AMSER Case of the Month January 2025

A 16-year-old female presents with chronic low back pain now radiating to the R thigh.

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Our Patient

HPI: A 16-year-old female with autism spectrum disorder presents to the orthopedic clinic with a one-year history of low back pain now radiating into the R thigh. Parents deny any recent travel or sick contacts. No incontinence.

Patient Presentation

HPI cont'd:

- Previous complaint of non-radiating low back pain led to PT referral
- Pain progressively worsened, started experiencing pain in R thigh
- Referred to outside hospital orthopedics, diagnosed with kyphosis and referred again to PT
- 3 months of PT and chiropractic therapy did not improve symptoms
- Pain is aggravated by prolonged sitting or walking
- Patient uses diclofenac gel daily and OTC ibuprofen as needed

Patient Presentation

PMH: Patient has a cognitive age of 7-8 years old and was recently evaluated by psychiatry for new behavioral changes suspected to stem from anxiety related to poor sleep.

ROS: (+) back pain, (+) gait disturbance

Vitals: WNL

Physical Exam: Thoracic kyphosis. Good muscle bulk and tone with 5/5 muscle strength of upper and lower extremities. Slight inversion of left foot. Gait with small steps but ambulates without assistance. Diminished patellar reflexes bilaterally and normal Achilles reflex. No tenderness to palpation of spine.

No pertinent lab findings.

What Imaging Should We Order?

ACR Appropriateness Criteria

Variant 3: **Subacute or chronic low back pain with or without radiculopathy. Surgery or intervention candidate with persistent or progressive symptoms during or following 6 weeks of optimal medical management. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI lumbar spine without IV contrast	Usually Appropriate	O
Radiography lumbar spine	May Be Appropriate	☢☢☢
MRI lumbar spine without and with IV contrast	May Be Appropriate	O
Bone scan whole body with SPECT or SPECT/CT complete spine	May Be Appropriate	☢☢☢
CT lumbar spine without IV contrast	May Be Appropriate	☢☢☢
CT myelography lumbar spine	May Be Appropriate	☢☢☢☢
MRI lumbar spine with IV contrast	Usually Not Appropriate	O
CT lumbar spine with IV contrast	Usually Not Appropriate	☢☢☢
Discography and post-discography CT lumbar spine	Usually Not Appropriate	☢☢☢
CT lumbar spine without and with IV contrast	Usually Not Appropriate	☢☢☢☢
FDG-PET/CT whole body	Usually Not Appropriate	☢☢☢☢

This imaging modality was ordered by the orthopedic surgeon in clinic.

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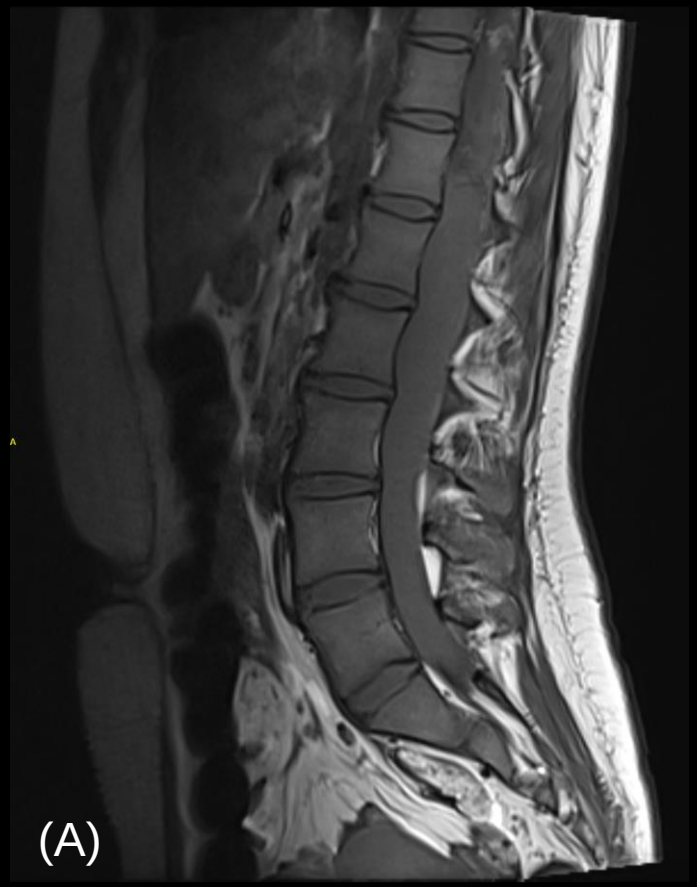
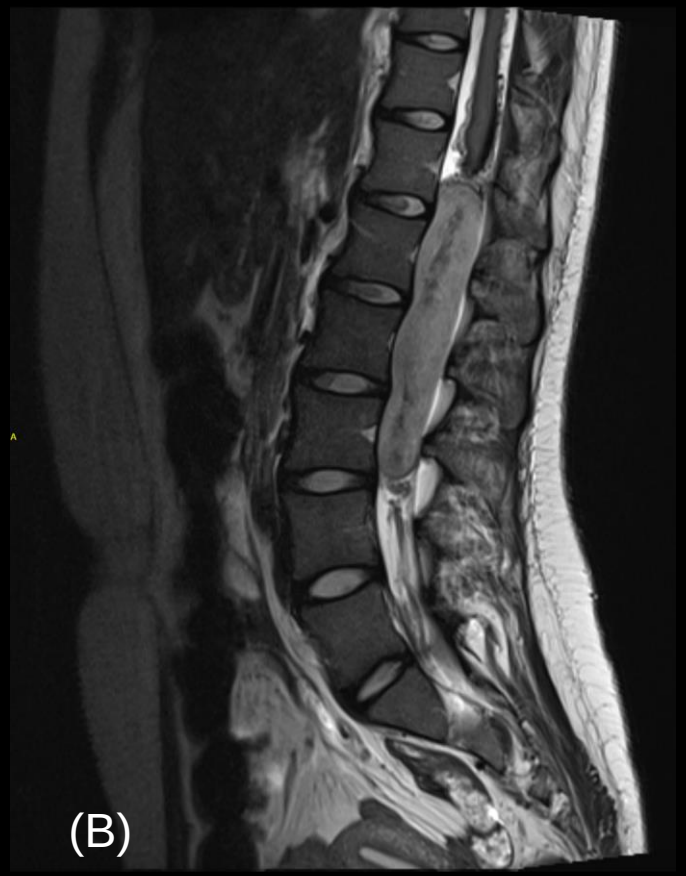
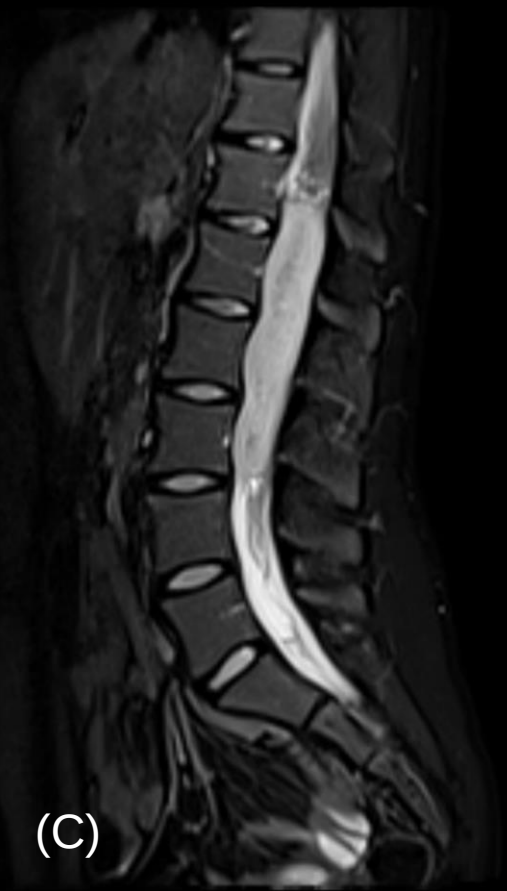
Findings







