# Vascular myelopathies

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# Disclosure

- Off label usage
  - IV alteplase (tPA)
- Financial relationships
  - None

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# Learning Objectives

- Identify 4 clinical categories of vascular myelopathies
- Understand typical clinical and radiographic characteristics of vascular myelopathies
- Apply spinal cord infarction diagnostic criteria

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# Introduction

- Vascular myelopathies are commonly misdiagnosed
  - Two large studies showed patients diagnosed with ITM frequently had alternative myelopathy diagnoses → vascular was common
    - 18% vascular
    - Unnecessary harm and costs
- Time to treatment important for outcomes

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1. Barreras P, Fitzgerald KC, Mealy MA, et al. Clinical biomarkers differentiate myelitis from vascular and other causes of myelopathy. *Neurology*. 2017.
2. Zalewski NL, Flanagan EP, Keegan BM. Evaluation of idiopathic transverse myelitis revealing specific myelopathy diagnoses. *Neurology*. 2018;90(2):e96-e102.

# Vascular myelopathy categories

- Arterial Ischemia
- Venous congestion/ischemia
- Hematomyelia
- Extraparenchymal hemorrhage

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# CASES

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# Arterial ischemia case 1

- 66 M
- PMH: unremarkable
- HPI:
  - Lifting equipment, severe neck pain
  - Generalized weakness that day
  - Morning
    - Generalized weakness still present
    - Nap → rapid paraplegia
    - Outside ED: near quadriplegia → respiratory distress

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# Arterial ischemia case 1

- PE
  - Ventilator (fully dependent)
  - Mild skew deviation
  - Severe quadriparesis (asymmetric)
  - Sensory level:
    - Pain/temp left C4, and right T3-T4

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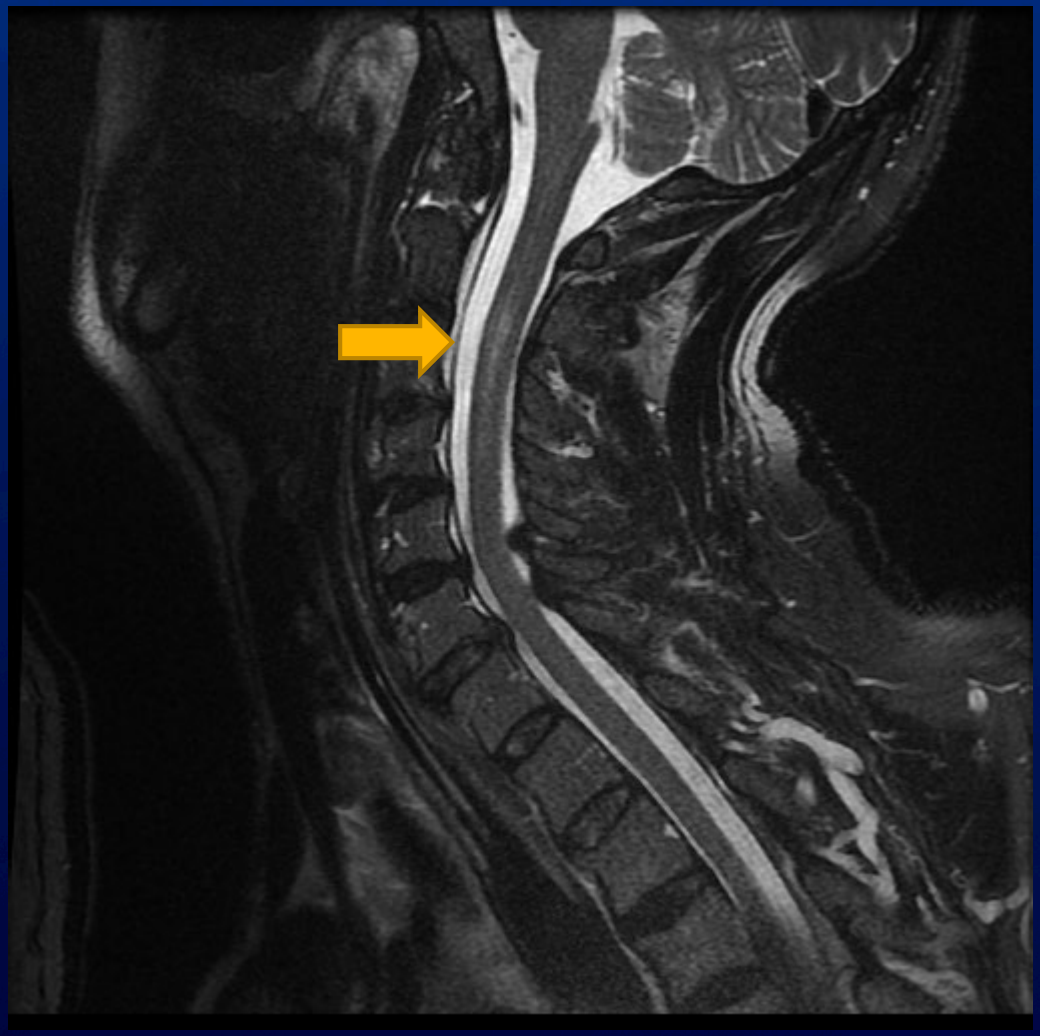

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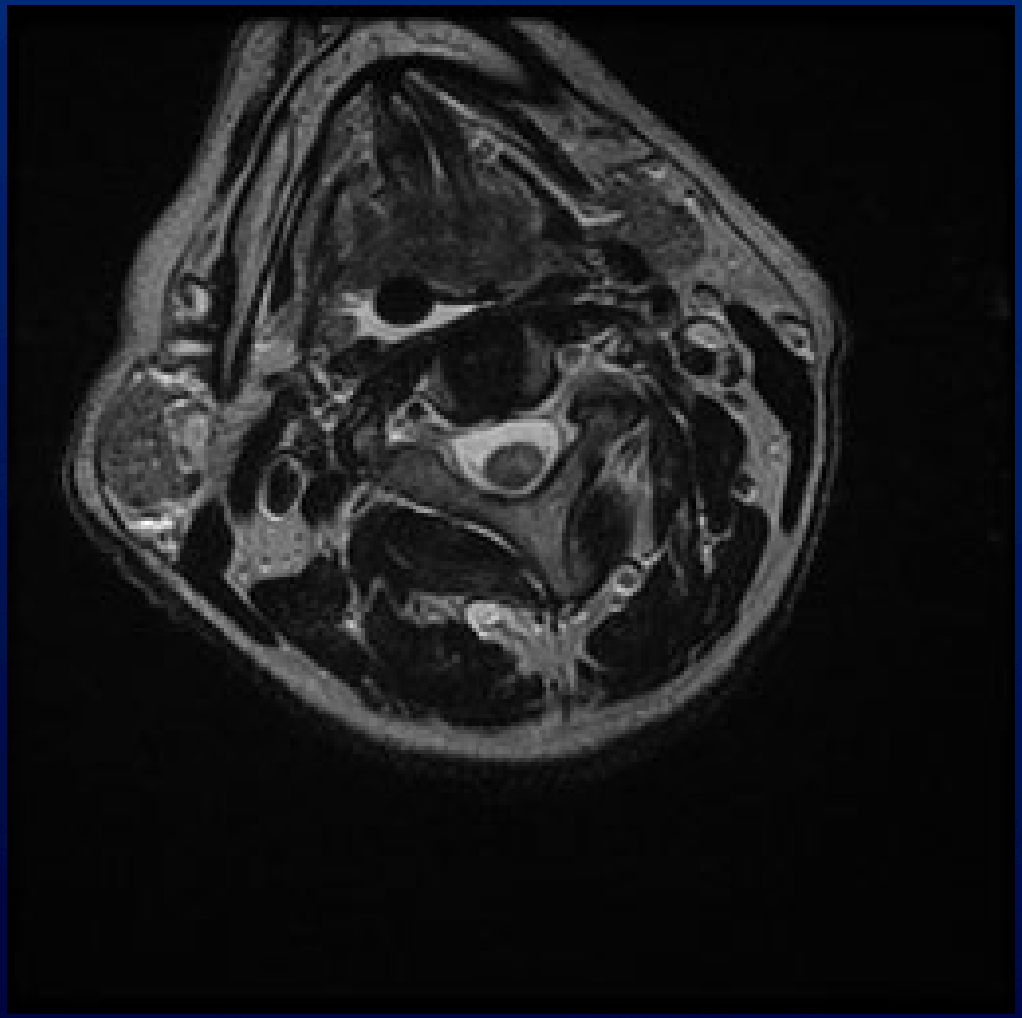

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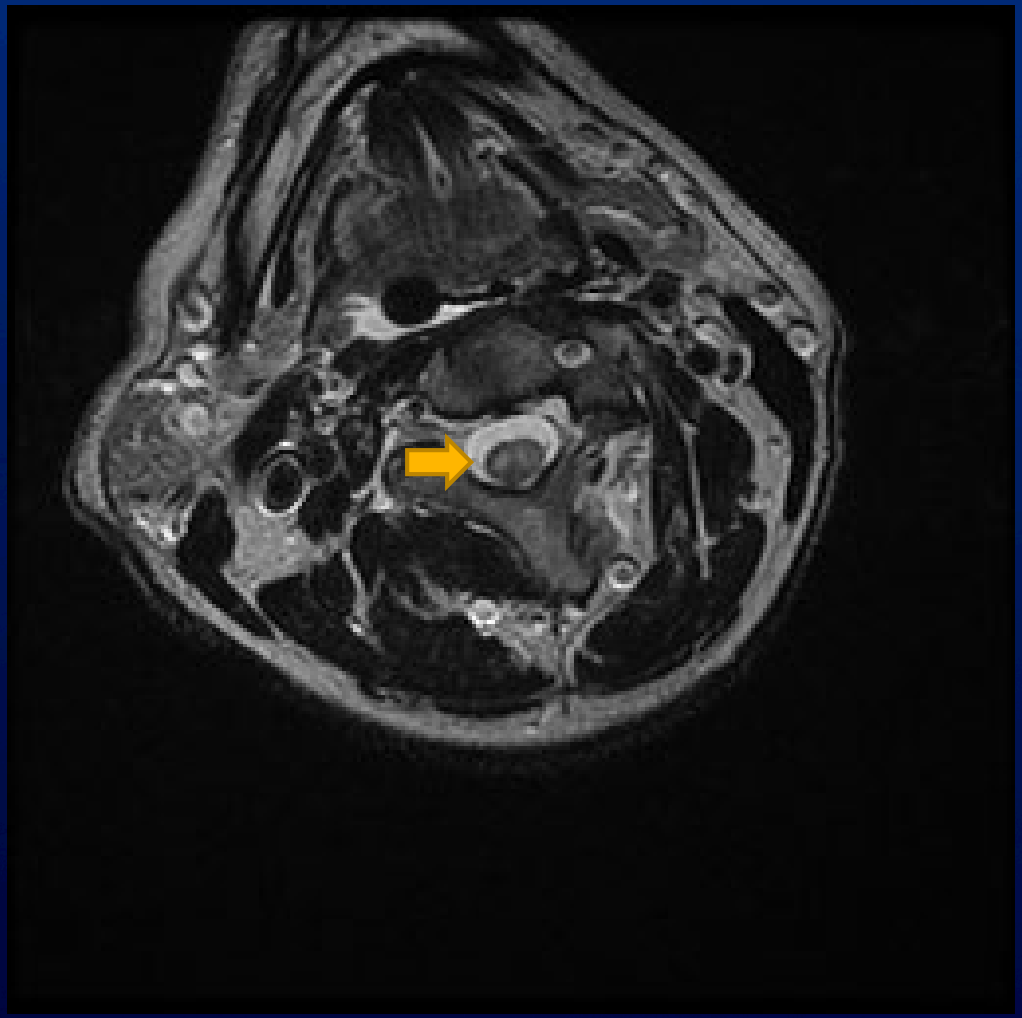

