

RADY 403: Case Presentation

Paralysis in an Adolescent

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Focused patient history and workup

- Patient is a 16 yo male with history of asthma who presented to outside hospital with sudden-onset paralysis. Yesterday, patient with complaints of stomachache and feeling "sluggish." Woke up this morning feeling normal. Throughout the day patient felt faint pain radiating down lateral aspect of legs, bilaterally. Later in the morning, patient found laying on ground unable to move legs.
 - Denies dysuria, hematuria, penile discharge, loss of sensation over GU area.
 - No recent history of trauma.
 - No recent URI, no fevers, or sick contacts. No recent travel. No history of walking in wooded area or tick bites.
 - UTD on all vaccines, including Covid-19.
 - Denies recreational drug, alcohol, or tobacco use. Never been sexually active.

Initial Physical Exam and Workup

Physical Exam

- Vital signs unremarkable, afebrile.
- PERRL, EOMI, CN II – XII intact.
- No skin rashes or bite marks appreciated on the abdomen or bilateral lower extremities.
- **Noticeably diminished pinprick sensation to 18-gauge blunt tip needle with a sensory level of T4-T5.**
 - above T4-T5, pinprick sensation seems to be intact
 - below this level he has only a pressure sensation with no sharp sensation.
- 5/5 strength bilateral upper extremities. **0/5 strength bilateral lower extremities.**

Labs

- CBC, CMP, Magnesium within normal limits.
- Lumbar Puncture: 0 WBC, 36 protein, 69 glucose, no WBCs or organisms on gram stain
- CSF culture = no growth.
- Infectious workup: COVID/flu negative, VDRL negative, Enterovirus CSF PCR negative, HSV 1,2 CSF negative, and RPP.
- **EKG unremarkable.**

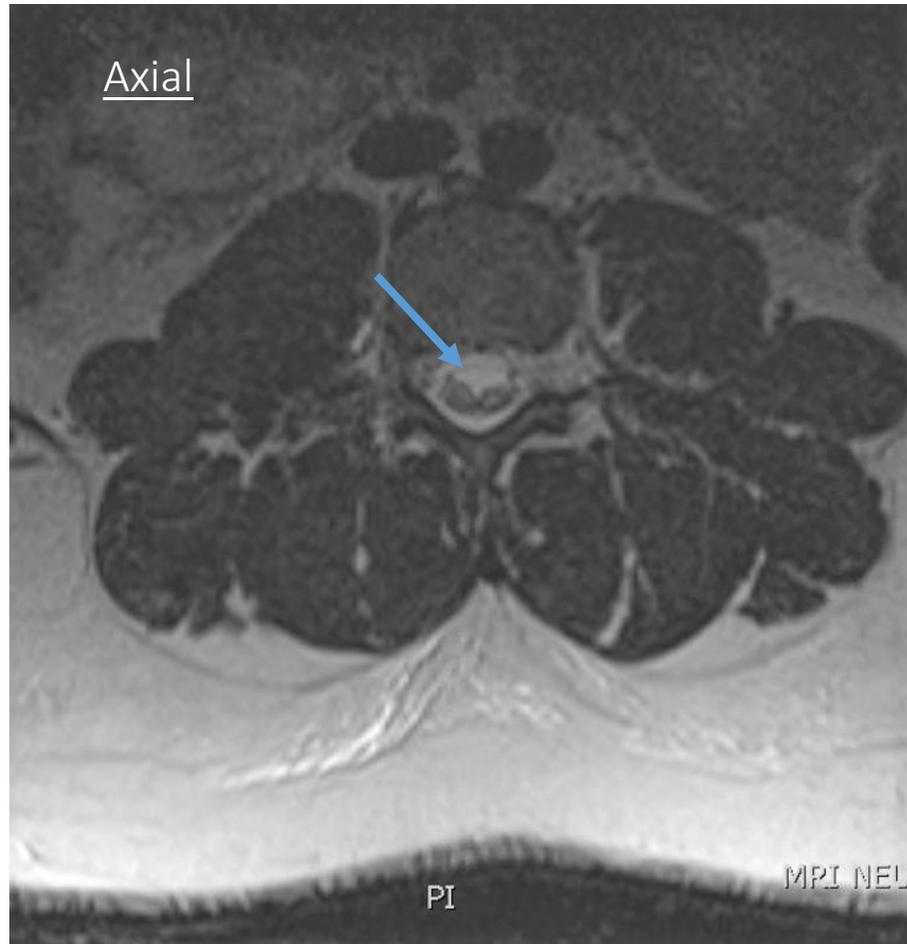
Differential Diagnosis

- Transverse Myelitis
- Acute Spinal Cord Infarction
- Compressive Myelopathy
 - CNS neoplasm
 - Epidural or Subdural hematoma
- Guillain-Barre Syndrome
- Aortic Dissection
- Tick Paralysis
- Intervertebral Disk Herniation

