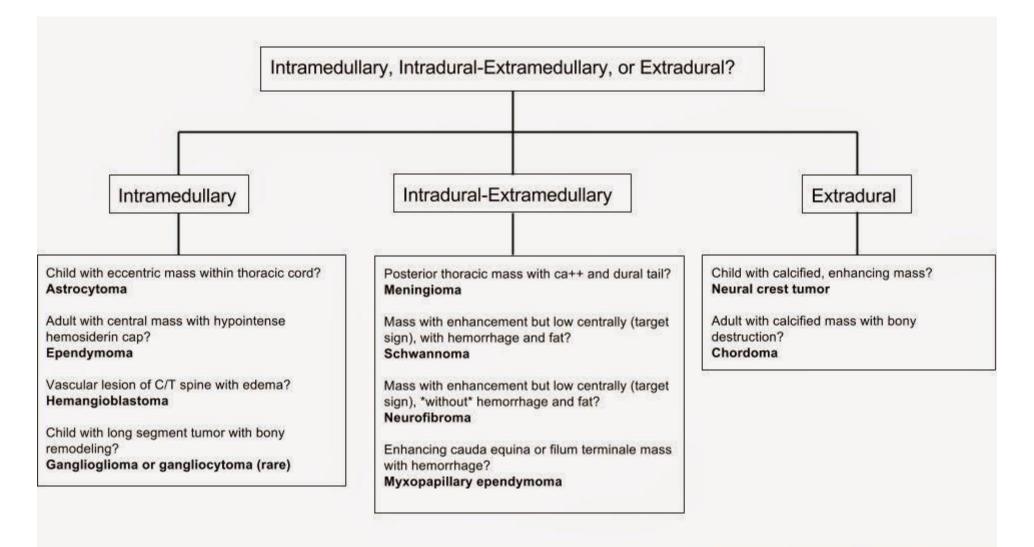
- Classically present with NF 1 >> NF2, but also sporadic
- Also arises for dorsal sensory root
 - Unlike schwannoma, invests (does not abut) into the nerve fibers
 - Not encapsulated
 - Generally unresectable unless nerve root is taken

Neurofibroma

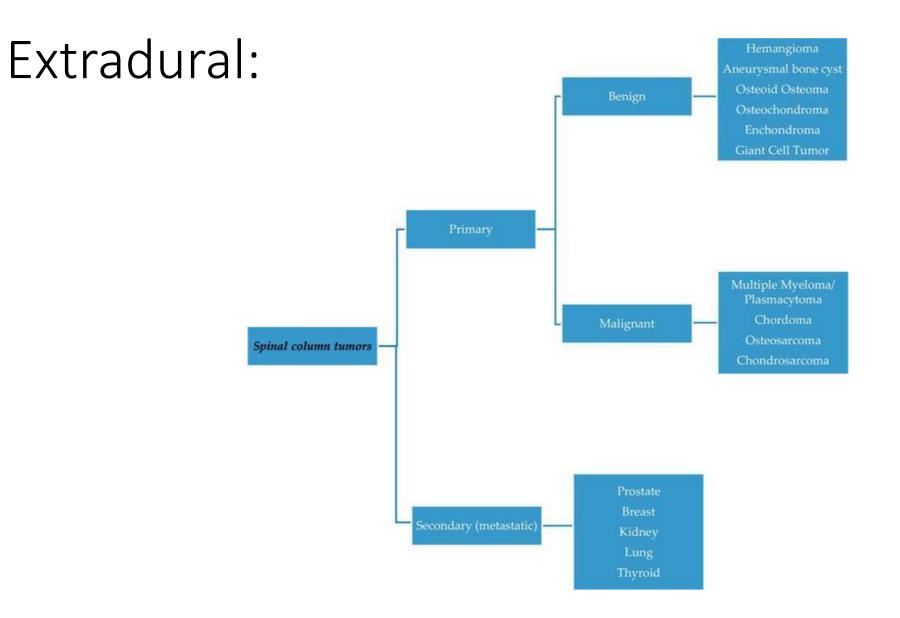
- 76 y/o M with RLE radicular pain and weakness in a L3-4 distribution
- Taken to OR for posterior and then lateral approach for resection
 - Nerve was completely intertwined with tumor regardless of approach
 - Tumor was unresectable











Vertebral hemangioma

- By far, the most common spinal bone "tumor"
 - Up to 30% on MRI
 - Benign
 - Generally discovered on routine MRI for degenerative changes
 - Almost always asymptomatic
 - "salt and pepper" appearance on MRI
 - Does not need neurosurgery intervention or consult