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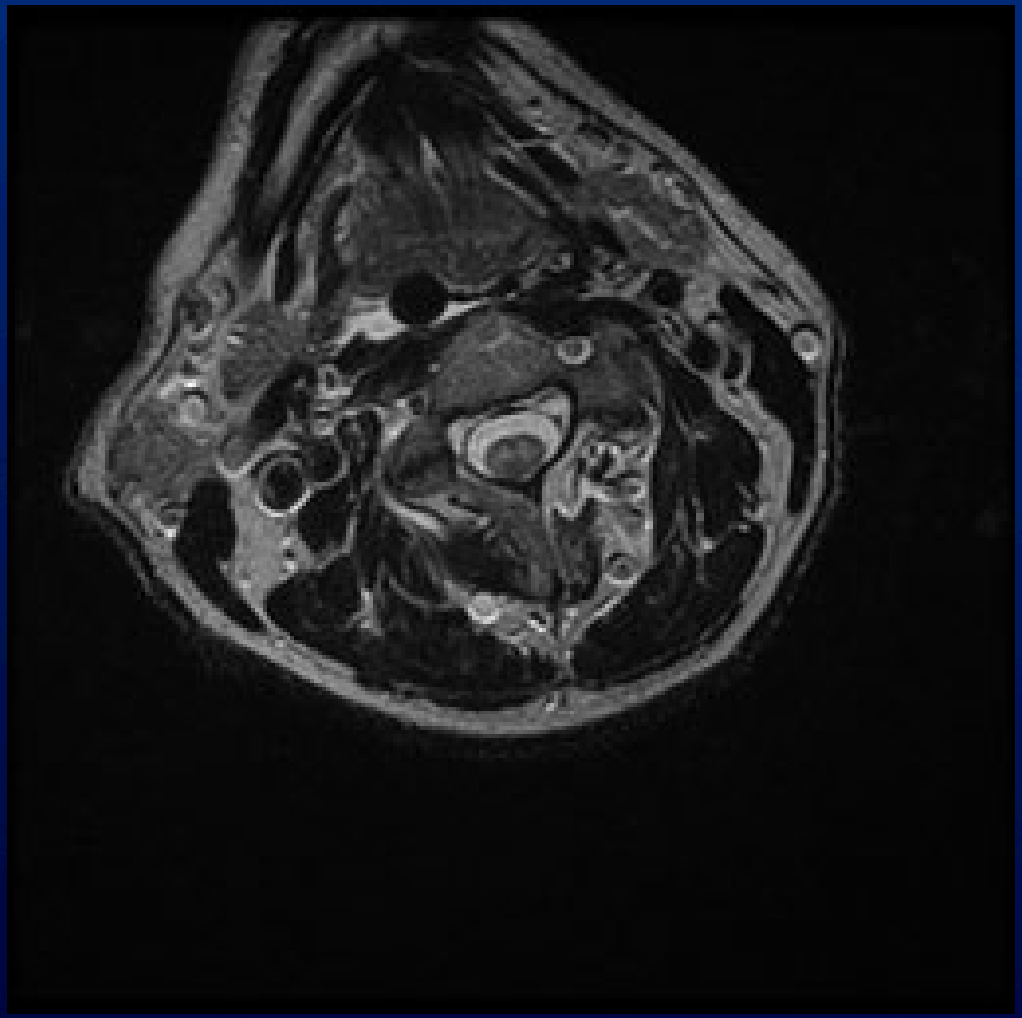

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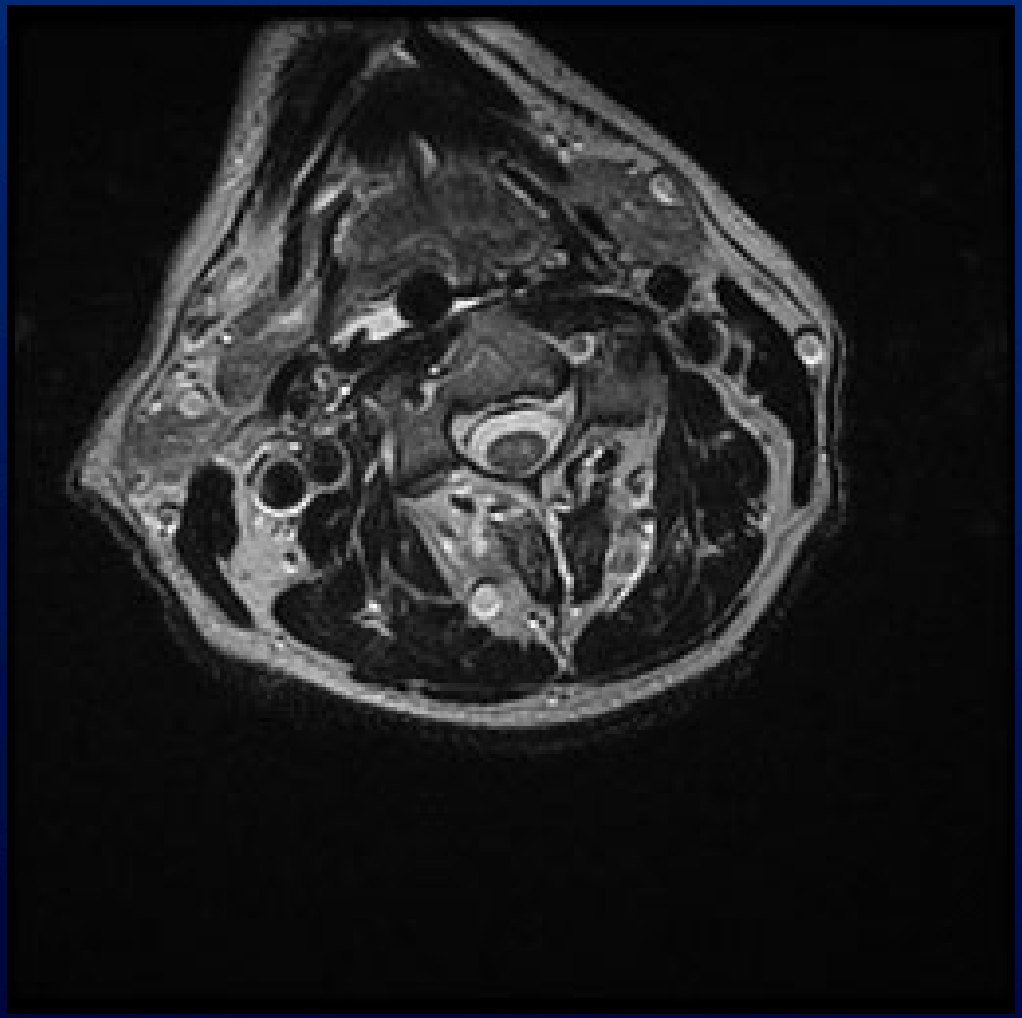

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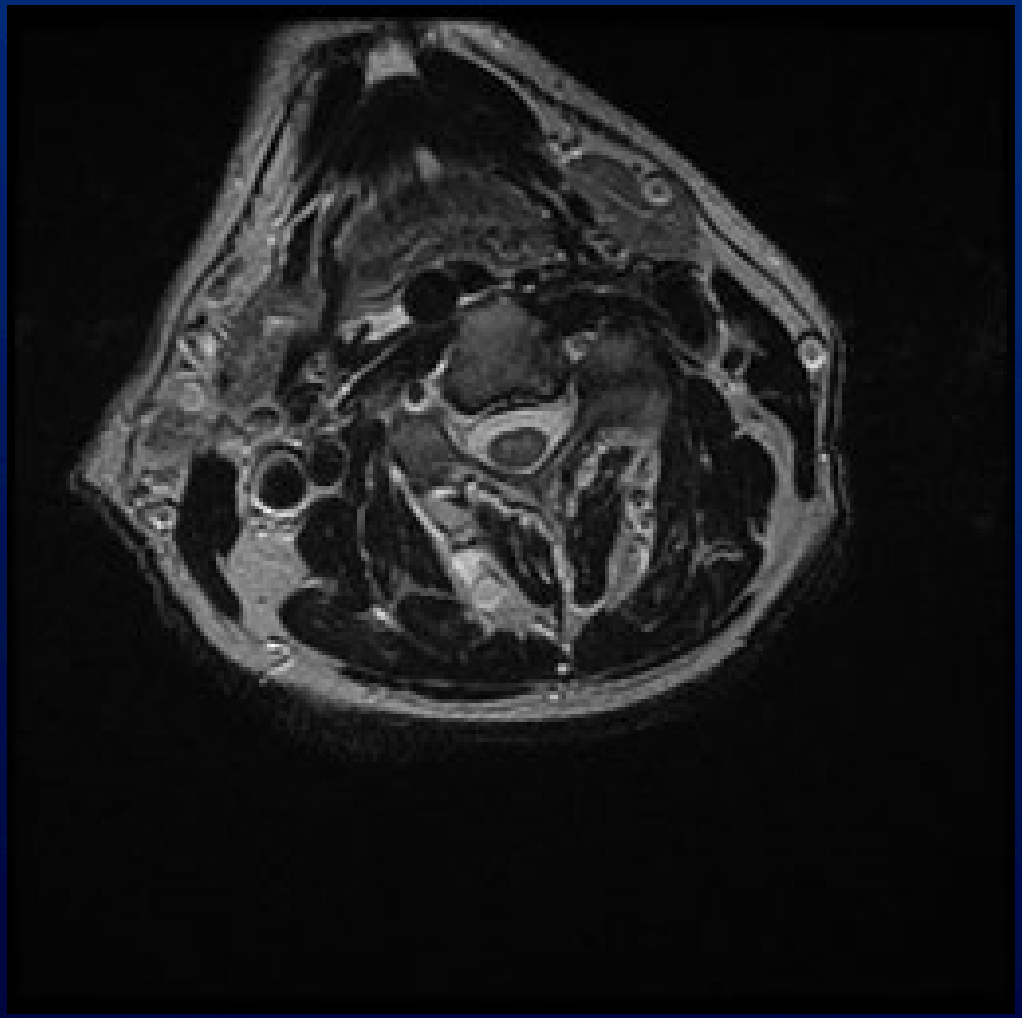

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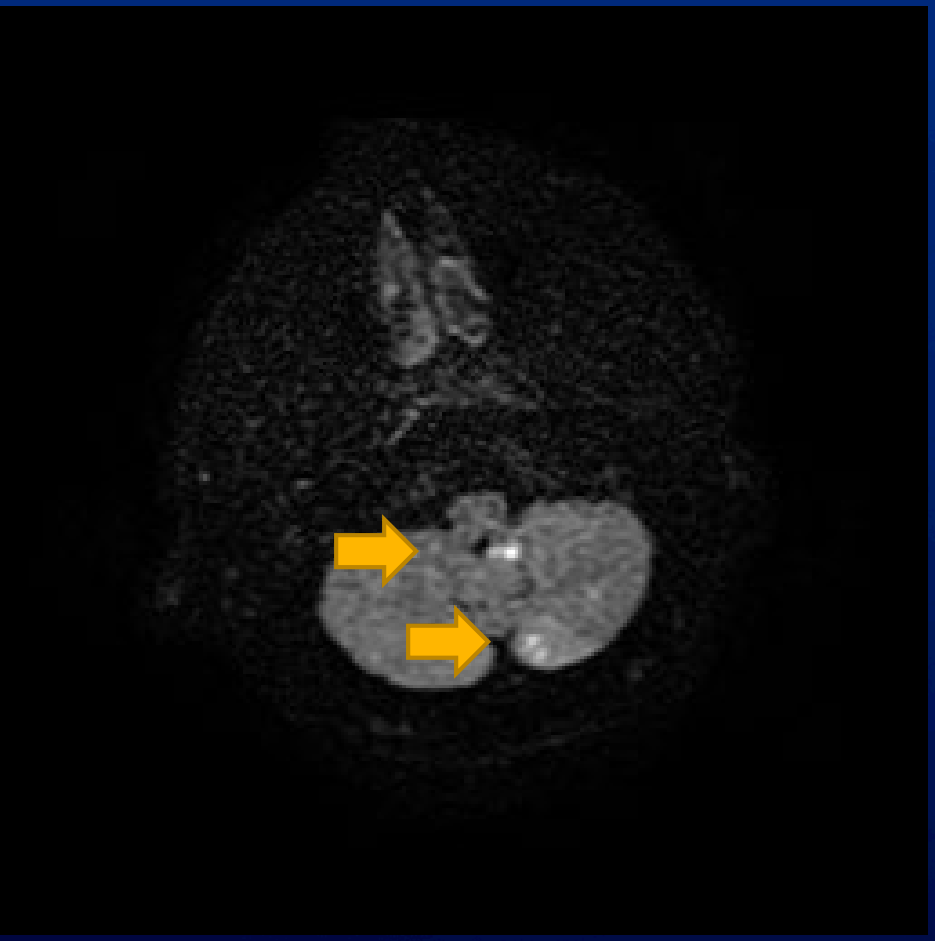

