Imaging of Disease of the Spinal Cord

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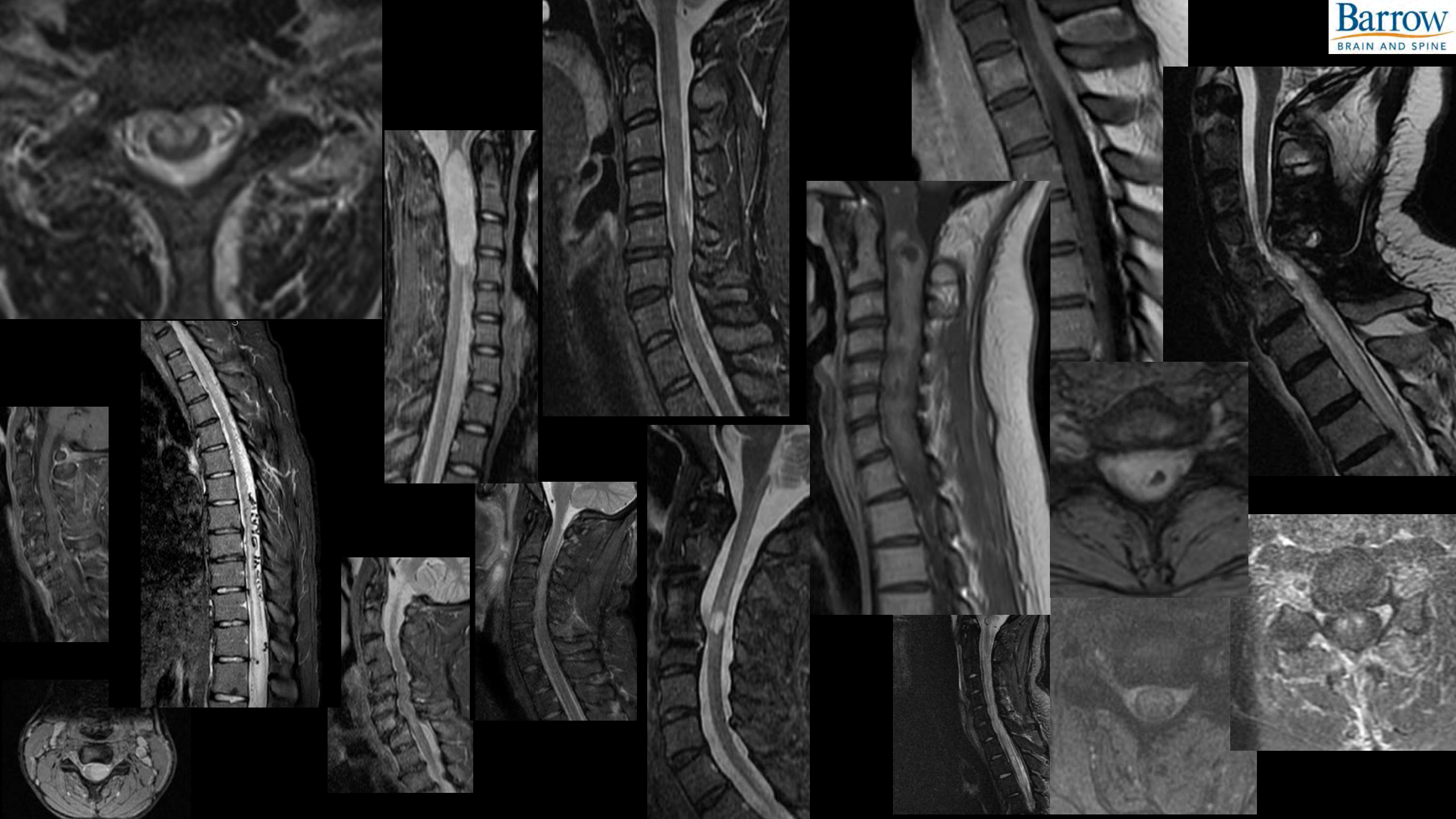
Assistant Professor
Barrow Neurological Institute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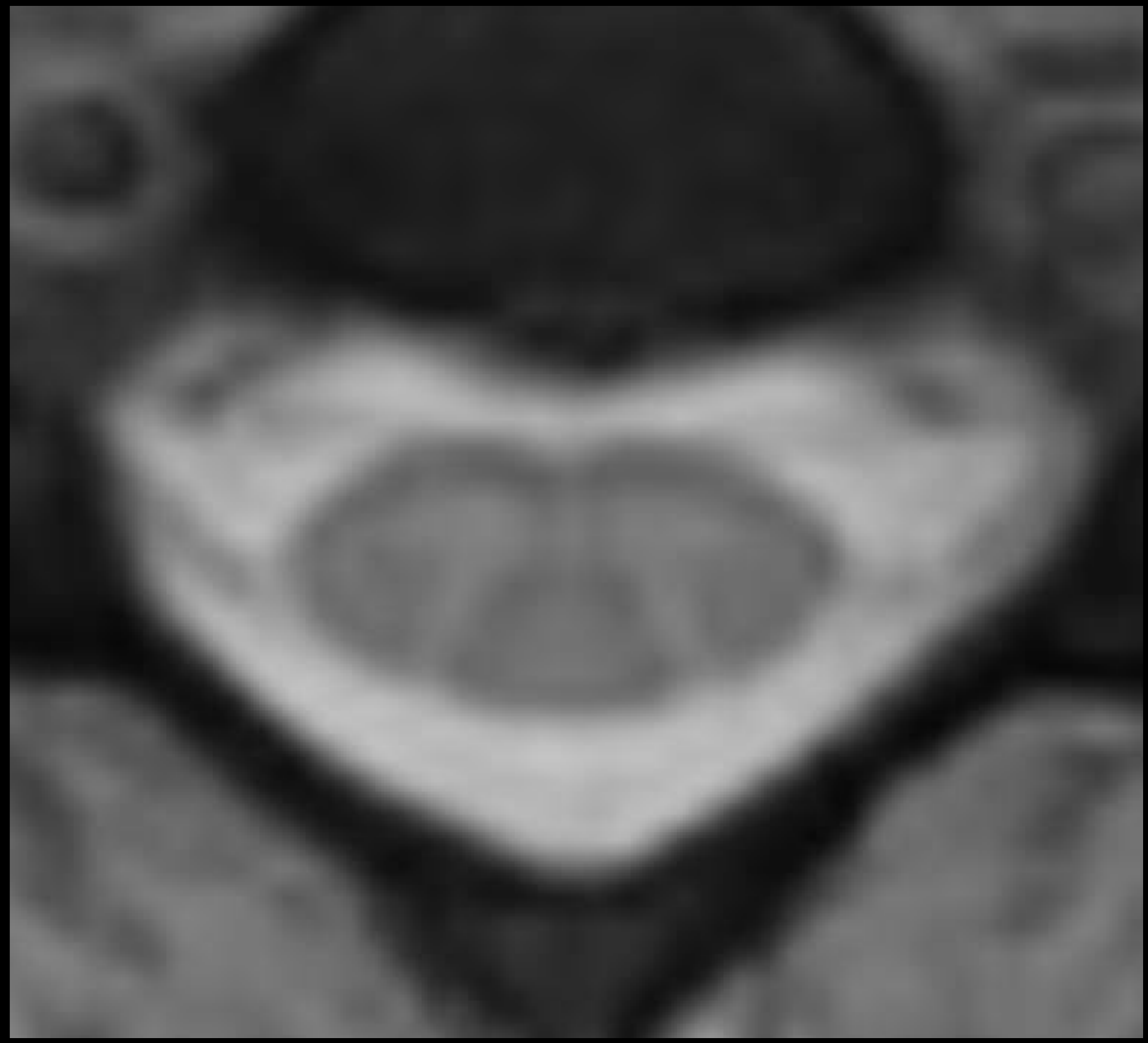
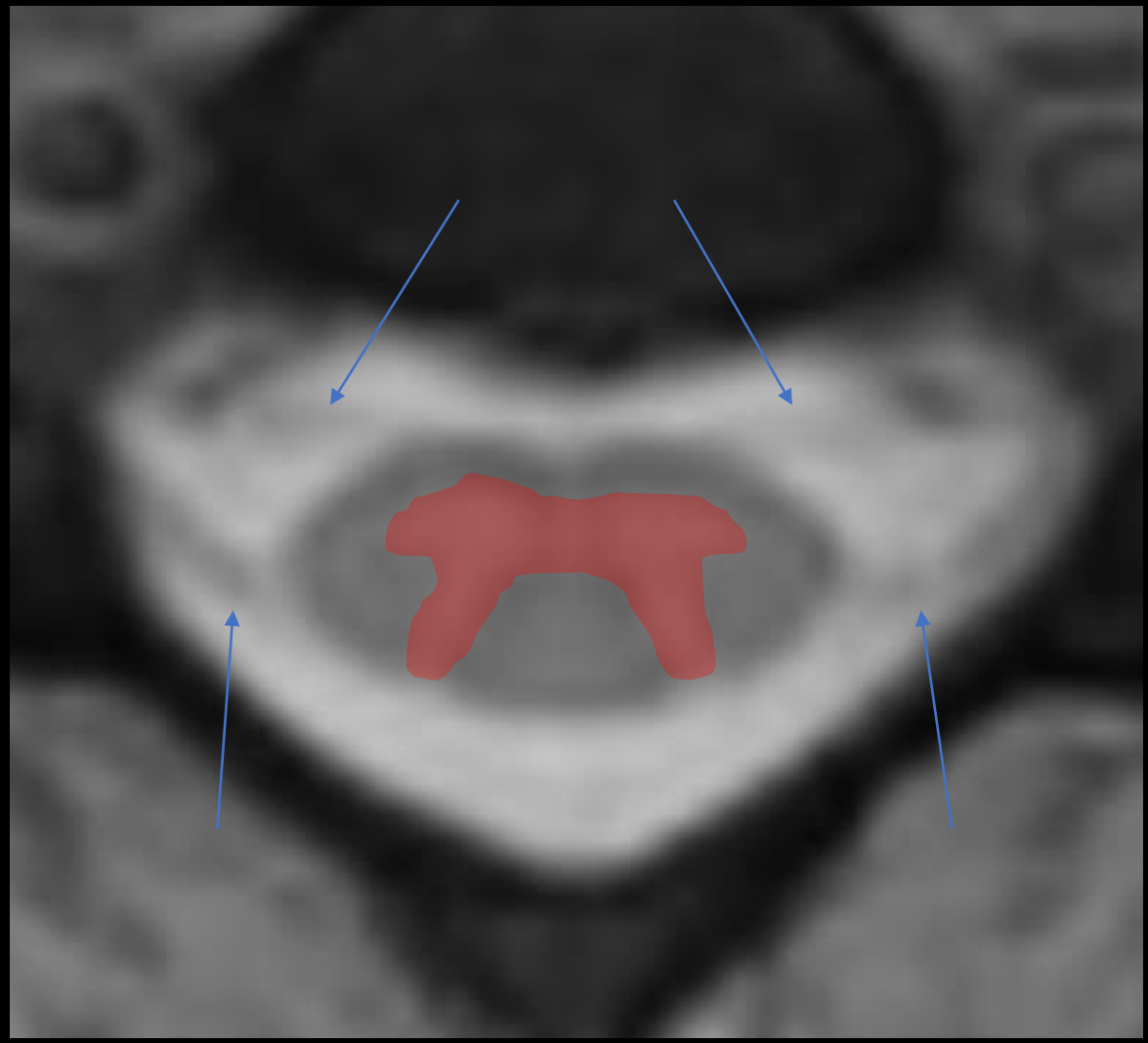
BRAIN AND SPINE

Objectives

- Understand pertinent imaging anatomy of the spinal cord.
- Understand the imaging appearance of common diseases of the spinal cord.
- Using a pattern based imaging approach be able to develop a differential diagnosis for spinal cord disease.