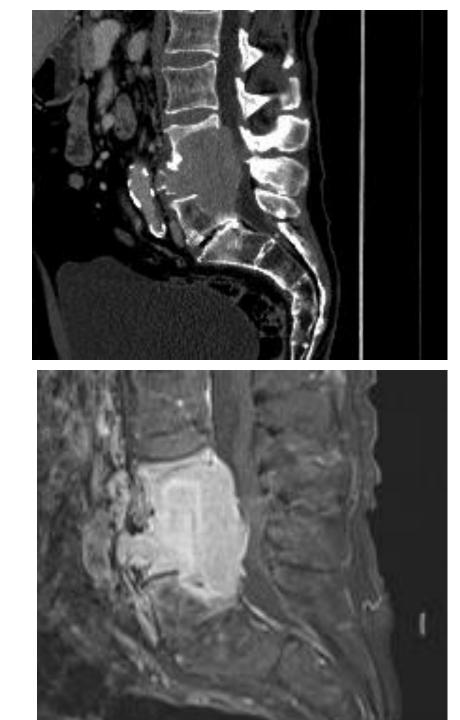


Multiple myeloma (plasma cell)

- Most common primary malignant bone neoplasm
 - Account for approximately 50% of all vertebral body tumors
- Arises from monoclonal proliferation of plasma cells of red marrow
- Arises in older adulthood (50-70)
 - M >> F (2:1)
- Can arise anywhere in axial skeleton, but vertebral column most common
- Presents with pain secondary to pathologic fracture
 - Anemia
 - Renal failure
 - Proteinuria
 - hypercalcemia

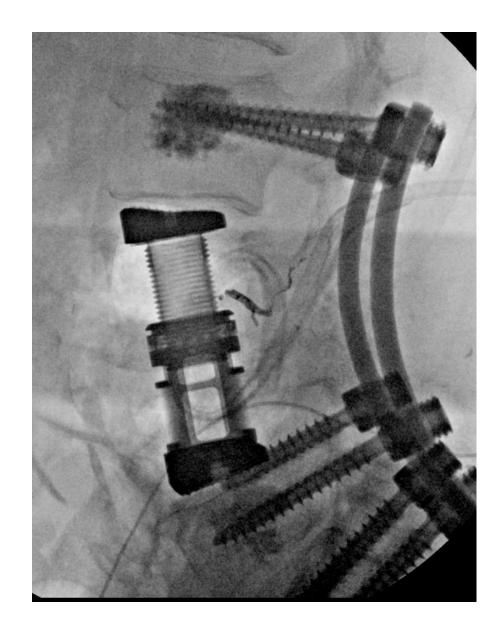
Boney/Extradural

- 74 y/o M with 1 year hx of LBP and LE radicular pain
 - Presented to ED
 - Neuro intact
 - Very lytic on CT
- Enhances brightly in MRI



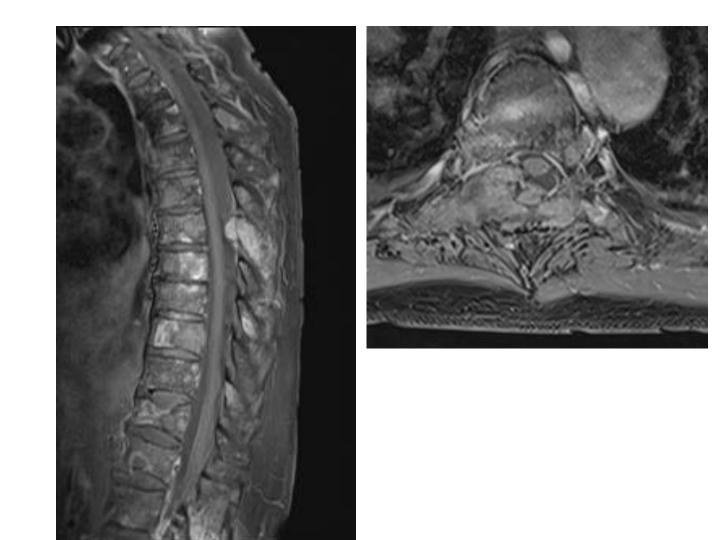
Boney/extradural

 Multiple myeloma is extremely radiation sensitive, and is often the first line treatment



Prostate

- When advanced, 90% prostate mets go to spine
- 81 y/o M with mid back pain
 - Legs feeling weaker
 - Requiring the use of a cane to ambulated



Spine Instability Neoplastic Score (SINS)

- SINS is used to assess the stability of the spine in patients with metastatic spinal cord compression
- It has near-perfect inter- and intraobserver reliability in determining three clinically relevant categories of stability.
- The sensitivity and specificity of SINS for potentially unstable or unstable lesions were 95.7% and 79.5% respectively
- Location
 - 3 points: Junctional (C0-C2, C7-T2, T11-L1, L5-S1)
 - 2 points: Mobile spine (C3-C6, L2-L4)
 - 1 point: Semi-rigid (T3-T10)
 - 0 points: Rigid (S2-S5)
- Pain relief with recumbency and/or pain with movement/loading of the spine
 - 3 points: Yes
 - 1 point: No (occasional pain but not mechanical)
 - 0 points: Pain free lesion
- Bone lesion
 - 2 points: Lytic
 - 1 point: Mixed (lytic/blastic)
 - 0 points: Blastic
- Radiographic spinal alignment
 - 4 points: Subluxation / translation present
 - 2 points: De novo deformity (kyphosis / scoliosis)
 - 0 points: Normal alignment
- Vertebral body collapse
 - 3 points: >50% collapse
 - 2 points: <50% collapse
 - 1 point: No collapse with >50% body involved
 - 0 points: None of the above
- Posterolateral involvement of the spinal elements (facet, pedicle or costovertebral joint fracture or replacement with tumor)
 - 3 points: Bilateral
 - 1 point: Unilateral
 - 0 points: None of the above