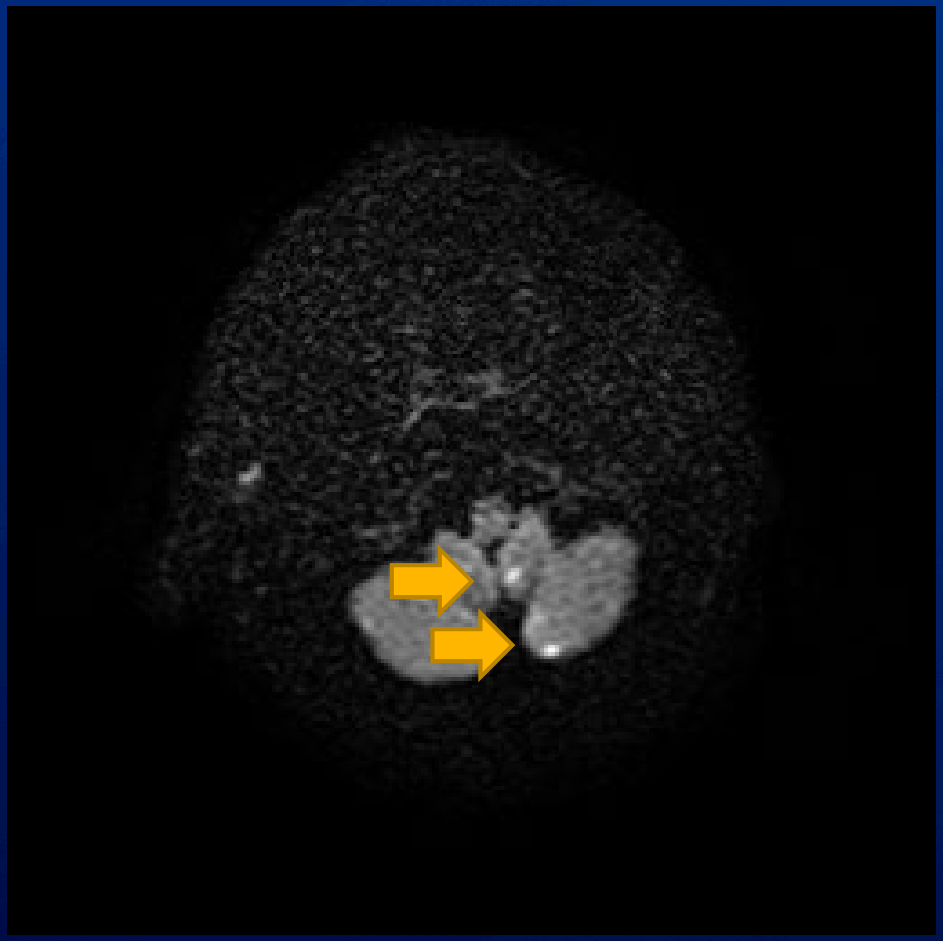

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# CTA: Vertebral artery occlusion; dissection at subclavian

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# Arterial case 1

- Dx: SCI and cerebellar stroke; vert dissection
- Treatment:
  - Pressors (phenylephrine, dopamine, midodrine)
  - Lumbar drain: 10 cc Q2H

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# 2 month follow-up MRI



 

# Arterial ischemia case 1

- Final outcome
  - Subtle left UMN weakness
  - Moderate-Severe pain/temp sensory loss

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# Spontaneous spinal cord infarction learning points

- Clinical
  - Very rapid decline (≤ 12 hrs)
  - DISSECTION (8%)
    - Aorta (5%)
    - Vertebral artery (3%)
  - Lumbar drain and MAP ⬆
  - Good outcomes possible
- Imaging
  - Small infarct, major deficits
  - Cervical cord (46%)

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Zalewski NL, Rabinstein AA, Krecke KN, et al. Characteristics of spontaneous spinal cord infarction and proposed diagnostic criteria. JAMA Neurol 2019;76(1):56-63.

# Arterial ischemia case 2

- 59 F
- CC: “transverse myelitis”
- PMH: HTN, HPL, Type 2 DM
- HPI:
  - Walking up stairs, acute buttock numbness
    - 2 min: entire legs
    - Fecal and urinary incontinence
  - 8 days later: paraparesis with electrical sensations BLE (mins)

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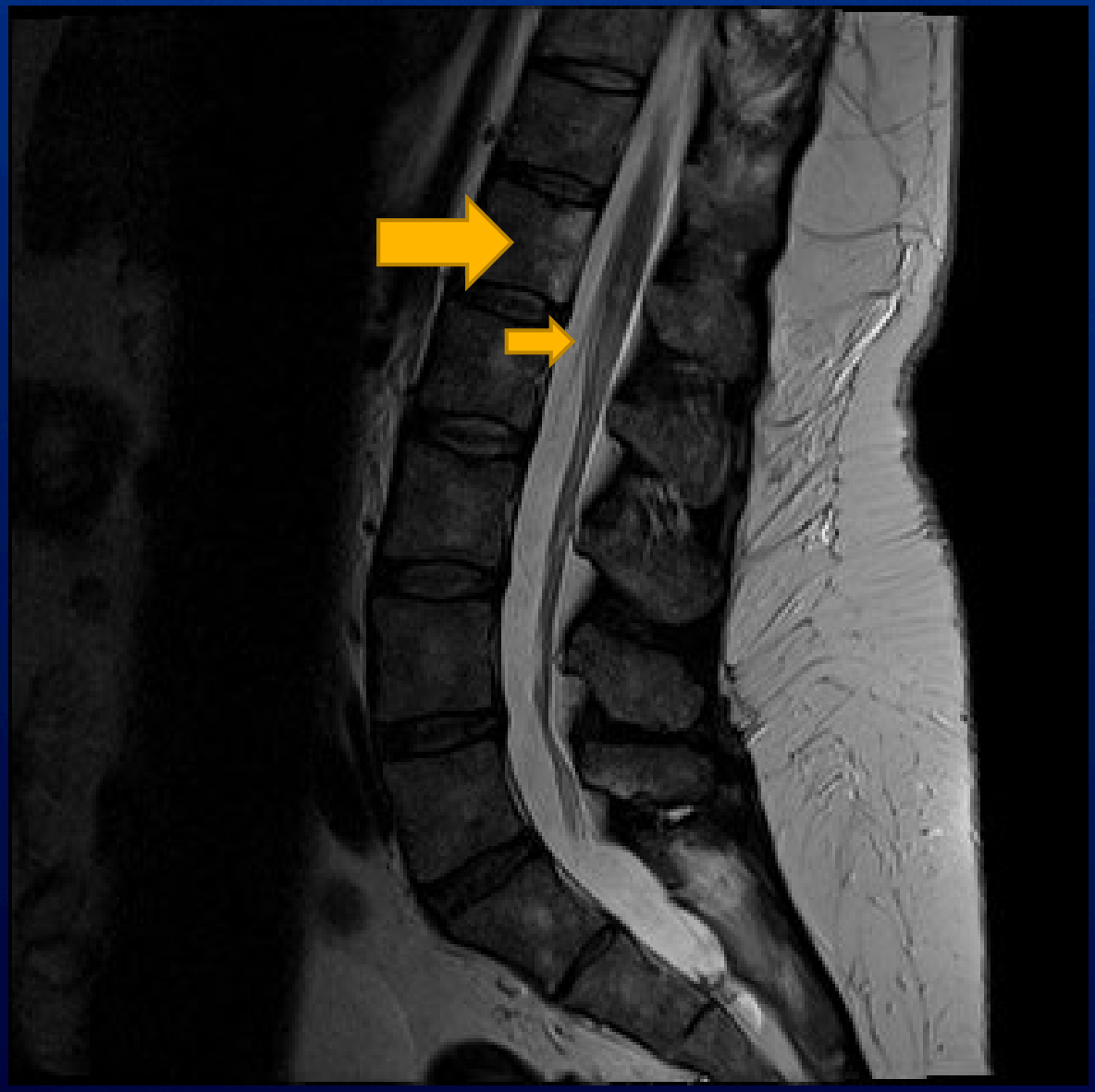
# Outside evaluation

- Serum:
  - Normal (including AQP4-IgG)
- CSF:
  - 2 WBCs, “elevated” protein, normal IgG index and 0 OCBs
  - Other normal
- MRI:





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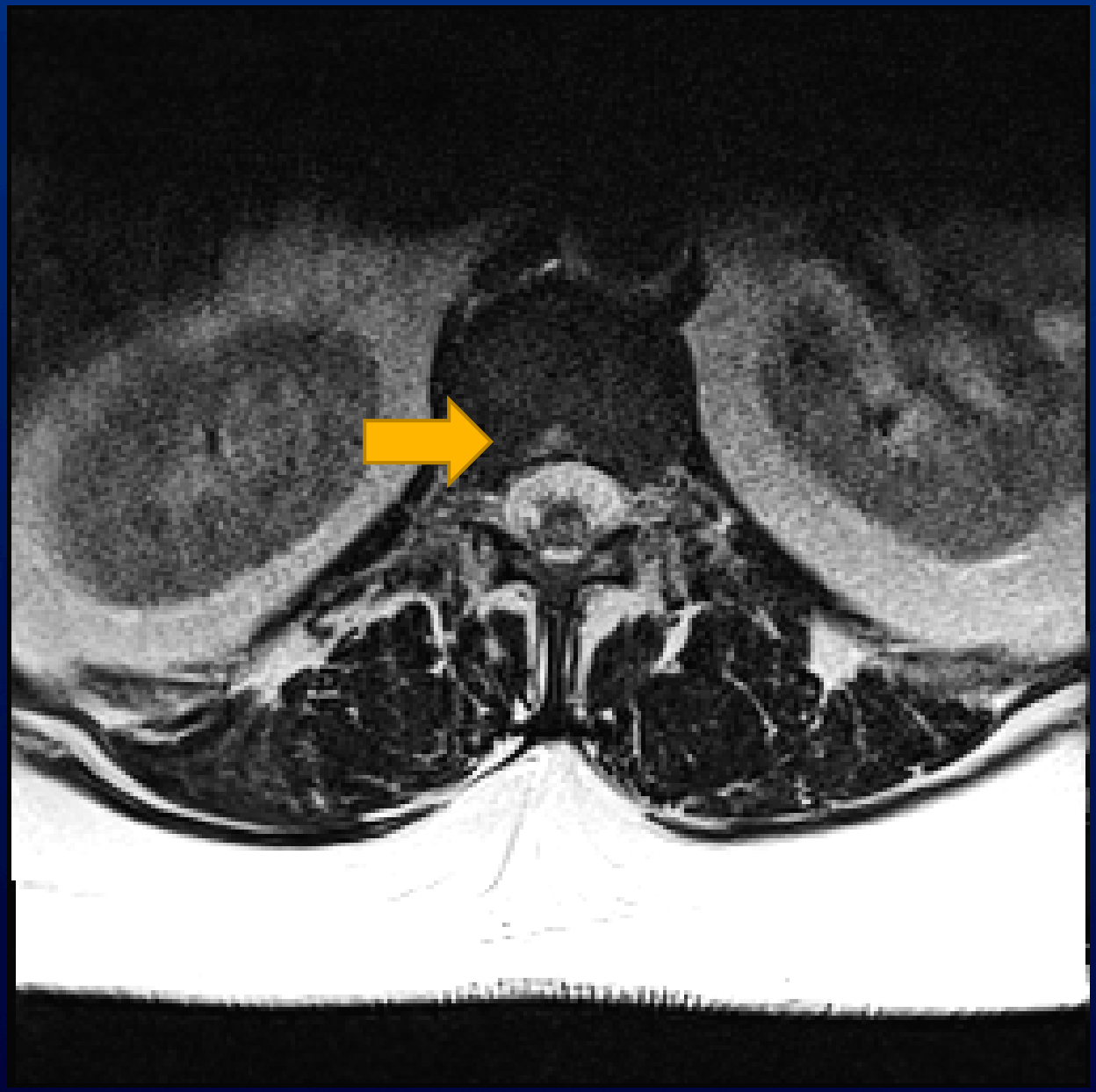