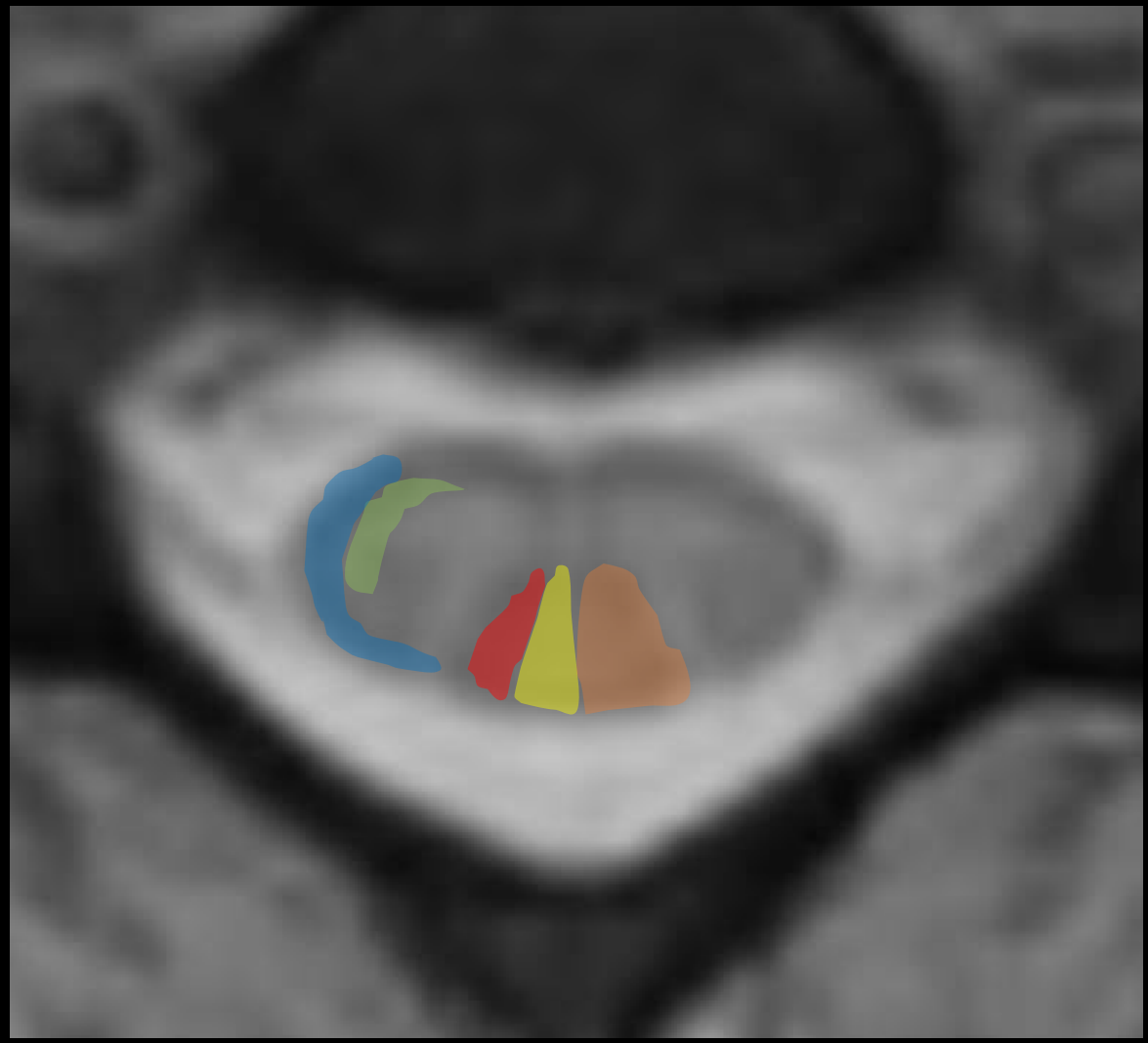




Central gray matter

Peripheral white matter

Major Descending Pathways



Major Ascending Pathways

Dorsal column system –
Fasciculus cuneatus and gracilis

Spinothalamic tract

Spinocerebellar tract

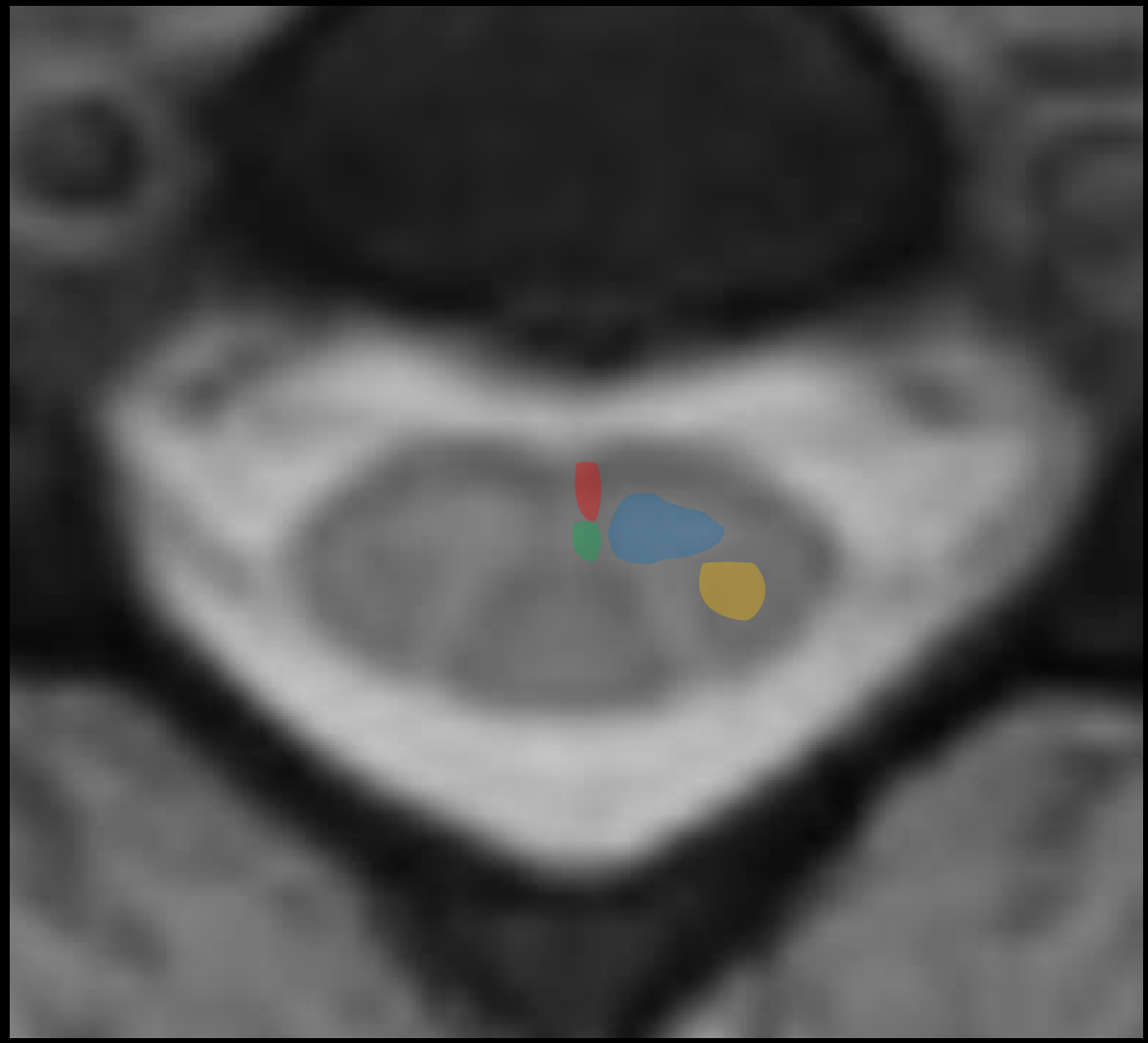
Barrow
BRAIN AND SPINE

Major Descending Pathways

Corticospinal tract, Anterior

Corticospinal tract, Lateral

Anterior horn cells



Medial longitudinal fascilulus

Major Ascending Pathways

Dorsal column system – Fasciculus cuneatus and gracilis

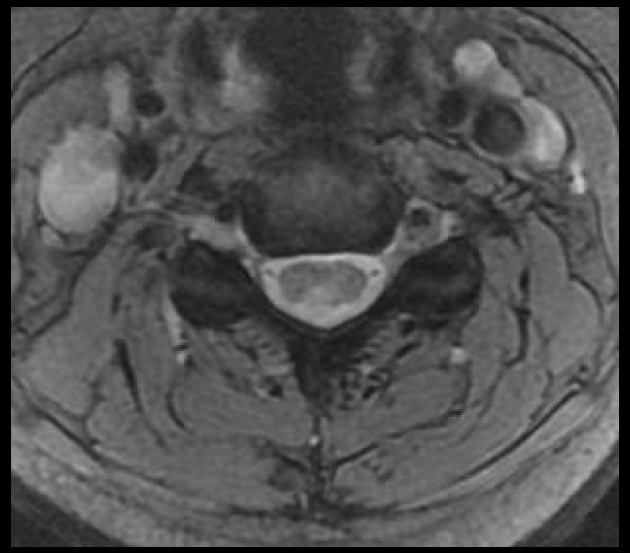
Spinothalamic tract

Spinocerebellar tract

Demyelinating

Extrinsic cord compression

Infectious



Toxic/metabolic

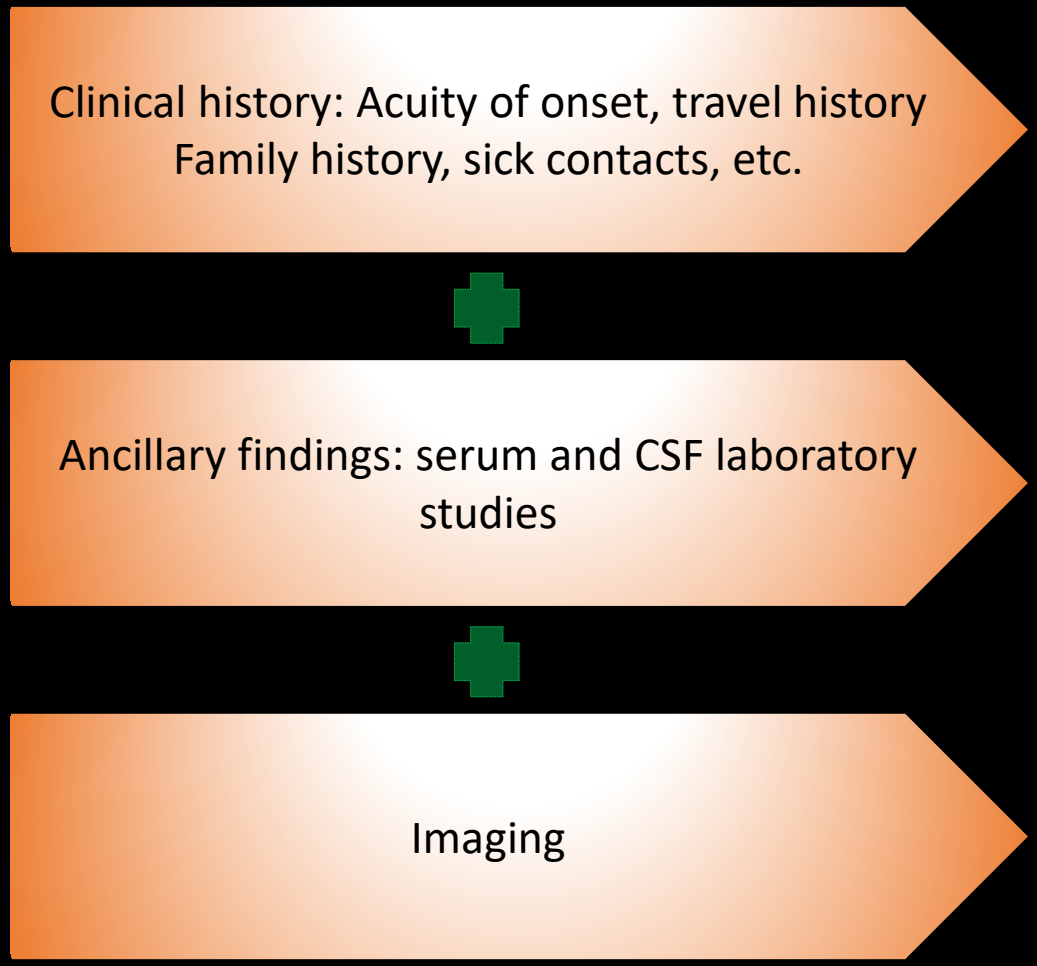
Tumor

Inflammatory

Vascular lesion

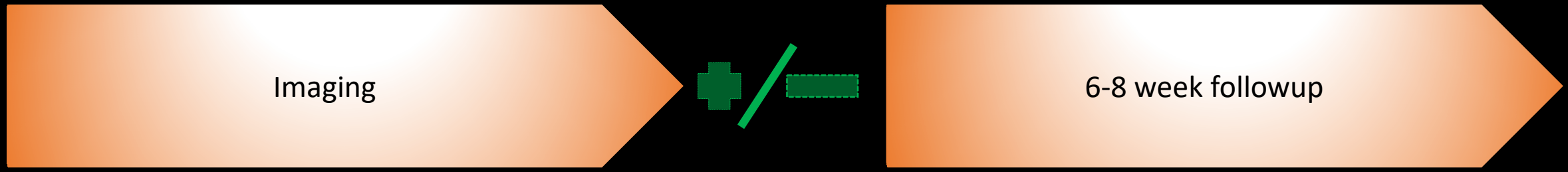
vasculitis

Diagnosing Disease of the Spinal Cord



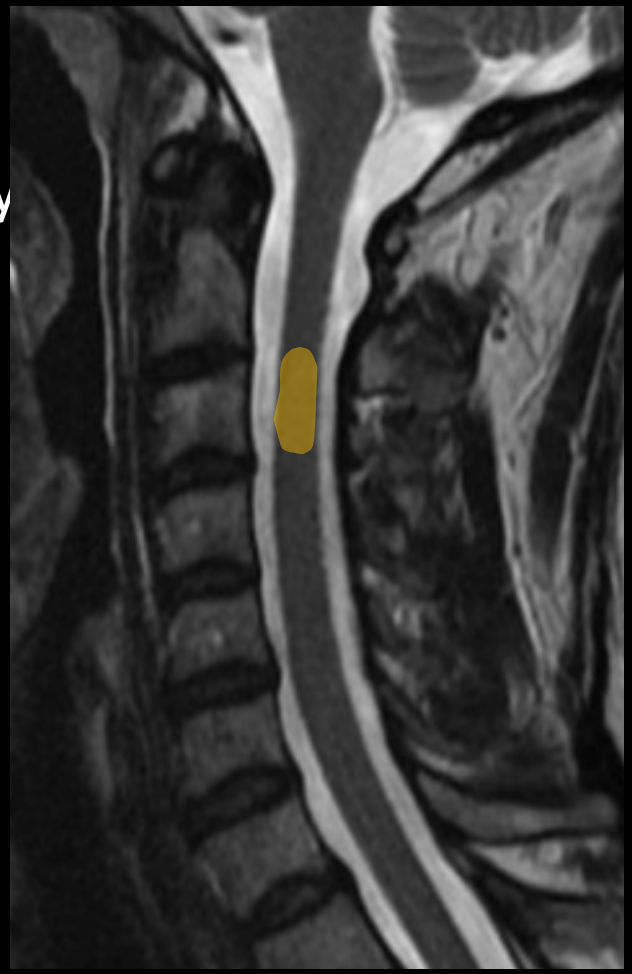
Diagnosing Disease of the Spinal Cord

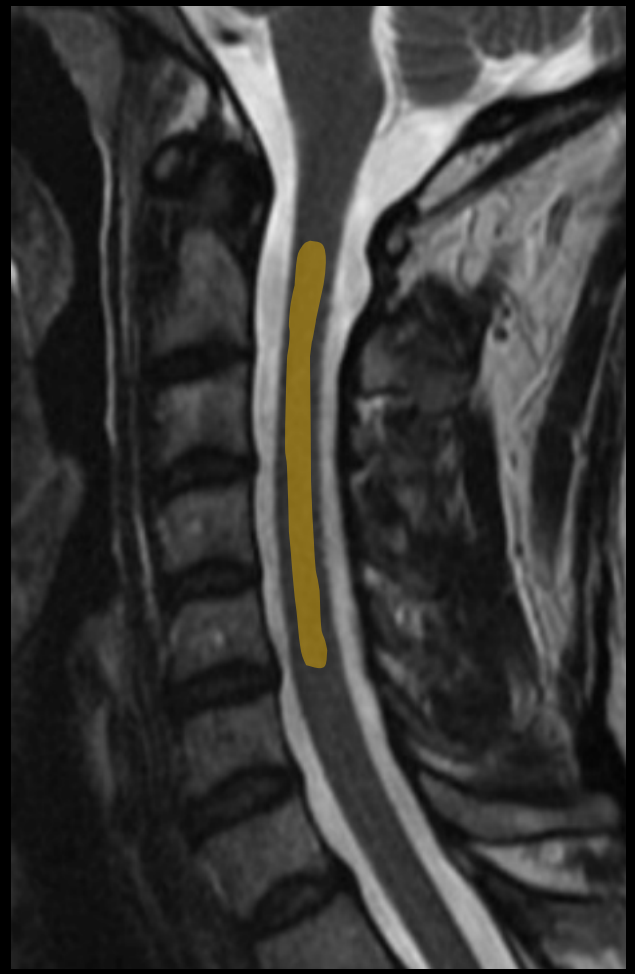
- At times after completion of workup the diagnosis remains elusive
  - 6-8 week follow up imaging
    - Resolution
      - Edema
        - Acute transverse myelitis
        - ADEM
      - Enhancement
        - Active MS plaque
    - Evolution
      - Atrophy
        - Spinal Cord infarct
    - Progression
      - Neoplasm



Imaging Patterns of Spinal Cord Disease

- Extent of involvement
  - Short segment signal abnormality
    - ≤ 2 vertebral bodies
  - Longitudinally extensive signal abnormality





Imaging Patterns of Spinal Cord Disease

- Extent of involvement
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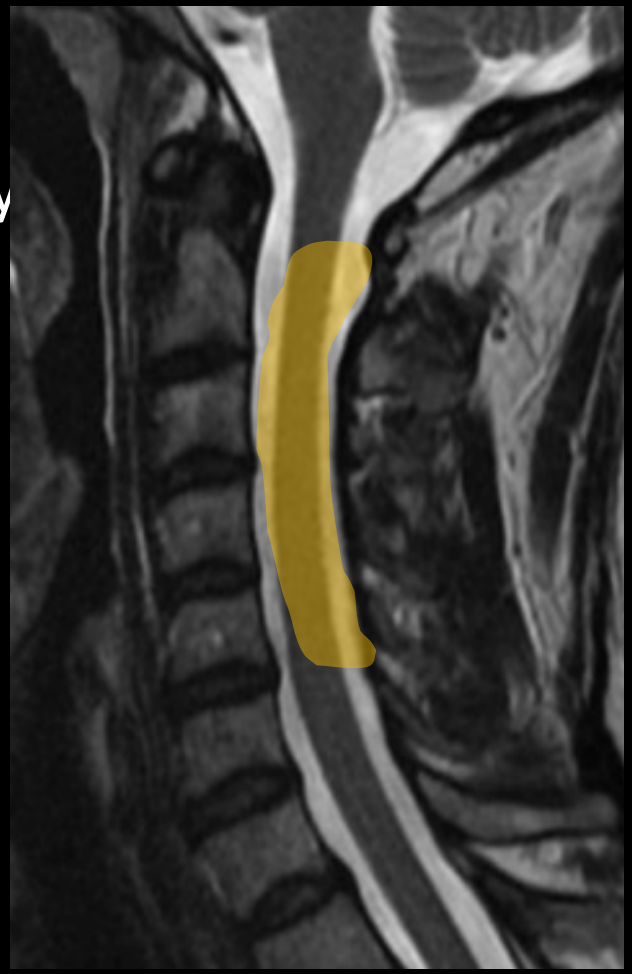
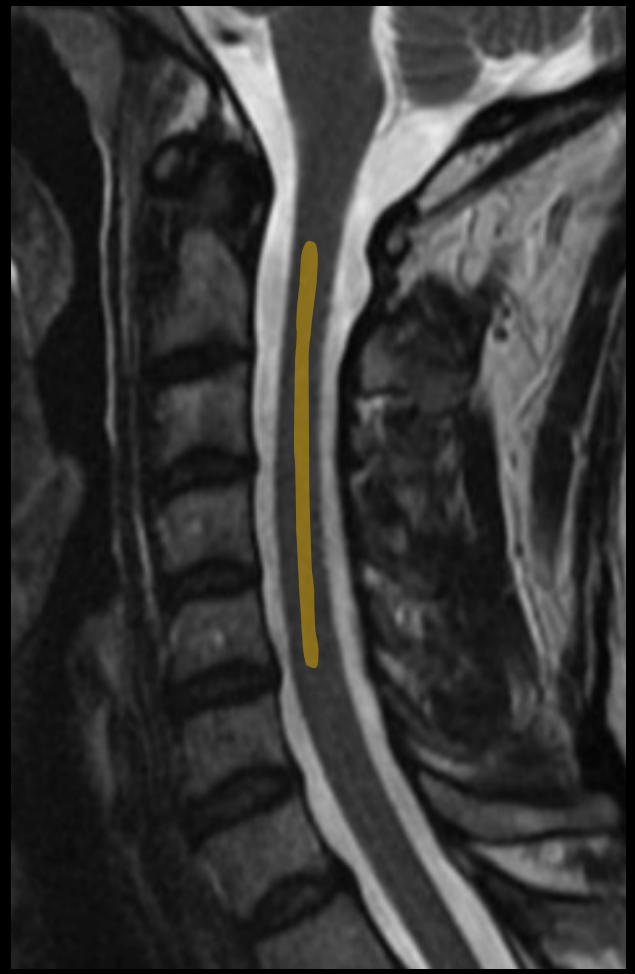
  - Longitudinally extensive signal abnormality
- Cord volume
  - Expansion
  - Atrophy





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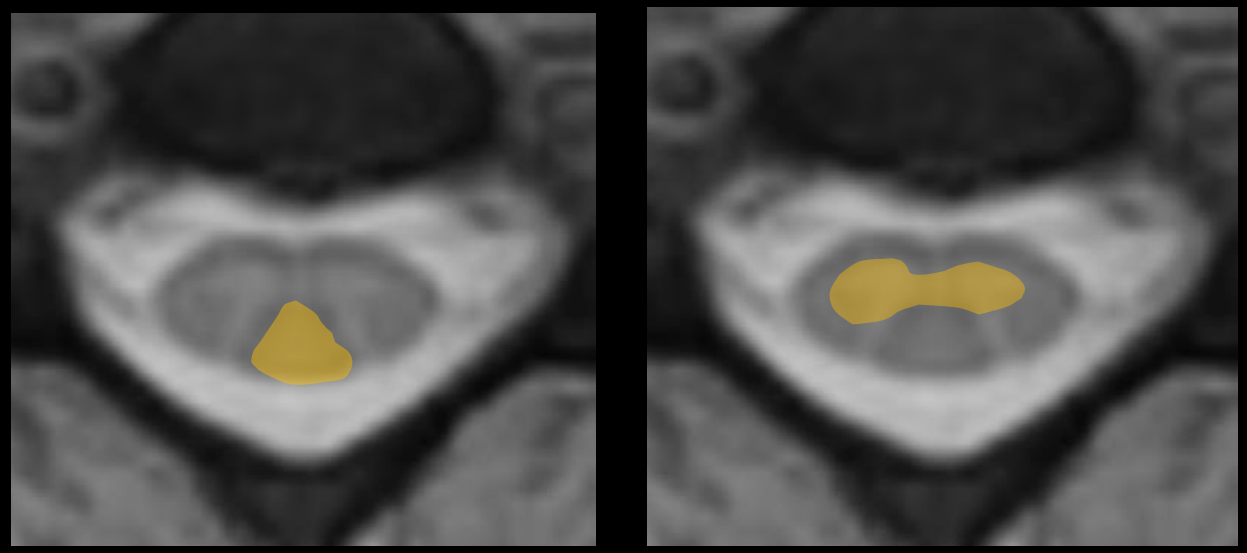
- Unique features
  - Anatomic distributions
    - Dorsal column
    - Anterior horn cell



