A Case-Based Review of Intradural, Extramedullary Tumors and Tumor Mimickers

Jared T. Verdoorn, MD

Mayo Clinic Department of Radiology
Rochester, MN

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DISCLOSURE

Relevant Financial Relationship(s)

None

Off Label Usage

None

MAYO CLINIC

Educational Objectives

1. Recognize frequently encountered intradural, extramedullary spine tumors and their imaging appearances
2. Understand imaging appearances of rare intradural, extramedullary tumors and features that may help distinguish them from more common tumors
3. Introduce several benign masses/mass-like processes that may be tumor mimickers
4. Develop a differential diagnosis for cauda equina region masses

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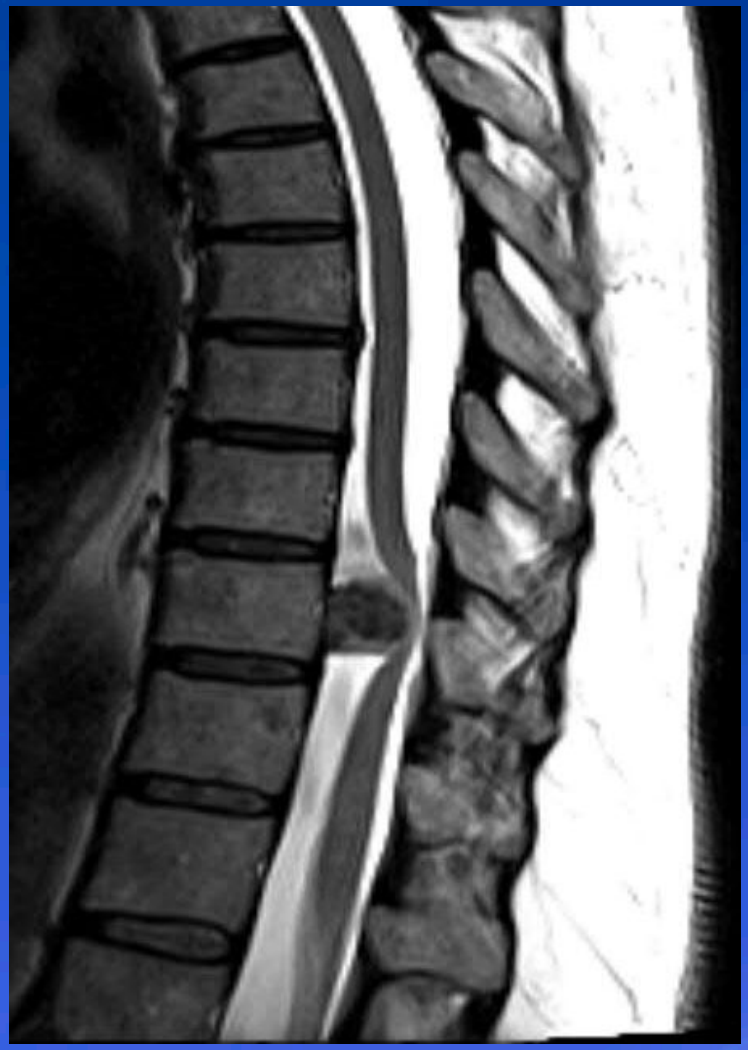
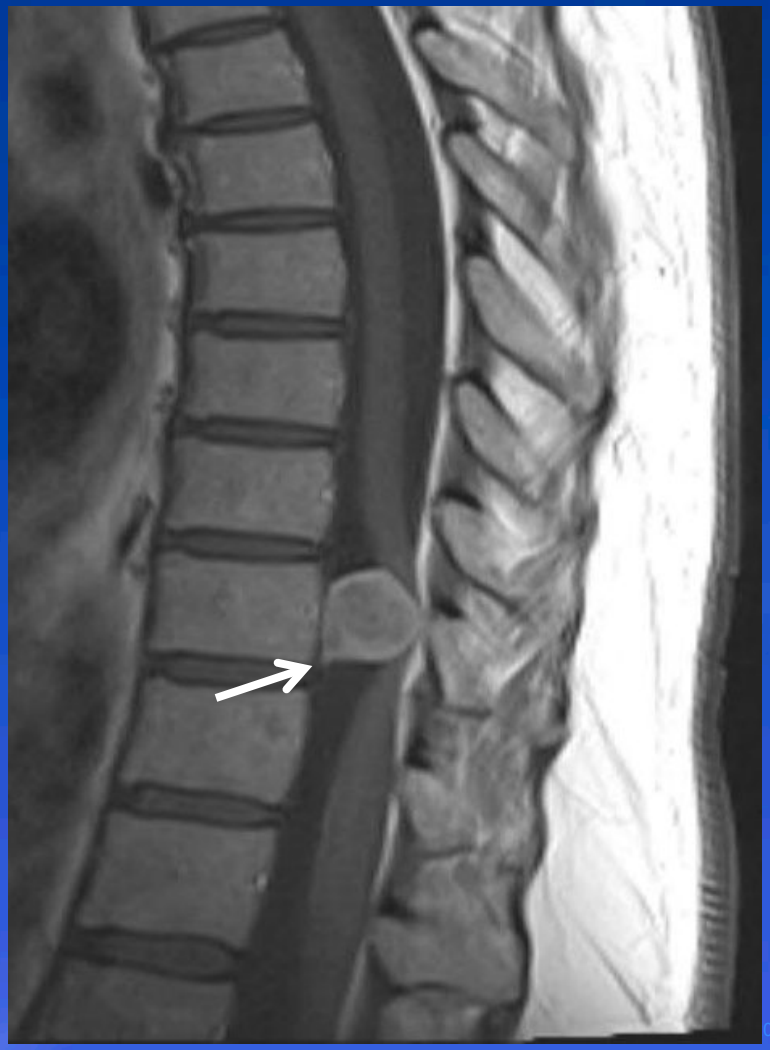
Case 1

56 yo female

Years of progressive lower abdominal and leg pain → worsening urinary incontinence, saddle anesthesia, and buttock/groin pain

Sag T2





Sag T1 Gad

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Meningioma

- 2nd most common intradural, extramedullary tumor
- Thoracic spine (80%)
- Middle aged women (80% female), myelopathy
- MRI
  - Variable signal characteristics, may be markedly T1 and/or T2 hypointense to cord if calcified
  - Round/ovoid > plaque-like
  - Homogeneous enhancement
    - Dural-based, dural tails, more common ventrally
    - Cystic degeneration rare
  - +/- calcification, less common than intracranial
  - No hyperostosis

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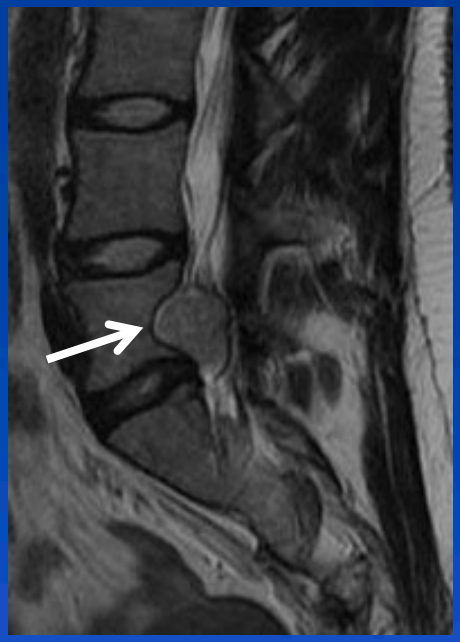
Case 2

24 yo female
Low back pain and left leg paresthesia



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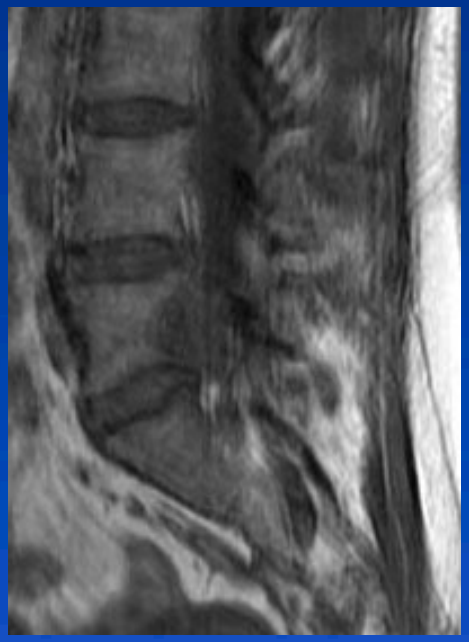

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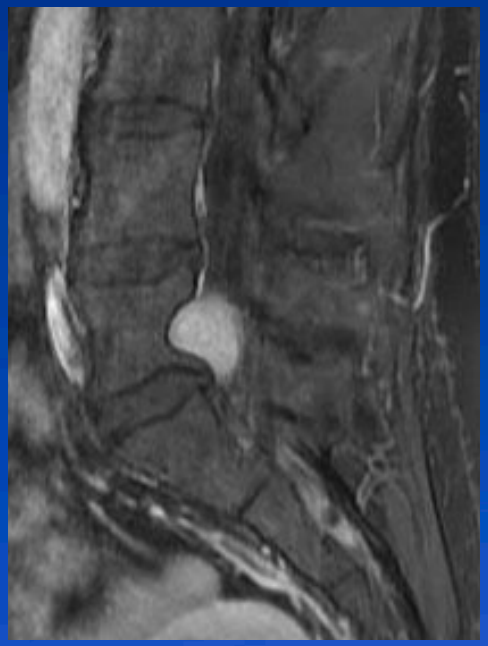

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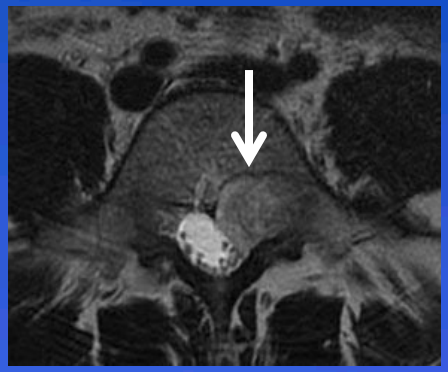