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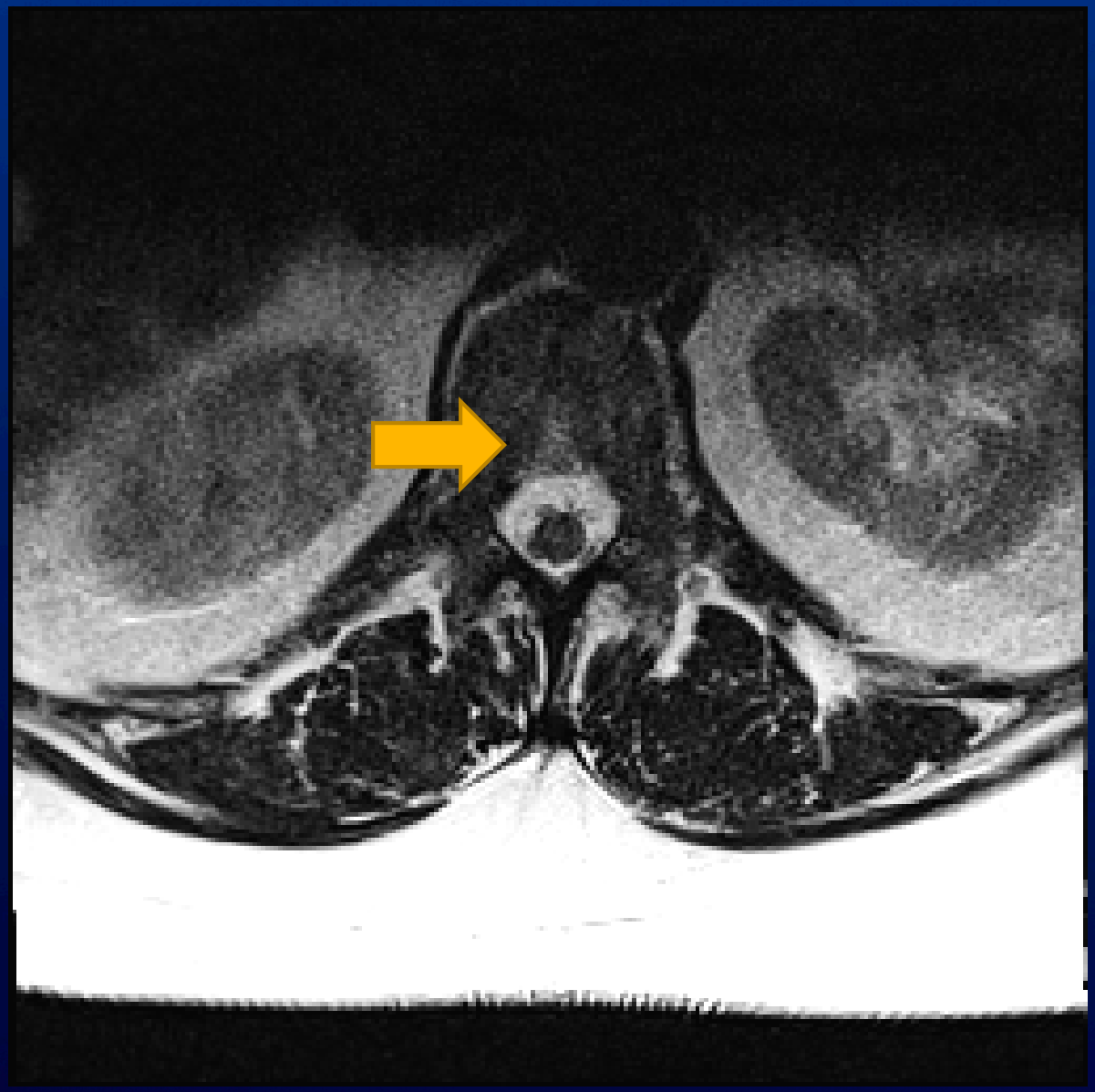

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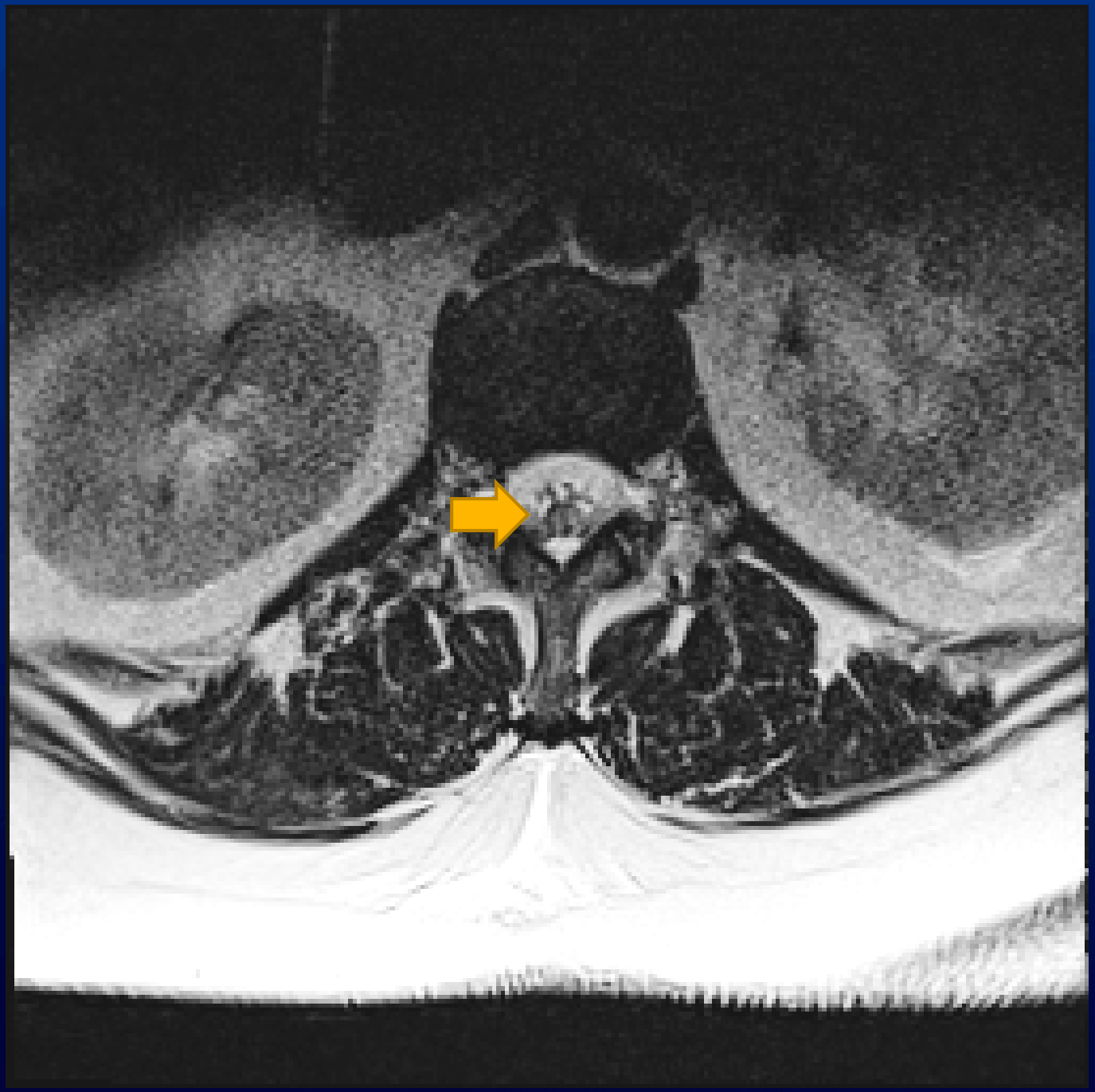

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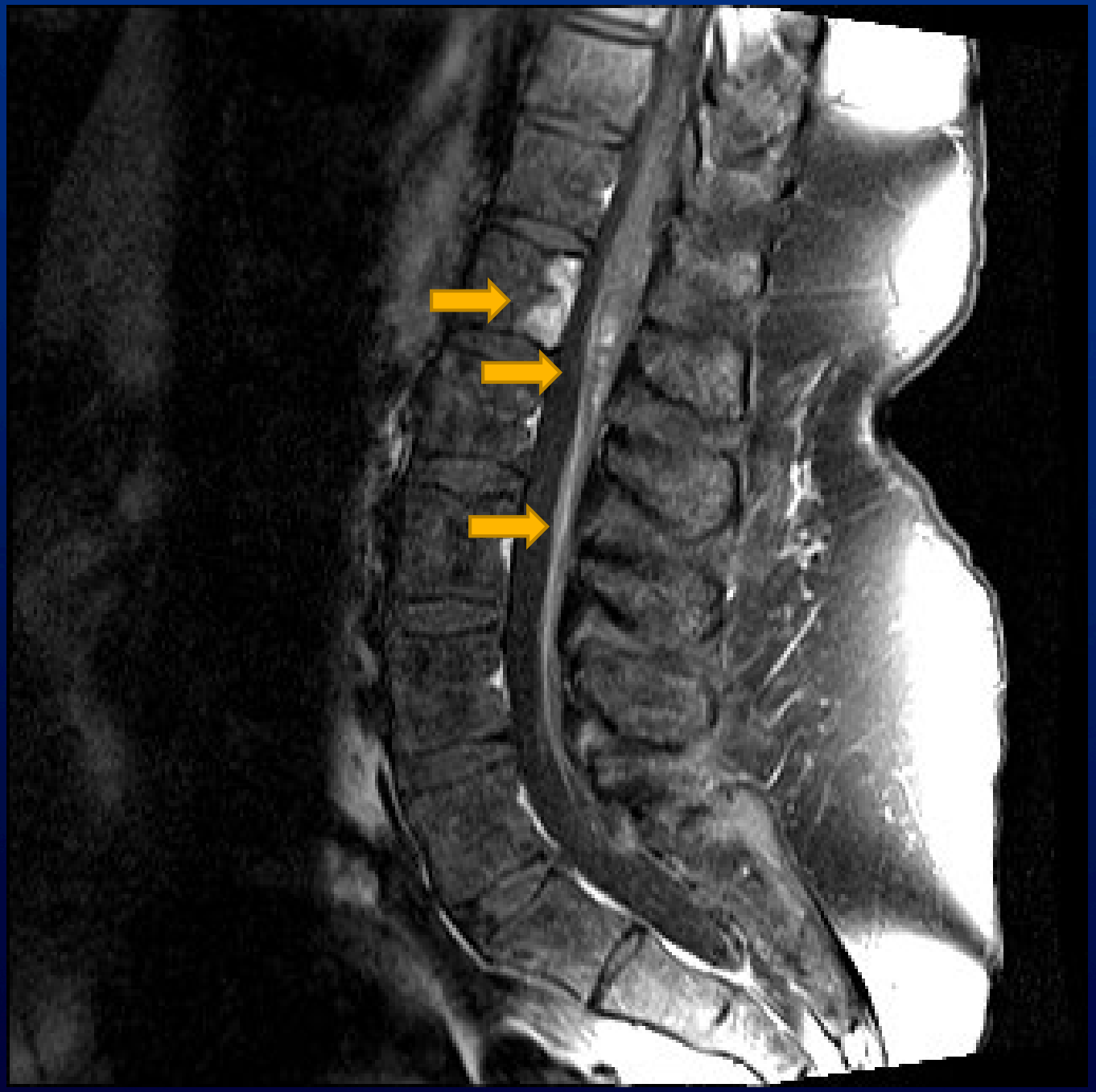