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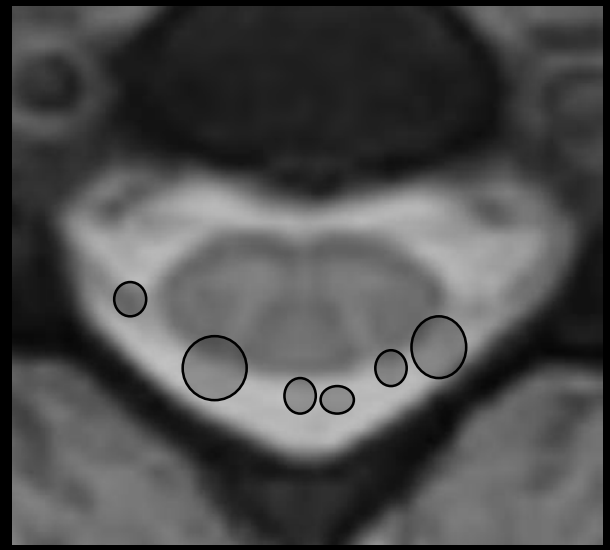

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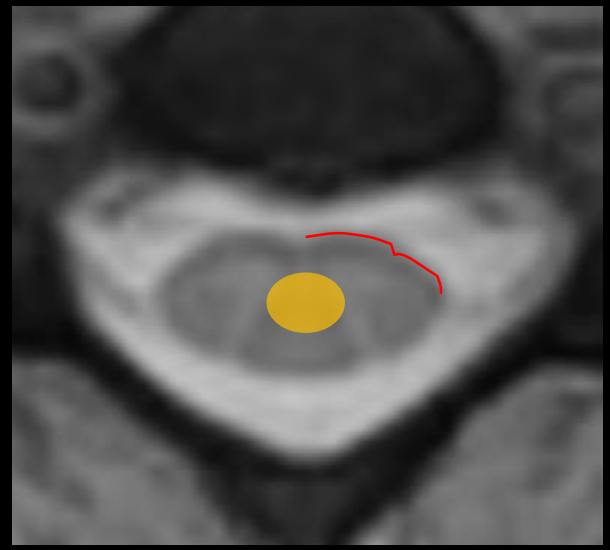
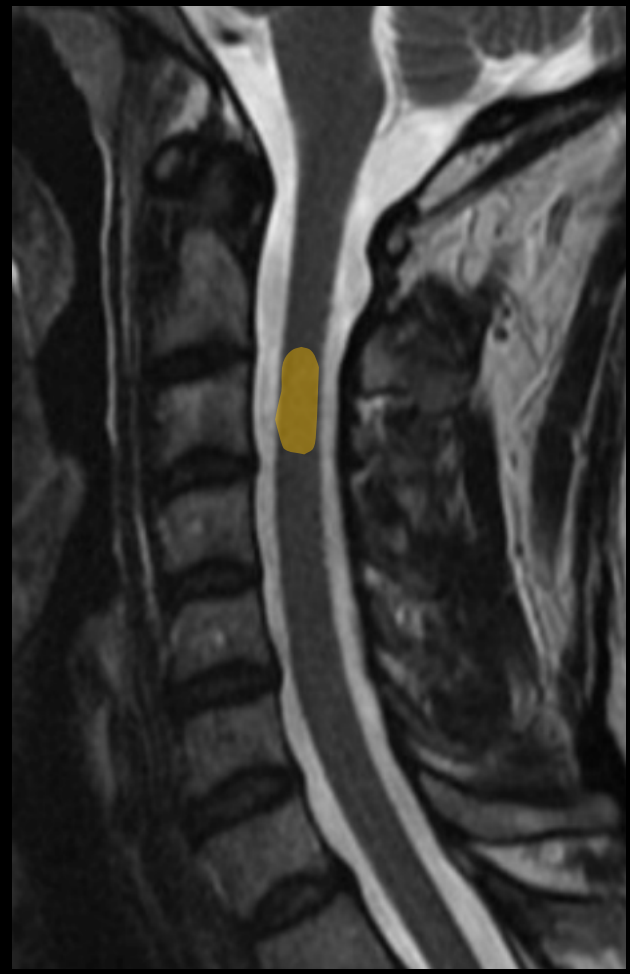
  - Flow voids
  - Enhancement pattern

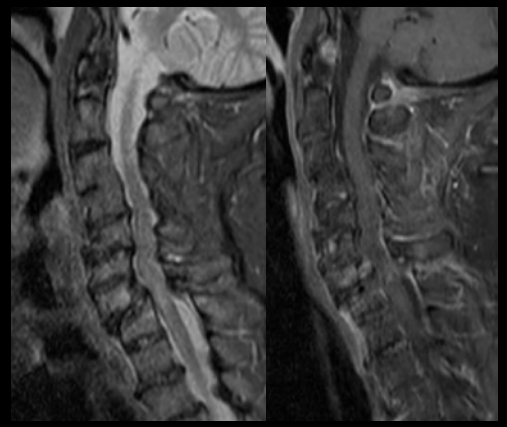


Short Segment Signal Abnormality

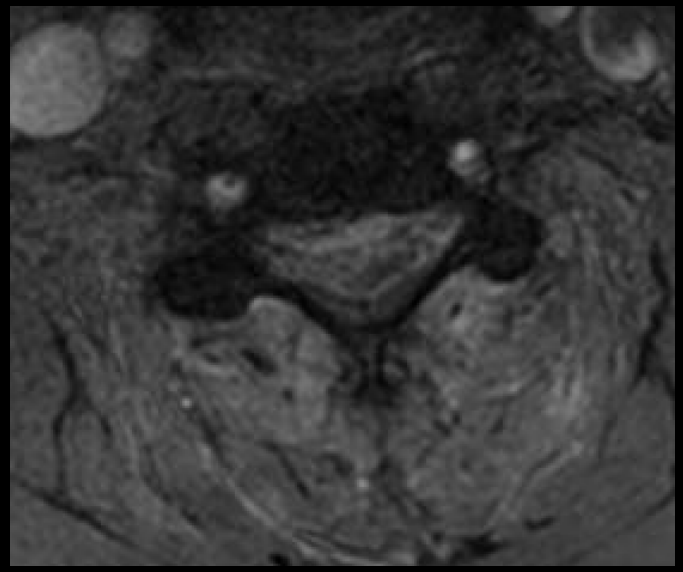
- Edema related to compressive lesions
  - Not typically a diagnostic dilemma
- Multiple sclerosis



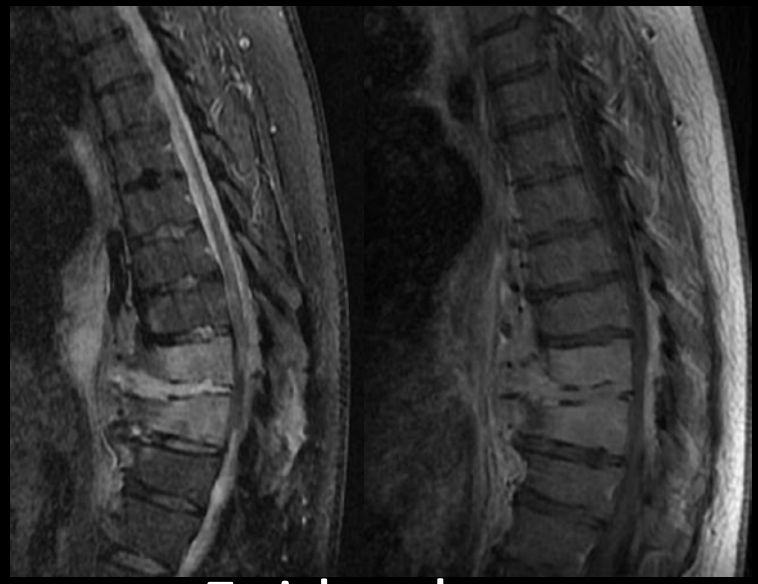
Short Segment Signal Abnormality – Cord Compression



- Spondylytic compressive edema
  - At or just inferior to compressive lesion (flexion)
  - May enhance



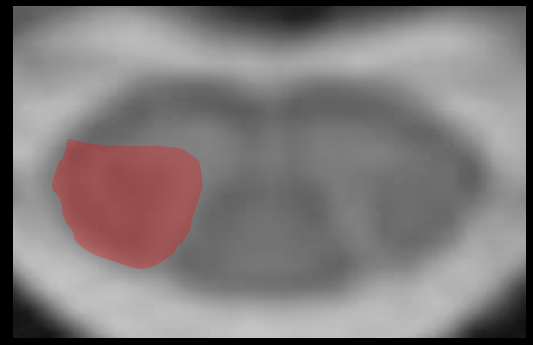
- Trauma
  - History
  - Ligamentous injury
  - Realignment following severe injury may appear relatively normal
  - Hemorrhage
    - Prognostic indicator



- Epidural compression
  - Tumor or infection

Short Segment Signal Abnormality

- Multiple Sclerosis
  - Autoimmune demyelinating disease
    - Common
      - Estimated 2.3 million people affected worldwide
      - 2nd most common cause of neurologic disability in US following trauma
    - Mean age at diagnosis – 35
    - F:M – 2:1
  - Diagnosis
    - Clinical and Imaging
    - McDonald Criteria
      - Dissemination in space and time
  - Clinical presentation related to spinal cord lesions
      - Limb sensory abnormality/paresthesias
      - Upper motor neuron signs
      - Urinary incontinence



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Diagnostic Criteria for Multiple Sclerosis: 2010 Revisions to the McDonald Criteria

TABLE 2: 2010 McDonald MRI Criteria for Demonstration of DIT

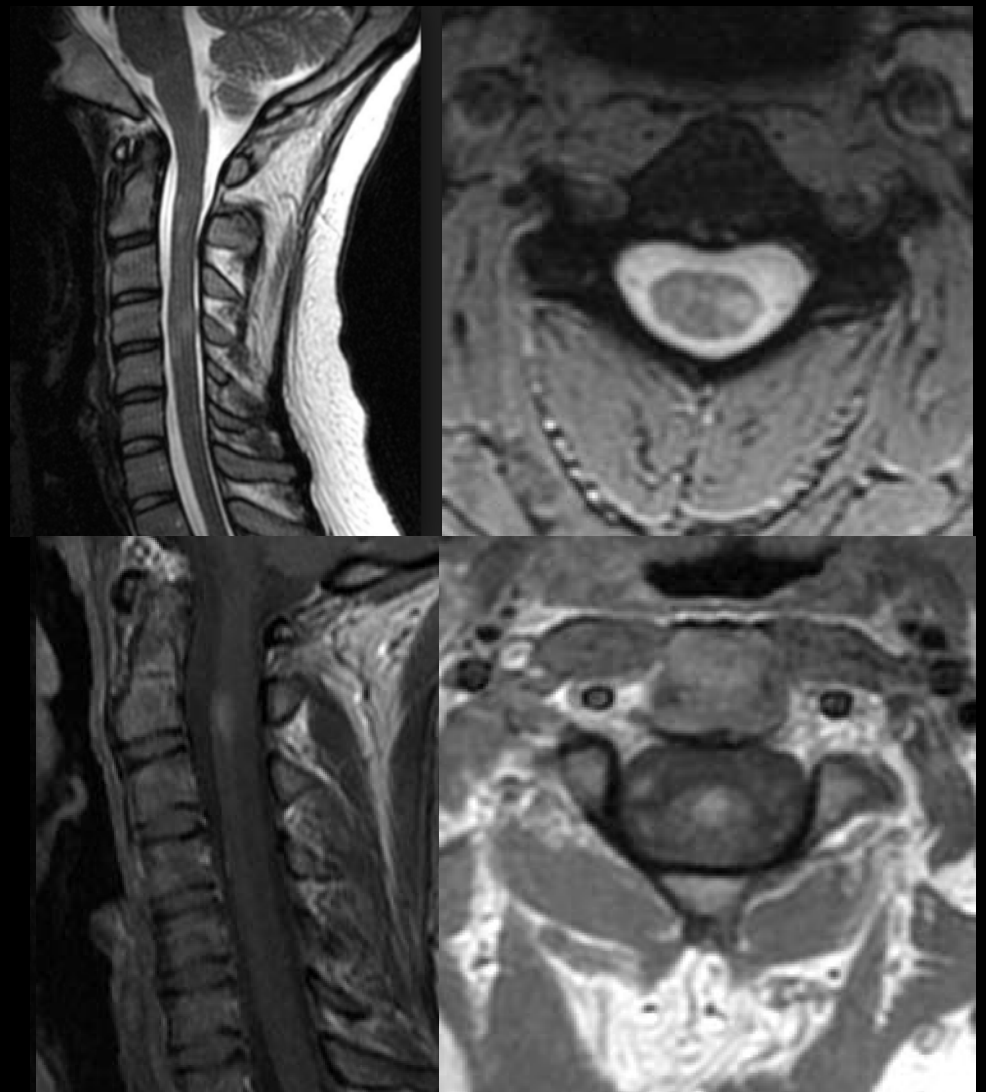
DIT Can Be Demonstrated by:
1. A new T2 and/or gadolinium-enhancing lesion(s) on follow-up MRI, with reference to a baseline scan, irrespective of the timing of the baseline MRI
2. Simultaneous presence of asymptomatic gadolinium-enhancing and nonenhancing lesions at any time

Based on Montalban et al 2010.[24]
MRI = magnetic resonance imaging; DIT = lesion dissemination in time.

ANN NEUROL 2011;69:292–302

Short Segment Signal Abnormality

- Multiple Sclerosis
  - Imaging
    - Cervical > Thoracic
    - Short segment
    - Prediliction for dorsolateral cord involvement
    - Enhancement
      - Active demyelination
    - Potential future biomarkers for disability
      - Lesions number/volume
      - Cord volume



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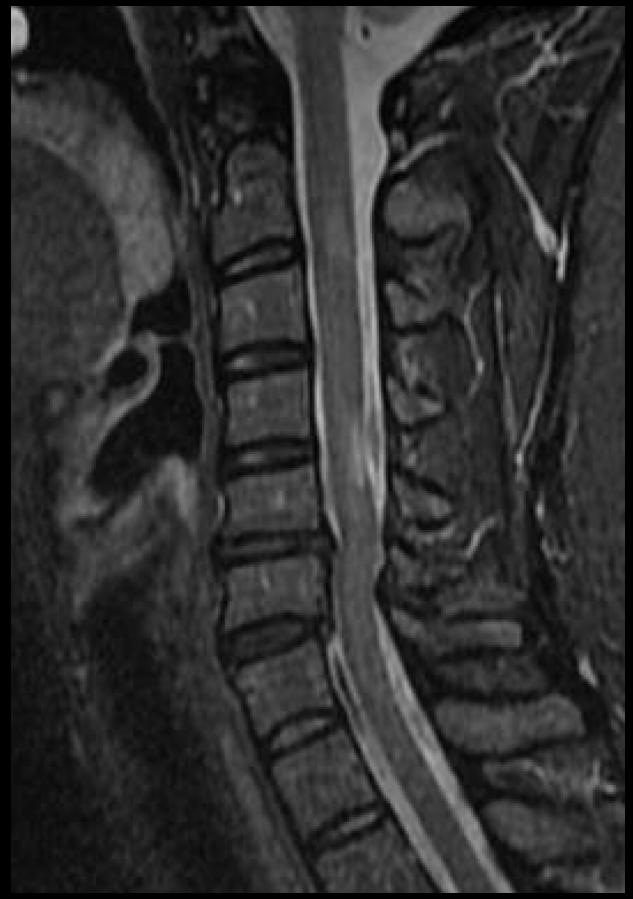
Spinal Cord Atrophy in Multiple Sclerosis: A Systematic Review and Meta-Analysis

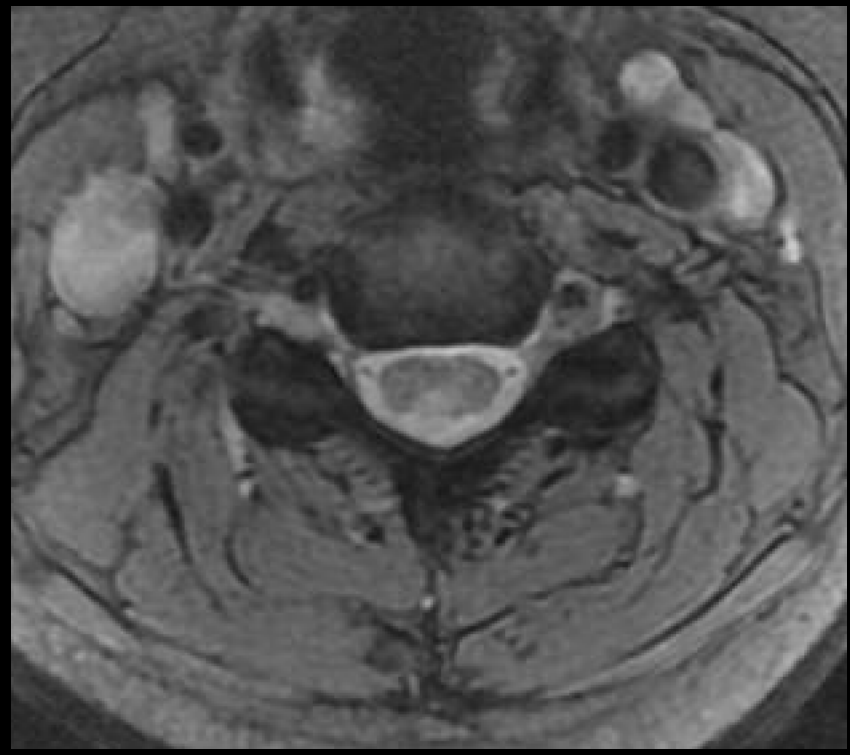
Courtney Casserly, Estelle E. Seyman, Paula Alcaide-Leon, Melanie Guenette, Carrie Lyons, Stephanie Sankar, Anton Svendrovski, Stefan Baral, Jiwon Oh

From the Division of Neurology, Department of Medicine, St Michael's Hospital, University of Toronto, Toronto, Ontario, Canada (CC, EES, MG, SS, AS, JO); Department of Neurology, London Health Sciences Centre, Western University, London, Ontario, Canada (CC); Division of Neuroradiology, Department of Medical Imaging, St. Michael's Hospital, University of Toronto, Toronto, Ontario, Canada (PA-L); Department of Epidemiology, Johns Hopkins School of Public Health, Baltimore, MD (CL, SB); and Department of Neurology, Johns Hopkins University, Baltimore, MD (JO).

