AMSER Case of the Month September 2023

47-year-old female presents with lower back pain and right lower extremity weakness

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Patient Presentation

- 47y/o Female with no significant past medical history
- Presents with lower back pain with right lower extremity weakness
- Vital:
  - BP 134/82, HR 70, RR 17, Temp 37.1, pulse Ox 98%
- Physical:
  - Right lower extremity pain, numbness and weakness on plantarflexion and dorsiflexion

What Imaging Should We Order?

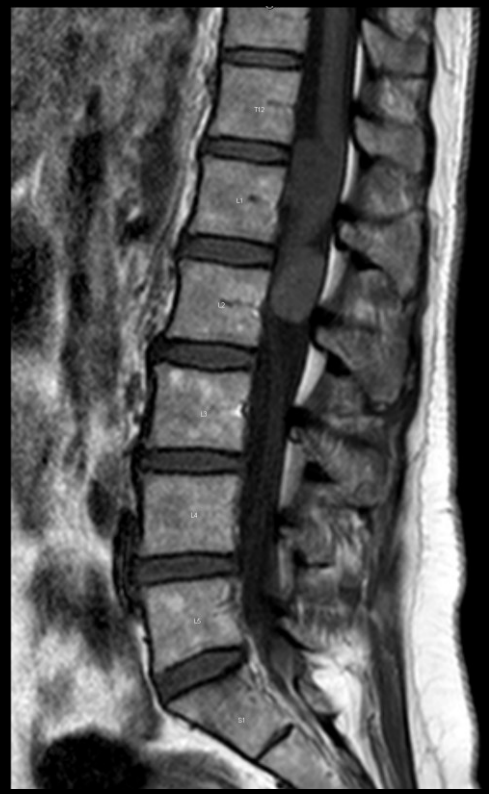
Select the applicable ACR Appropriateness Criteria

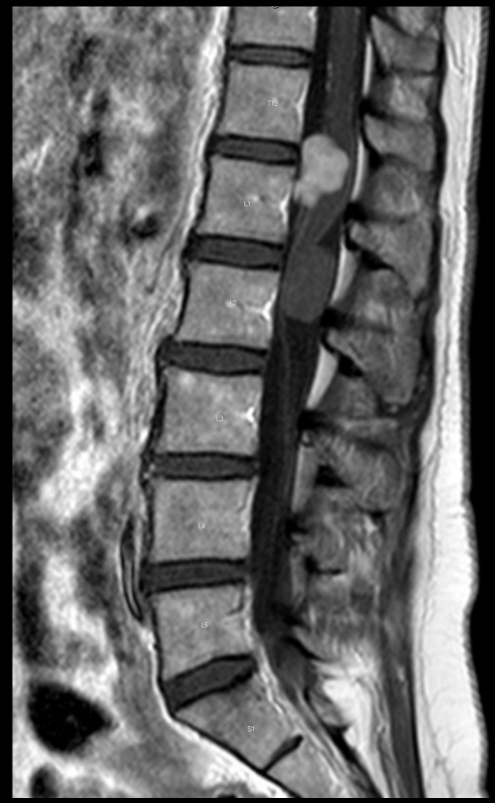
Variant 2: **Chronic or progressive myelopathy. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI spine area of interest without and with IV contrast	Usually Appropriate	O
MRI spine area of interest without IV contrast	Usually Appropriate	O
CT myelography spine area of interest	May Be Appropriate	Varies
CT spine area of interest with IV contrast	May Be Appropriate	Varies
CT spine area of interest without IV contrast	May Be Appropriate	Varies
Arteriography spine area of interest	Usually Not Appropriate	Varies
Radiography spine area of interest	Usually Not Appropriate	Varies
MRA spine area of interest with IV contrast	Usually Not Appropriate	O
MRA spine area of interest without and with IV contrast	Usually Not Appropriate	O
MRA spine area of interest without IV contrast	Usually Not Appropriate	O
MRI spine area of interest with IV contrast	Usually Not Appropriate	O
CT spine area of interest without and with IV contrast	Usually Not Appropriate	Varies
CTA spine area of interest with IV contrast	Usually Not Appropriate	Varies