Sagittal MRI: (A) T1, (B) T2, (C) T2-STIR

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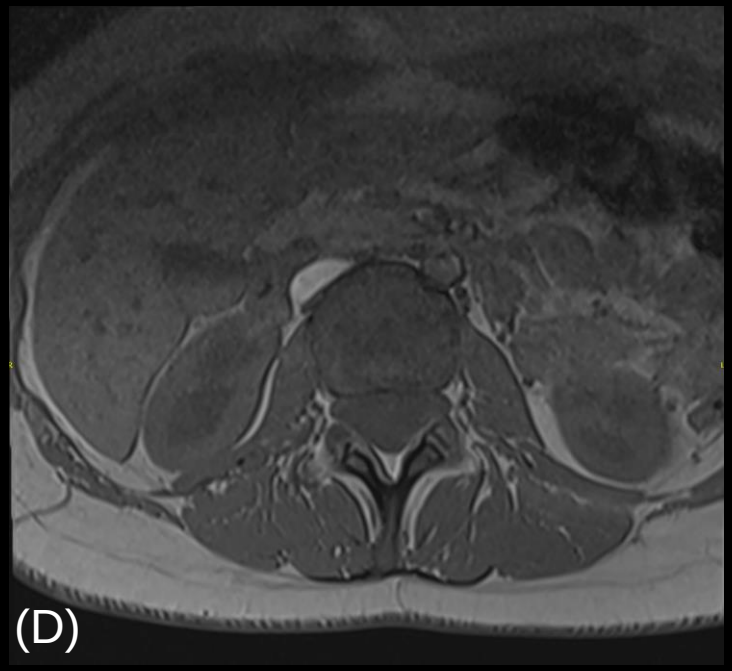
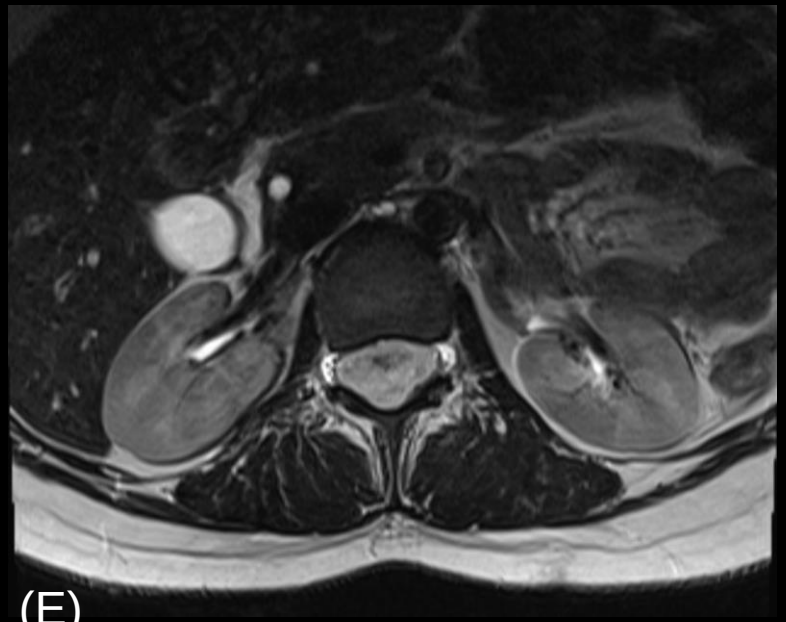
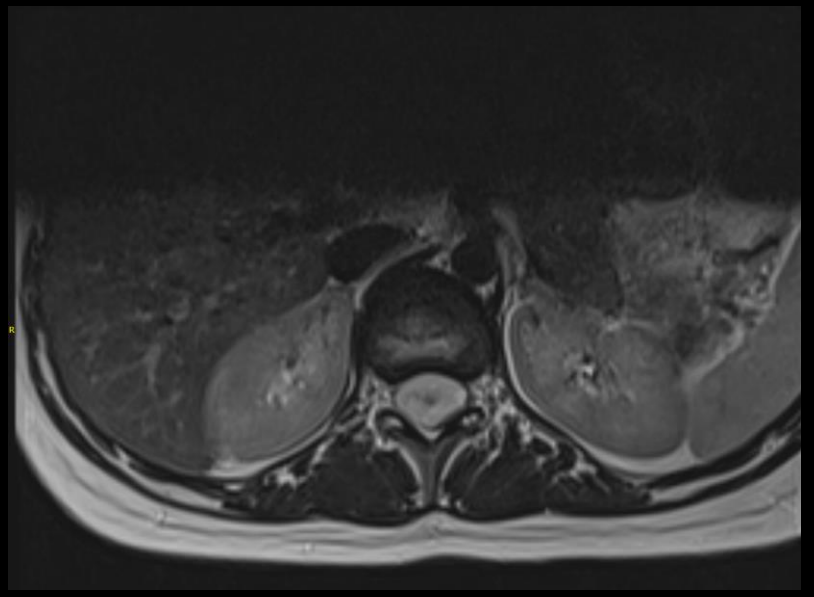
Findings