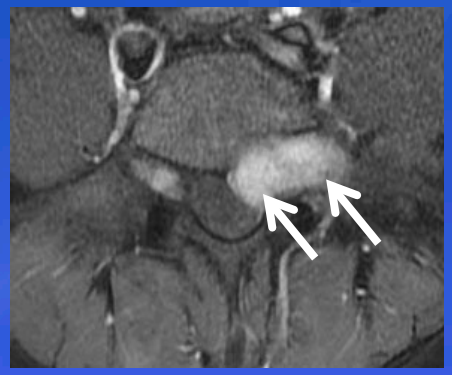

Schwannoma

- Most common intradural, extramedullary tumor
  - Intradural/extradural (dumbbell, ~15%)
  - Entirely extradural (extraforaminal, ~15%)
- MRI
  - T1 iso and T2 hyperintense to cord
  - Round/ovoid, well circumscribed
  - Typically homogeneous enhancement
  - Rarely calcify or hemorrhage (10%)
  - Can have cystic change if large
  - Difficult to differentiate from neurofibroma
    - Fusiform shape/“target sign” (T2 hyperintense rim surrounding dense, low signal stroma) → neurofibroma
    - Hemorrhage/cystic change → schwannoma

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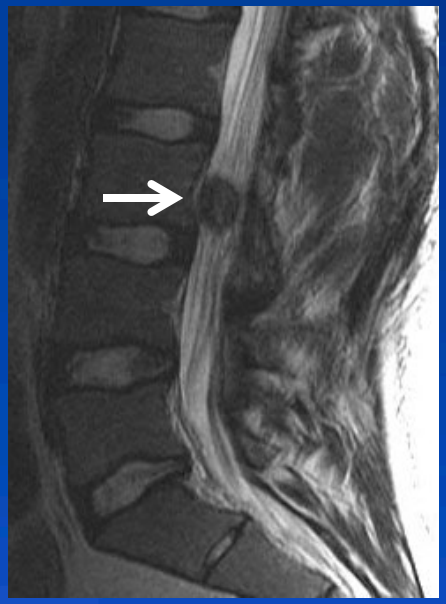
Case 3

32 yo male
4 years of pain/paresthesia in left low back, buttock, and posterior thigh



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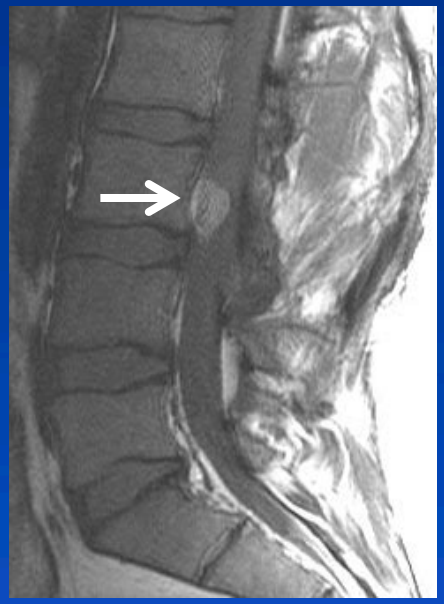

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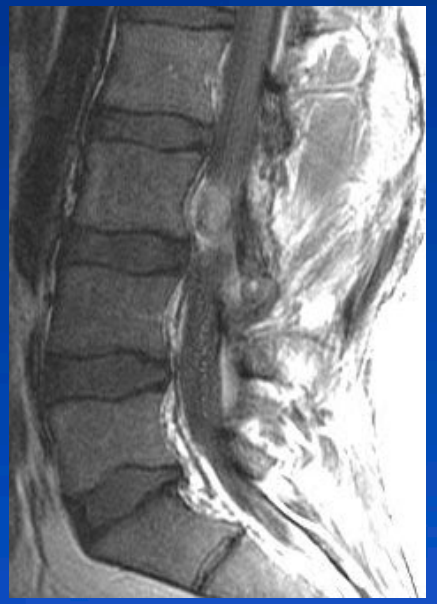

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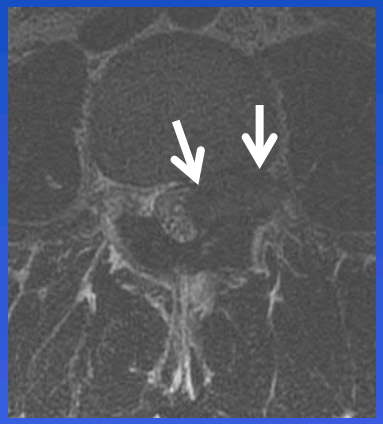

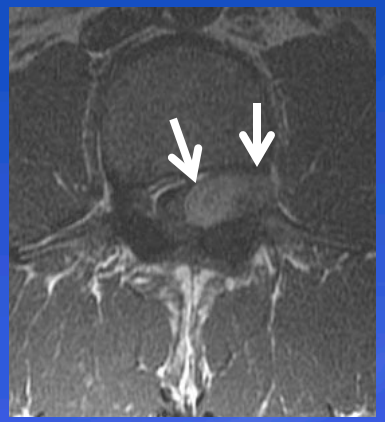

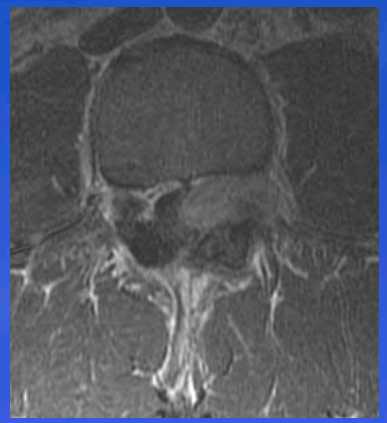

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Melanotic Schwannoma

- Schwannoma containing melanin
- MRI
  - Intrinsically T1 hyperintense
  - T2 hypointense relative to typical schwannoma
- May locally recur or metastasize (10-40%)
  - Gross total resection and close follow-up
- Carney Complex (present in 50%)
  - Melanotic schwannomas, myxomas, endocrine tumors, skin pigment abnormalities

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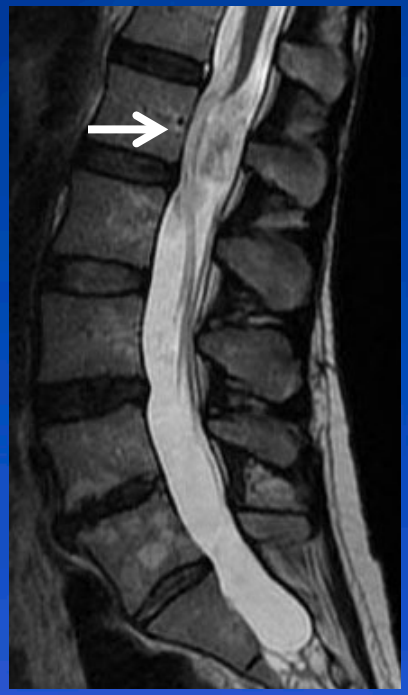
Case 4

47 yo female
Months of bilateral leg heaviness and left leg numbness and tingling



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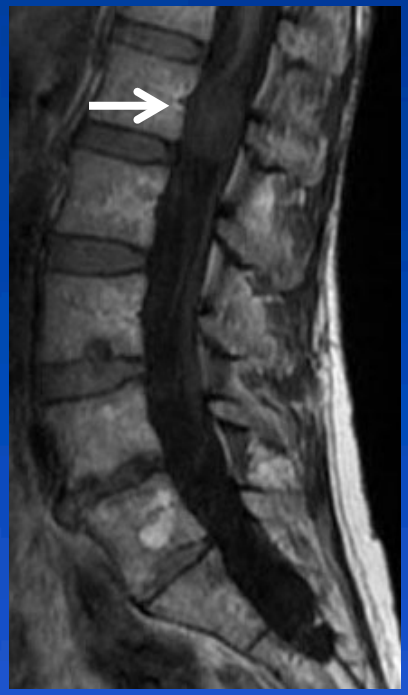

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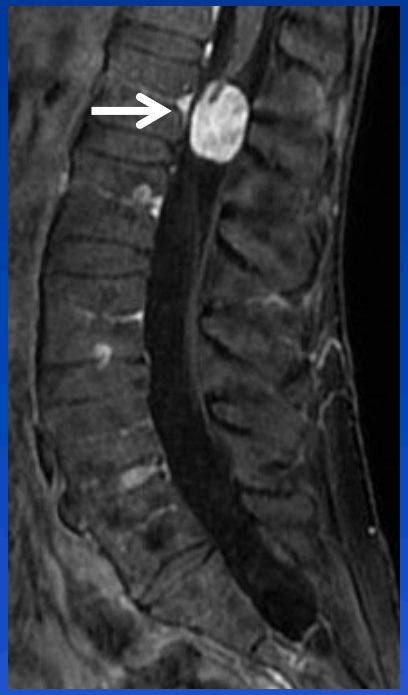

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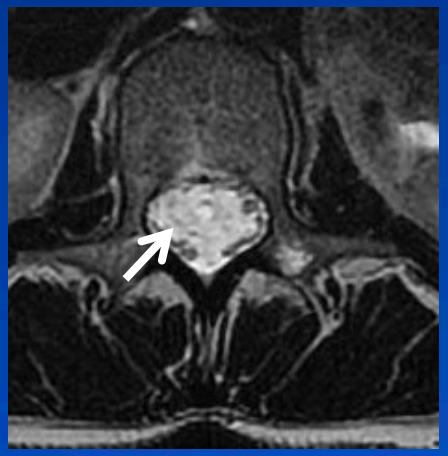

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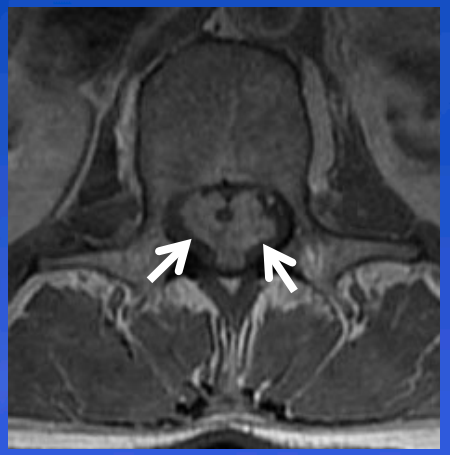

T1 Gad

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Myxopapillary Ependymoma

- Most common primary tumor of conus/cauda equina
- Arises from ependymal cells of filum terminale
- Slow onset of pain, neural compression symptoms
- MRI
  - Ovoid or “sausage-shaped”
  - T2 hyper and T1 iso/hyperintense to cord (mucin)
    - +/- T2 hypointense hemosiderin peripherally
  - Homogeneous enhancement
  - May fill/expand thecal sac with osseous remodeling
  - Nerve roots splayed around the mass
- WHO Grade I but may have CSF dissemination