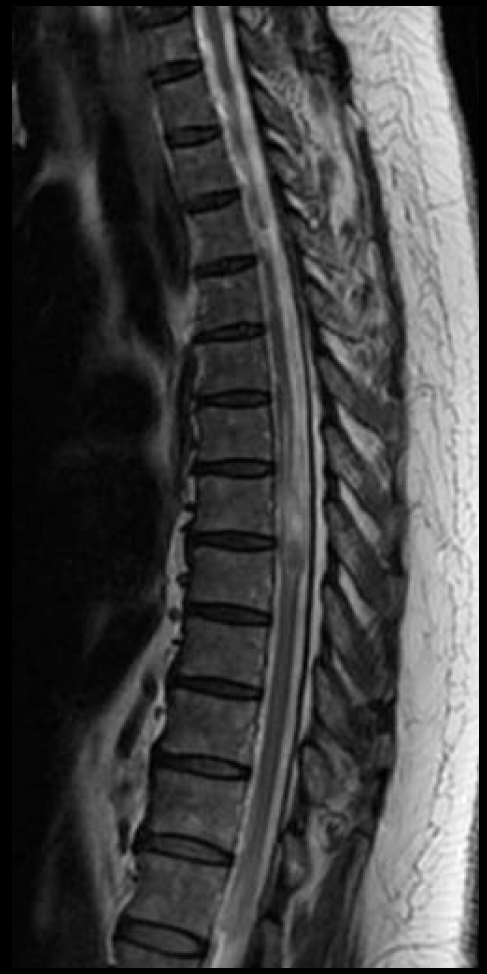
CONCLUSIONS: **The SC is atrophied in MS. The magnitude of SCA is greater in progressive versus relapsing forms and correlates with clinical disability. The pooled estimate of annual rate of SCA is greater than reported rates of brain atrophy in MS. These results demonstrate that SCA is highly relevant as an imaging outcome in MS clinical trials.**

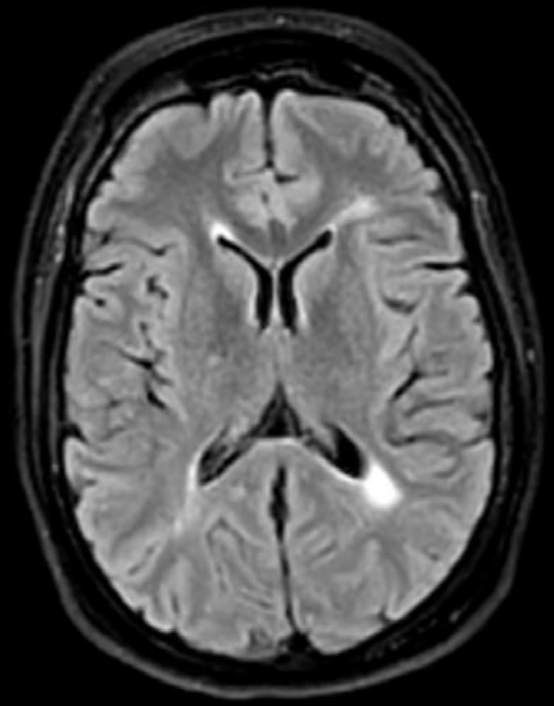
Short Segment Signal Abnormality

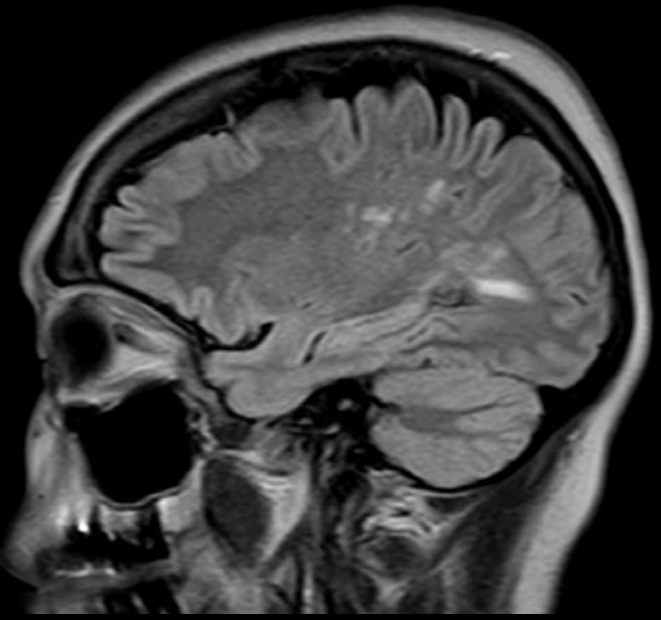




Short Segment Signal Abnormality

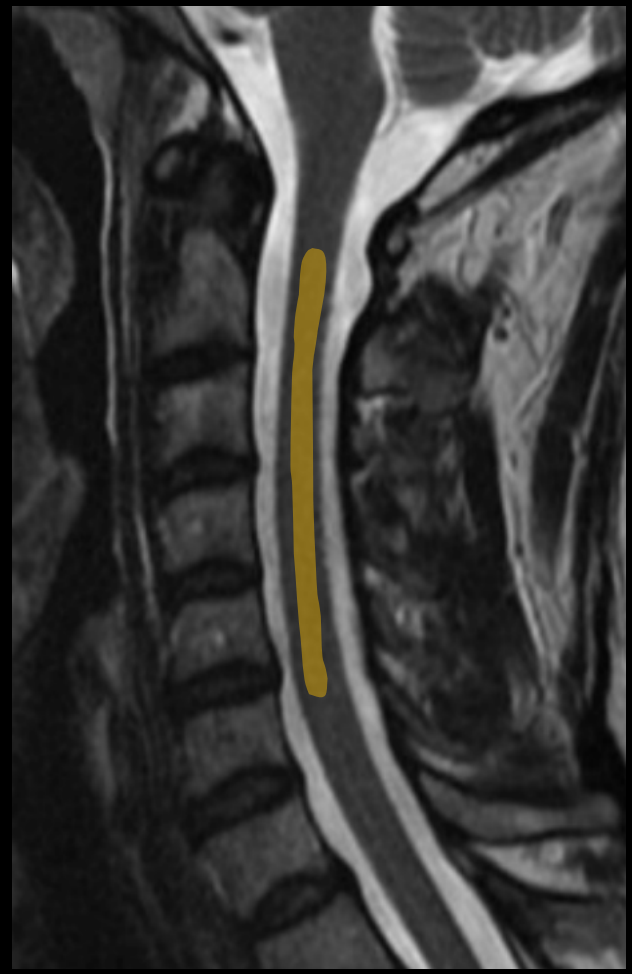






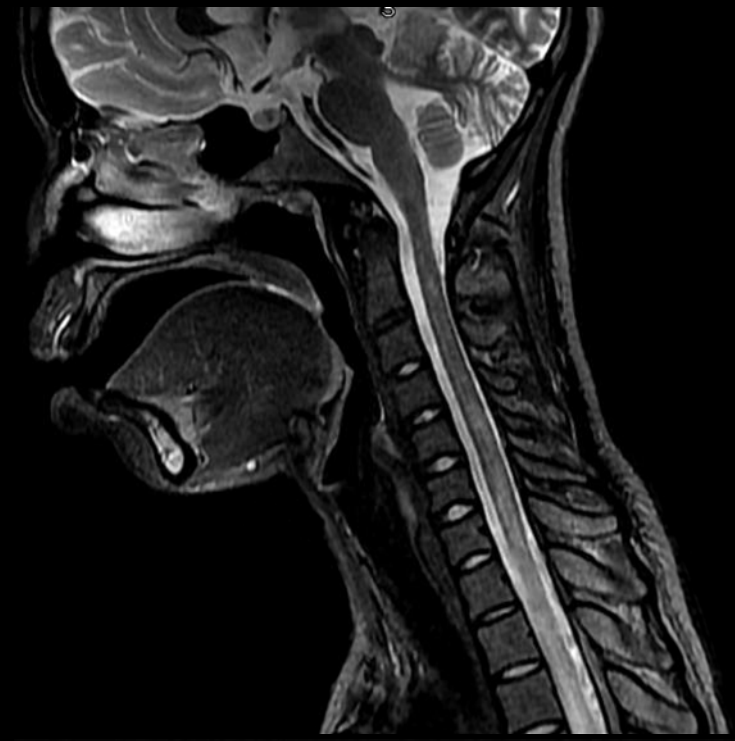
Longitudinally Extensive Signal Abnormality

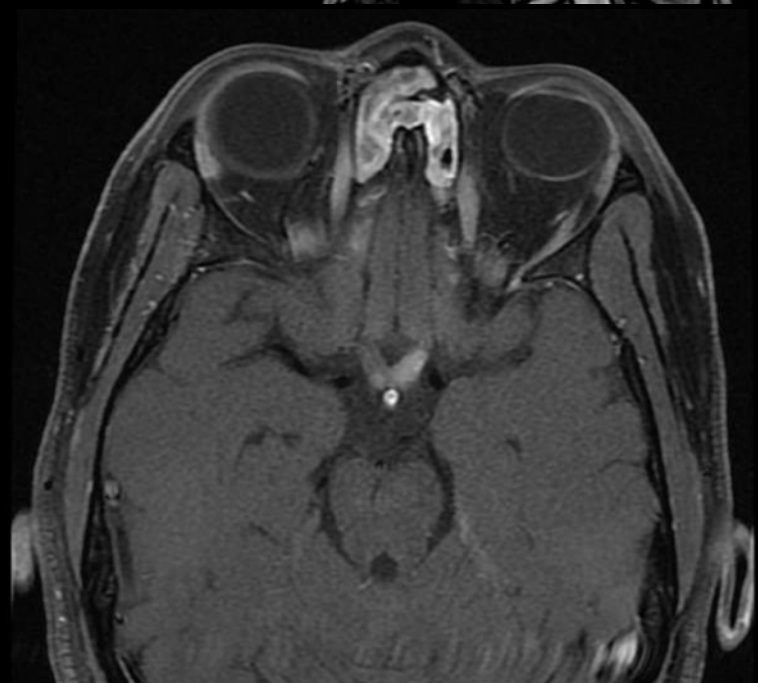
- Variable cord cross sectional involvement
  - NMO
  - Transverse myelitis
  - Acute disseminated encephalomyelitis (ADEM)
  - Vasculitis
  - Presyrinx edema



NMO – Devic's Disease

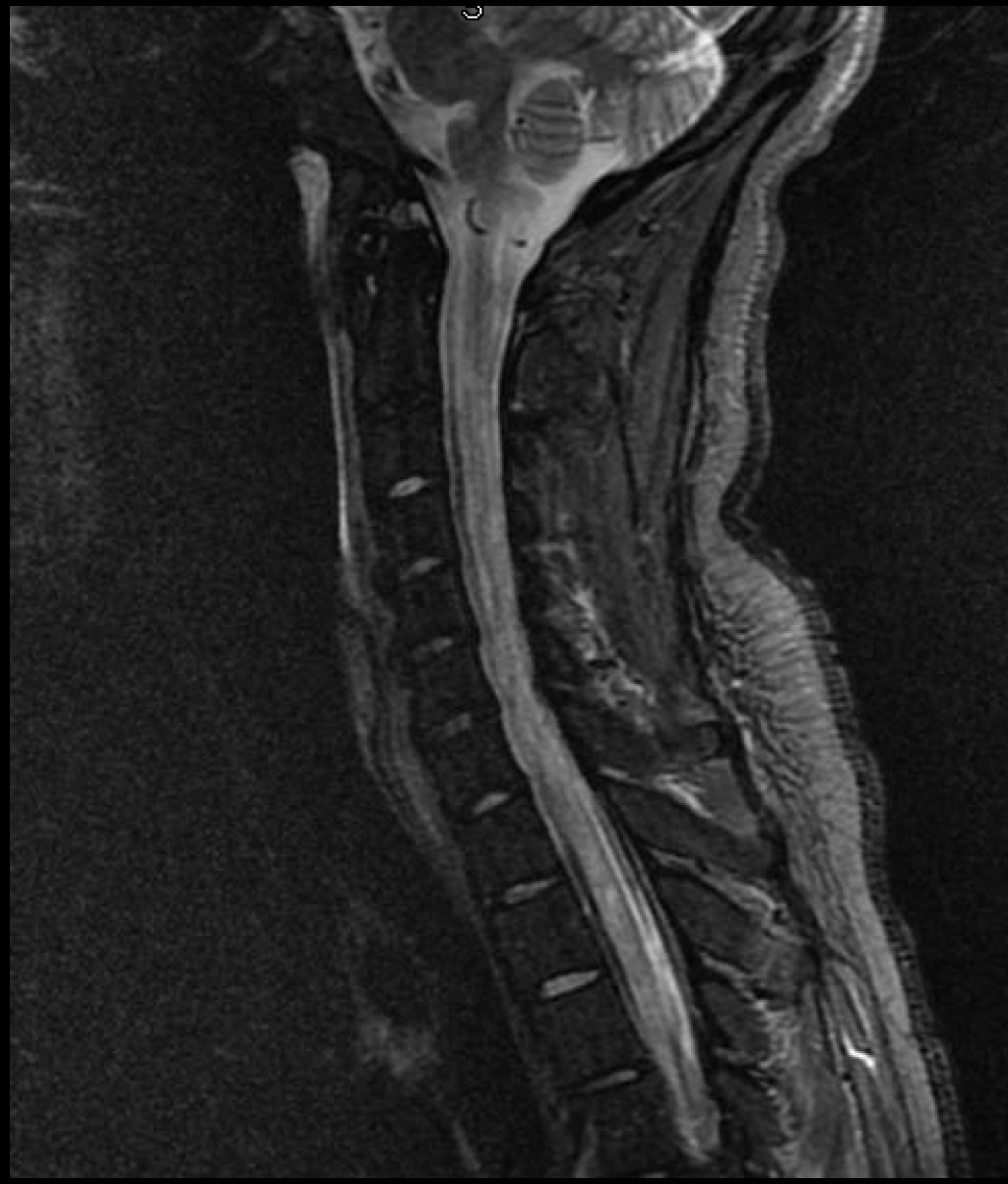
- Autoimmune demyelinating disease
- Preferentially involves the spinal cord and optic nerves
  - May involve brain
- Induced by specific antibody
  - Aquaporin-4 (Aq-4, Anti-NMO)
- Female:Male – 9:1
- Formerly thought to be a subtype of MS
  - Now known to be a distinct entity