List of imaging studies

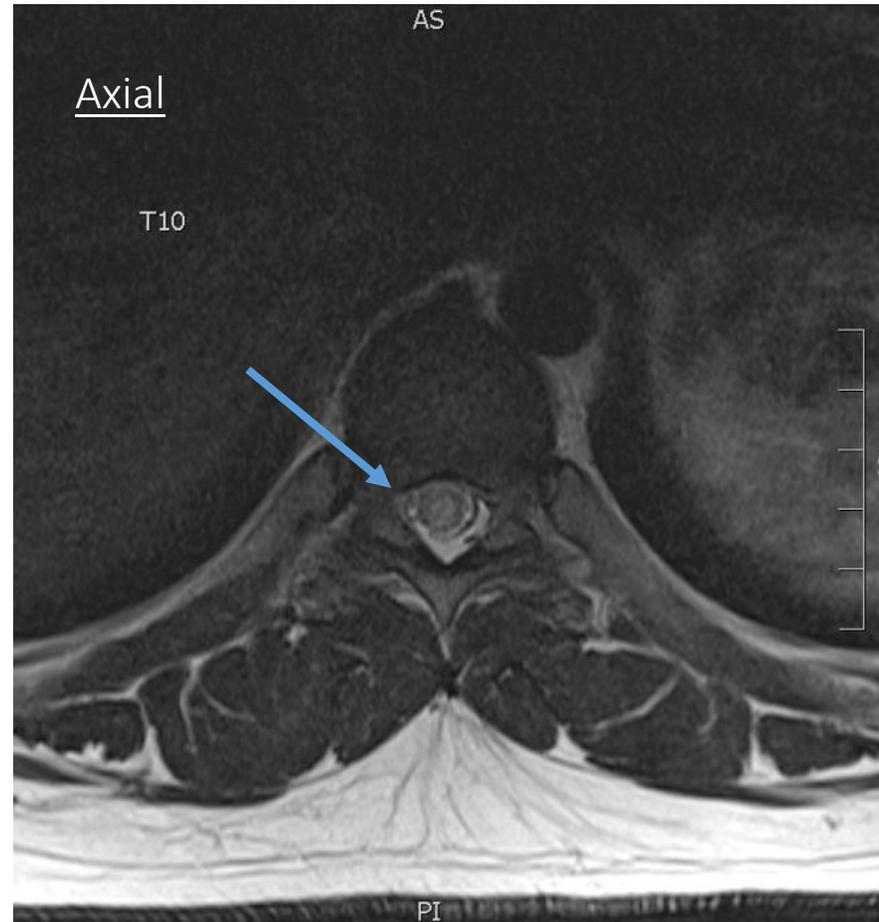
- **In the ED** – outside hospital
 - MRI w and w/o contrast brain
 - MRI w and w/o contrast cervical, thoracic, and lumbar spine
- **UNC Medical Center** – follow-up imaging
 - MRI w and w/o contrast thoracic and lumbar spine (9 days post-initial imaging)
 - CTA chest
 - Echocardiogram
 - PVL duplex bilateral lower extremities

MRI images post contrast



Abnormal T2 signal within ventral cord T3– T4 vertebral bodies -> conus medullaris
Impression: Likely represents Transverse Myelitis; spinal cord infarct less likely.

MRI images post contrast 9 days later



Abnormal T2 signal within ventral cord T3– T4 vertebral bodies -> conus medullaris
Impression: Same as prior, most consistent with transverse myelitis, with spinal cord infarct less likely

Patient treatment and outcome

- Initial MRI showed long anterior spinal cord abnormality concerning for transverse myelitis.
- Peds neurology consulted -> patient started on 1 gram of Solumedrol x 2 days with no improvement.
- LP unremarkable, no evidence of inflammation, no CSF growth. Infectious work up = negative.
- Clinical picture best represents anterior spinal infarct.
 - Normal Echo, PVL duplex lower extremities w/o DVTs, CTA aorta w/o evidence of aortic dissection or vascular anomaly.
 - Negative thrombophilic workup (guided by Hematology) -> PT/INR, PTT, Factor V/VIII activity, protein C/S, ATIII, lupus anticoagulant, Factor II/V DNA panel. Hbg electrophoresis.
- ***“Despite initial etiologic uncertainty, [patients’] clinical presentation highly consistent with anterior spinal infarct, as opposed to infectious myelitis or other inflammatory central nervous system disease.”***
- Treatment = expectant management. PM&R, PT, OT consulted and heavily involved in care.
- Patient ultimately discharged to acute inpatient rehabilitation facility. At time of discharge patient had minimal recovery of lower extremity neurologic function.

Anterior Spinal Infarct

- Very rare !!! Especially in pediatric population.
- Initial presentation typically acute paraparesis or quadriparesis.
 - Paresis depends on level of spinal cord involvement!
- Abrupt onset – 77% patients onset to nadir deficit within 12 hours.
- Diagnosis generally made clinically, with confirmation with neuroimaging.
- Anterior spinal artery syndrome
 - Loss of motor function and pain/temperature sensation
 - Relative sparing of proprioception and vibratory sense below level of lesion
 - Stages: Flaccidity -> loss of DTRs -> spasticity and hyperreflexia = days to weeks
 - Autonomic dysfunction may be present (hypotension, sexual dysfunction, bowel/bladder dysfunction)
- Treatment and prognosis associated with severity of neurological deficits.
 - Long-term rehabilitation – deficits likely to be chronic.