(D)



(E)

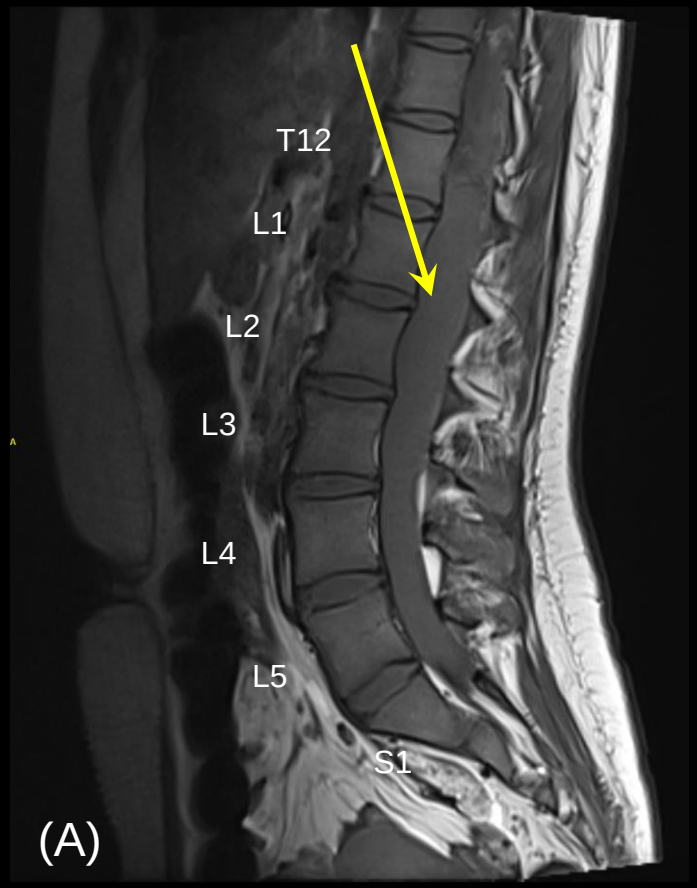


(F)