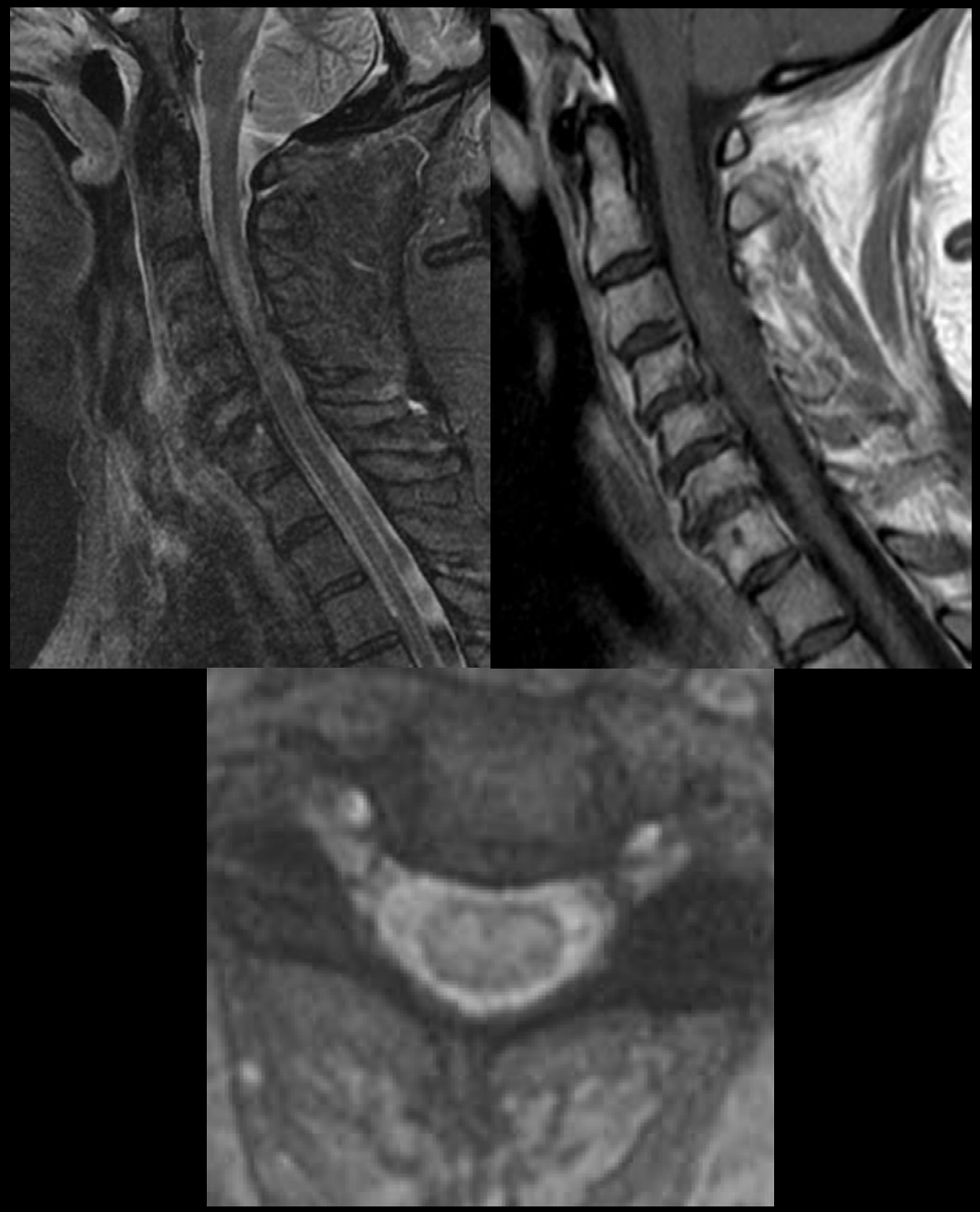
NMO – Devic's Disease

- Aggressive demyelination
  - Bright spotty lesions
    - Areas of very bright cystic T2 signal on a background of homogeneous edema
    - MS – 3%
    - NMO – 54%
- Brain involvement
  - Confluent periventricular
    - Disease directed against AQ-4 antibodies at ventricular margin
    - Often larger and more irregular than typical MS plaques
  - +/- Patchy enhancement



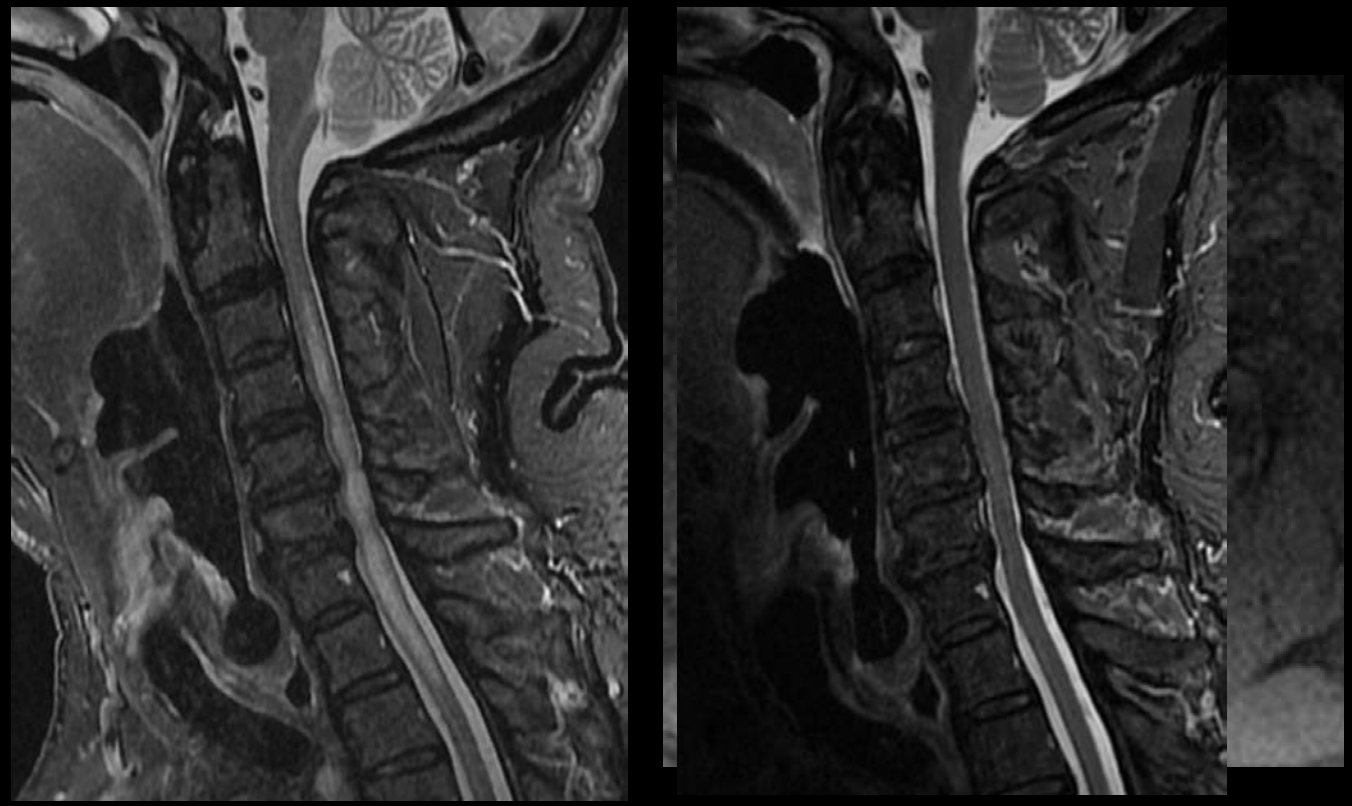
Transverse Myelitis

- Clinical Syndrome
  - Acutely ascending or static loss of sensory and motor function which affects both halves of the spinal cord.
  - Not a unique pathologic entity
  - Many etiologies
    - Infection, vaccination, Immune disorders, SLE, MS, vascular disease, paraneoplastic
    - Often remains undiagnosed
      - Idiopathic – Diagnosis of exclusion
  - 1/3 rule
    - 1/3 no long term sequela, 1/3 moderate, 1/3 severe



Transverse Myelitis

- Imaging
  - T2 hyperintense signal
    - Typically long segment involvement
    - Rarely short segment
      - May progress to MS
  - +/- cord expansion
  - +/- enhancement
  - Follow up
    - Resolution or myelomalacia



Infectious Myelitis

ELSEVIER

Seminars in ULTRASOUND CT and MRI

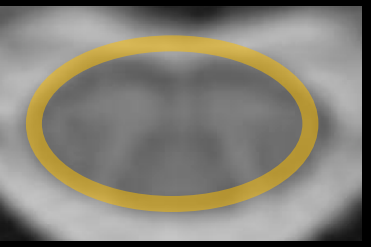
An Imaging-Based Approach to Spinal Cord Infection

CrossMark

Jason F. Talbott, MD, PhD,[†,‡] Jared Narvid, MD,[†] J. Levi Chazen, MD,[§] Cynthia T. Chin, MD,[*,†,‡,§] and Vinil Shah, MD[*,†]

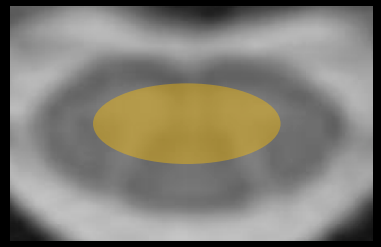
Semin Ultrasound CT MR. 2016 Oct;37(5):411-30.

Infectious Myelitis



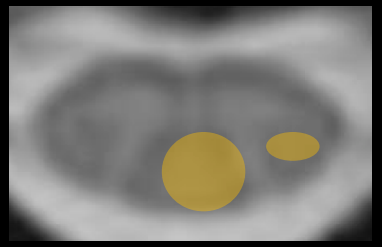
Extramedullary

Pyogenic/
Fungal
meningitis



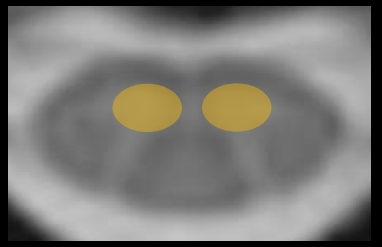
Centromedullary

EBV
HCV
Parainfectious
Lyme



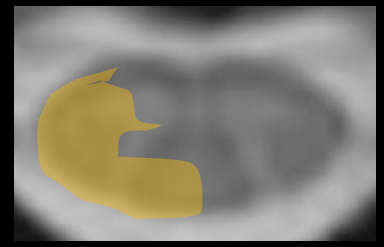
Eccentric

Tract Specific
Syphilis
VZV
HTLV-1
HIV



Frontal horns

Polio
West nile
Acute flaccid
Myelitis
Enterovirus D-68
Coxsackie virus



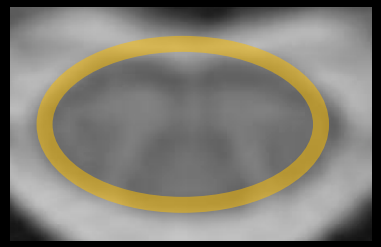
Irregular

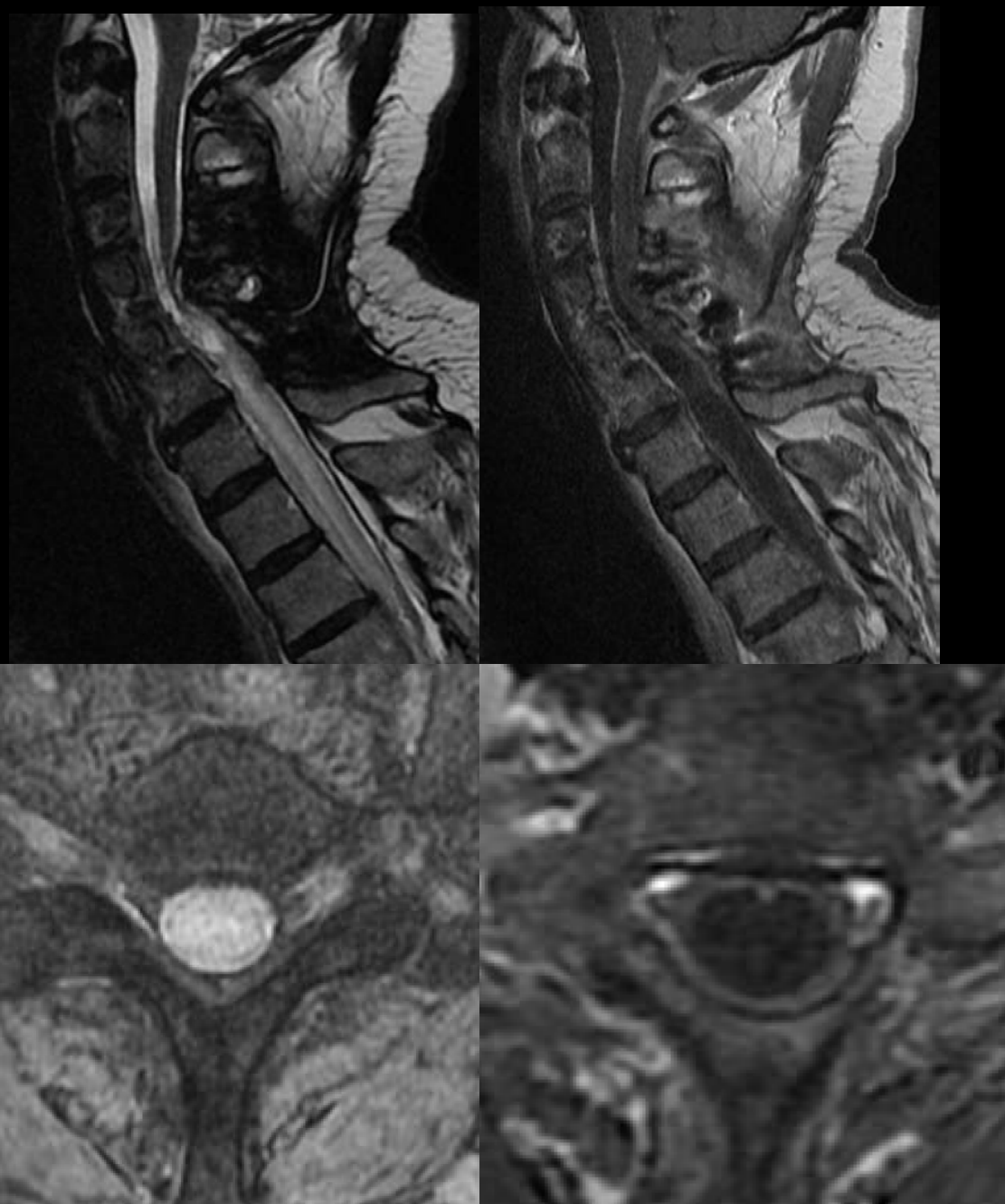
Pyogenic
Absces
Schistosomiasiss

Semin Ultrasound CT MR. 2016 Oct;37(5):411-30.

Infectious Myelitis

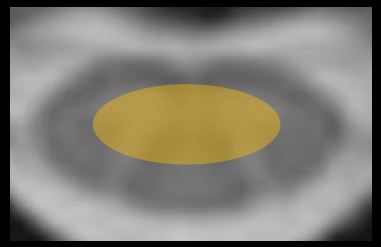
- Extramedullary pattern
  - Disseminated coccimeningitis

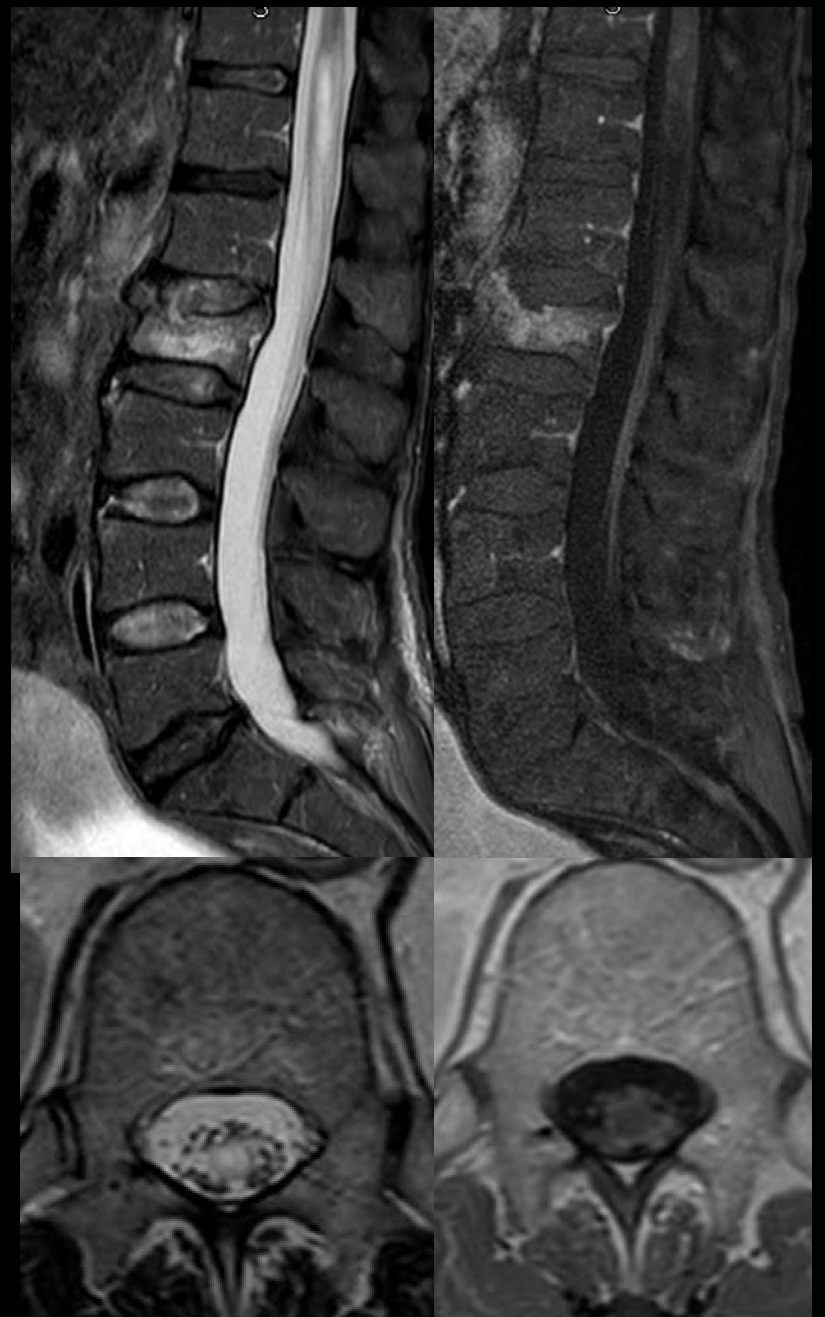




Infectious Myelitis

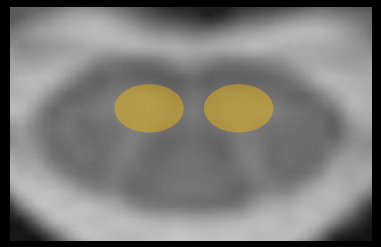
- Centromedullary pattern
  - Suspected Lyme myelitis
    - Positive titers
    - Not confirmed