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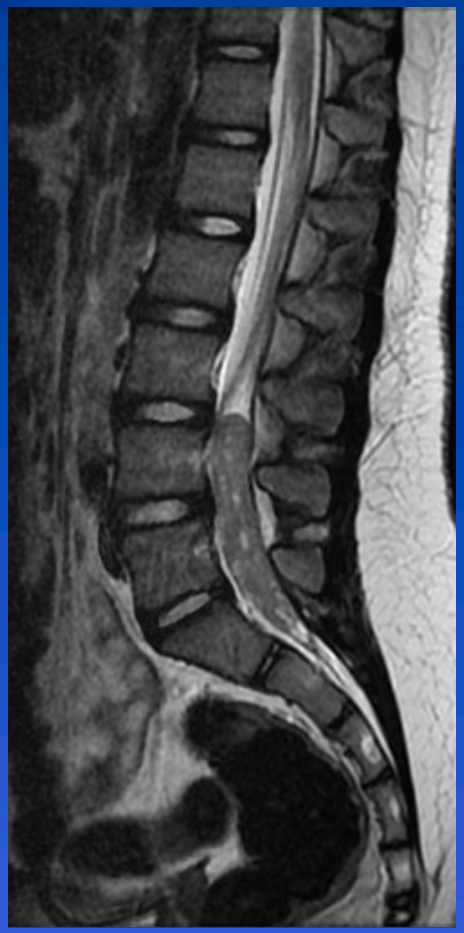
Companion Case 4-1

13 yo female with progressive low back pain



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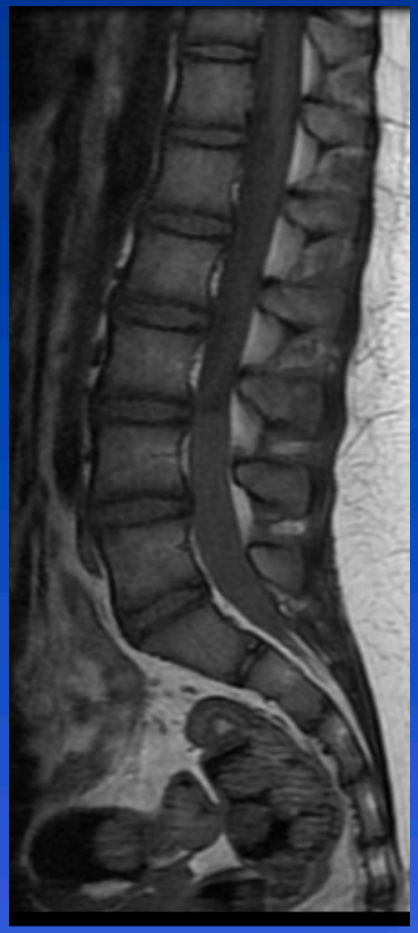

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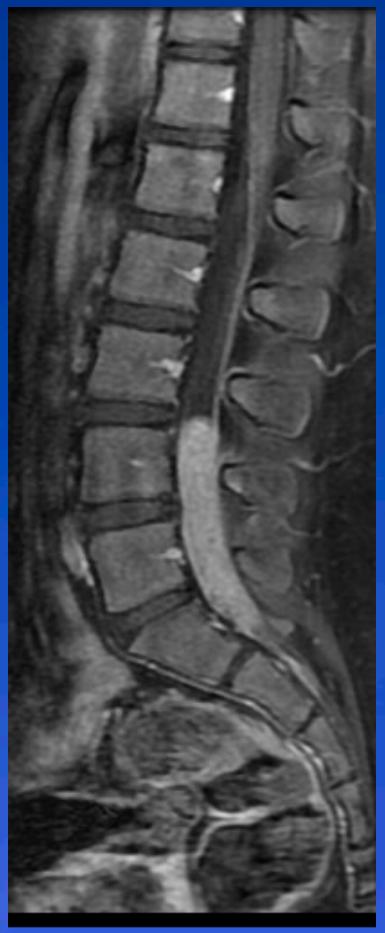

T1 Gad

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Ewing Sarcoma

- Primary extraosseous Ewing sarcoma can (very rarely) present as an intradural mass
- Main differential diagnostic consideration is myxopapillary ependymoma (MPE)
  - Note the relative T2 hypointensity is atypical for MPE, which should raise possibility of alternative considerations

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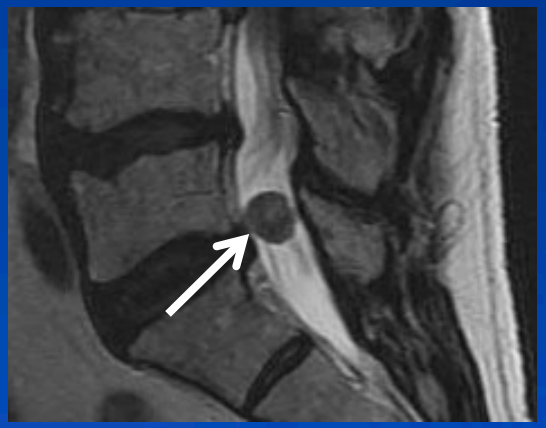
Case 5

60 yo male
Chronic radicular low back pain, history of suspected schwannoma
More recent intermittent slurred speech and tremulousness

5 yrs ago



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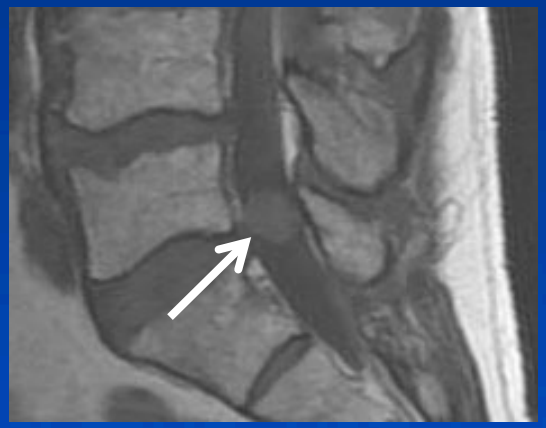

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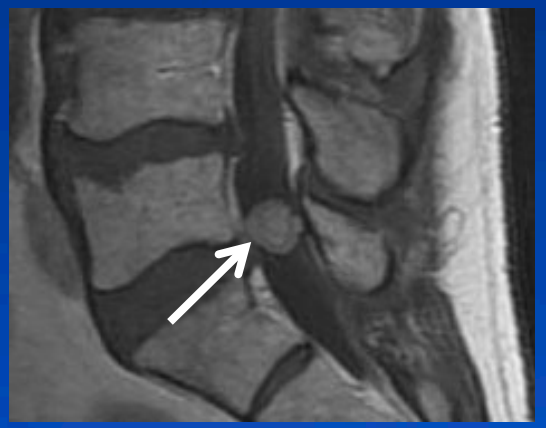
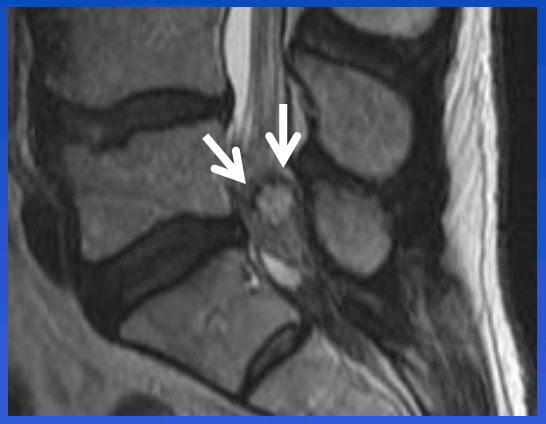
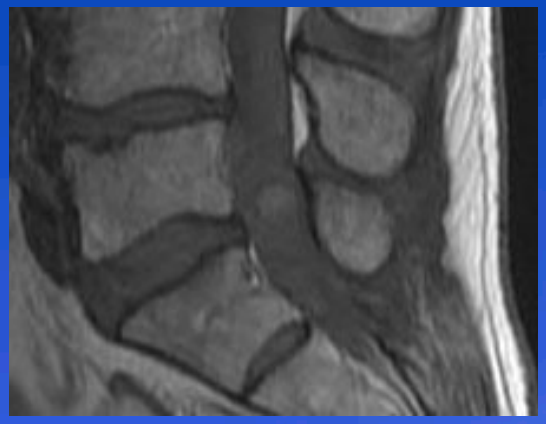
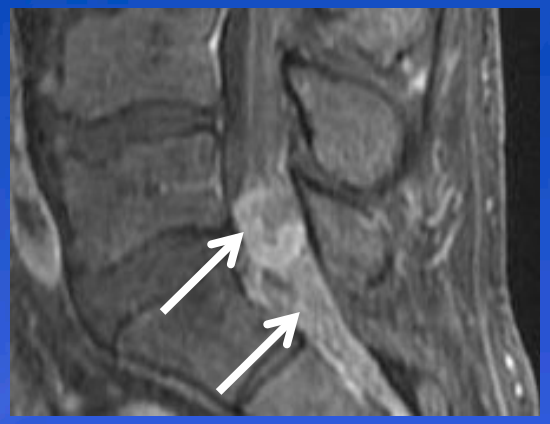


T1 Gad

Now







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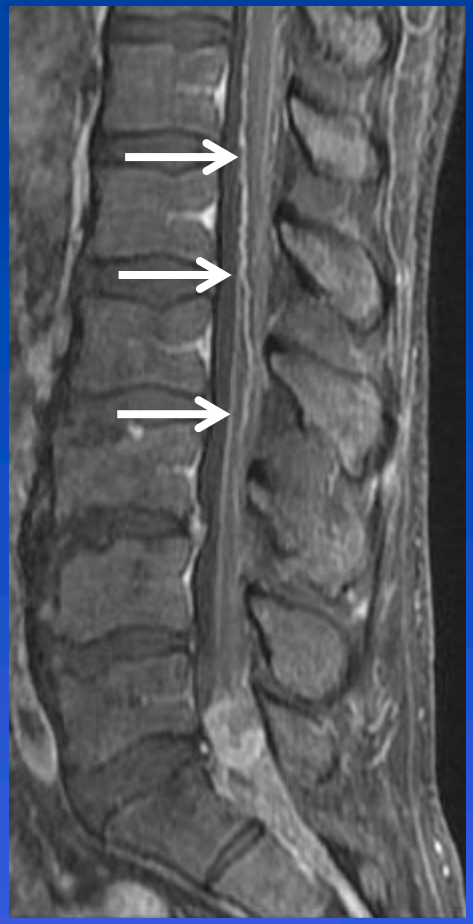
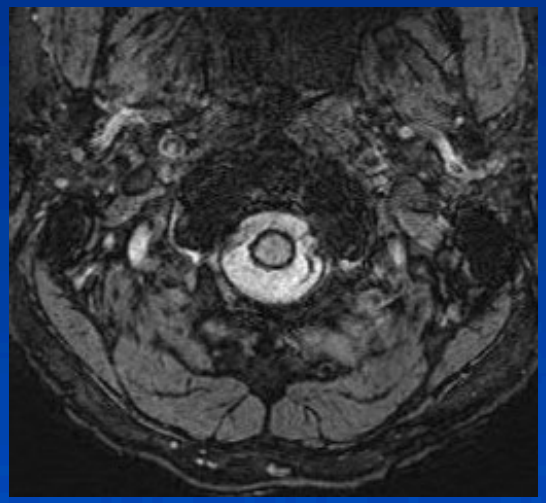
Case 5

