# AMSER Case of the Month November 2022

# 33 y/o F with bilateral paresthesias of the hands and wrists

#### Thomas Esber Ohio University Heritage College of Osteopathic Medicine

Dr. Deborah Brahee, MD Cleveland Clinic Foundation



#### Patient Presentation

- 33 y/o female with persistent paresthesias of the hands and wrists bilaterally
- PMH: Hepatitis C and IV drug use
- PSH: Cholecystectomy
- Nerve conduction studies showed bilateral median & ulnar n. changes
  - C5, C6
    - chronic active right sided radiculopathies & ongoing denervation of the right biceps brachii & pronator teres
    - chronic active left sided radiculopathies & ongoing degeneration of the left deltoid & biceps brachii
  - C7, C8
    - demonstrated evidence of inactive radiculopathies bilaterally



#### Differential diagnosis:

- Multiple Sclerosis
- Carpal Tunnel
- Malignancy
- Degenerative Disk Disease
- Nerve Compression, osseous
- Spinal Ischemia



### What Imaging Should We Order?



#### Select the applicable ACR Appropriateness Criteria

Variant 2: New or increasing nontraumatic cervical radiculopathy. No "red flags." Initial imaging.		
Procedure	Appropriateness Category	Relative Radiation Level
MRI cervical spine without IV contrast	Usually Appropriate	0
CT cervical spine without IV contrast	May Be Appropriate	⊕⊕⊕
Radiography cervical spine	May Be Appropriate (Disagreement)	<b>*</b> *
contrast	Usually Not Appropriate	0
Radiographic myelography cervical spine	Usually Not Appropriate	♥♥♥
CT myelography cervical spine	Usually Not Appropriate	€€€€
CT cervical spine with IV contrast	Usually Not Appropriate	<del>ଡ</del> ଡଡ
CT cervical spine without and with IV contrast	Usually Not Appropriate	€€€
CTA neck with IV contrast	Usually Not Appropriate	€€
Discography cervical spine	Usually Not Appropriate	<del>8</del> 8
Facet injection/medial branch block cervical spine	Usually Not Appropriate	**
MRA neck with IV contrast	Usually Not Appropriate	0
MRA neck without IV contrast	Usually Not Appropriate	0
MRI cervical spine with IV contrast	Usually Not Appropriate	0
Bone scan whole body with SPECT or SPECT/CT neck	Usually Not Appropriate	€€€

This imaging modality was ordered by the chiropractic physician



#### Findings (unlabeled)



**MSER** 

#### Findings (labeled)



Loss of lordosis

Degenerative facet changes



#### Select the applicable ACR Appropriateness Criteria

Variant 1: Acute onset myelopathy. Initial imaging.		
Procedure	Appropriateness Category	<b>Relative Radiation Level</b>
MRI spine area of interest without and with	Usually Appropriate	0
MRI spine area of interest without IV contrast	Usually Appropriate	0
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	0
MRA spine area of interest without and with IV contrast	Usually Not Appropriate	0
MRA spine area of interest without IV contrast	Usually Not Appropriate	0
MRI spine area of interest with IV contrast	Usually Not Appropriate	0
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 due to continued symptoms & neurologist recommendation



#### Findings (unlabeled)





#### Findings (labeled)

Intramedullary isointense mass on T1 Intramedullary hyperintense mass on T2 Ill defined hyperintensity in the adjacent spinal cord

Susceptibility artifact rim inferiorly

STIR

#### Final Dx:

#### Intramedullary Spinal Cord Ependymoma (WHO Grade 2)



#### **Characteristic Imaging Findings**

- Plain film
  - Canal widening
  - Vertebral body scalloping
  - Pedicle erosion
  - Laminar thinning
- CT
  - Non-specific canal widening
  - Mass may be a similar or lower attenuation than white matter
  - Mass enhancement with IV contrast



#### Characteristic Imaging Findings

- MRI
  - Circumscribed Intramedullary (within the spinal cord) mass
  - T1: isointense to hypointense, or heterogenous in signal intensity
  - T2/STIR: isointense to hyperintense
    - peritumoral edema (as seen in this case)
      - Hyperintense signal around the mass
    - Hemosiderin cap sign (as seen in this case): susceptibility artifact from prior hemorrhage/ hemosiderin deposition, a common finding
      - Susceptibility artifact: disruption of magnetic fields causing image distortions appearing hypointense on all sequences

**MASER** 

- Other findings
  - Syringohydromyelia
    - Fluid in the central canal of the spinal cord
  - Tumoral cysts and non-tumoral cysts

#### Case Discussion

- Epidemiology:
  - Most common intramedullary neoplasm in adults
  - Second most common in children
  - Peak incidence is the 4<sup>th</sup> decade of life, male > female
  - Association with NF2
- Presentation:
  - Pain, weakness, sensory deficits
- Pathology:
  - Arise from embryonic rests of ependymal tissue
  - Perivascular pseudorosettes
  - MYCN amplification association
  - 60% infratentorial, 30% supratentorial & 10% in spinal cord (as seen in this case)
    - Infratentorial tend to be solid, supratentorial tend to be cystic



## Case Discussion: WHO Grading System

- Grade I:
  - Myxopapillary
    - Slow growing, good long-term survival
    - Exclusively in conus-cauda-filum-terminale region  $\rightarrow$  low back pain presentation
  - Subependymoma
    - Slow growing, noninvasive, usually does not present in the spinal cord
- Grade II:
  - Four variants: cellular, papillary, clear cell, and tancytic
  - Pseudorossettes and less commonly ependymal rossettes
  - GFAP or EMA differentiation
  - RELA positive can be grade II or III
- Grade III:
  - Anaplastic, microvascular proliferation, central necrosis
  - Decreased T2 signal on MRI



#### Case Discussion: Treatment and Prognosis

- Slow growing, compressing spinal cord
- Complete curative resection achieved in 50% of patients
  - 5-year survival = 85%
  - Incomplete resection: 57% 5-year survival
- Metastatic spread is rare
- Radiation therapy can be used for high grade tumors or incomplete resection
- MYCN-amplification are aggressive tumors and spread to central nervous system and have high reoccurrence



#### References:

- Myelopathy. American College of Radiology. ACR Appropriateness Criteria. Available at https://acsearch.acr.org/docs/69484/Narrative. Accessed on August 16, 2022.
- Cervical Neck Pain or Cervical Radiculopathy. American College of Radiology. ACR Appropriateness Criteria. Available at https://acsearch.acr.org/docs/69426/Narrative/. Accessed on August 23, 2022.
- Smith, Alice Boyd, et al. "Radiologic-pathologic correlation of pediatric and adolescent spinal neoplasms: Part 1, Intramedullary spinal neoplasms." *American Journal of Roentgenology* 198.1 (2012): 34-43.
- Koeller, Kelly K., R. Scott Rosenblum, and Alan L. Morrison. "Neoplasms of the spinal cord and filum terminale: radiologic-pathologic correlation." *Radiographics* 20.6 (2000): 1721-1749.
- Kobayashi, Kazuyoshi, et al. "MRI characteristics of spinal ependymoma in WHO grade II: a review of 59 cases." *Spine* 43.9 (2018): E525-E530.
- Reni, Michele, et al. "Ependymoma." *Critical reviews in oncology/hematology* 63.1 (2007): 81-89.
- Mermuys, Koen, et al. "Supratentorial ependymoma." *Radiographics* 25.2 (2005): 486-490.
- Shih, Robert Y., and Kelly K. Koeller. "Intramedullary masses of the spinal cord: radiologic-pathologic correlation." *Radiographics* 40.4 (2020): 1125-1145.