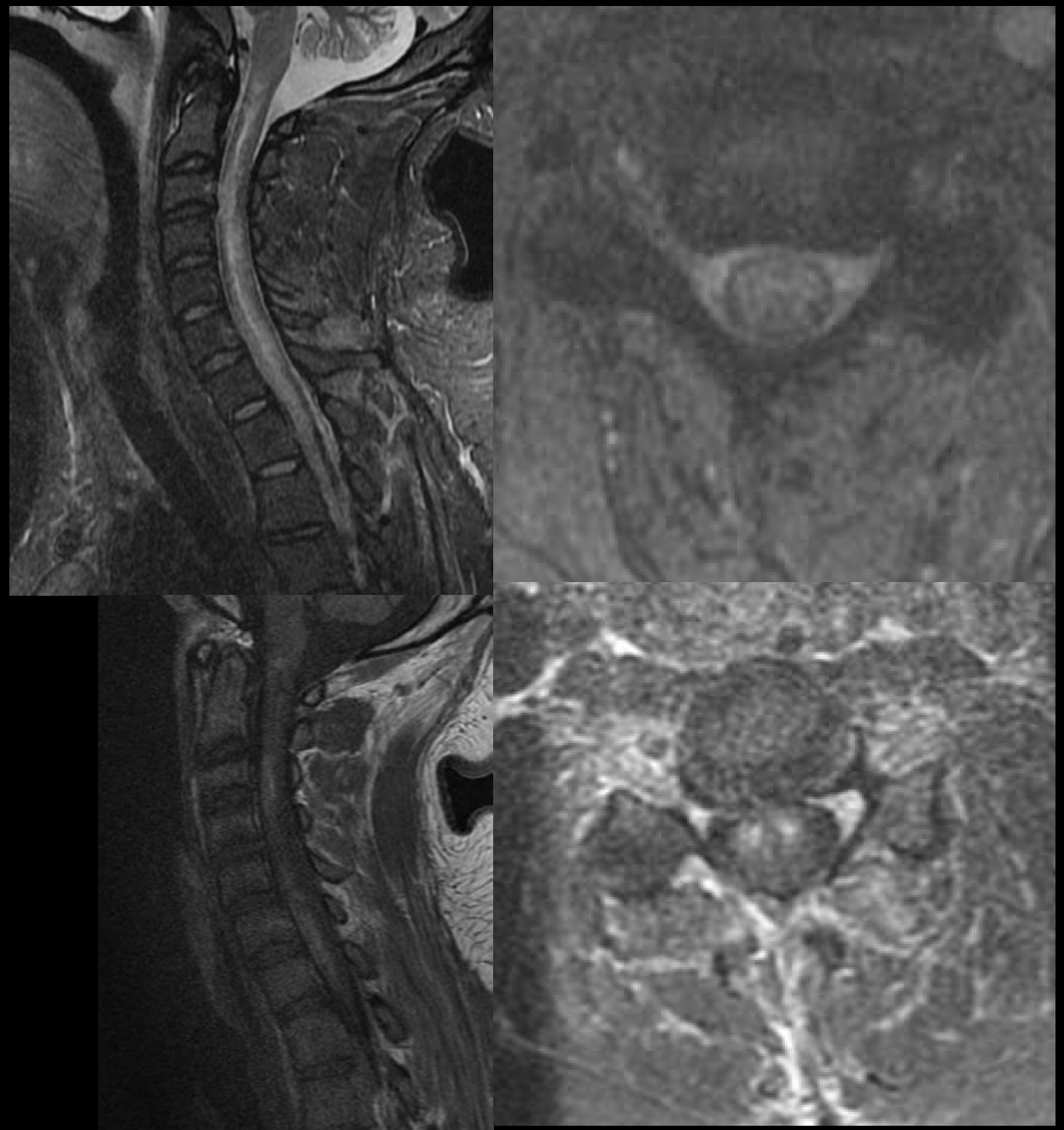




Infectious Myelitis

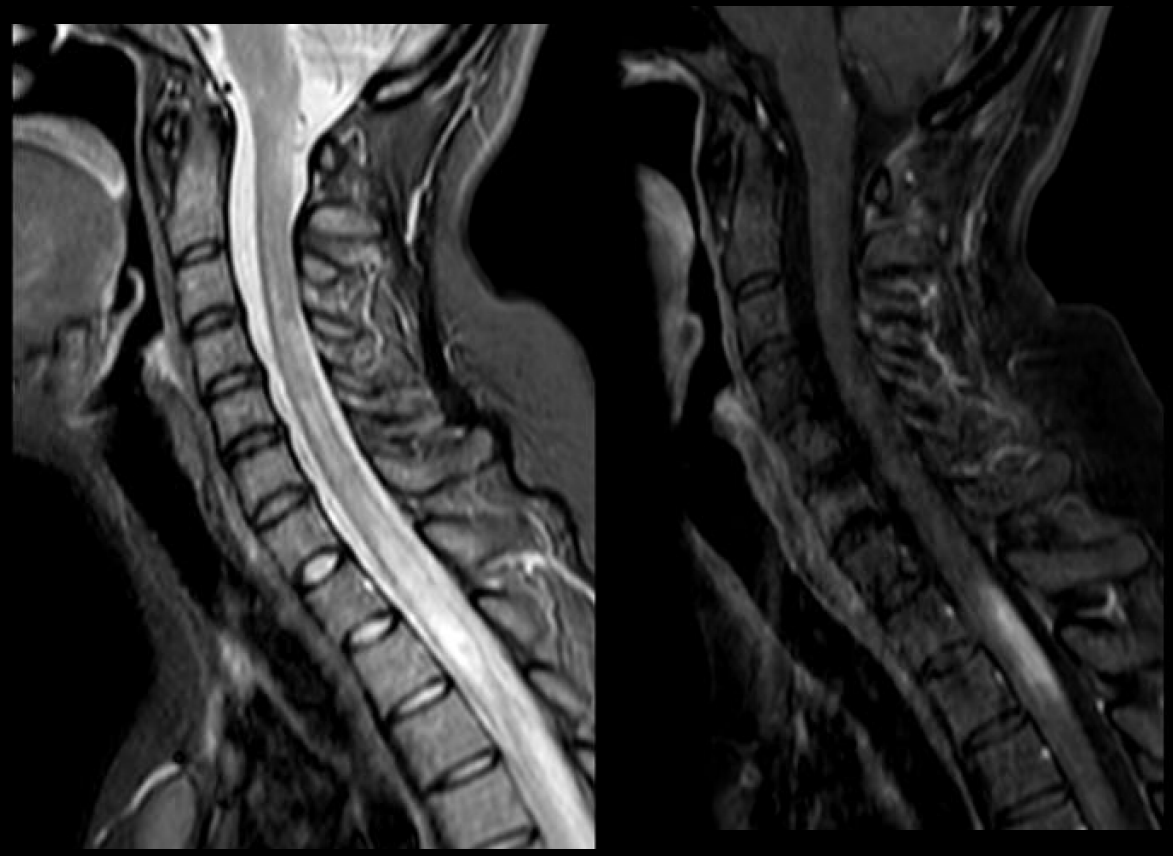
- Frontal horn pattern
  - Confirmed Coxsackie virus





Acute Disseminated Encephalomyelitis (ADEM)

- Post-viral or post-infectious inflammatory demyelination
- May involve brain and/or spinal cord
- Monophasic (>90%)
- Anti-MOG IgG (myelin oligodendrocyte glycoprotien)
  - + in 50% of ADEM cases
- May occur at any age
  - More common in younger patients



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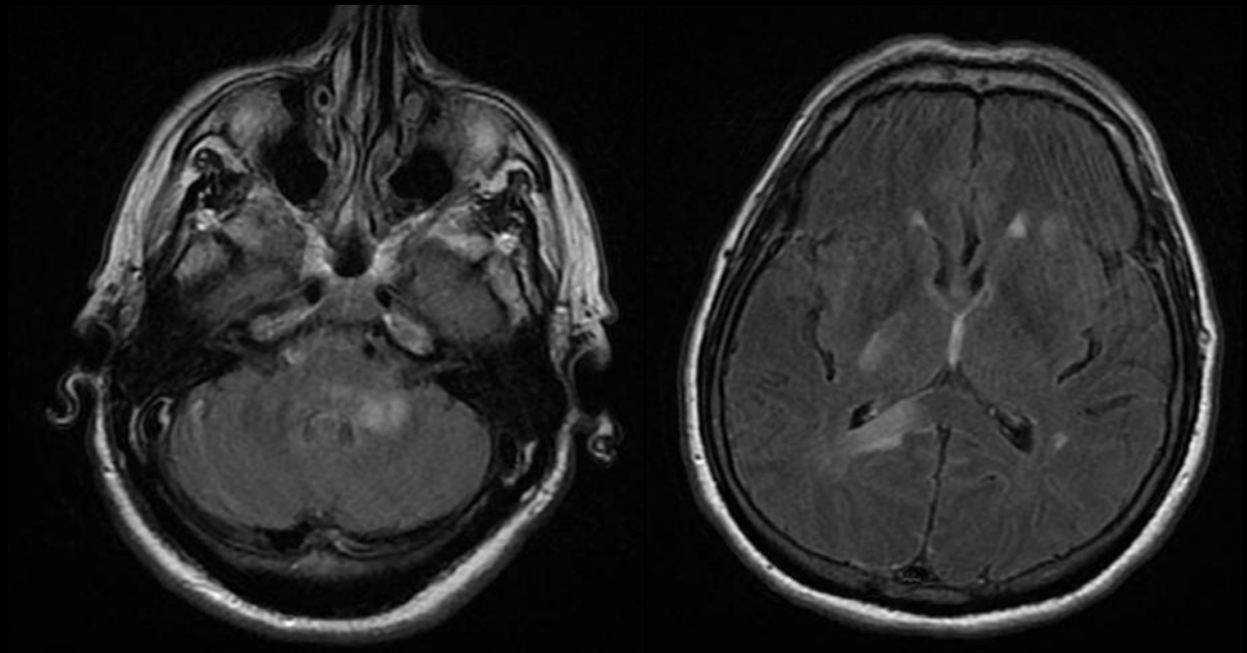
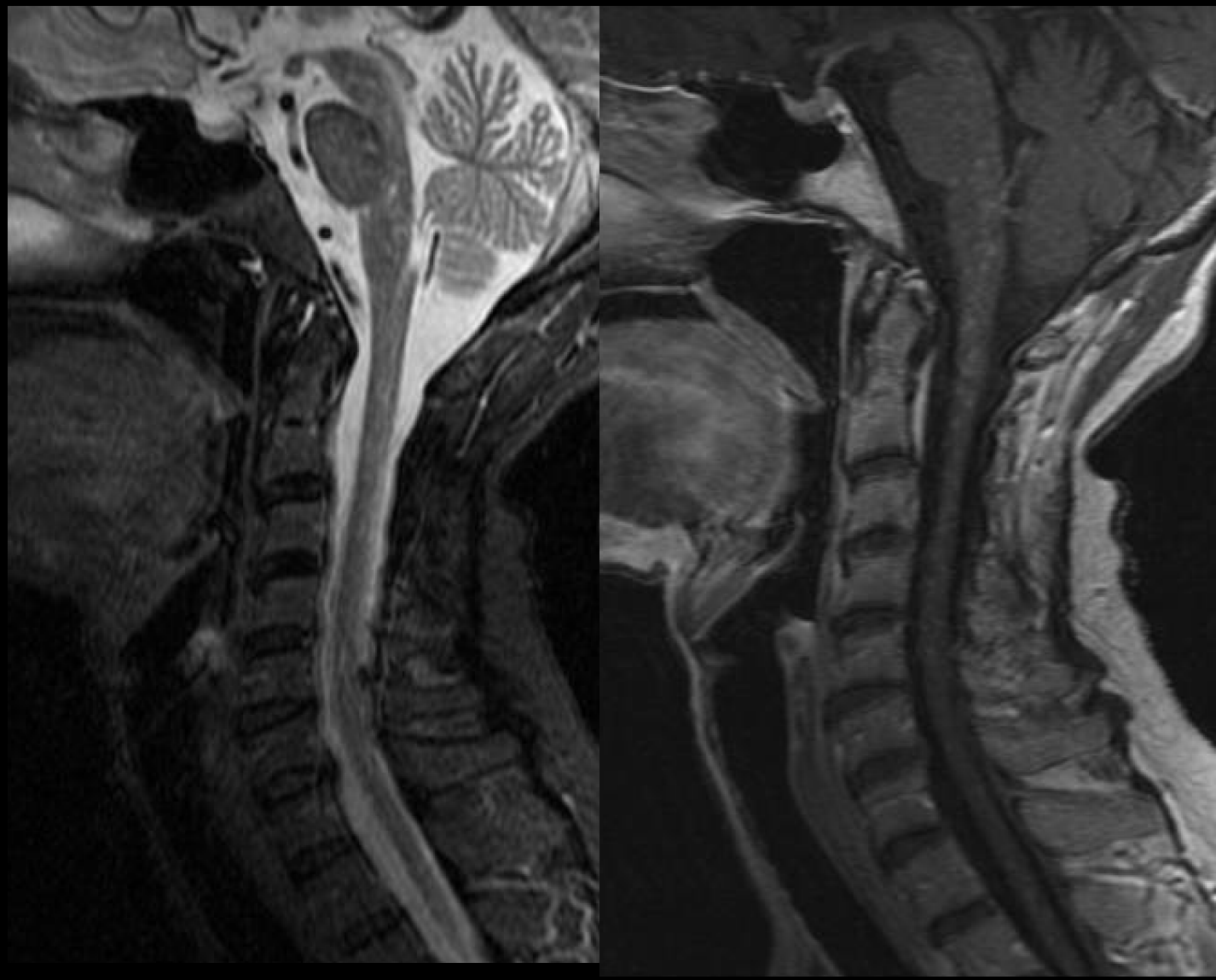


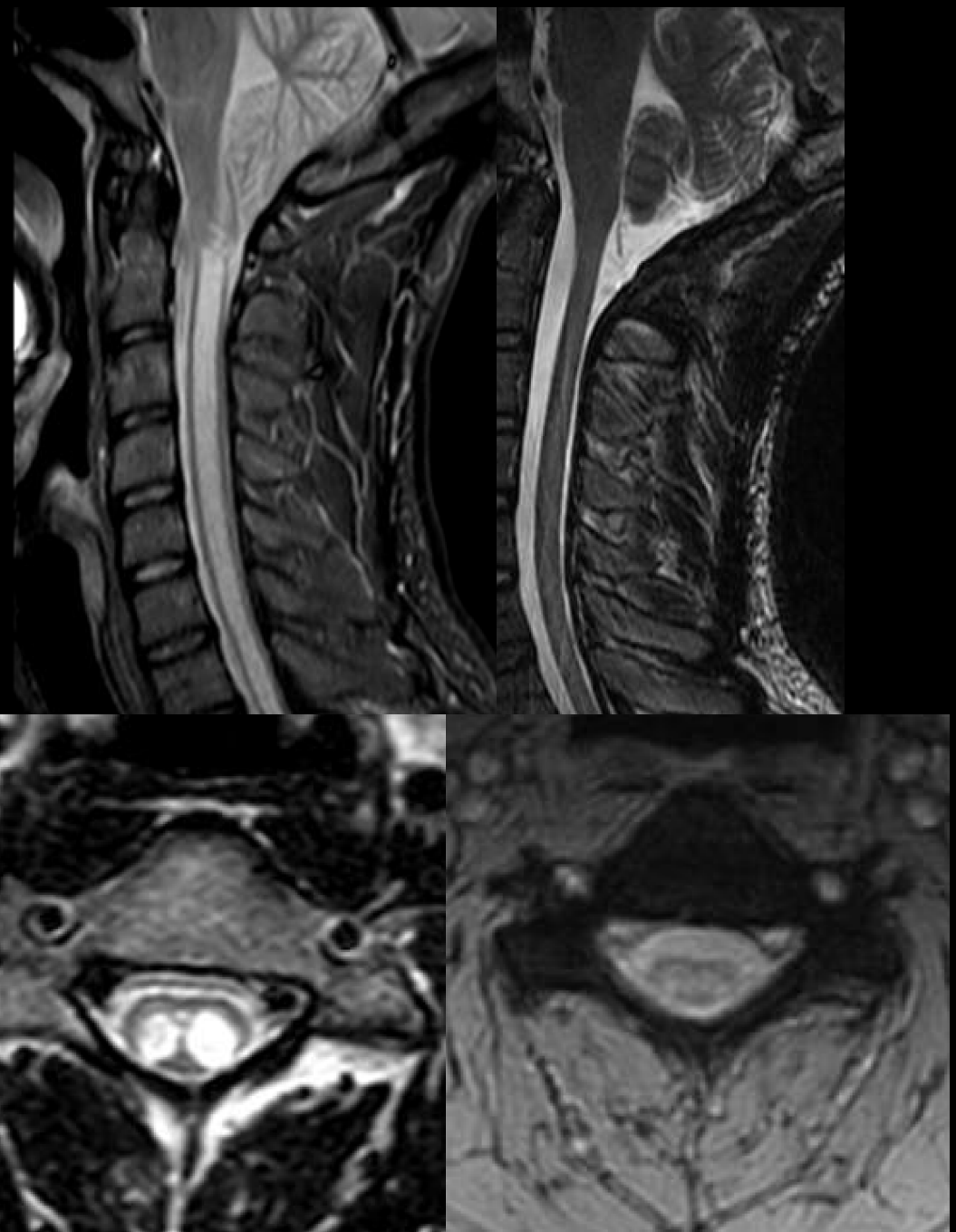
Vasculitis

- CNS vasculitis
  - Primary
    - Isolated to brain and SC
  - Rare – 2.4 cases per million
    - Spinal cord involvement in 5-14%
  - Secondary vasculitis
    - Systemic disease
      - SLE, Sjogren's, Behcet's, etc.
- Imaging
  - Patchy diffuse T2 signal abnormality
  - Parenchymal and/or leptomeningeal enhancement
  - Angiographic imaging may be normal



Presyrinx Edema

- Syrinx
  - Altered CSF flow dynamics
    - Chiari malformation, trauma, arachnoiditis, neoplasm
- Presyrnix edema
  - Initially described by Fischbein in 1999
  - Only one case showed progression to syrinx
  - Relationship not sufficiently validated
    - Rarity
    - Surgical decompression