Vascularity



T1 Gad

Siderosis



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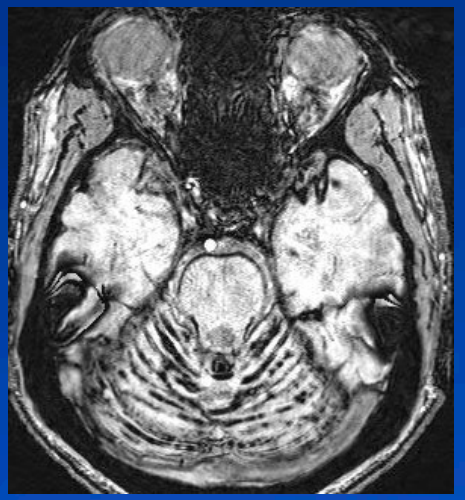

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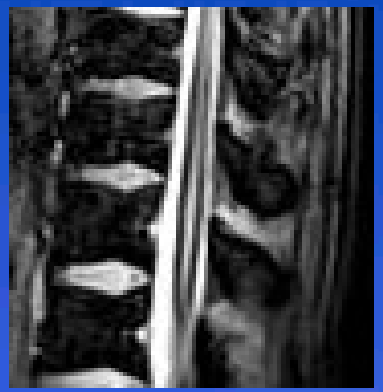

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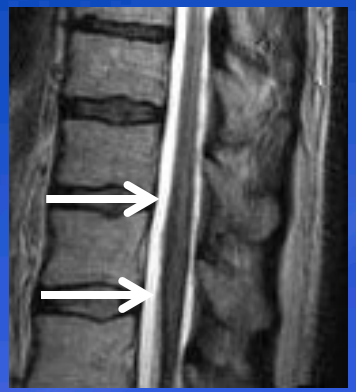

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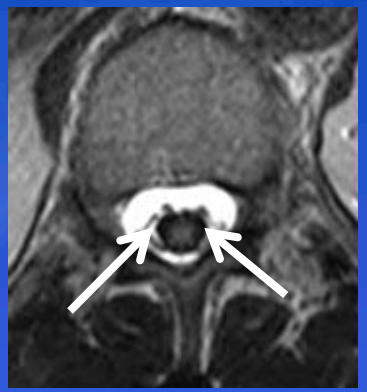

T2

Paraganglioma

- Rare intradural, extramedullary mass
- Arises in filum terminale; cauda equina location
- Slow onset back pain, neural compression
  - Catecholamine activity rare (1-3%)
- Imaging
  - T1 iso and T2 hyper to cord
    - Hypo if prior hemorrhage (hemosiderin)
  - Avidly enhancing, usually positive on MIBG scan
  - Prominent vascularity/draining veins
  - +/- cystic change and hemorrhage, siderosis
- DDx: myxopapillary ependymoma, schwannoma, mets, hemangioblastoma

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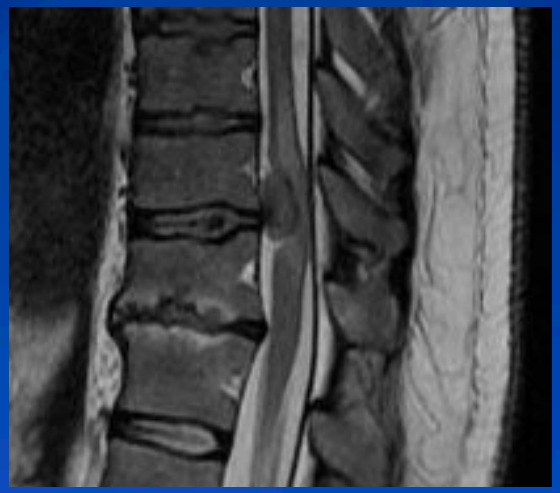
Case 6

43 yo female
1 year of urinary urgency, intermittent fecal incontinence, leg weakness and balance difficulties with falls



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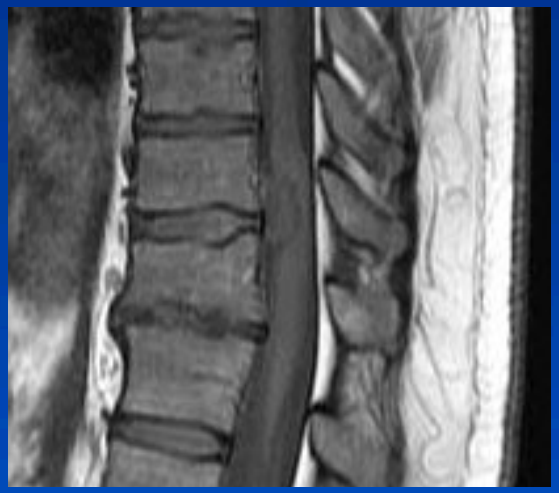

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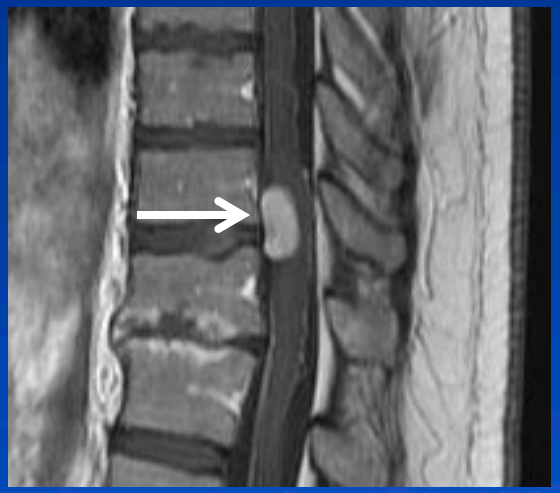

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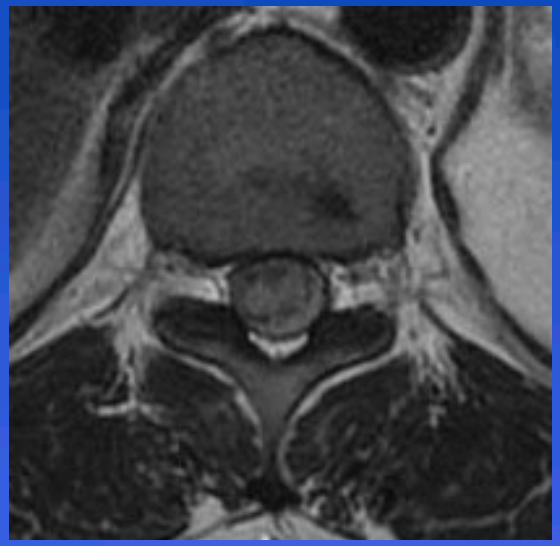

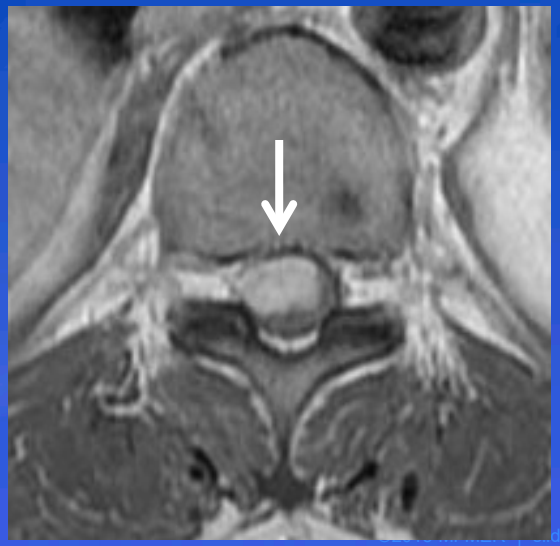

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Solitary Fibrous Tumor / Hemangiopericytoma

- Rare intradural, extramedullary tumor
  - Extradural, paraspinal locations also
- 60% thoracic, slow onset pain/compressive symptoms
- WHO 2016 classification
  - Same tumor genetically with differing phenotypes
    - SFT: Grade I, resection +/- radiation
    - HPC: Grade II-III, resection + radiation, recurrence/mets more common
- MRI
  - Similar to meningioma if small intradural, extramedullary
  - Osseous destruction, paravertebral soft tissue mass, more heterogeneous/vascular if large, higher grade

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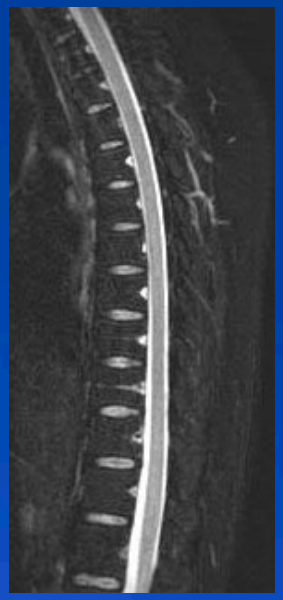
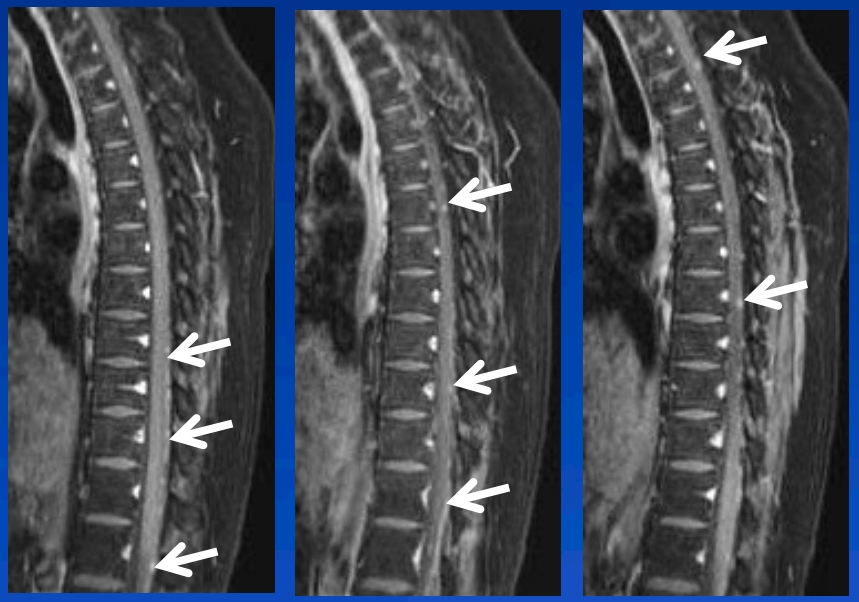
Case 7