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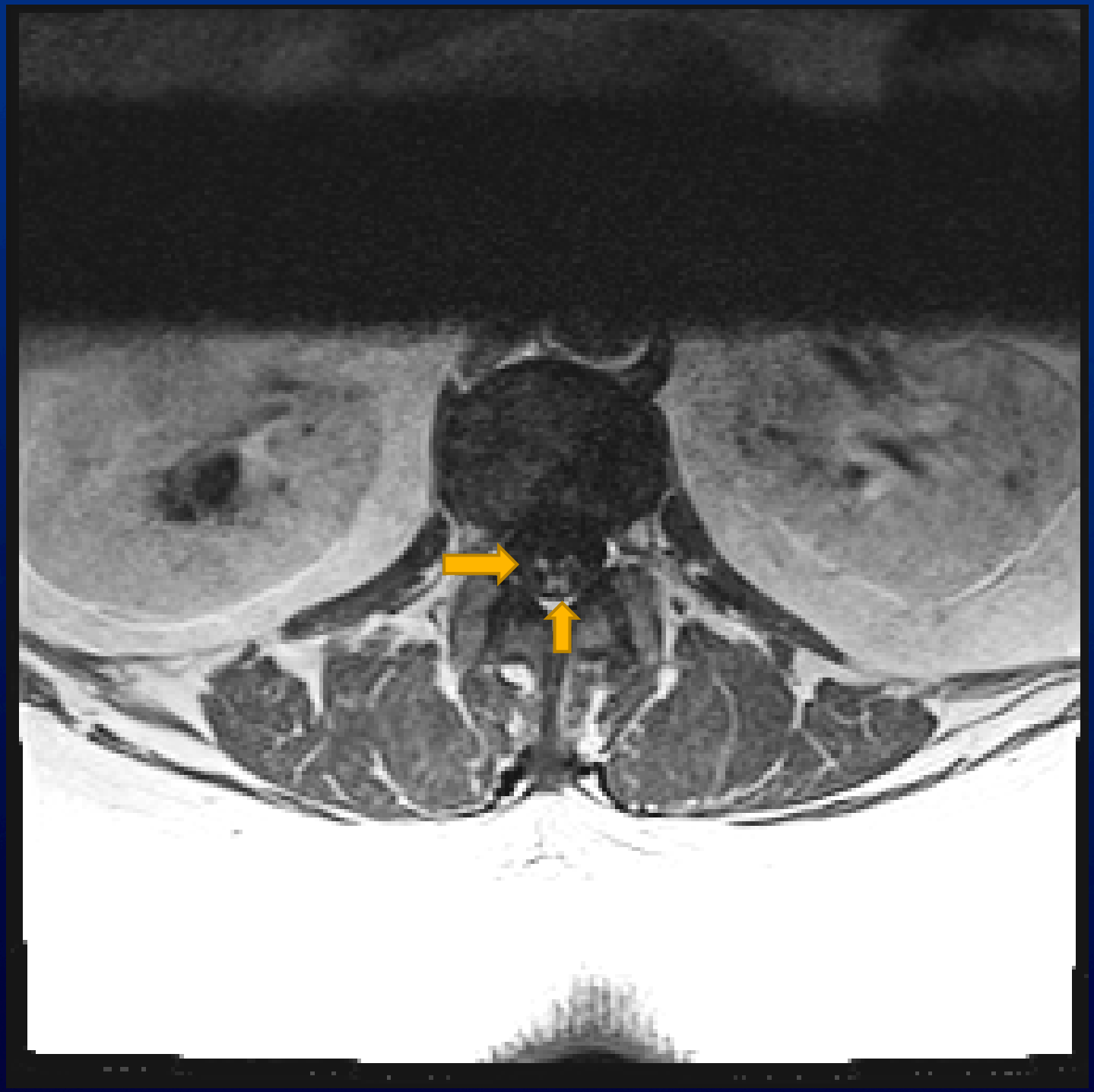

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# Arterial ischemia case 2

- MRA spinal canal
  - Normal

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# Spontaneous SCI Learning points

- Clinical
  - Prolonged time to nadir (8 days)
    - Rapid stepwise/stuttering (23%)
  - Pain at/before onset (~70%)
  - Vascular risk factors common
- Imaging
  - Confirmed vertebral body infarction (9%)
  - SCI enhancement (39%)
  - Ventral root enhancement (8 pts)

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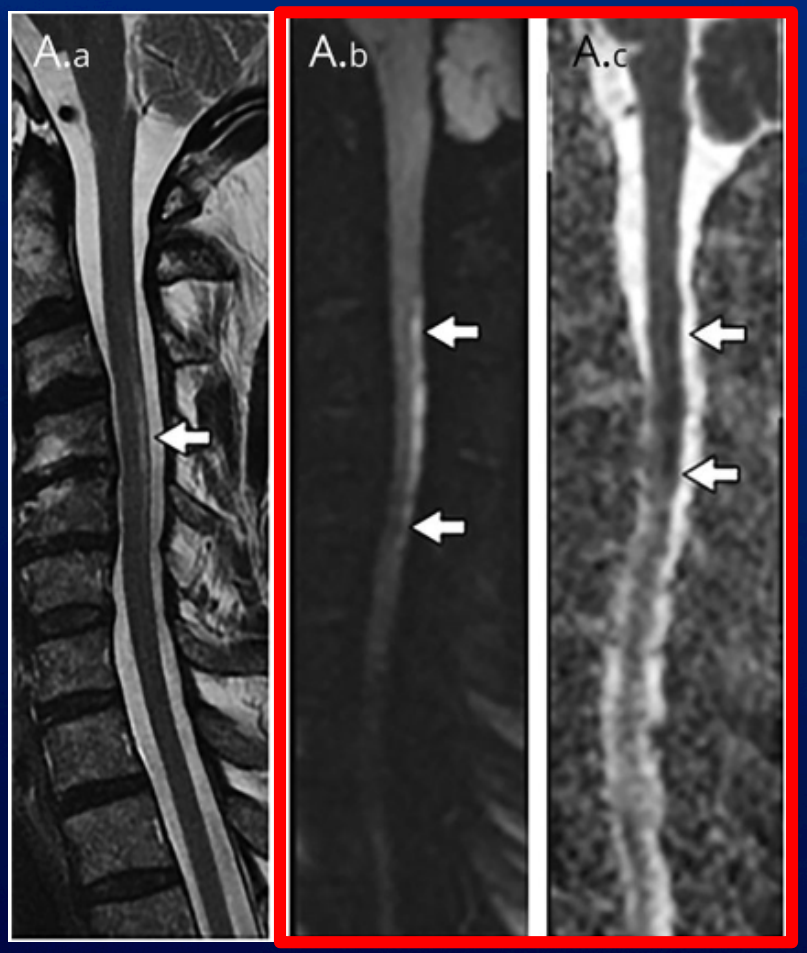
# DWI Spinal cord infarction

- Sensitivity: 50-70%




Picture:Zalewski NL, Rabinstein AA, Wijdicks EFM, Petty GW, Pittock SJ, Mantyh WG, Flanagan EP. Spontaneous posterior spinal artery infarction: An under-recognized cause of acute myelopathy. Neurology. 2018 Aug 28;91(9):414-417.

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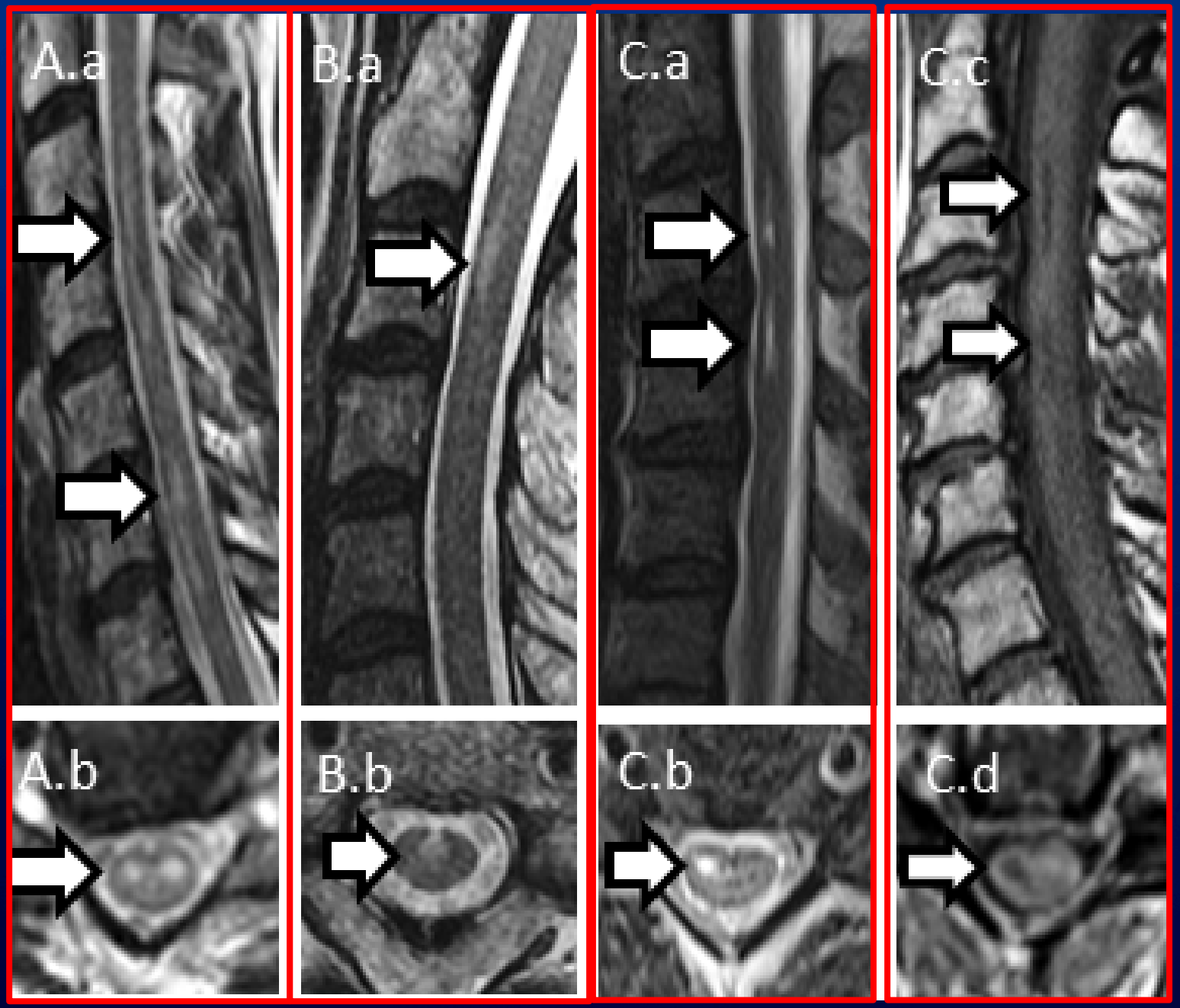