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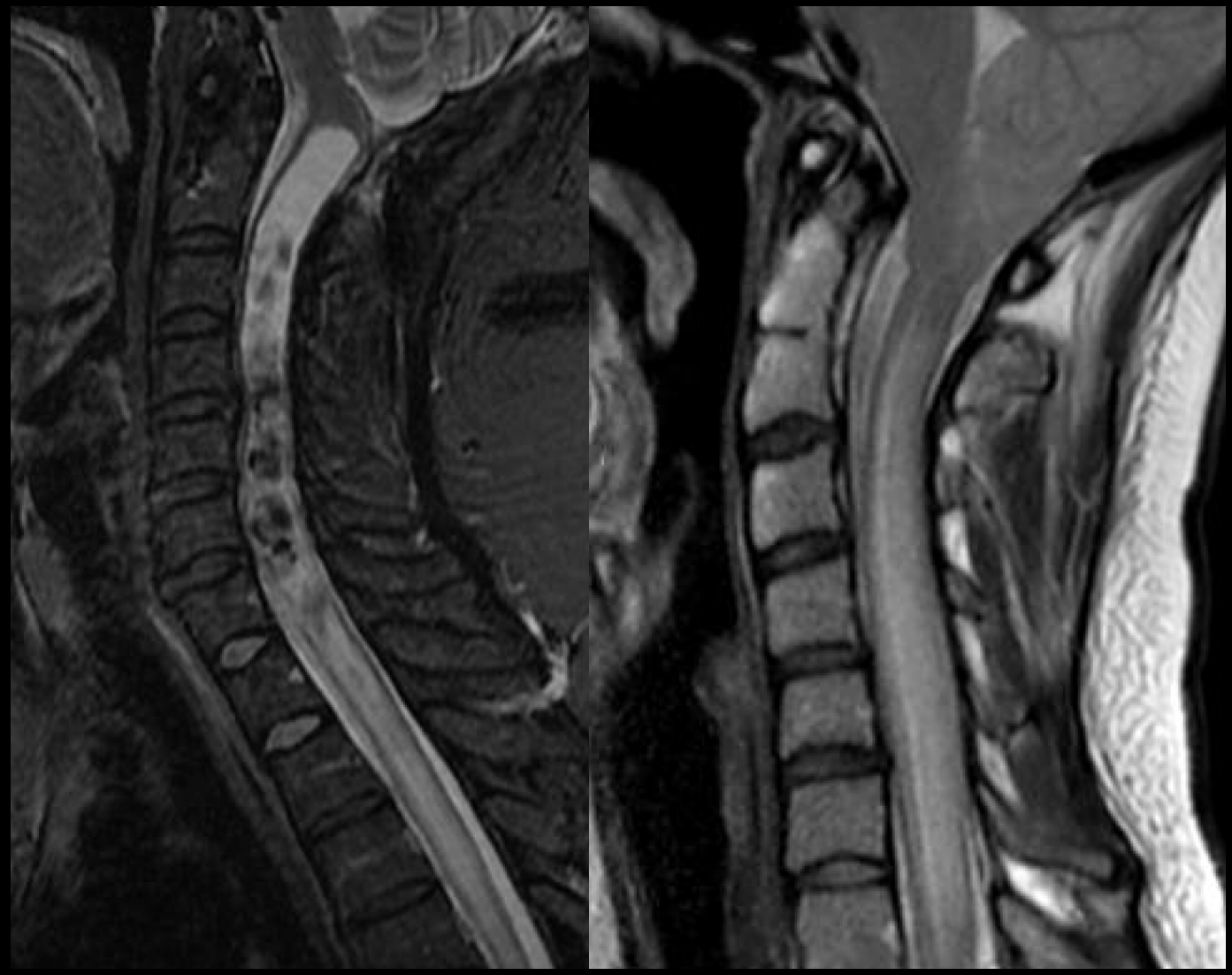
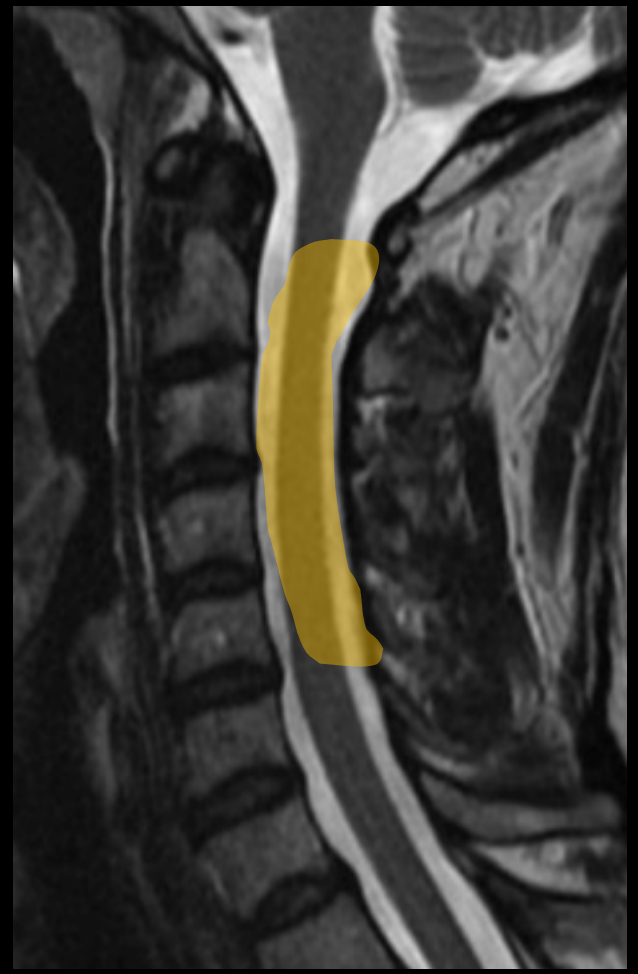


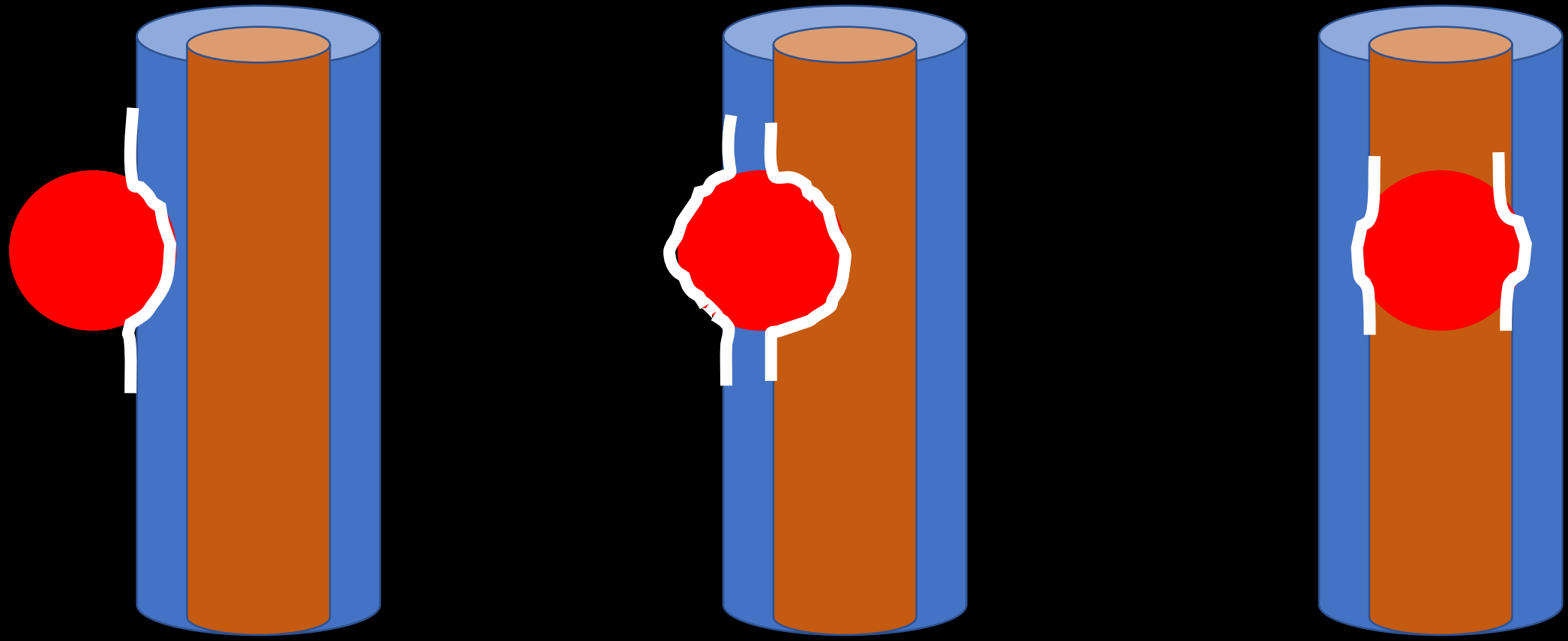
Spinal Cord Expansion

- Acute edematous demyelinating/inflammatory process
  - Transverse myelitis
  - ADEM
  - NMO
- Tumor



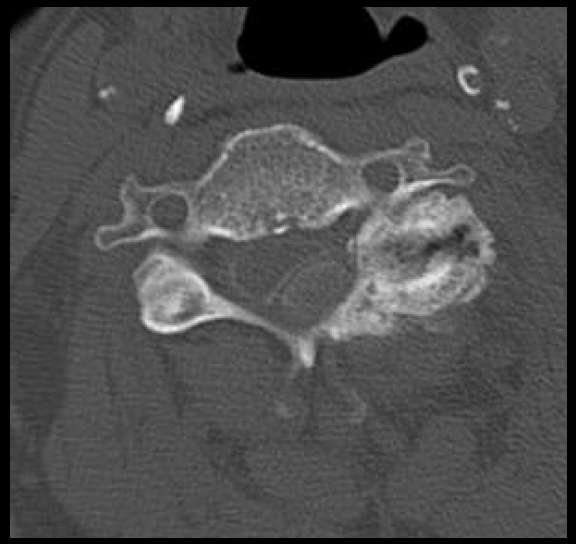
Spinal Tumors

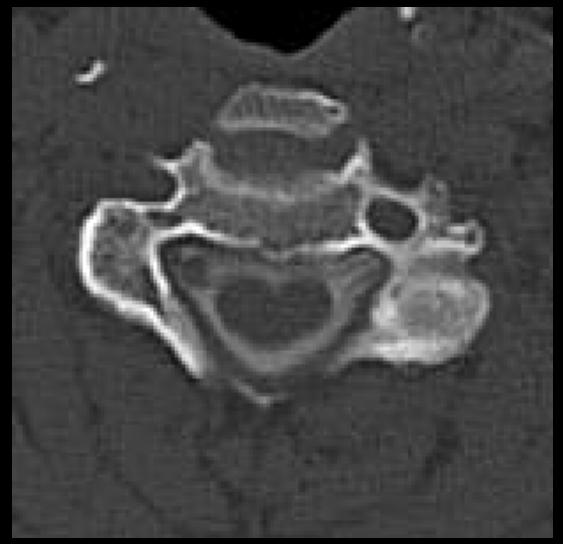
- Extradural
- Intradural Extramedullary
- Intradural Intramedullary

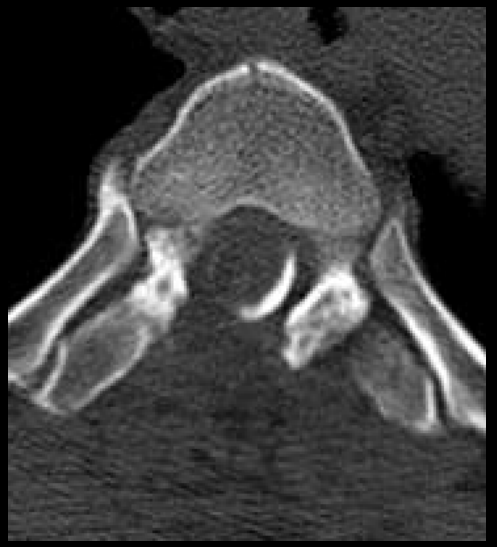


Spinal Tumors

- Extradural
- Intradural Extramedullary
- Intramedullary

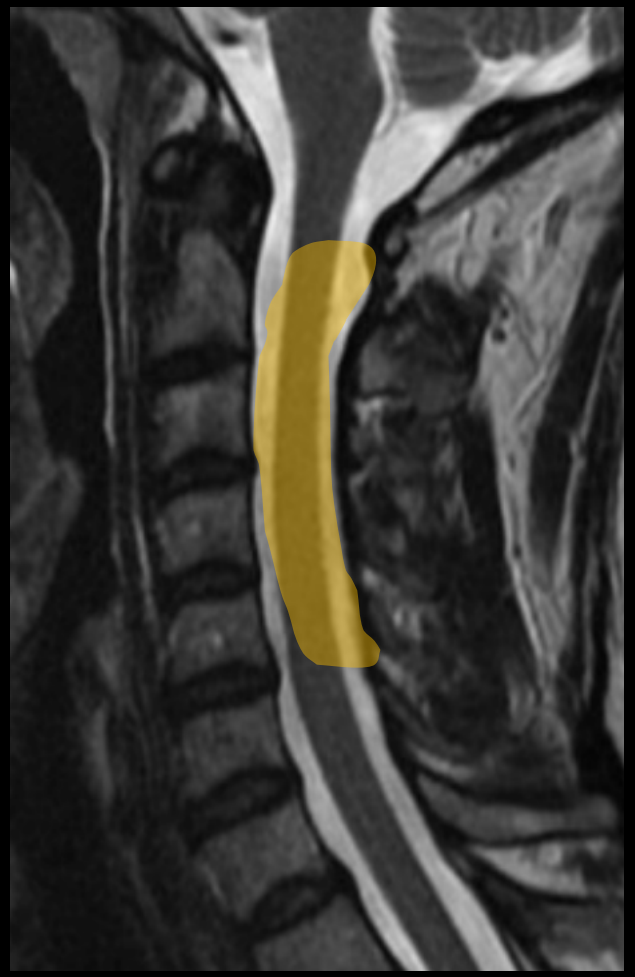






Spinal Cord Tumors

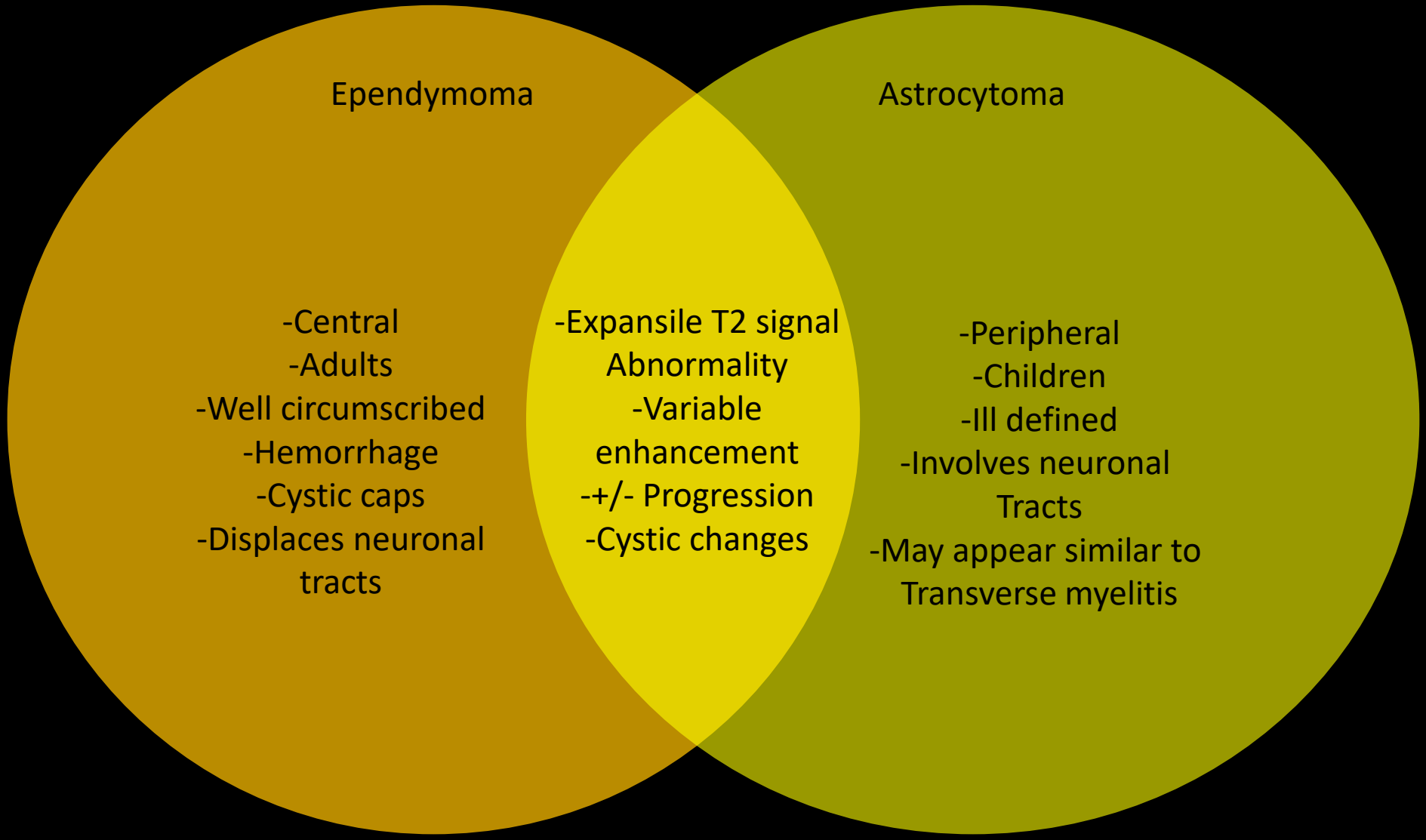
- Intramedullary tumors
  - Ependymoma
  - Astrocytoma
  - Hemangioblatoma
  - Ganglioglioma
  - Metastatic disease