35 yo female

History of triple negative breast cancer with new headaches and right leg weakness

T2





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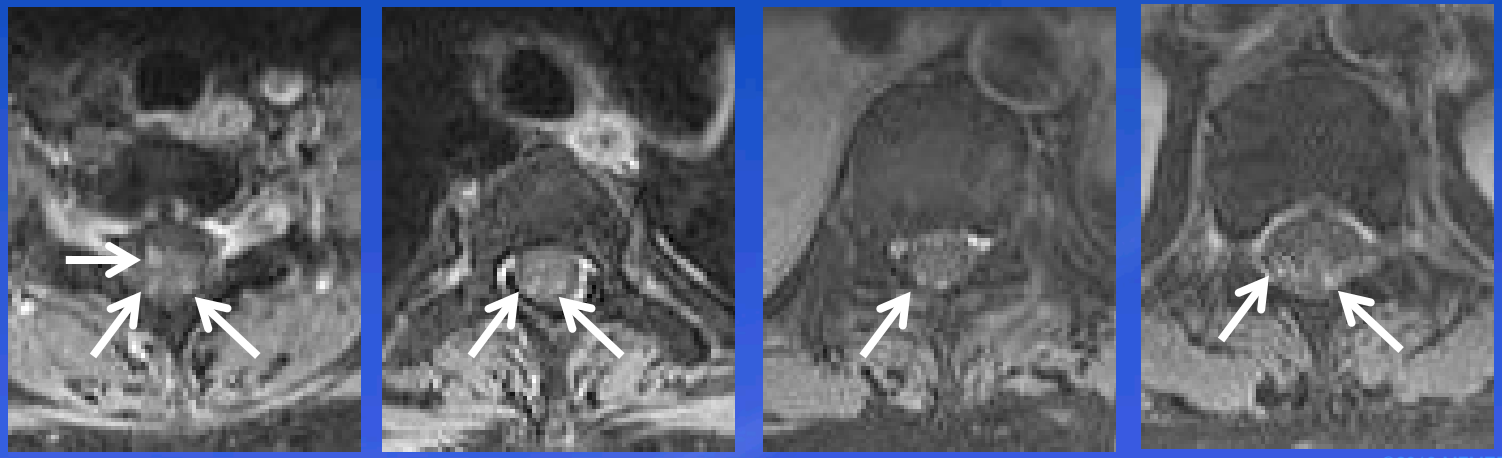

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Leptomeningeal Metastases

- May occur along entire neuraxis
- Typically advanced disease with other mets – history key
  - Poor survival - months
- Common tumors
  - Solid: hematogenous spread
    - Breast, lung, melanoma
  - Primary CNS: “drop” mets
    - Medulloblastoma, ependymoma, germinoma most common
  - Lymphoma and leukemia
- False negative CSF cytology in 40%
  - MRI prior to surgery and prior to LP if possible
- MRI
  - May be subtle, careful scrutiny
  - Varied appearances
    - Typically multifocal
    - Nodular enhancement more specific
    - “Sugar-coating” of cord/nerve roots
    - Diffuse thickening or nodularity of cauda equina
- PET-CT
  - May see increased FDG uptake, non-specific

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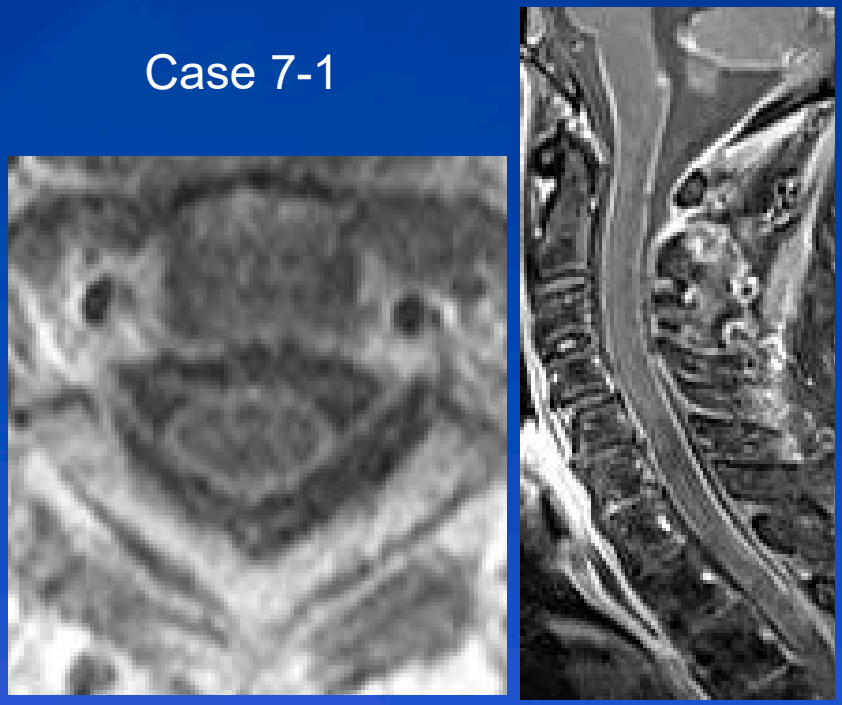
Companion Cases 7-1 and 7-2

Case 7-1



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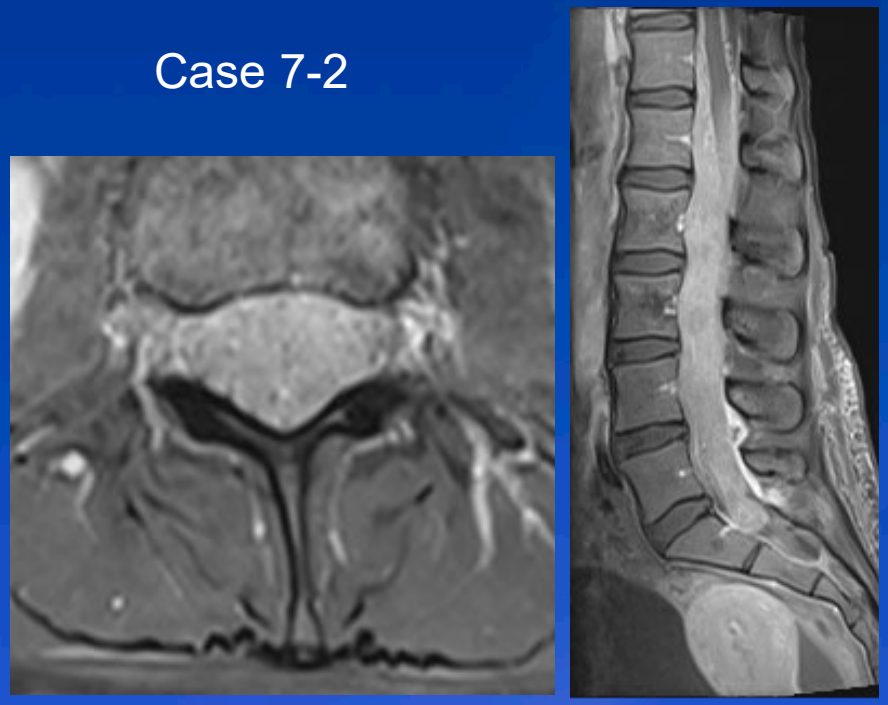
Case 7-2



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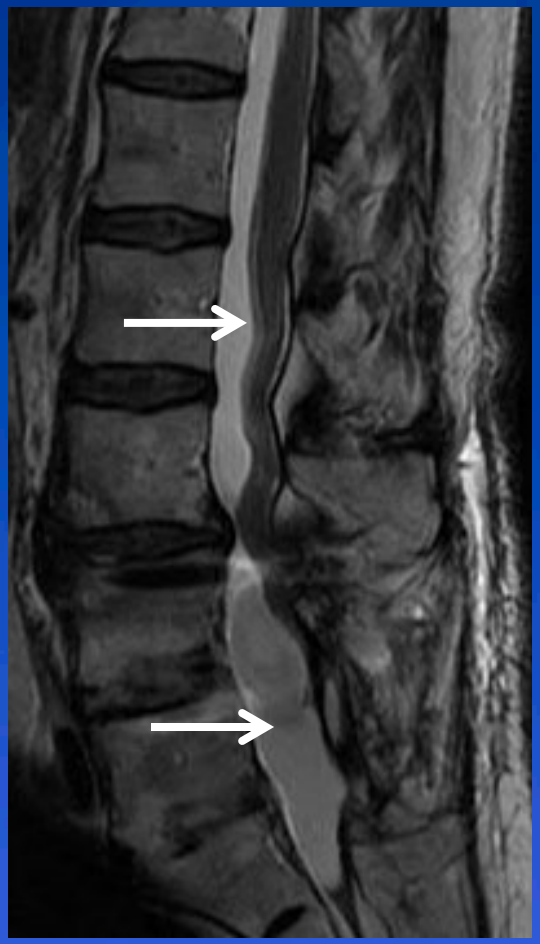
Additional appearances of leptomeningeal metastases including “sugar coating” (Case 7-1) and diffuse cauda equina nerve root thickening (Case 7-2).