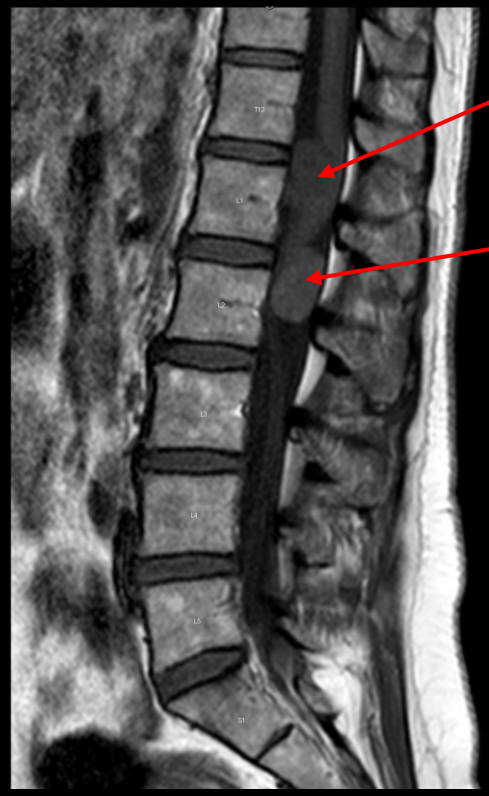
This imaging modality was ordered

Findings (Unlabeled)





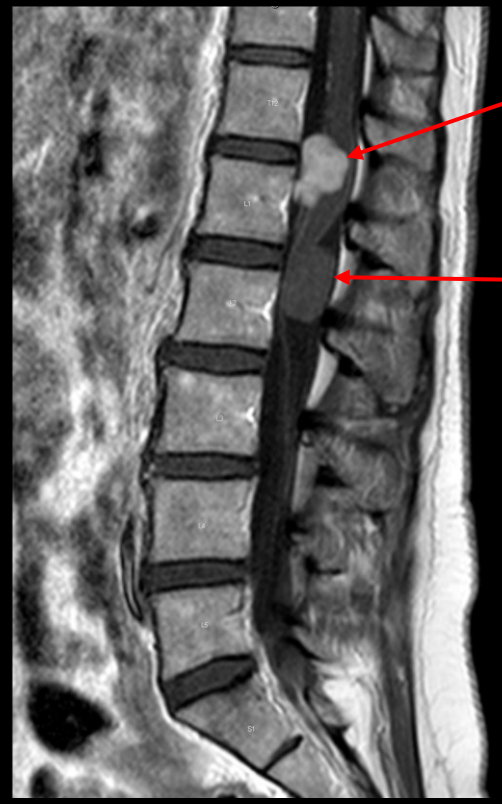
Findings (labeled)



Anterior intradural extramedullary mass at T12-L1 spinal level demonstrates homogenous hypointense T1 signal relative to the adjacent vertebral body

Posterior intradural extramedullary mass at L1-L2 spinal level demonstrates homogenous hypointense T1 signal relative to the adjacent vertebral body

T1 precontrast



Anterior intradural extramedullary mass at T12-L1 spinal level demonstrates avid contrast enhancement.

Posterior intradural extramedullary mass at L1-L2 spinal level demonstrates no significant contrast enhancement