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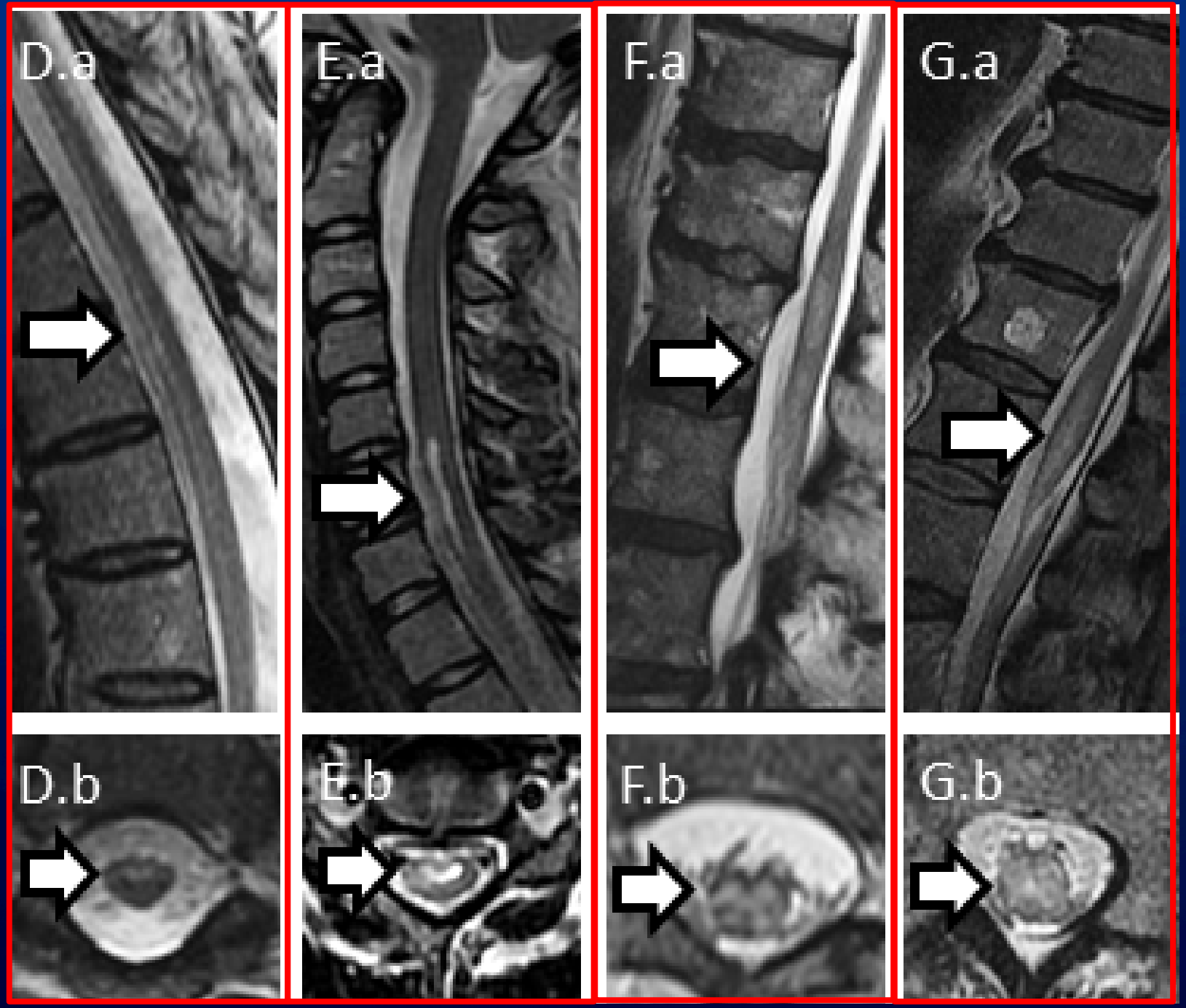
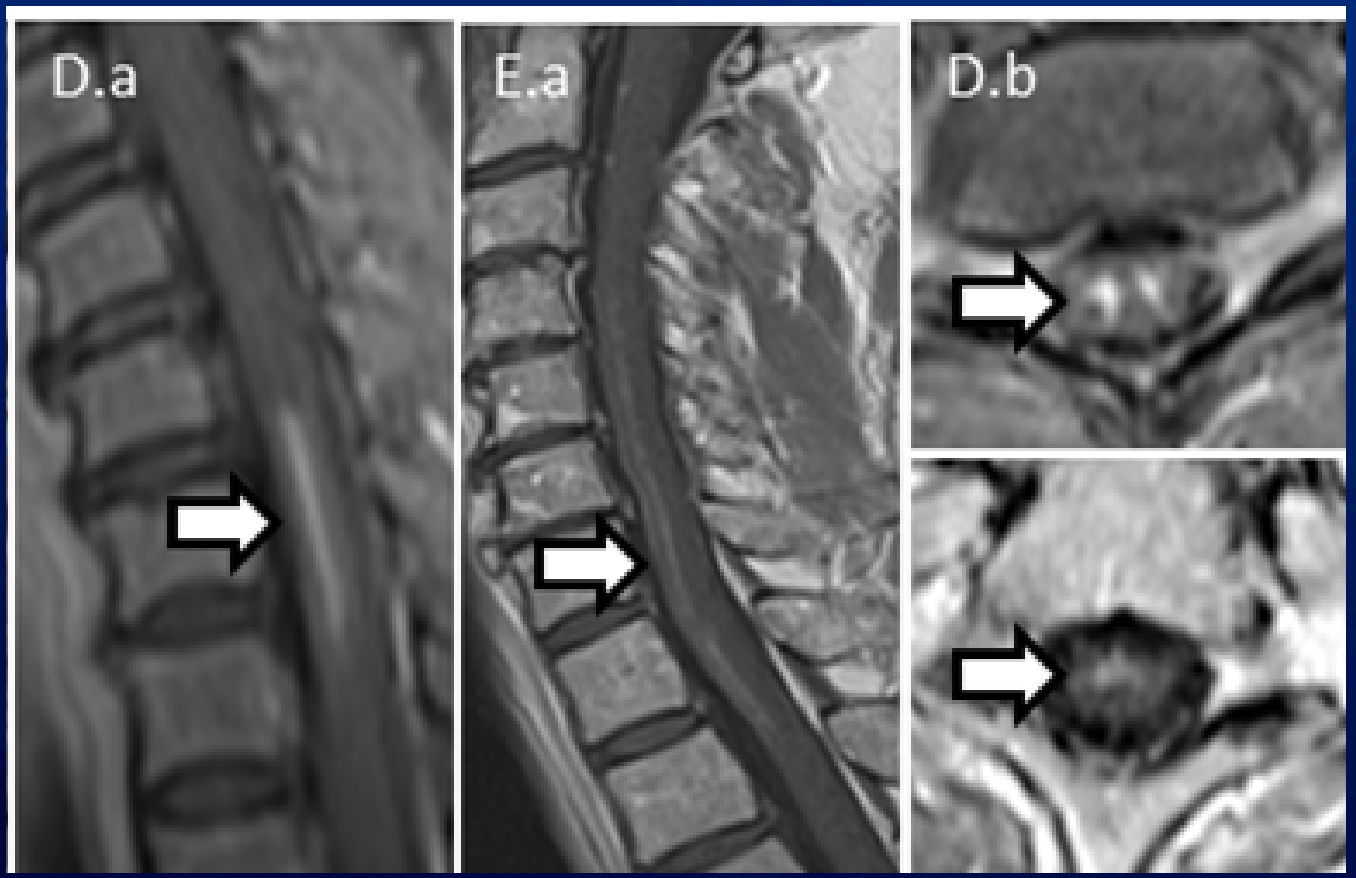


# SCI Enhancement



# SCI Diagnostic Criteria

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JAMA Neurology | Original Investigation

# Characteristics of Spontaneous Spinal Cord Infarction and Proposed Diagnostic Criteria

Nicholas L. Zalewski, MD; Alejandro A. Rabinstein, MD; Karl N. Krecke, MD; Robert D. Brown Jr, MD; Eelco F. M. Wijdicks, MD; Brian G. Weinshenker, MD; Timothy J. Kaufmann, MD; Jonathan M. Morris, MD; Allen J. Aksamit, MD; J. D. Bartleson, MD; Giuseppe Lanzino, MD; Melissa M. Blessing, DO; Eoin P. Flanagan, MBBCh

**IMPORTANCE** Spinal cord infarction (SCI) is often disabling, and the diagnosis can be challenging without an inciting event (eg, aortic surgery). Patients with a spontaneous SCI are often misdiagnosed as having transverse myelitis. Diagnostic criteria for SCI are lacking, hindering clinical care and research.

**OBJECTIVE** To describe the characteristics of spontaneous SCI and propose diagnostic criteria.

**DESIGN, SETTING, AND PARTICIPANTS** An institution-based search tool was used to identify patients evaluated at Mayo Clinic, Rochester, Minnesota, from January 1997 to December 2017 with a spontaneous SCI. Patients provided written consent to use their records for research. Participants were 18 years and older with a diagnosis of spontaneous SCI (n = 133), and controls were selected from a database of alternative myelopathy etiologies for validation of the proposed diagnostic criteria (n = 280).

**MAIN OUTCOMES AND MEASURES** A descriptive analysis of SCI was performed and used to propose diagnostic criteria, and the criteria were validated.

**RESULTS** Of 133 included patients with a spontaneous SCI, the median (interquartile range) age at presentation was 60 (52-69) years, and 101 (76%) had vascular risk factors. Rapid onset of severe deficits reaching nadir within 12 hours was typical (102 [77%]); some had a stuttering decline (31 [23%]). Sensory loss occurred in 126 patients (95%), selectively affecting pain/temperature in 49 (39%). Initial magnetic resonance imaging (MRI) spine results were normal in 30 patients (24%). Characteristic MRI T2-hyperintense patterns included owl eyes (82 [65%]) and pencil-like hyperintensity (50 [40%]); gadolinium enhancement (37 of 96 [39%]) was often linear and located in the anterior gray matter. Confirmatory MRI findings included diffusion-weighted imaging/apparent diffusion coefficient restriction (19 of 29 [67%]), adjacent dissection/occlusion (16 of 82 [20%]), and vertebral body infarction (11 [9%]). Cerebrospinal fluid showed mild inflammation in 7 of 89 patients (8%). Diagnostic criteria was proposed for definite, probable, and possible SCI of periprocedural and spontaneous onset. In the validation cohort (n = 280), 9 patients (3%) met criteria for possible SCI, and none met criteria for probable SCI.

**CONCLUSIONS AND RELEVANCE** This large series of spontaneous SCIs provides clinical, laboratory, and MRI clues to SCI diagnosis. The diagnostic criteria proposed here will aid clinicians in making the correct diagnosis and ideally improve future care for patients with SCI. The validation of these criteria supports their utility in the evaluation of acute myelopathy.

Supplemental content

CME Quiz at jamanetwork.com/learning

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*JAMA Neurol*. 2019;76(1):56-63. doi:10.1001/jamaneurol.2018.2734
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**1. Acute non-traumatic myelopathy** *(no proceeding progressive myelopathy)*

- Onset to nadir severe deficit(s)* ≤ 12 hours
- If onset to nadir > 12 hours:
  - Severe stepwise decline(s) ≤ 12 hours

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**2. Supportive MRI**

- T2-hyperintense cord lesion (Box 1b) *(non-compressive, no tortuous flow voids)*

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**3. CSF**

- Non-inflammatory (normal cell count, IgG index and no OCBs)

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**4. Specific MRI**

One of:

- DWI/ADC restriction
- Associated vertebral body infarction
- Acute arterial dissection/occlusion adjacent to lesion

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**5. Alternative diagnoses**

- Alternative diagnosis is not more likely (Box 1c)

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**DEFINITE** Spontaneous SCI: 1, 4, 5. Periprocedural SCI: 1, 4, 5 or 1, 2, 5
**PROBABLE** Spontaneous SCI: 1, 2, 3, 5. Periprocedural SCI: 1, 5
**POSSIBLE** Spontaneous SCI: 1, 2 and 5

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# Mechanism of SCI?

- Idiopathic/atherosclerotic: 68%
- Fibrocartilaginous embolism: 14%
  - 53% nearby/adjacent disc extrusions
- Arterial dissection: 8%
- Others

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Zalewski NL, Rabinstein AA, Krecke KN, et al. Characteristics of spontaneous spinal cord infarction and proposed diagnostic criteria. JAMA Neurol 2019;76(1):56-63.