MAYO CLINIC

Case 8

73 yo female
History of remote L4-S1 decompression and fusion
Persistent low back pain and lower extremity numbness for years



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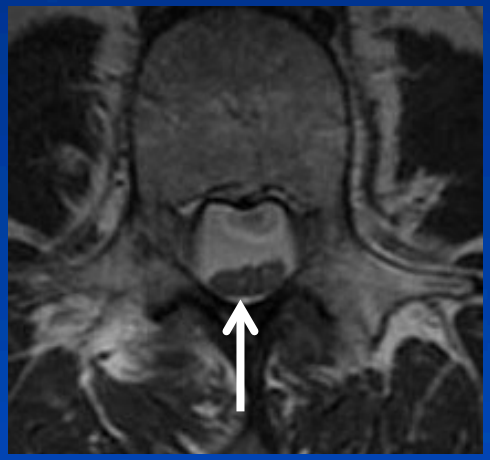
L2 level



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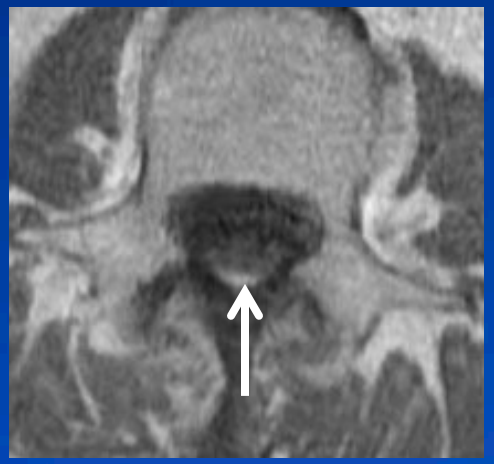
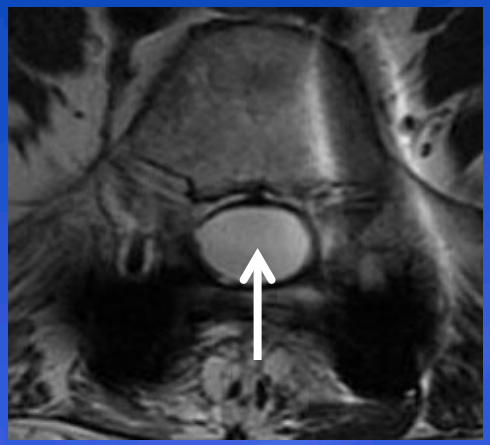
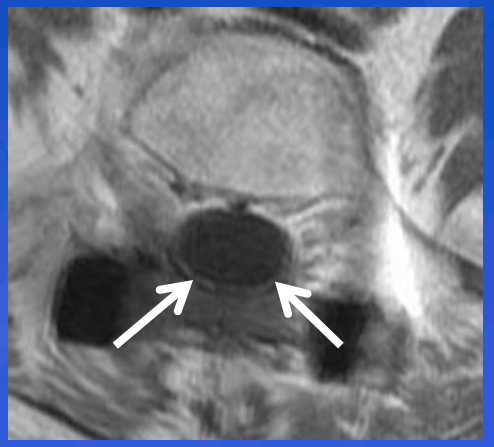


T1 Gad

L5 level





MAYO CLINIC

Arachnoiditis

- Inflammatory changes in the arachnoid
- Etiology
  - Post-op (up to 1 in 6 patients) > prior trauma or infection
- Symptoms variable: none to severe
- MRI
  - Peripheralized or centrally clumped nerve roots
    - "Empty sac" and "pseudo-mass" signs (seen in the example case at L5 and L2 levels, respectively)
  - +/- nerve root and dural thickening
  - +/- focal adhesions, CSF loculations
  - +/- mild enhancement (smooth or nodular)
    - Enhancement does not correlate with symptoms
  - Arachnoiditis ossificans – rare - calcification

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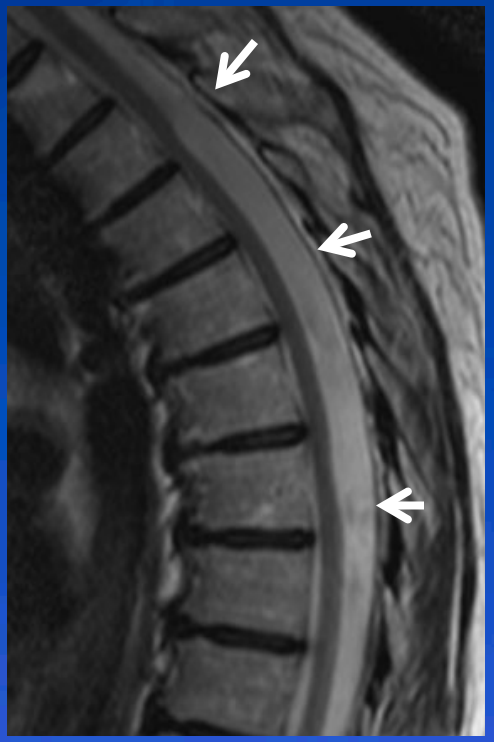
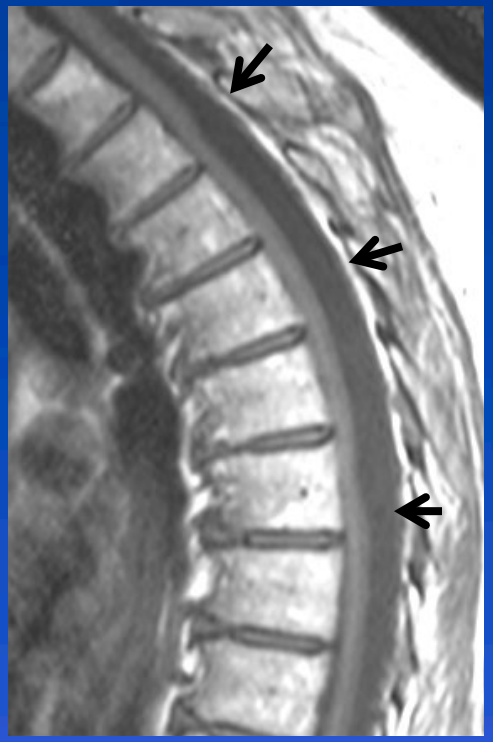
Case 9

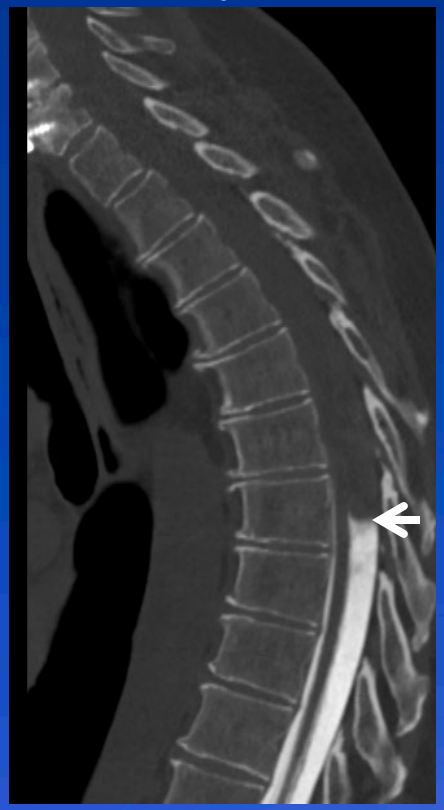


T2

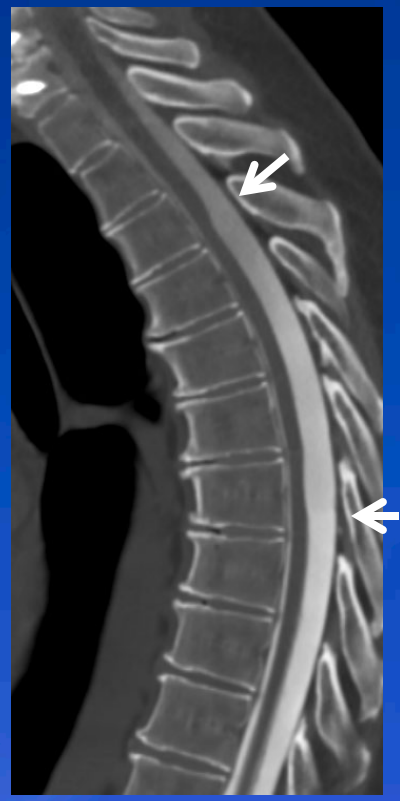


T1

Dynamic CT Myelogram



Before roll and head-down tilt



After roll and head-down tilt*

*Note that the cyst fills quickly and is virtually indistinguishable at this point and would therefore be difficult to identify on conventional CTM.

Arachnoid Cyst

- aka Meningeal cyst
- Extramedullary, loculated CSF collection, may be intradural or extradural
- MRI
  - Focal dorsal mass effect/cord deformity, may be subtle
  - Similar appearance to “scalpel sign” of arachnoid web; these entities may exist on a continuum
  - High resolution 3D sequences such as CISS/FIESTA may allow visualization of cyst walls
- CT myelogram
  - Can observe delayed/incomplete contrast filling of the cyst relative to the subarachnoid space, but often the cyst fills relatively quickly and modified dynamic technique should be considered as in this example case
- Treatment is laminectomy with complete cyst wall excision if possible (> marsupialization, shunt, or fenestration)

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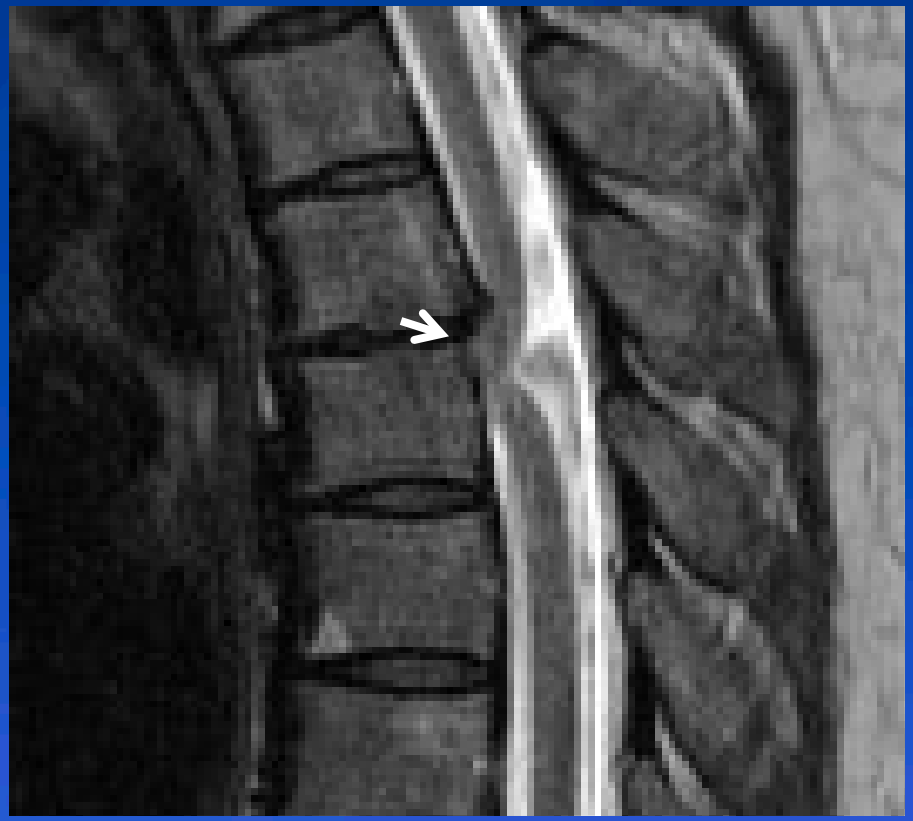
Companion Case 9-1