T1 postcontrast

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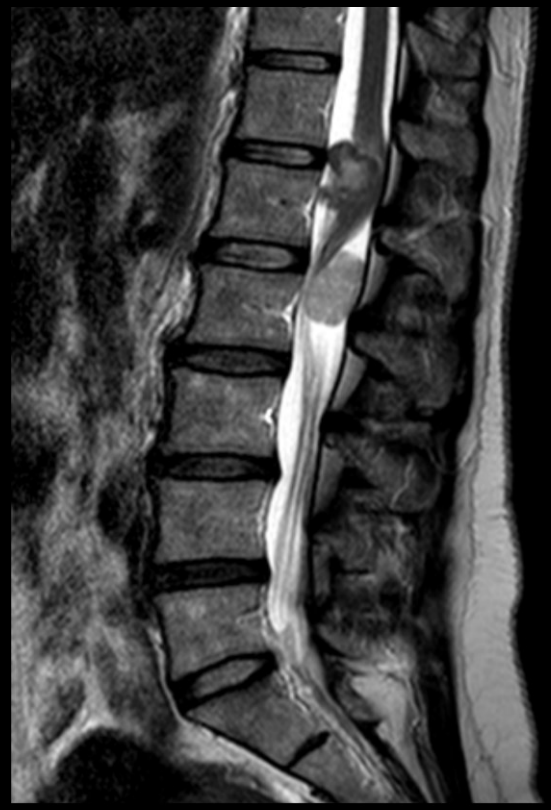
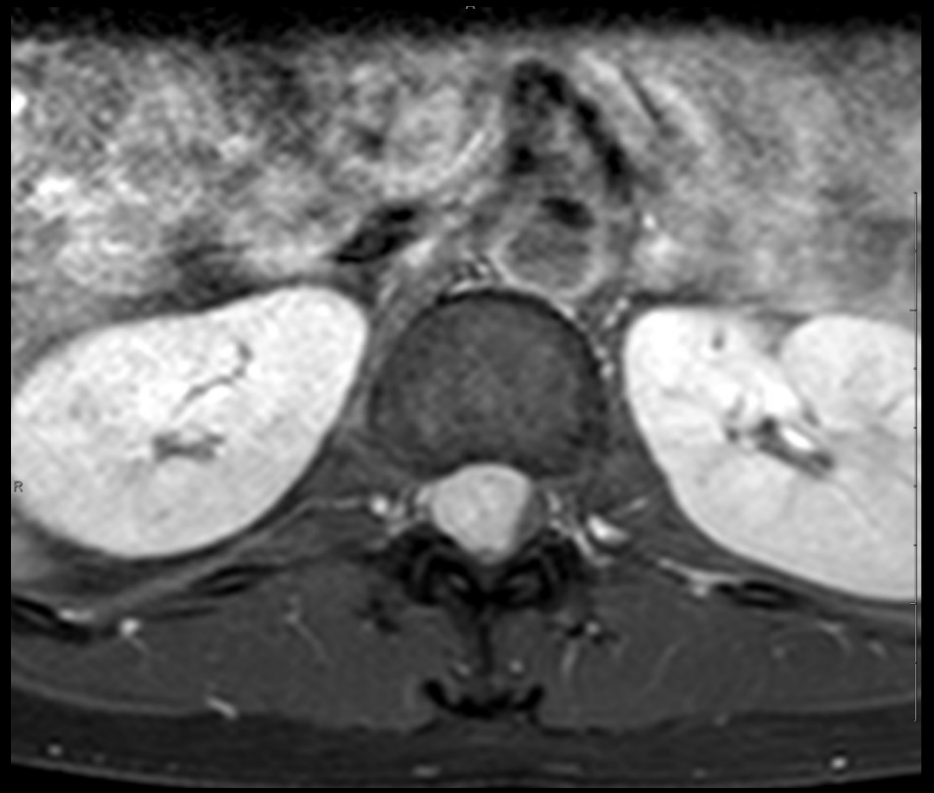
Findings: (Unlabeled)