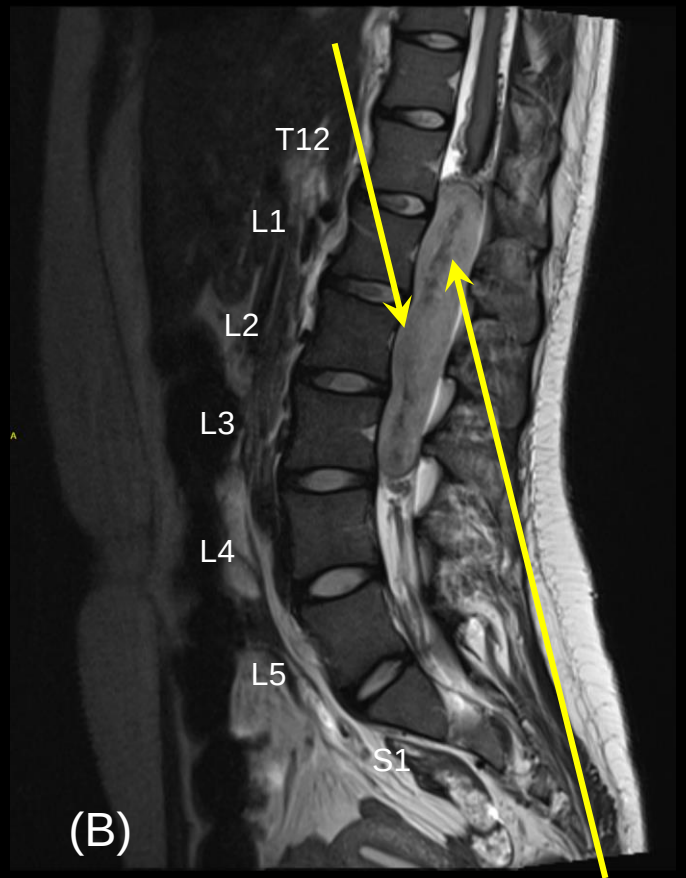
Axial MRI: (D) T1, (E) T2, (F) T2

Findings: Labeled

T1 isointense to cord

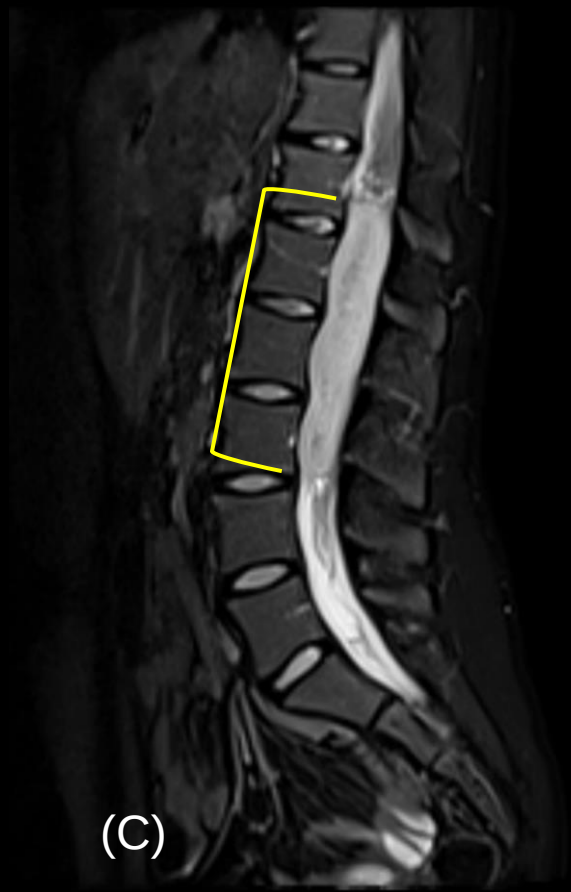


T2 hyperintense to cord



Intralesional hypointensity suggestive of hemosiderin

STIR hyperintense to cord



Sagittal MRI: (A) T1, (B) T2, (C) T2-STIR

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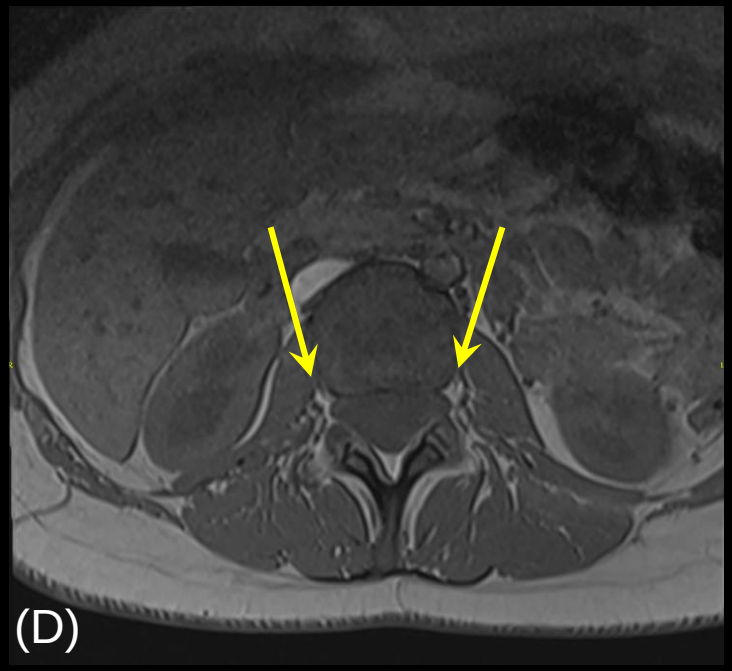
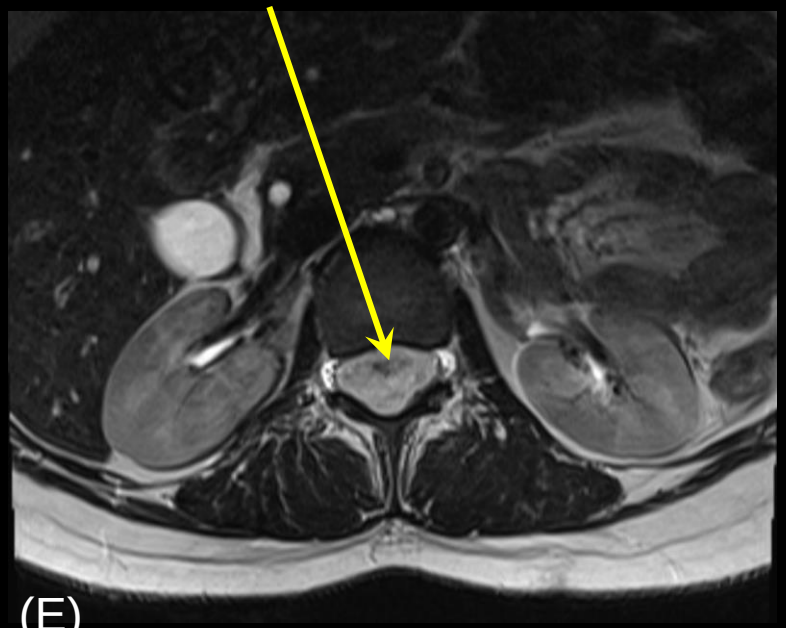
Findings: Labeled