# Treatment

- Acute treatment SCI?
  - tPA? At least 13 cases published w/ no harm[1]
  - Lumbar drain and/or MAP ⬆
- Secondary stroke prevention
  - Treat mechanism/risk factors

1. Zalewski NL. Vascular myelopathies. Continuum: Lifelong Learning in Neurology—Spinal Cord Disorders, Volume 27, Issue 1 In Progress



 

# Neurofilament lesion Area Ratio in SCI

- ≥0.35 pg/(mL·mm$^2$) NAR, 95% sens, 86% spec

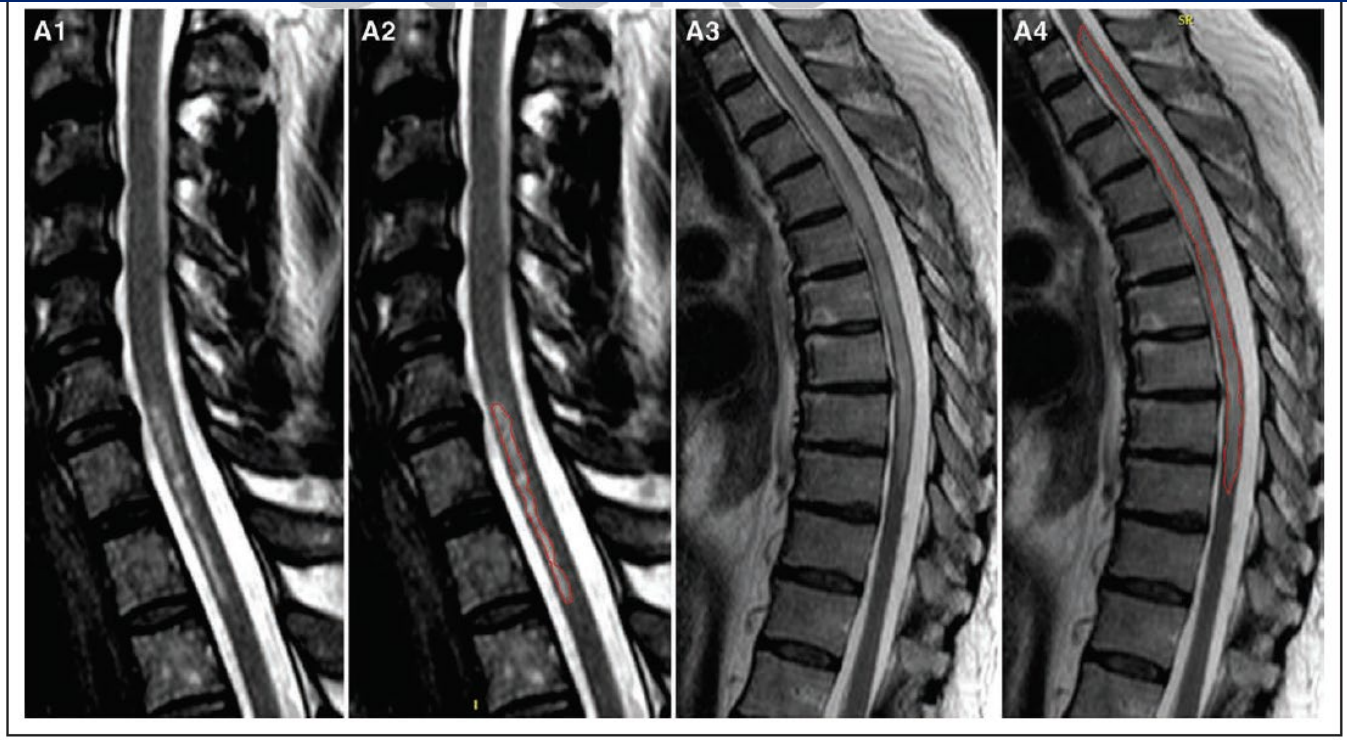


**Figure 1. Manual assessment of lesion area on spinal cord magnetic resonance imaging (MRI).**
Sagittal T2-weighted MRI in a patient with spinal cord infarction (SCI; **A**1) and AQP4 (aquaporin 4)-IgG-associated myelitis (**B**1), with their respective sagittal lesion areas manually outlined (**A**2, **B**2; red tract).

Sechi E, Mariotto S, McKeon A, Krecke KN, Pittock SJ, Ferrari S, Monaco S, Flanagan EP, Zanzoni S, Rabinstein AA, Wingerchuk DM, Nasr DM, Zalewski NL. Serum Neurofilament to Magnetic Resonance Imaging Lesion Area Ratio Differentiates Spinal Cord Infarction From Acute Myelitis. Stroke. 2021 Jan 11:STROKEAHA120031482. doi: 10.1161/STROKEAHA.120.031482. Epub ahead of print. PMID: 33423516.

 

# Spontaneous SCI learning points

- Acute severe myelopathy
  - ≤ 12 hours
- MRI
  - Variable T2-hyperintensity patterns
  - Gad enhancement (linear/grey/arterial)
  - Specific findings
    - DWI (*incomplete sensitivity*)
    - Vertebral body infarct
    - Vessel occlusion/dissection
- CSF
  - Non-inflammatory (*mild abnormalities*)
- *Others: Neurofilament? (NAR)*

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# Venous congestion/ischemia case

- 62 M
- PMH: L5 laminectomy, HTN
- HPI:
  - Progressive walking difficulty x 3 years
  - Weakness + sensory loss in BLE, bowel/bladder incontinence, back + leg pain
  - Episodes of severe paraparesis
- Exam
  - Moderate paraparesis, severe mixed sensory loss to waist, reflexes diminished, plantars ⬆

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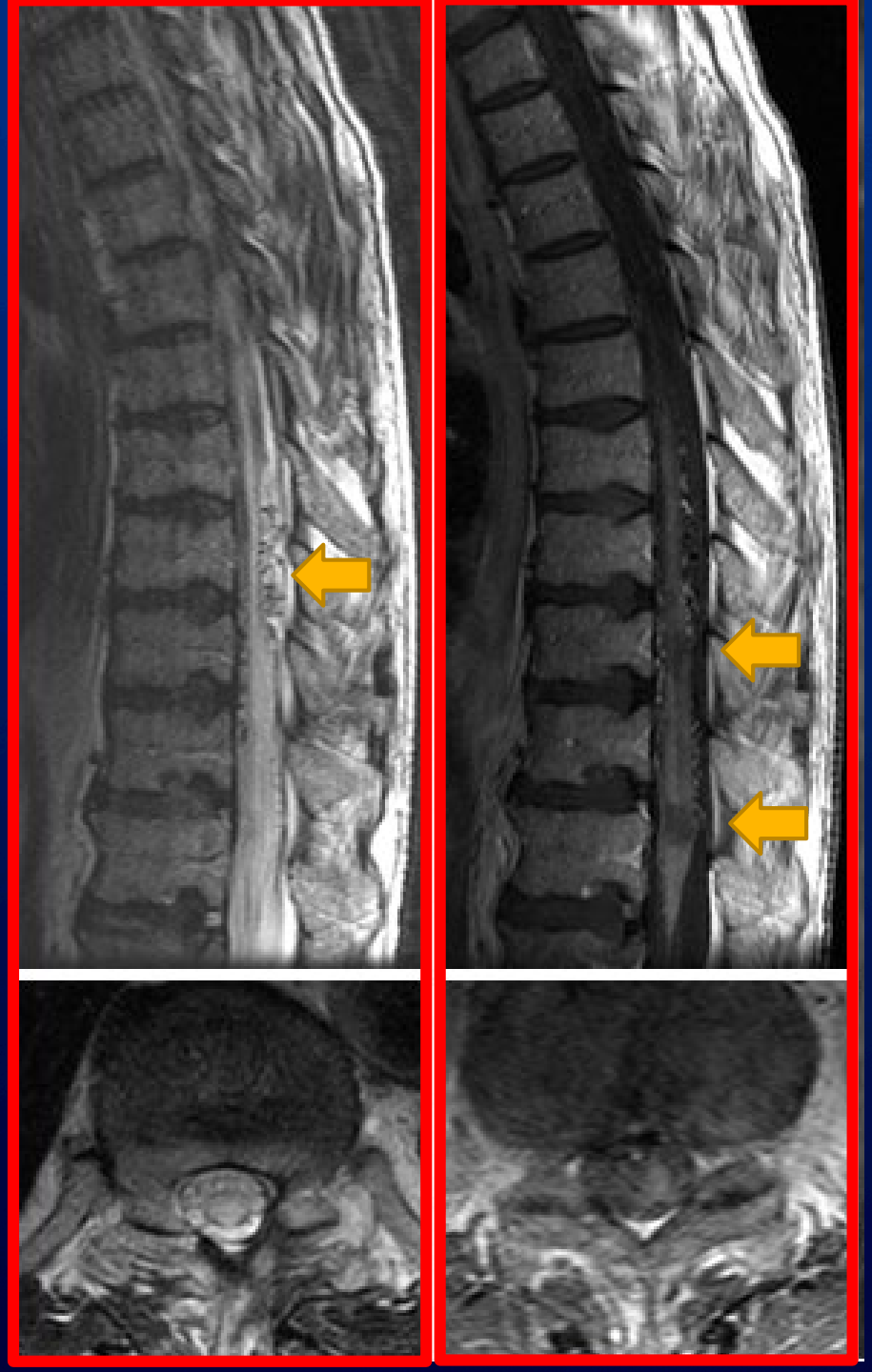

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# Venous congestion/ischemia case

- Administered steroids outside (dx myelitis)
  - Abrupt decline in function
- Referral → angiogram: spinal DAVF
  - Tx: embolization

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# Venous congestion/ischemia learning points

- sDAVF
  - Progressive thoracic myelopathy/conus
    - Conus 90%
    - Episodic worsening
    - Steroids worsening
  - UMN and LMN features
  - Flow voids on T2 and/or T1 + Gad imaging
    - ~ 80%[1,2,3]
  - Enhancement common
    - 65-85%

1. Muralidharan R, Saladino A, Lanzino G, et al. The clinical and radiological presentation of spinal dural arteriovenous fistula. Spine (Phila Pa 1976) 2011;36(25):E1641-E1647. doi:10.1097/BRS.0b013e31821352dd
2. Toossi S, Josephson SA, Hetts SW, et al. Utility of MRI in spinal arteriovenous fistula. Neurology 2012;79(1):25-30. doi:10.1212/WNL.0b013e3182582f07
3. Krings T, Geibprasert S. Spinal dural arteriovenous fistulas. AJNR Am J Neuroradiol 2009;30(4):639-648. doi:10.3174/ajnr.A1485

# Venous congestion/ischemia teaching points

- MRA spinal canal estimate fistula level → helps angiographer
- Tx:
  - Embolization: ~70-80% effective
  - Surgical disconnection: 98%[1,2,3]
- Follow-up MRI (~ 3 months)
  - Possible T2-hyperintense signal in cord, absent flow voids

1. Nasr DM, Brinjikji W, Rabinstein AA, Lanzino G. Clinical outcomes following corticosteroid administration in patients with delayed diagnosis of spinal arteriovenous fistulas. J Neurointerv Surg 2017;9(6):607-610. doi:10.1136/neurintsurg-2016-012430
2. Steinmetz MP, Chow MM, Krishnaney AA, et al. Outcome after the treatment of spinal dural arteriovenous fistulae: a contemporary single-institution series and meta-analysis. Neurosurgery 2004;55(1):77-87; discussion 87-78. doi:10.1227/01.neu.0000126878.95006.0f
3. Open and endovascular treatment of spinal dural arteriovenous fistulas: a 10-year experience. J Neurosurg Spine 2017;26(4):519-523. doi:10.3171/2016.9.SPINE16394

# Other venous congestion/ischemia

- Spinal epidural and pial AVFs
- Behçet disease
- Inferior vena cava thrombosis
- Others (Spinal cord compression from an enlarged vena cava, extraspinal arteriovenous fistulas, prothrombotic state with venous thrombosis (eg, pelvic vein thrombosis, cancer, sepsis), epidural infection leading to venous thrombosis, nitrogen emboli of the venous system)
- Workup for others: CTV chest/abdomen/pelvis