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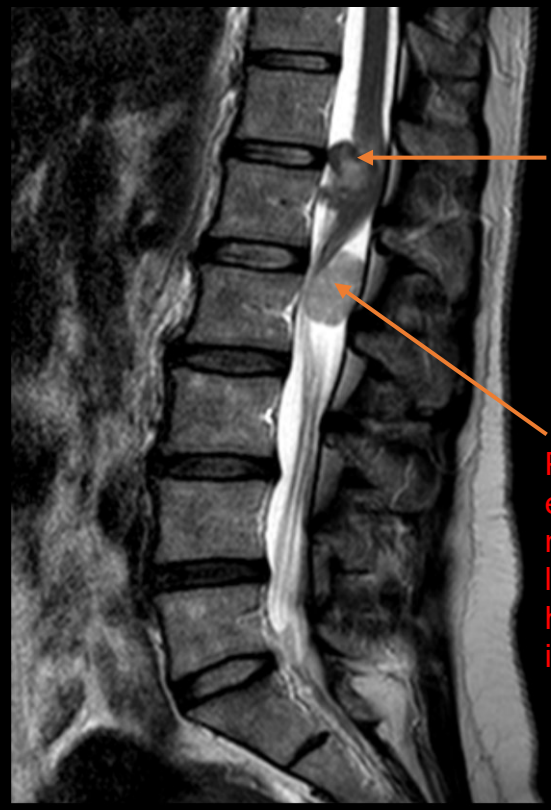
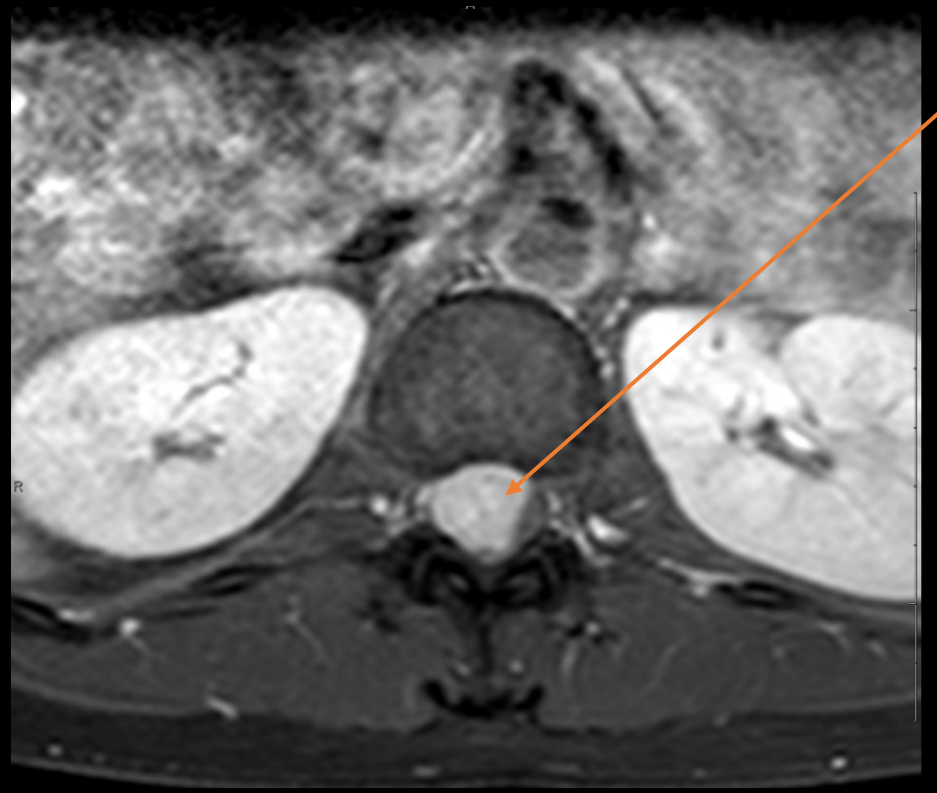


Anterior intradural extramedullary mass at T12-L1 spinal level demonstrates heterogenous T2 signal which is centrally hyperintense and peripherally hypointense

Posterior intradural extramedullary mass at L1-L2 spinal level demonstrates homogenous intermediate T2 signal

T2 weighted



Avidly enhancing intradural extramedullary mass occupying majority of the spinal canal at the T12-L1 spinal level