(D)

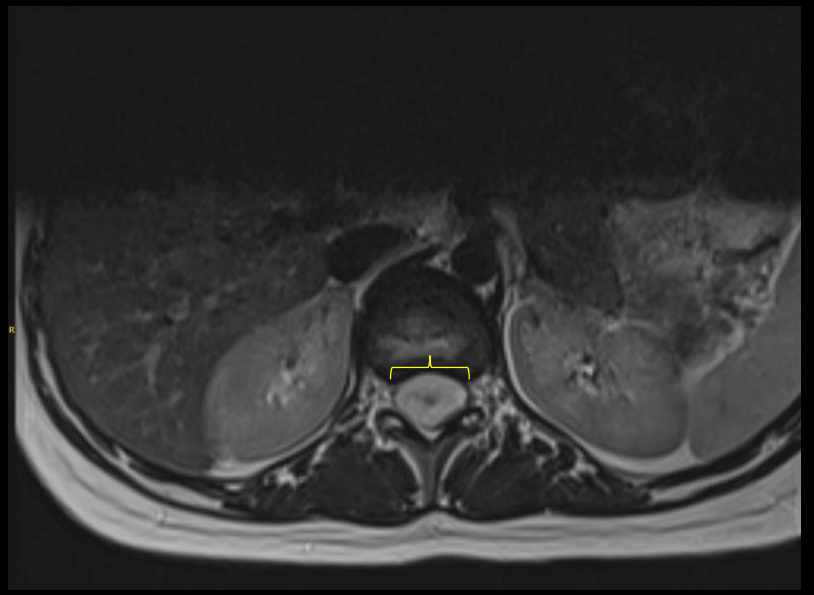
Spinal canal at L2-L3 entirely occupied by T1 isointense mass, limiting evaluation of exiting and descending nerve roots.

Central T2 hypointensity at approximate origin of filum terminale



(E)

Spinal canal at L1-L2 entirely occupied by T2 hyperintense mass, limiting evaluation of exiting and descending nerve roots.



(F)

Spinal canal at T12-L1 also entirely occupied by mass.

Axial MRI: (D) T1, (E) T2, (F) T2

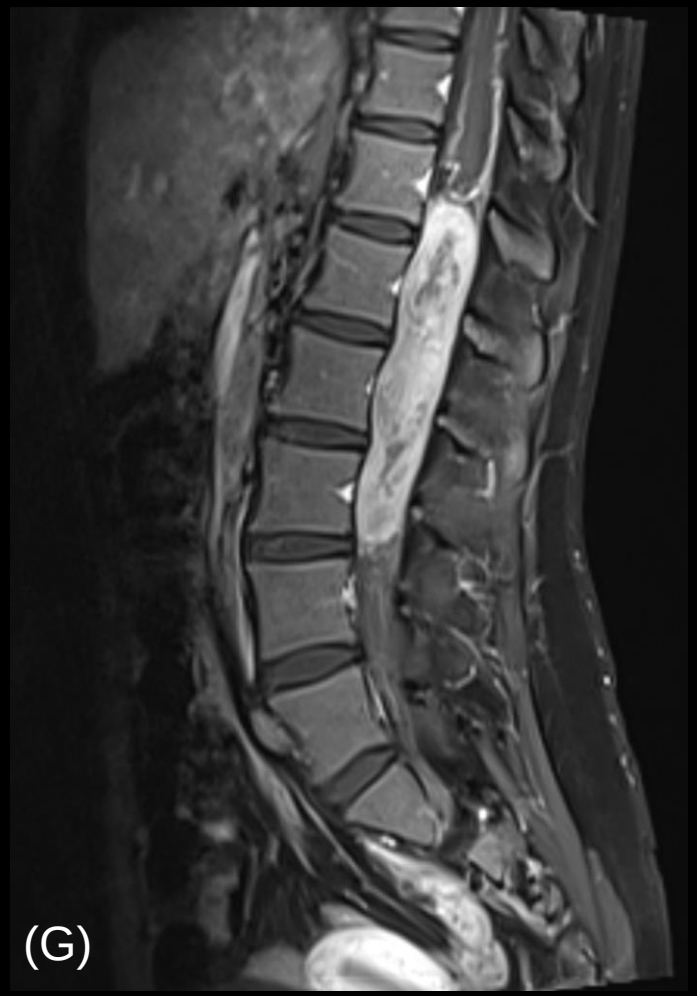
Differential Diagnoses

1. Myxopapillary ependymoma
2. Schwannoma (nerve sheath tumor)
3. Paraganglioma
4. Chordoma
5. Intradural metastasis ie. Metastatic adenocarcinoma
6. Meningioma