**American College of Radiology
ACR Appropriateness Criteria®
Myelopathy**

Variant 1: Acute onset myelopathy. Initial imaging.

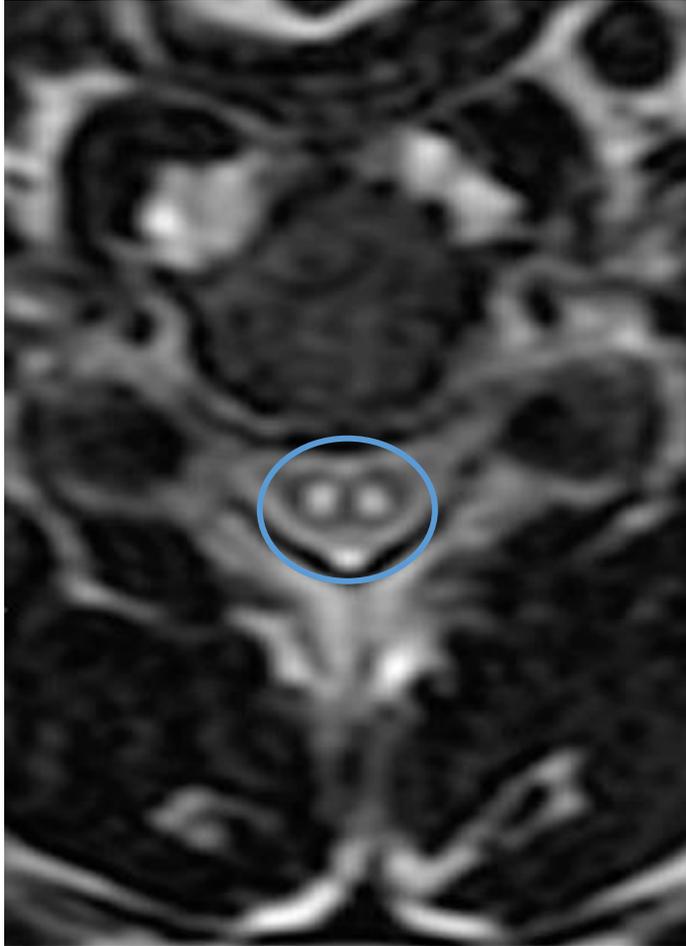
Procedure	Appropriateness Category	Relative Radiation Level
MRI spine area of interest without and with IV contrast	Usually Appropriate	O
MRI spine area of interest without IV contrast	Usually Appropriate	O
CT myelography spine area of interest	May Be Appropriate	Varies
CT spine area of interest with 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

Seidenwurm, David J., et al. "ACR Appropriateness Criteria® Myelopathy." *Journal of the American College of Radiology* 9.5 (2012): 315-324. Revised 2020.

Get an MRI!

- MRI is required for diagnosis of anterior spinal infarction.
 - Rule-out alternative diagnosis!!
- With and without contrast
- Limited sensitivity (45 to 73 percent) depending, in part, on timing of scan from onset of symptoms
 - If initially normal, plan follow-up MRI scans
- Spinal cord ischemia findings:
 - T2-weighted and short-tau inversion recovery image hyperintensities – more specific if hyperintense signal restricted to spinal vascular territory or ventral horns (“owl’s eye sign”)
- **Important note:** T2- weighted abnormalities are not specific for spinal cord ischemia as this finding can be found in transverse myelitis and other intrinsic cord pathologies
- May help determine underlying etiology of infarction – intervertebral disc disease, spondylotic disease (compression?), spinal cord vascular malformations

“Owl’s Eye Sign”



- Also known as “snake-eyes sign” or “fried-eggs sign.”
- Represents bilateral, symmetric, circular T2-weighted hyperintense foci in the anterior horn cells of spinal cord.
- This sign is more specific for spinal cord infarct as it illustrates T2-hyperintensities within the vascular territories.

Image not from this case; image cited in references slide

TOP THREE

1. Keep a **broad** differential diagnosis!
2. Take into account the entire clinical picture.
3. T2-signal abnormal hyperintensities is suggestive, but **not** diagnostic of spinal infarction.

References

- Zalewski NL, Rabinstein AA, Krecke KN, et al. Characteristics of Spontaneous Spinal Cord Infarction and Proposed Diagnostic Criteria. JAMA Neurol 2019; 76:56.
- Image: The owl's eyes sign. Radsource. (2020, October 13). Retrieved June 17, 2022, from <https://radsource.us/the-owls-eyes-sign/>
- Seidenwurm, David J., et al. "ACR Appropriateness Criteria® Myelopathy." Journal of the American College of Radiology 9.5 (2012): 315-324. Revised 2020.