T1 contrast axial MRI

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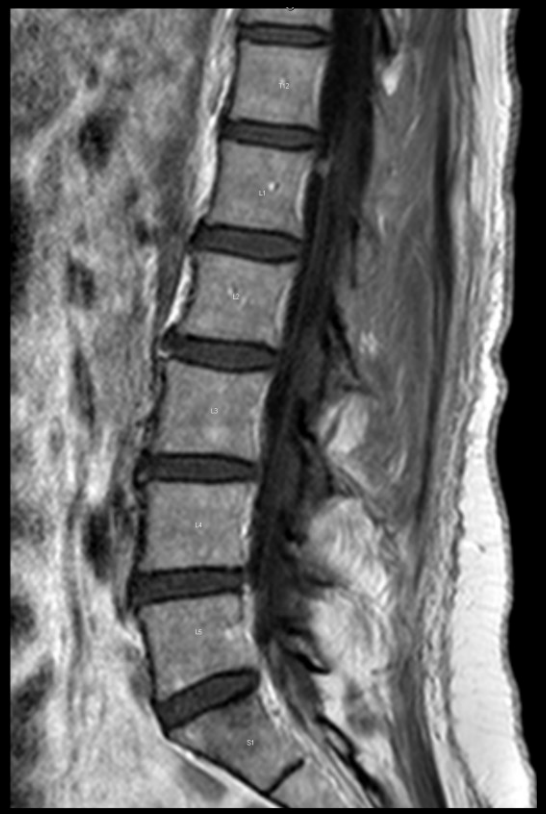
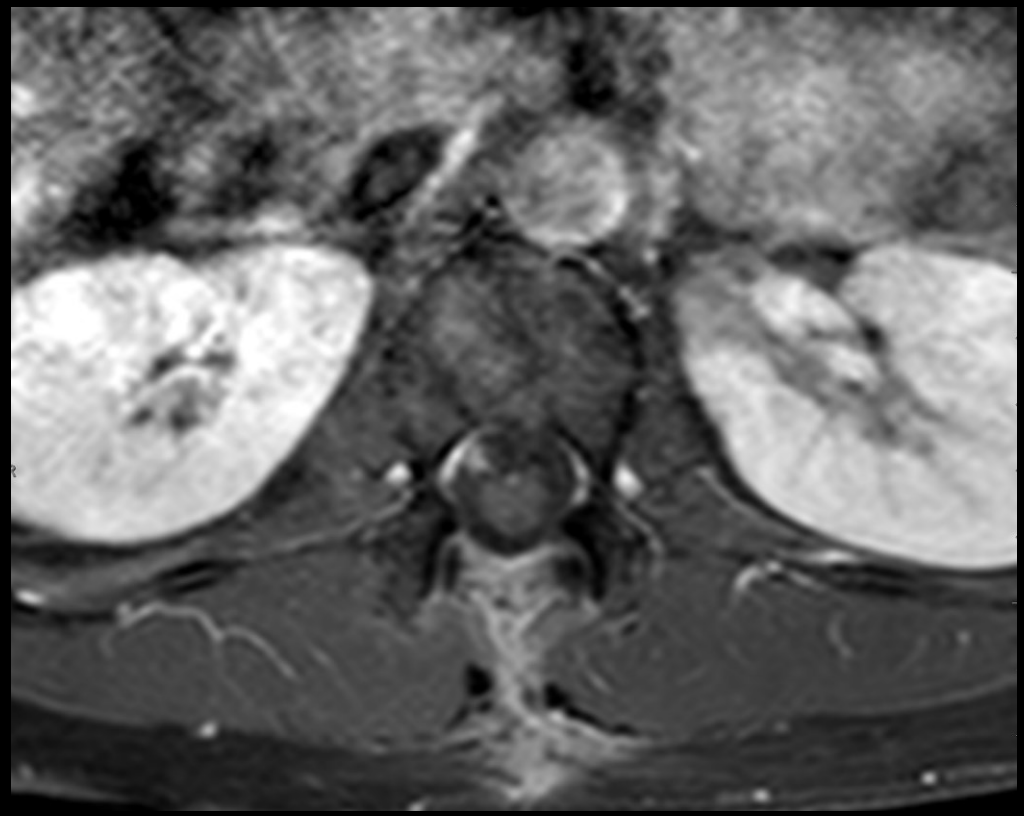
Findings: (Unlabeled)