Investigating Further

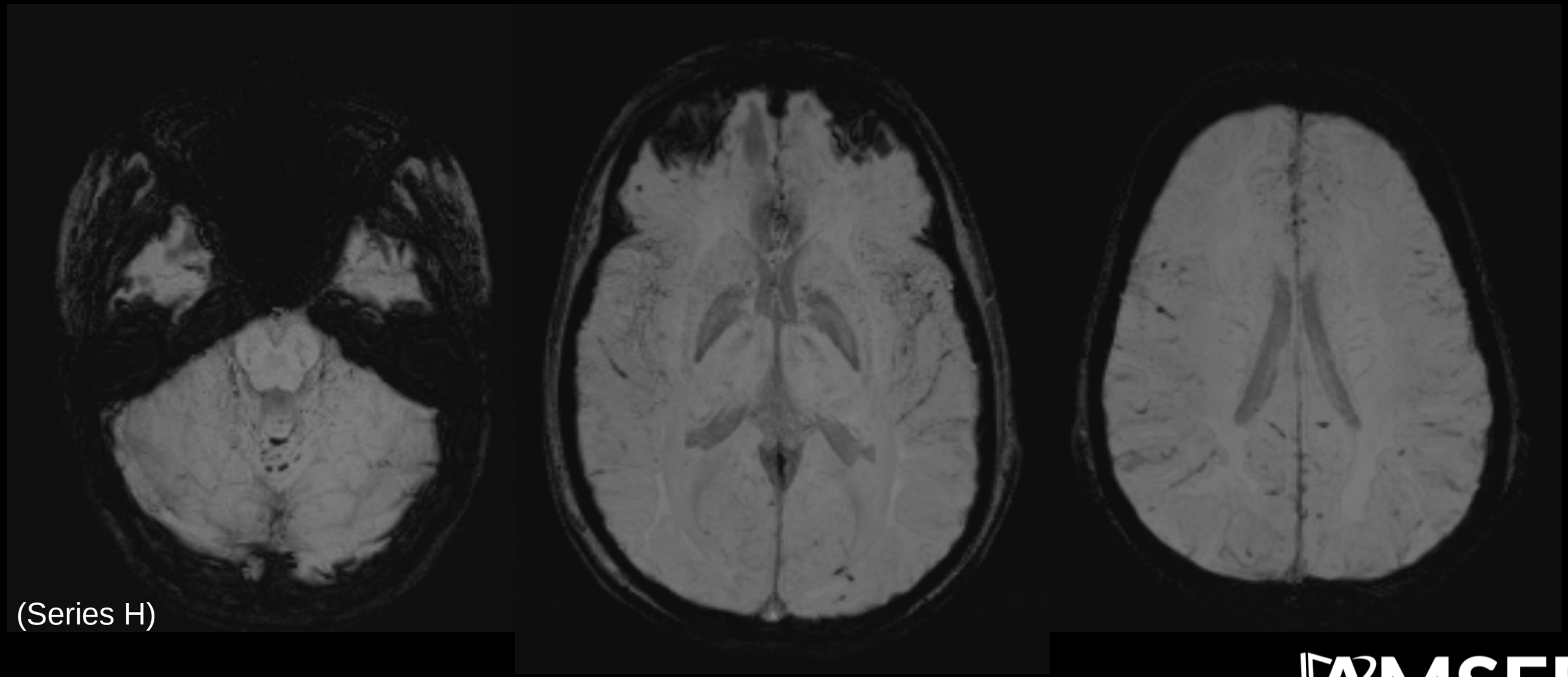
MRI Brain, C-, T-, and L-spine w/wo contrast

Additional Relevant Findings



(G) Sagittal post-contrast T1

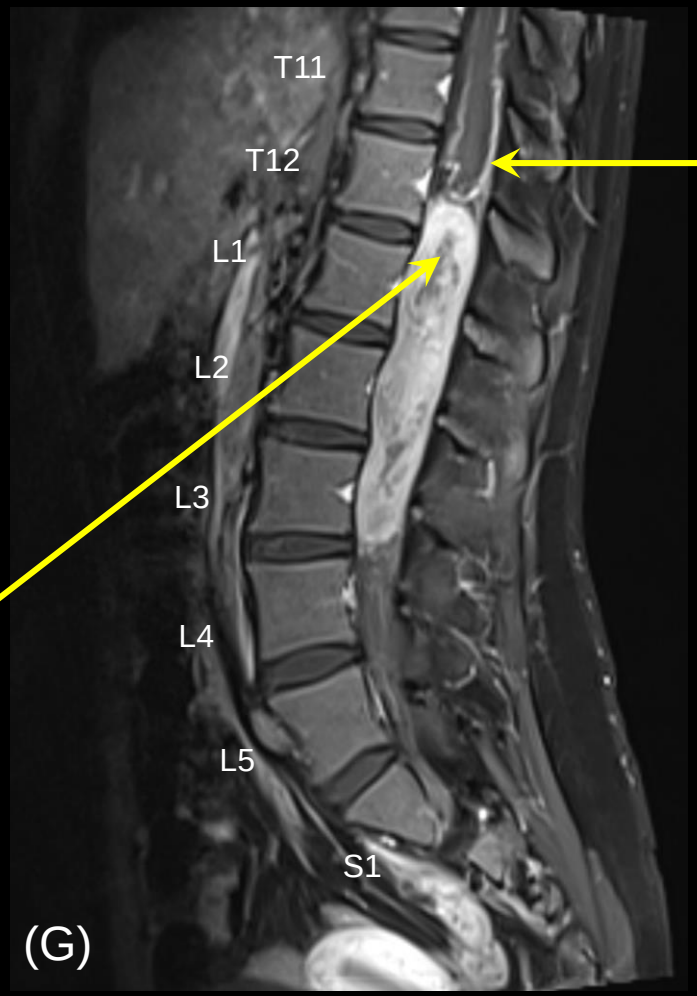
Additional Relevant Findings



(Series H)

(H) Axial SWI

Additional Relevant Findings: Labeled



Curvilinear enhancement along thoracic cord suggestive of prominent vessels vs. leptomengingeal tumor spread

Intralesional areas of hypointensity may suggest hemosiderin deposition within tumor vs. non-enhancing tumor