Interpretation of the SINS score

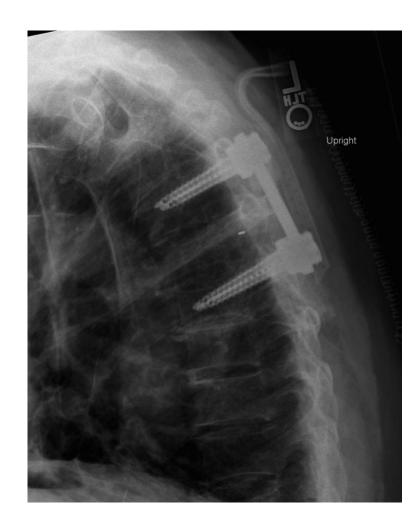
- sum score 0-6: stable
- sum score 7-12: indeterminate (possibly impending) instability
- sum score 13-18: instability
- SINS scores of 7 to 18 warrant surgical consultation.

Reference

 Fisher CG, DiPaola CP, Ryken TC, Bilsky MH, Shaffrey CI, Berven SH, et al. A novel classification system for spinal instability in neoplastic disease: an evidence-based approach and expert consensus from the Spine Oncology Study Group. Spine 2010; 35:E1221-9.

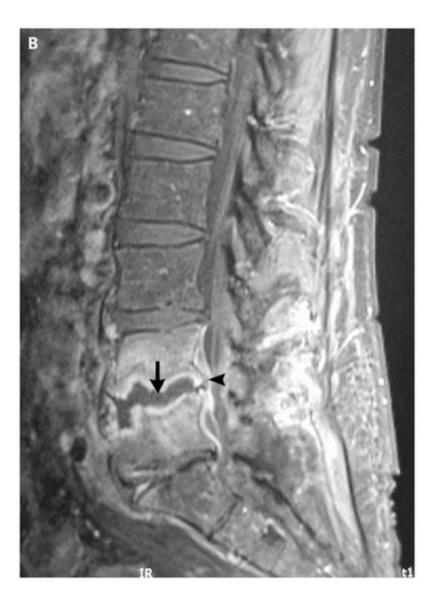
Epidural abscess

- Did well post op, with improvement of LE function
- Will receive radiation



Epidural abscess

- Generally presents with back pain
 - +/- radiculopathy
- Most common location in lumbar spine
 - If pain only, not a neurosurgical emergency (or even operative indication)
 - Treat with Abx
- Often associated with discitis/osteomyelitis





Osteomyelitis

Metastatic disease

Infection vs Metastatic disease

Sample distribution based on magnetic resonance imaging abnormalities

A 1	Spondylitis (n=22)		Metastasis (n=13)	
Abnormality	Available	Not available	Available	Not available
End plate irregularity	19 (86.4)	3 (13.6)	0	13 (100.0)
Narrowing of discs	18 (81.8)	4 (18.2)	0	13 (100.0)
Paravertebral mass	20 (90.9)	2 (9.09)	12 (92.3)	1 (7.7)
Paravertebral abscess	19 (86.4)	3 (13.6)	0	13 (100.0)
Spinal canal stenosis	22 (100.0)	0	13 (100.0)	0
Abnormalities of spinal medulla signal	22 (100.0)	0	13 (100.0)	0
Skip Lesion	3 (13.6)	19 (86.4)	8 (61.5)	5 (38.5)
Involvement of sequential vertebral segment	19 (86.4)	3 (13.6)	5 (38.5)	8 (61.5)

Values are presented as number (%).

<u>Asian Spine J.</u> 2015 Oct; 9(5): 776–782.

Compartment	Malignant	Benign	Nontumorous Growths
Extradural	Metastases	Hemangioma	
	Myeloma	Aneurysmal bone cyst	
	Lymphoma	Giant cell tumor	
	Ewing sarcoma	Osteoid osteoma	
	Osteosarcoma	Osteoblastoma	
	Chordoma	Osteochondroma	
	Leukemia	Eosinophilic granuloma	
	Chondrosarcoma		
Intradural–extradural	Metastases	Nerve sheath tumors	Lipoma
	Malignant nerve sheath tumors	Schwannoma	Epidermoid
		Neurofibroma	Dermoid
		Meningioma	Arachnoid cyst
		Hemangiopericytoma	
		Paraganglioma	
Intramedullary	Astrocytoma	Astrocytoma	
	Metastases	Hemangioblastoma	
		Ependymoma	

Table 12.1 Differential Diagnosis of Spine Lesion by Anatomic Compartment

Thank you!

- Questions?
- Katie Krause- Neurosurgeon
- <u>Katie.Krause@virginiamason.org</u>
- 262-224-7070