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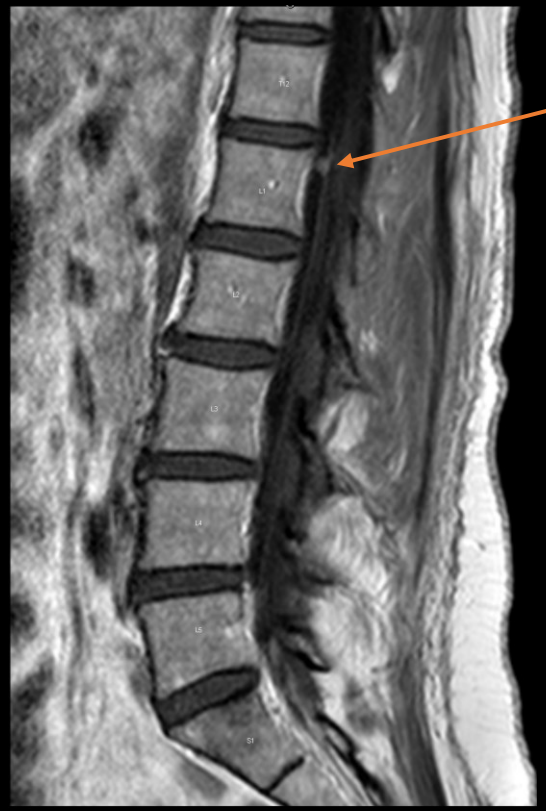
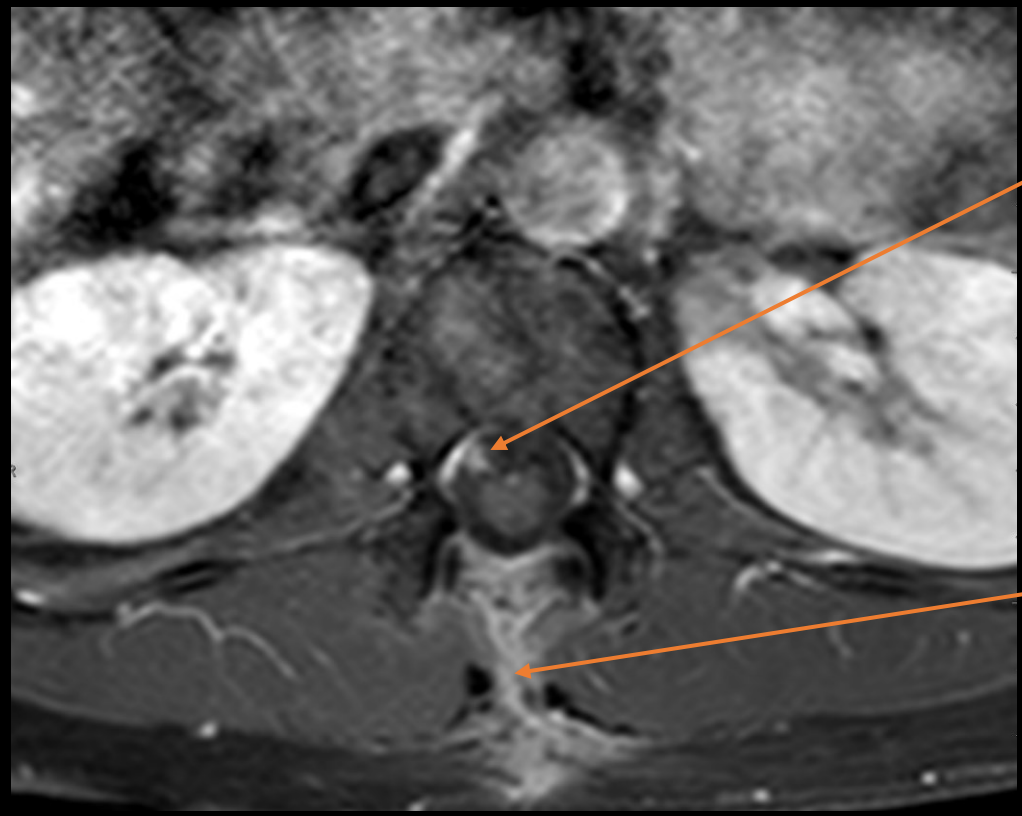