(G) Sagittal post-contrast T1

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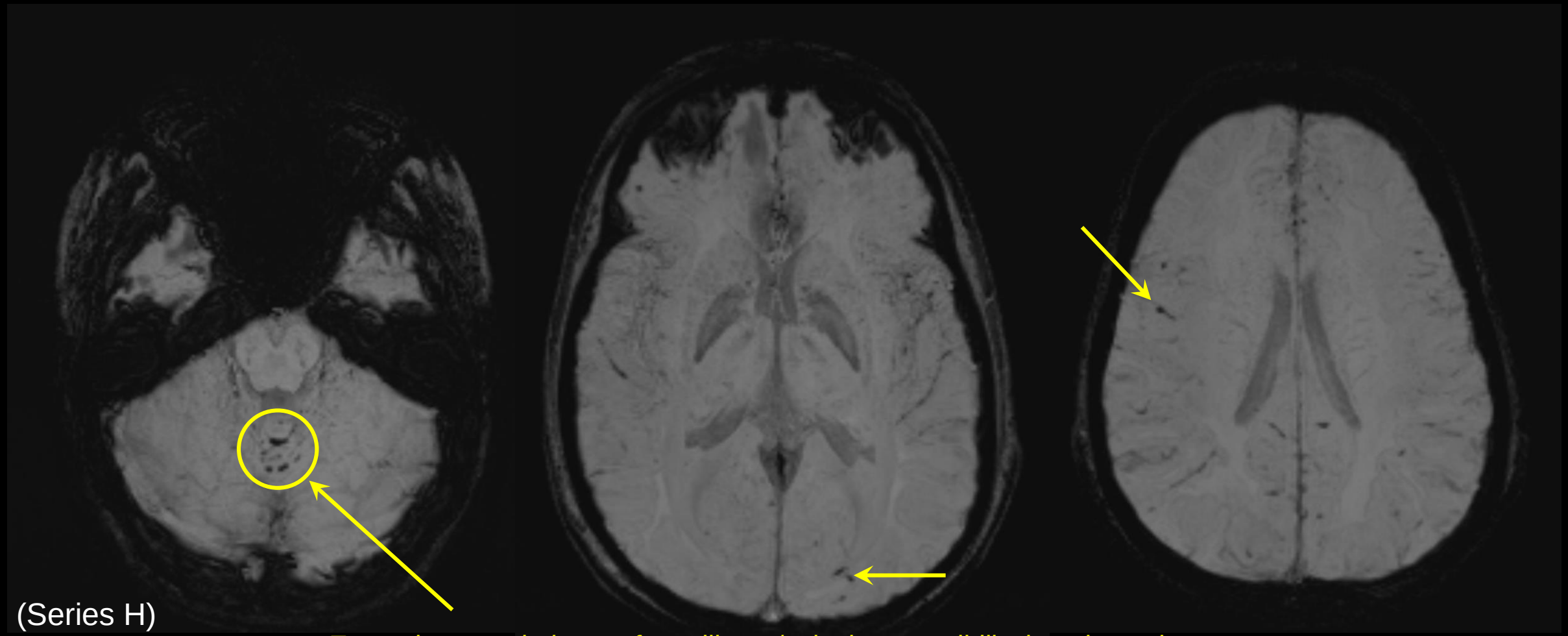
Additional Relevant Findings: Labeled



(Series H)

(H) Axial SWI

Examples noted above of curvilinear/sulcal susceptibility hypointensity along the bilateral cerebral hemispheres and cerebellar vermis, suggestive of superficial siderosis.

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Summary of Findings

- Avidly enhancing intradural and extramedullary mass
- T1 isointense, T2 hyperintense, and T2-STIR hyperintense to cord
- Completely occupies the spinal canal from T12-L3
- Involvement of the conus medullaris/cauda equina and filum terminale measuring up to 9.8 cm (craniocaudal)
- Possible regional vascular congestion
- Possible intralesional hemosiderin deposition
- Scattered foci of susceptibility hypointensity suggestive of superficial siderosis along bilateral cerebral hemispheres, cerebellar vermis

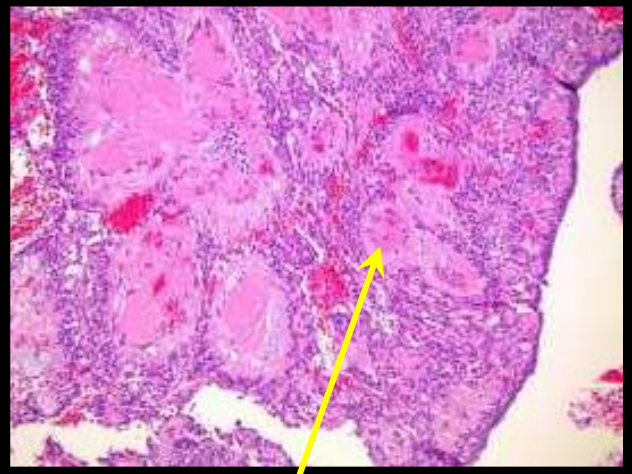
Final Dx:

Myxopapillary ependymoma

Histopathology: Overview

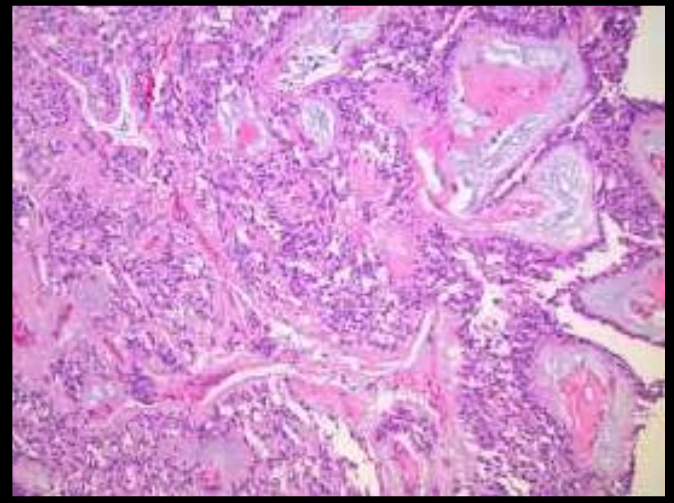
- Gross specimen: often encapsulated, red/purplish, soft consistency
- Papillary architecture with blood vessels surrounded by cuboidal and/or spindled glial cells, perivascular myxoid changes
- Can see mucin-rich microcysts and perivascular pseudorosettes
- Scarce mitotic figures, no necrosis
- Pathology will distinguish from chondrosarcoma/chordoma:[1]
  - Positive for GFAP > S-100 is typical; often positive for CD99, CD56, COX-2
  - Negative for EMA and Olig2
  - Example: chondrosarcoma would be negative for GFAP, chordoma positive for EMA and negative for GFAP

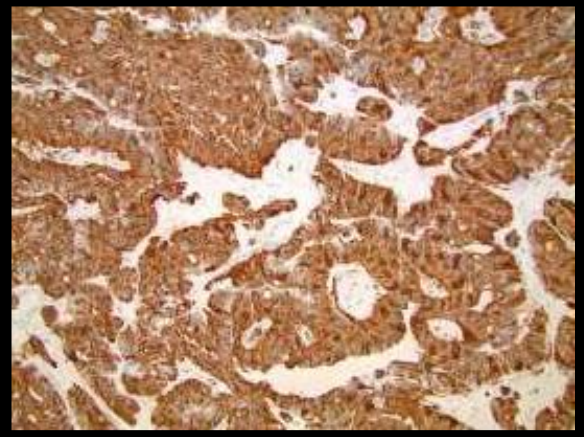
Histopathology: Our Patient



Pseudorosette

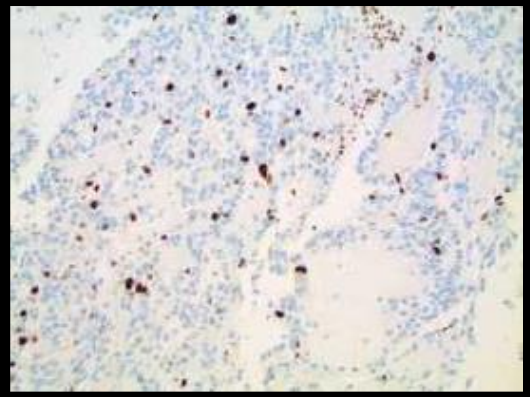
Myxoid and papillary features





Strongly GFAP positive

Olig2 negative



Myxopapillary ependymomas are WHO Grade 2, considered low-grade but more likely to recur than Grade 1.

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Case Discussion: Epidemiology

- Slow-growing (WHO Grade 2) glioma with predilection for lumbosacral spine, arising from ependymal cells of the conus, cauda equina, and filum terminale[2]
  - Occasionally lateral and fourth ventricles, cervicothoracic spinal cord, brain
- 1-5% of all spinal tumors, 13% of spinal ependymomas, and 90% of tumors of the conus[2]
- Mean age of onset is 35 years old with bimodal distribution in 20's and 40's, with slightly higher incidence in men[3]
- No consistent, associated genetic alterations

Case Discussion: Radiographic Findings

- Tends to span several vertebral segments and fills thecal sac[4]
  - Can erode pedicles and extend into neural foramina
- “Sausage”-shaped
- T1 typically isointense to cord (mucin can cause hyperintensity)
- T2 almost always hyperintense to cord
  - Hypointensity can suggest hemosiderin deposition
- Post-contrast T1 shows intense enhancement
- Possible to have local seeding, subarachnoid dissemination → important to image entire spine and brain
- If unencapsulated, possible to see tumor necrosis

Case Discussion: Radiographic Findings

- Associated findings include acute subarachnoid hemorrhage and superficial siderosis, which is the hemosiderin deposition that can be seen with recurrent subarachnoid hemorrhage[2]
  - Susceptibility hypointensity on SWI, as found in our patient on brain MRI
  - Usually, normal T1
  - Possible linear low signal along cord and nerve root surfaces on T2
  - T2*-GRE similar to T2, but more pronounced findings
  - Post-contrast T1 shows no leptomeningeal enhancement at areas of hemosiderin deposition

Case Discussion: Treatment & Prognosis

- Our patient: total resection
  - Hemorrhage was noted within the tumor inferiorly, tumor significantly adherent to nerve roots but complete surgical resection was achieved
  - T12-L3 laminectomies and laminoplasties, L4 partial laminectomy
  - No residual symptoms at less than one-month post-op
  - No further treatment required at this point, surveillance MRI q3 months in the first year
- Subtotal resection can be followed with radiotherapy[5]
- Long-term survival is excellent with gross total resection, which occurs in > 85% of cases with overall 10-year survival > 90.6%[6]
  - Distant metastasis uncommon; CSF dissemination more likely if tumor capsule is disrupted; anaplastic variants more aggressive

References

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6. Pratiti Bandopadhayay, V. Michelle Silvera, Pedro, et al. Myxopapillary ependymomas in children: imaging, treatment and outcomes. *Journal of Neuro-Oncology*. 2015;126(1):165-174. doi:https://doi.org/10.1007/s11060-015-1955-2

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