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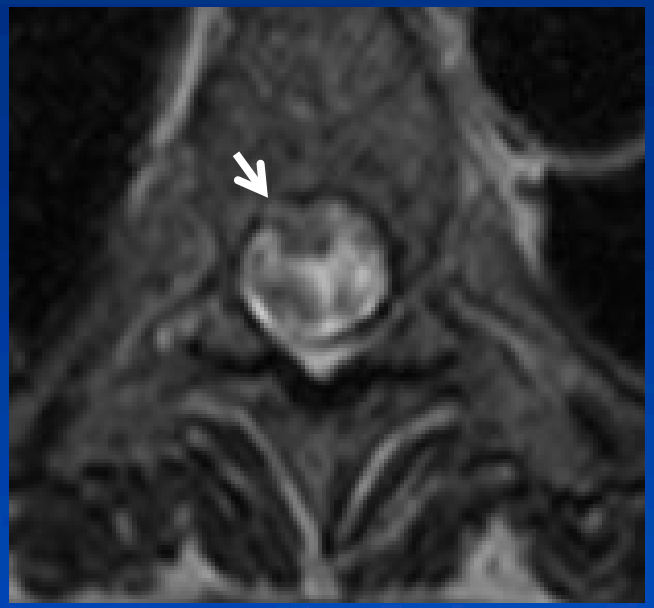
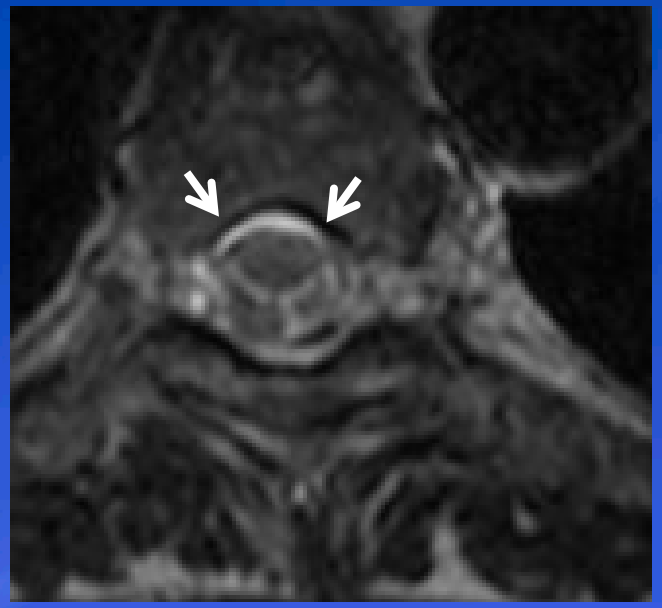


T2



T2

Note the thin ventral extradural fluid indicated by the arrows on the bottom right image, compatible with dural defect.

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Idiopathic Ventral Cord Herniation

- Herniation of the spinal cord through a dural defect
  - Dura may be weak congenitally or secondarily due to disc disease, CSF pulsation, or underlying adhesion
- Symptoms often chronic leg pain, weakness
- MRI
  - Focal cord deformity and anterior displacement
    - Need to differentiate from arachnoid web/cyst
  - Mid thoracic spine most common
  - Focal ventral extradural fluid collection may be hint
  - CT myelography often confirmatory
    - Distorted cord outside of dural margin +/- extradural contrast through defect
- Treatment is surgical repair

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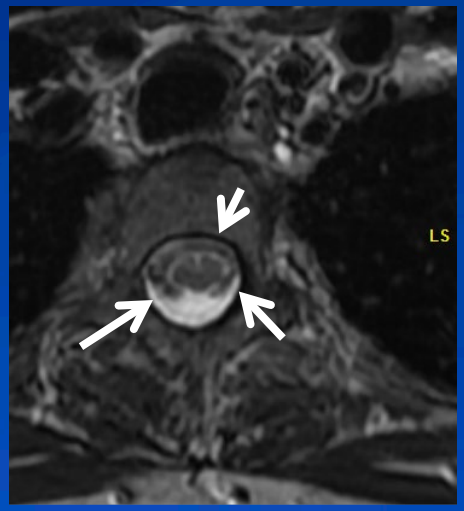
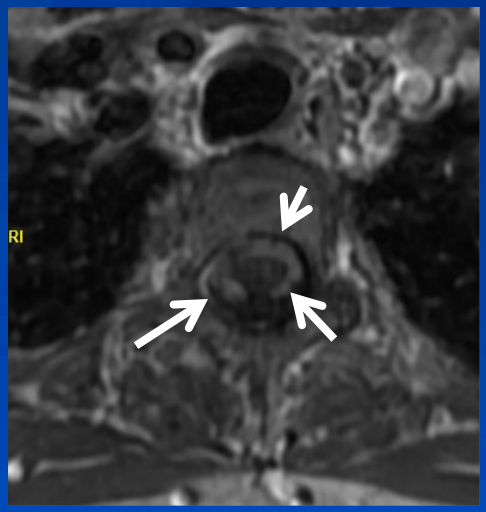
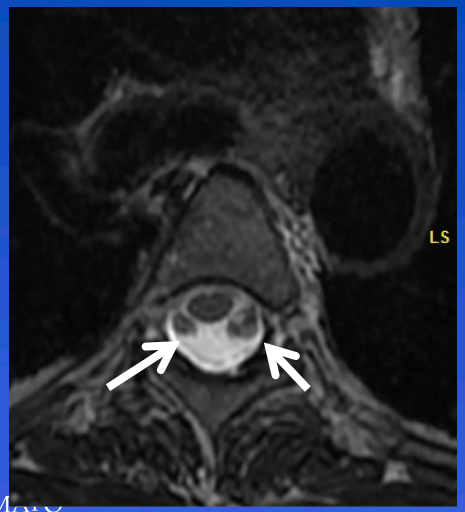
Case 10

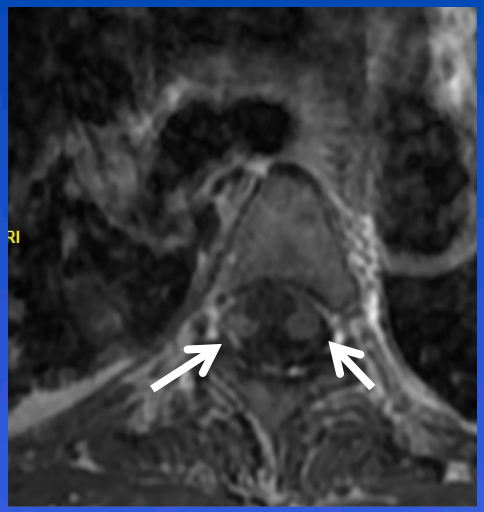
Outside referral for “dural mass” biopsy



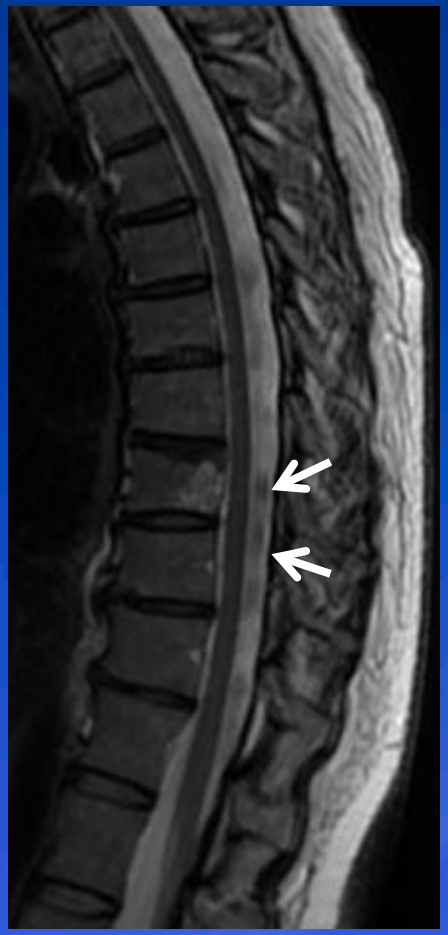




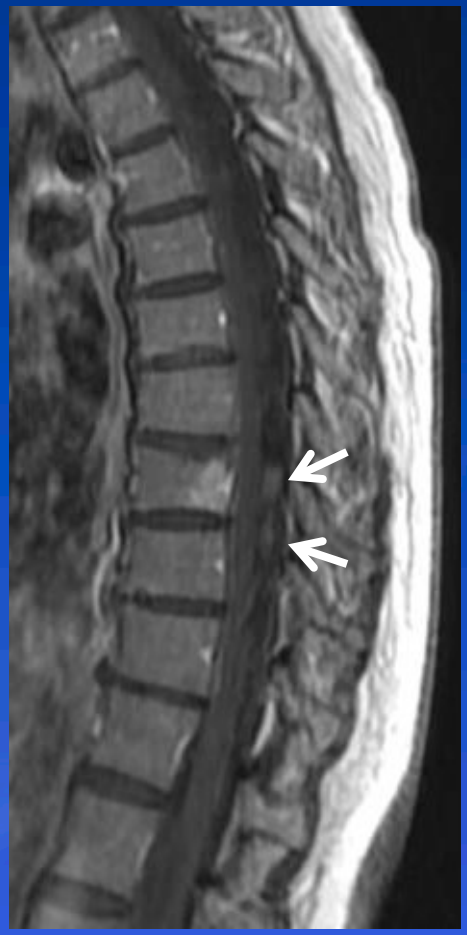
T2



T1 Gad



T2



T1 Gad

CSF Pulsation Artifact

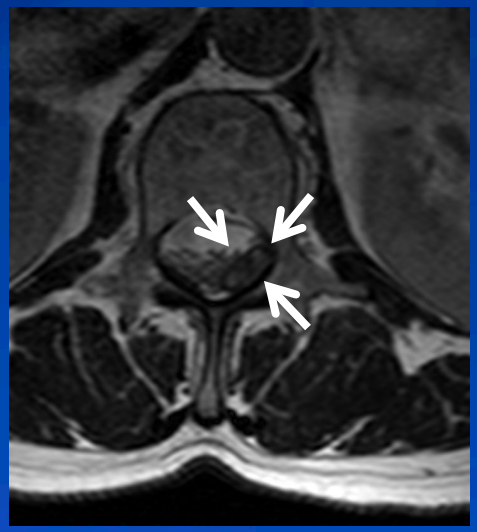
- Problem
  - Artifactual “enhancement” on post-gad T1 sequences (especially axial) is common
  - Can be mistaken for intradural, extramedullary pathology
  - Typically worst in the thoracic spine due to lower ratio of cord to canal diameters
- How to Avoid
  - Exclude true lesion by reviewing orthogonal planes and all sequences
  - Look for corresponding flow voids on T2 sequences
  - Non-FSE sequences such as sagittal GRE, FIESTA, or 3D T1 can provide additional reassurance as they are less prone to CSF pulsation artifact