Postop exam demonstrates a tiny focus of contrast enhancement along the anterior spinal canal at the superior aspect of L1 compatible with residual tumor

T1 contrast enhanced



Postop exam demonstrates a tiny focus of contrast enhancement along the anterolateral spinal canal compatible with residual tumor with significantly decreased mass effect

Mild edema along the surgical tract

T1 contrast enhanced

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Pertinent Labs

- CBC: WNL except WBC (3.1)

- Immunohistochemistry:
  - GFAP + (suggests Myxopapillary Ependymoma)
  - S100 & Sox10 + (suggests Schwannoma)

Final Dx:

T12-L1 Spindle Cell Schwannoma
L1-L2 Grade 2 Myxopapillary Ependymoma

Case Discussion (1-3 slides)

- Management:
  - T12-L2 laminectomy and resection of intradural extramedullary tumors was performed
  - Frozen section consistent with Schwannoma and myxopapillary ependymoma
  - Patient was placed on tapering steroid postop
  - Physical therapy was initiated postop

Case Discussion (1-3 slides)

- Schwannoma:
  - Benign well encapsulated, slow growing nerve sheath tumors composed of Schwann cells from the neural crest
  - Can present as Antoni A (hypercellular variant) or Antoni B, a hypocellular myxomatous variant that can mimic myxopapillary ependymoma
- Myxopapillary ependymoma:
  - Rare subtype of spinal cord ependymoma with predilection for lumbosacrum
  - An intradural , extramedullary benign and slow growing grade 2 neoplasm
  - Believed to originate at conus medullaris or filum terminale
  - Presents with nonspecific back pain, motor, sensory, urinary and gait problem

Case Discussion (1-3 slides)

- Differential diagnoses:
  - Spinal Paraganglioma – neuroendocrine tumor found around conus medularis
  - Meningioma- intradural extramedullary tumor originating from the meninges
- Imaging:
  - Best modality for myxopapillary ependymoma and schwannoma is MRI (T1 hypo/isointense, T2 hyperintense)
- Treatment:
  - Surgical excision is the treatment of choice for Myxopapillary ependymoma
  - Treatment of Schwannoma also involves surgical resection or radiotherapy

References:

- Ottenhausen, Malte, et al. “Intradural Spinal Tumors in Adults—Update on Management and Outcome.” *Neurosurgical Review*, vol. 42, no. 2, 2018, pp. 371–388, https://doi.org/10.1007/s10143-018-0957-x.

- Rege, Shrikant V., et al. “Spinal Myxopapillary Ependymoma with Interval Drop Metastasis Presenting as Cauda Equina Syndrome: Case Report and Review of Literature.” *Journal of Spine Surgery*, vol. 2, no. 3, 2016, pp. 216–221, https://doi.org/10.21037/jss.2016.08.06.

- Shih, Robert Y., and Kelly K. Koeller. “Intramedullary Masses of the Spinal Cord: Radiologic-Pathologic Correlation.” *RadioGraphics*, vol. 40, no. 4, 2020, pp. 1125–1145, https://doi.org/10.1148/rg.2020190196.

- Trivedi, Rishika, and Pankaj Trivedi. “Multisegment Intradural Extramedullary Ependymoma.” *Cureus*, 2021, https://doi.org/10.7759/cureus.20329.

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