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# Hematomyelia case

- 50 F
- HPI:
  - Abrupt neck pain and upper extremity weakness
  - Pain became progressive over days, extending into her shoulders
  - Over the next few years of observation, she developed recurrent episodes

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# Hematomyelia case

- Dx: cavernous malformation
- Tx: surgical resection
  - Good improvement

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# Cavernous malformation teaching points

- Likely most common etiology of nontraumatic hematomyelia
- Annual hemorrhage as low as 0.8% incidental, 10% symptomatic
- Back pain, deficits hours → days
- MRI
  - A well-defined lobulated masslike lesion
  - Heterogenous T1- and T2-weighted signal intensity surrounded by a well-defined dark T2-hypointense rim (classic popcorn appearance)
  - GRE/SWI dark
  - T1-hyperintense signal and perilesional edema ➡ recent hemorrhage
  - Less distinct border acutely

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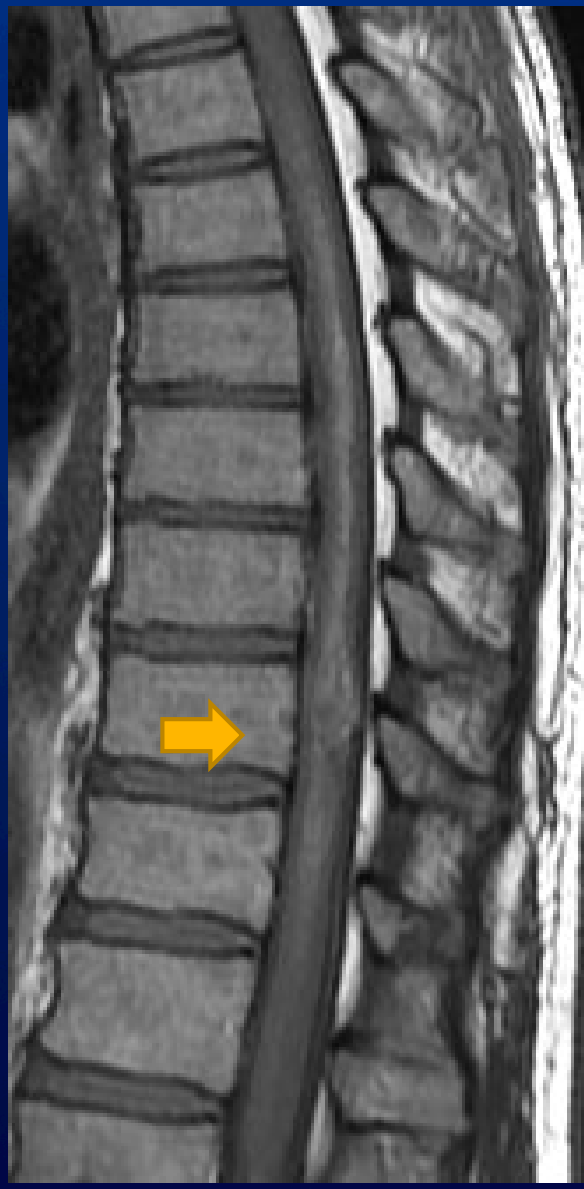

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# AVM

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# AVM

- sDAVF (type I) 70% of AVS
- 30% of cases types II-V: “AVM”
- Predominant presentation: hematomyelia (50%).
- Annual hemorrhage 4%, 10% for recurrent
- MRI intramedullary flow voids, mixed T1, T2 features
- 70% of patients deteriorate within 4 years, early surgery recommended
- Endovascular embolization, surgical resection, and radiosurgery are options

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# Other hematomyelia

| Hematomyelia etiologies | Helpful features |
|---|---|
| **Trauma** | Most common cause of hematomyelia |
| **Cavernous malformation** | Common cause of hematomyelia; young male patient; heterogeneous lobular mass, popcorn appearance with dark rim of T2 hypointensity; pain frequently precedes myelopathy deficits |
| **Arteriovenous malformation** | Common cause of hematomyelia, young patient, intramedullary flow voids |
| **Bleeding diatheses and medications** | Prior history of bleeds, family history; contributing medications or supplements |
| **Neoplasm** | Focal masslike component not typical of intramedullary spinal cord cavernous malformation, history of primary neoplasm, contrast enhancement |
| **Miscellaneous** (spinal cord aneurysm, intrasyringal hemorrhage, vasculitis, radiation, infection [eg, postinfluenza, varicella-zoster virus, herpes simplex virus type 2, acute hemorrhagic leukoencephalitis, checkpoint inhibitor]) | Rare, typical radiographic and clinical features for respective disorders |
| **Idiopathic** | Common diagnosis of exclusion in hematomyelia, hypertension as typical risk factor |

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# Extraparenchymal hemorrhage case

- 73 F
- PMH: A Fib, HTN, HPL, smoking
- HPI:
  - Minor slip and fall down on ground/ice, minor pain
  - Severe back pain over 1-2 hrs → BLE and hand weakness, sensory loss

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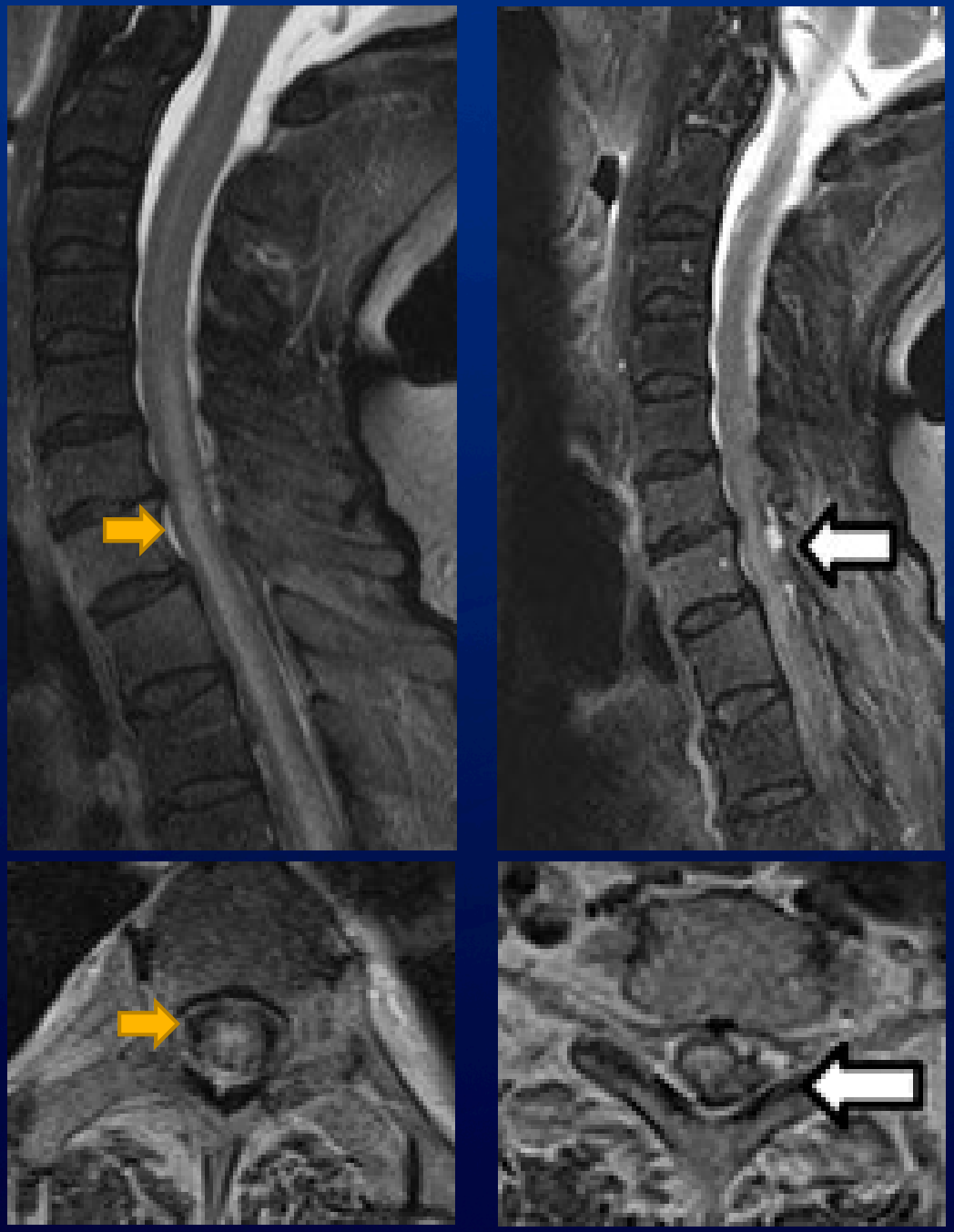

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# Extraparenchymal hemorrhage case

- Dx: Spinal subdural hematoma, SAH
- Tx:
  - Reverse anticoagulation
  - Emergent subdural evacuation
  - Control blood pressure
- Outcome:
  - Improvement over several weeks

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# Extraparenchymal hemorrhage case

- Spinal subdural hematoma
  - Acute back pain, myelopathy, or radicular
  - Traumatic/anticoag most common
  - Tx: similar to epidural hematoma (emergent decompression symptomatic)
- Spontaneous Spinal SAH:
  - Rare (1% SAH)
  - Severe neck or back pain, variable myelopathy, other SAH features
  - Tx: conservative, mechanism
  - HTN (common), AVM, aneurysm, vascular neoplasm, cavernoma, sDAVF, blood dyscrasia, others

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# Extraparenchymal hemorrhage

- Epidural hematoma
  - Back pain + hemiparesis common
  - Reverse
    - Coagulopathy
    - Meds
  - BP control
  - Emergent surgery
    < 12 hrs[1]

1. Lawton MT, Porter RW, Heiserman JE, Jacobowitz R, Sonntag VK, Dickman CA. Surgical management of spinal epidural hematoma: relationship between surgical timing and neurological outcome. Journal of neurosurgery. 1995;83(1):1-7
2. Kim et al. Spontaneous spinal epidural hematoma of the thoracic spine after herbal medicine: a case report. BMC Complement Altern Med. 2018

 

# Extraparenchymal hemorrhage

- Epidural hematoma
  - Spontaneous (non-traumatic): 0.1 per 100,000 person-years
  - Secondary to coagulopathy (eg, anticoagulation, liver disease, or portal hypertension)
  - Asymptomatic/minimal deficits → conservative management option
    - May acutely deteriorate

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# SUMMARY

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# Vascular Myelopathy Summary

- Vascular category:
  - Arterial ischemia
  - Venous congestion/ischemia
  - Hematomyelia
  - Extraparenchymal hemorrhage
- SCI diagnostic criteria

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Thank you

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# Diagnostic Evaluation of Suspected Spontaneous Spinal Cord Infarction

**MRI cervical/thoracic spine** (with DWI/ADC and gadolinium, +/- brain MRI)
**MRA cervical spine** (if cervical level affected, with T1 fat sat for dissection)
**Blood**
- **HbA1c**
- **Fasting lipids, glucose**
- **CBC**
- **PT/aPTT**

**CSF** (can defer if very low suspicion alternative and/or concern of LP risks)
- **Cell count, glucose, protein, IgG index, oligoclonal bands**

***Consider:**

Additional CSF studies
- -VZV PCR
- -VDRL
- -Lyme PCR/serology
- -Cryptococcus

Additional blood studies
- -AQP4/MOG
- -Syphilis serology
- -Lyme serology
- -Hypercoaguable profile
- -ESR, CRP
- -ANA, ENA, ANCA, dsDNA
- -Paraneoplastic evaluation
- -Toxicology screen

Additional imaging
- -MRI brain (evaluate for concurrent stroke, other lesions)
- -MRA spinal canal or spinal angiogram (if clinical/radiographic concern for vascular malformation or vasculitis)
- -CT chest/abdomen/pelvis (if suspicion for underlying malignancy)

Additional cardiac evaluation
- -Echocardiogram
- -Holter monitoring

Other
- -EMG (if suspicion GBS)

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