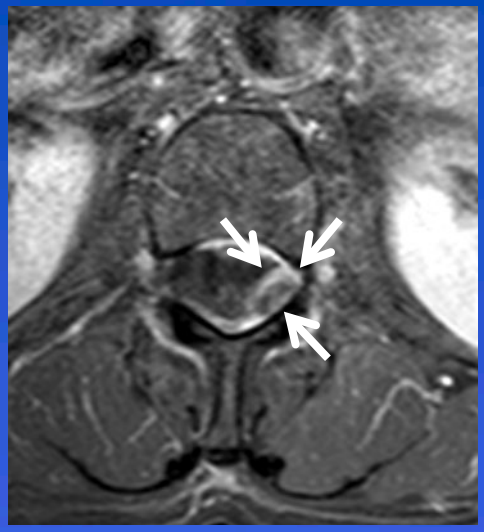
MAYO CLINIC

Case 11

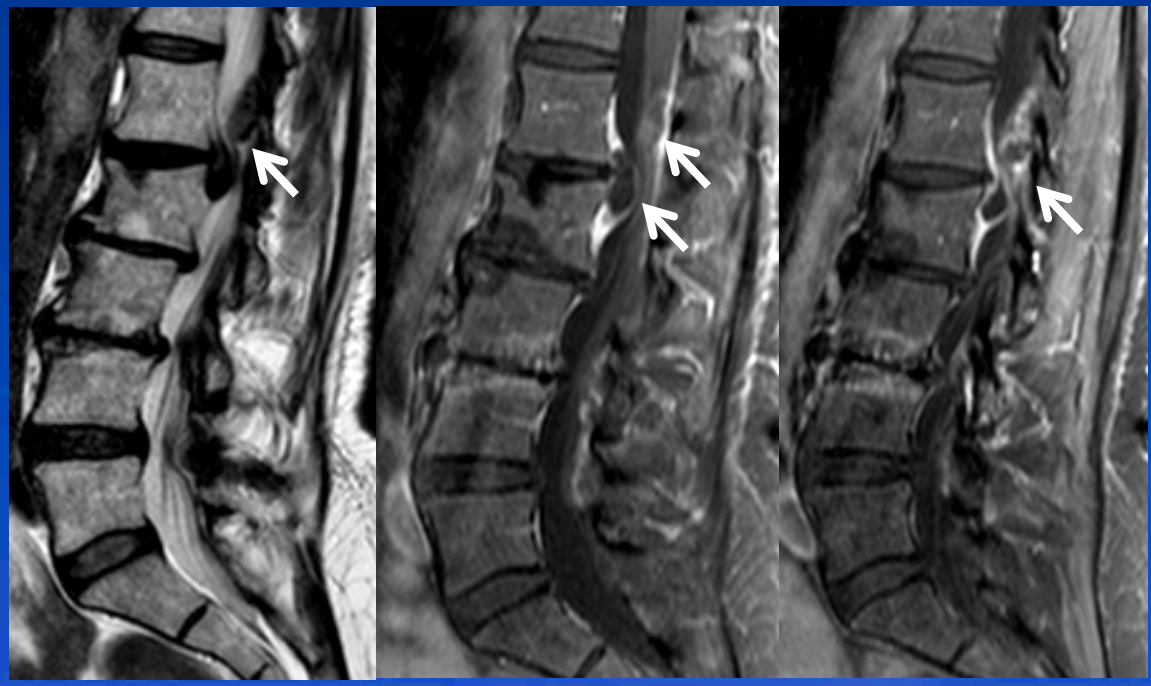
68 yo female with chronic low back pain

T2





T1 Gad



T2 T1 Gad T1 Gad

Intradural Disc Herniation

- Atypical disc herniation locations
  - Dorsal epidural
    - Often acute symptoms
    - Migration of disc material along the dorsal thecal sac
  - Intradural
    - Often chronic symptoms
    - May have beak-like morphology on axial
  - Can be difficult to distinguish on imaging alone
- Additional imaging characteristics
  - Ring enhancement is typical
  - May be able to see contiguous involvement with the disc
- Can mimic other mass lesions

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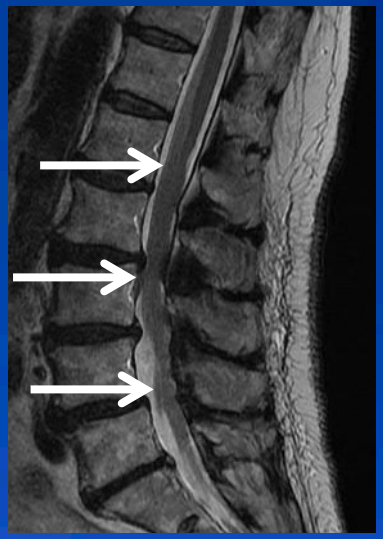
Case 12

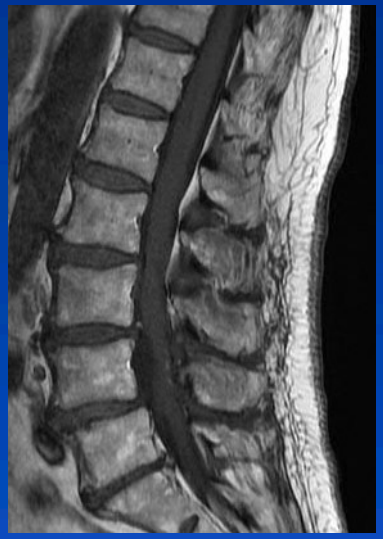
83 yo female

History of “injections” outside the US to treat a variety of conditions including macular degeneration, osteoporosis, prior strokes, and depression/dementia 1 yr ago

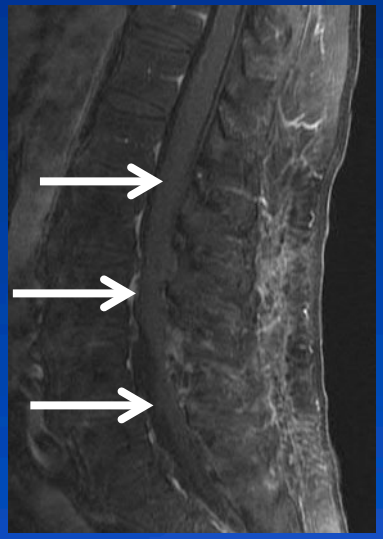
New leg pain and balance/gait difficulty



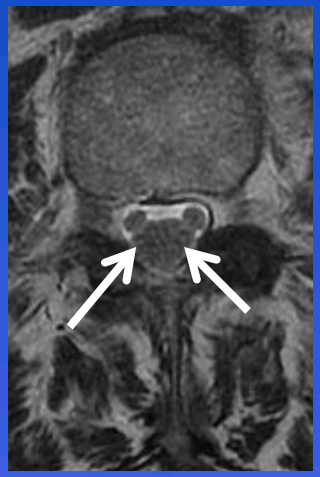
T2

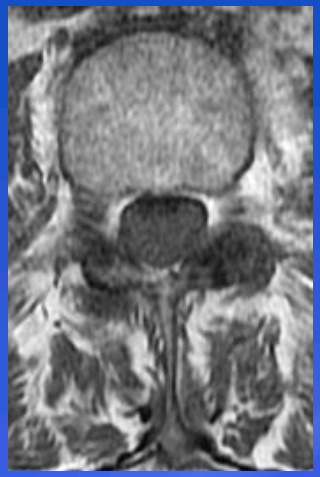


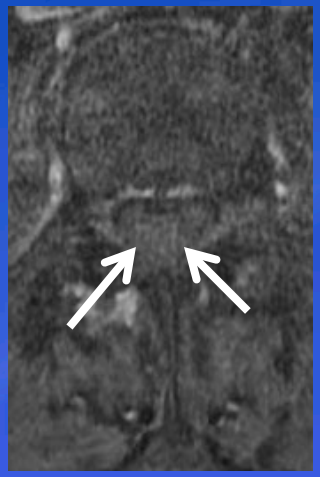
T1



T1 Gad







MAYO CLINIC

Intrathecal Stem Cell Injection

- Case reports with biopsy suggest inflammatory and/or "glioproliferative" changes as cause
- Sensory and motor symptoms months after injection
- MRI
  - Marked diffuse cauda equina nerve root enlargement
  - Nerve root clumping
  - +/- mild enhancement
- History key
  - Stem cell "tourism"
  - Clinical trials
- Can mimic CIDP, hereditary motor sensory neuropathy, or arachnoiditis

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Summary

- Intradural, extramedullary tumors
  - Meningioma, schwannoma, mets most common
  - Less common tumors may be mimickers
    - Paraganglioma, melanotic schwannoma, solitary fibrous tumor
- Non-tumor pathology may also mimic
  - May be excluded or evaluated by additional MRI sequences (e.g. CSF pulsation artifact) or studies (e.g. dynamic CT myelography for arachnoid cyst)
  - History may be key (stem cell injection, prior surgery for arachnoiditis, etc.)

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