Ependymoma Vs. Astrocytoma

- Ependymoma
  - Arises from cells of the central canal of the spinal cord
  - Most common intramedullary tumor (60%)
  - Cervical > Thoracic > Conus
  - Associated with NF 2 (MISME)
  - Imaging
    - Heterogenous signal and enhancement
    - ++ Hemorrhage
    - Centrally located
    - Peritumoral caps or syrinx
- Astrocytoma
  - Arises from astrocytic glial cells within the spinal cord
  - 2nd most common intramedullary tumor
  - Most common intramedullary tumor in children and young adults
  - Imaging
    - Hemorrhage rare
    - Peripherally located
    - May have intratumoral cysts

Spinal Cord Tumor - Intramedullary



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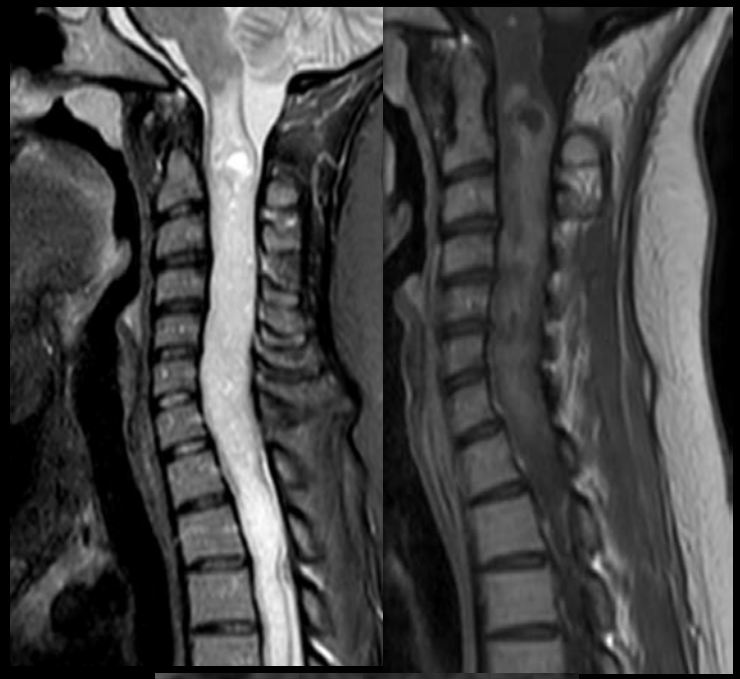
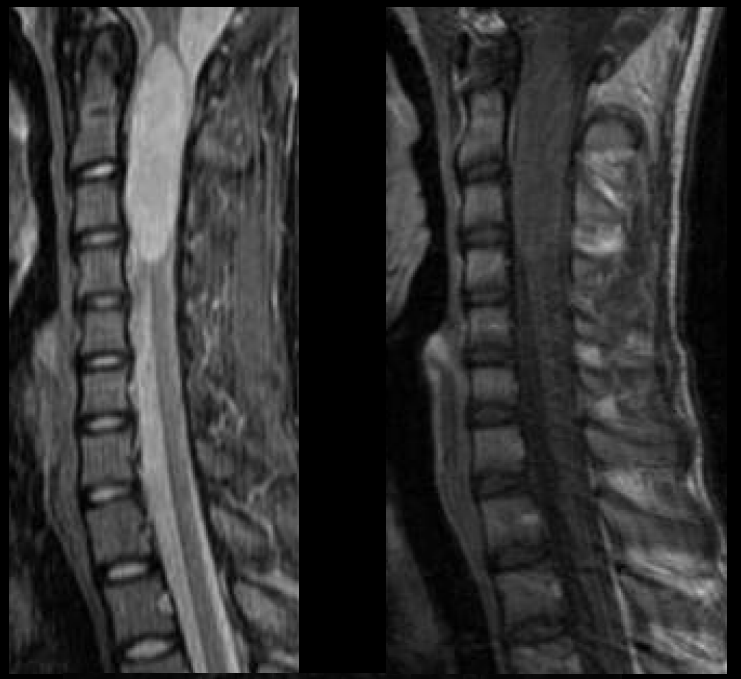
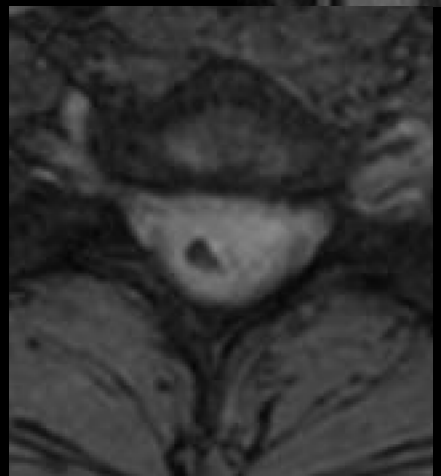
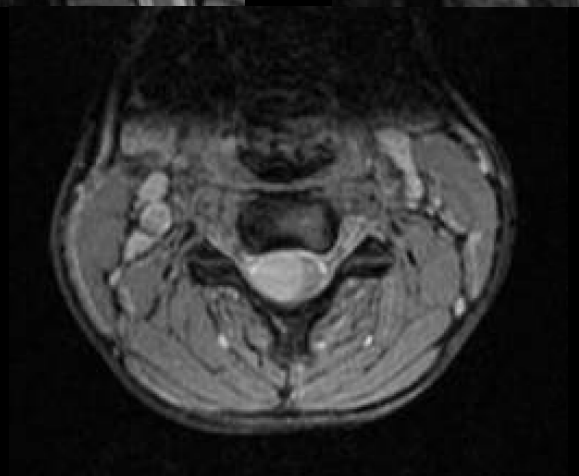


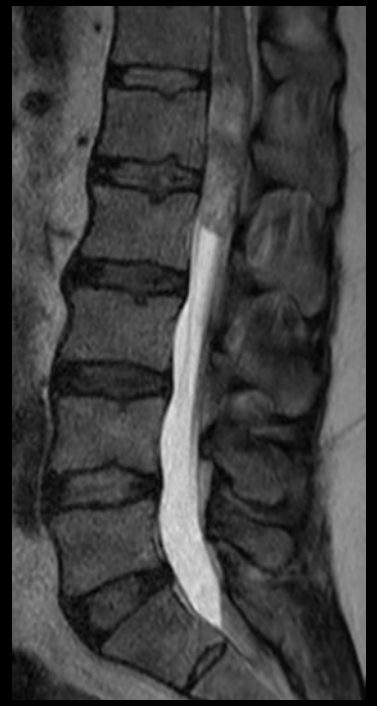
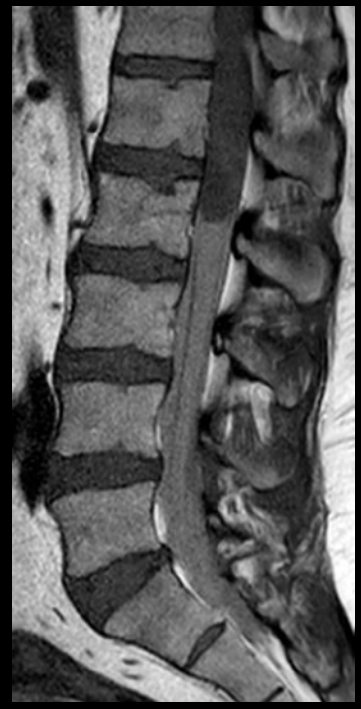
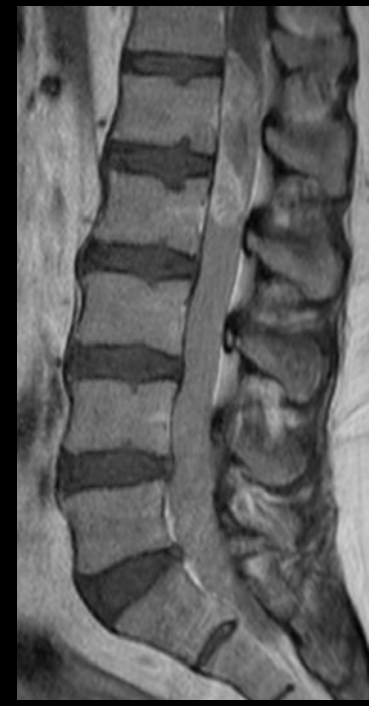
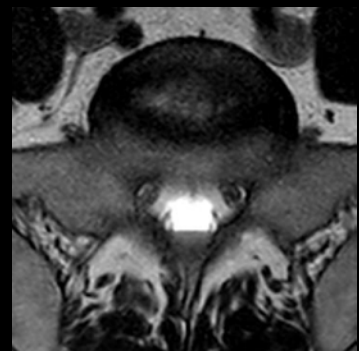
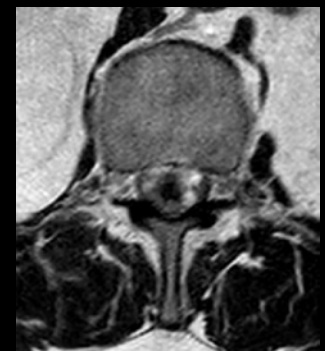


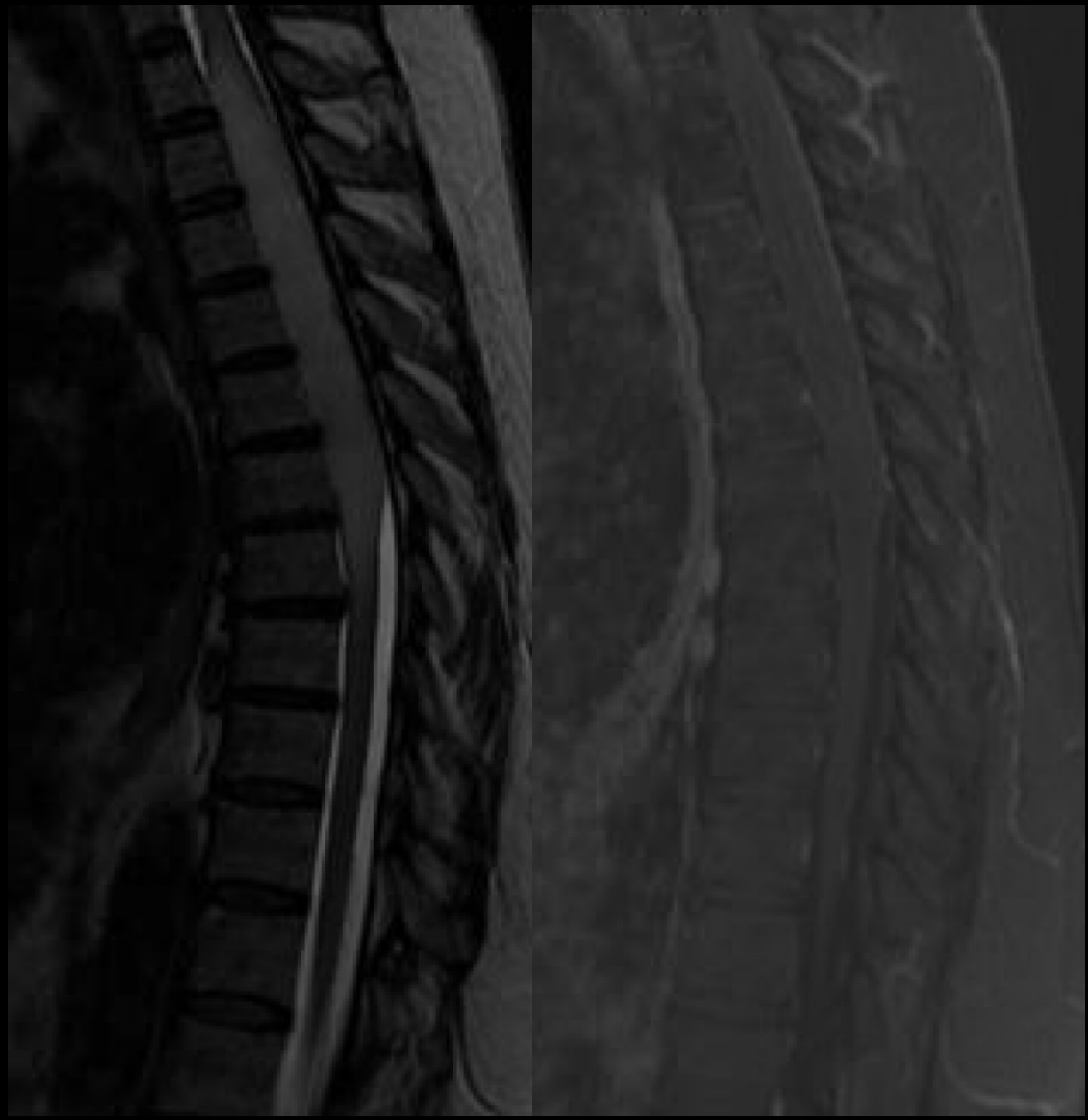




Myxopapillary Ependymoma

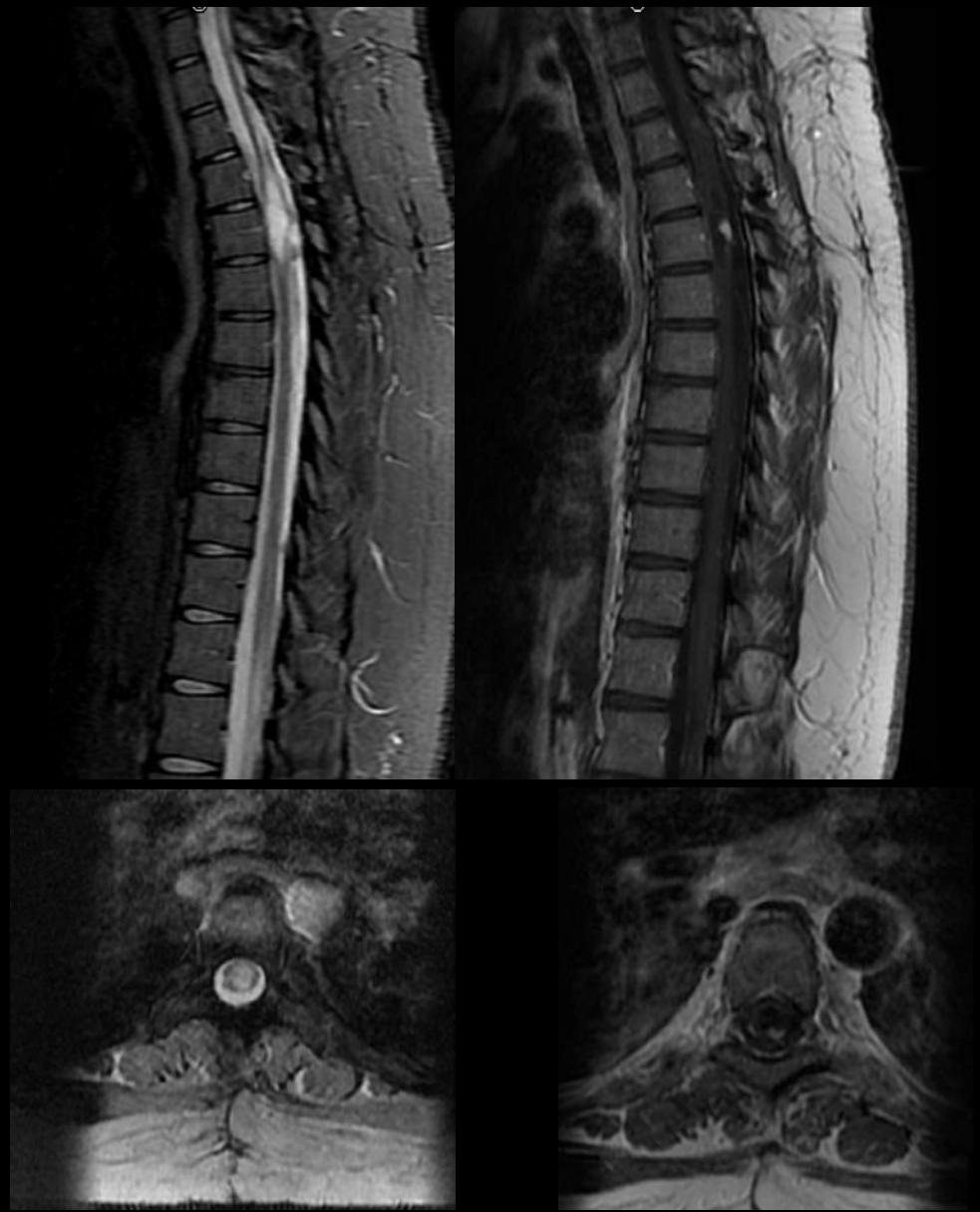
- Variant of ependymoma
  - 13% of all ependymomas
- Occur near conus/filum
- Average age at presentation – 35 yo
- WHO grade I
  - CSF dissemination may occur
  - More aggressive in children
- Imaging
  - Background of T2 hyperintense signal
  - T2 hypointense foci
    - Calcium and/or hemorrhage
  - Typically homogeneous enhancement however pattern variable



Spinal Cord Tumor

- Hemagnioblastoma
- Rare
  - 1-6% of spinal tumors
- Sporadic or familial
  - VHL
- Histopathologically cerebellar HBL similar to spinal HBL
- Hypervascular tumors
- Most intramedullary
  - Abut pial surface
  - May occur along nerve roots
- Enlarged arteries and veins associated with HBL
  - May simulate AVM
- Associated edema and syrinx
  - May simulate astrocytoma
  - Syrinx more common in HBL than other spinal tumors



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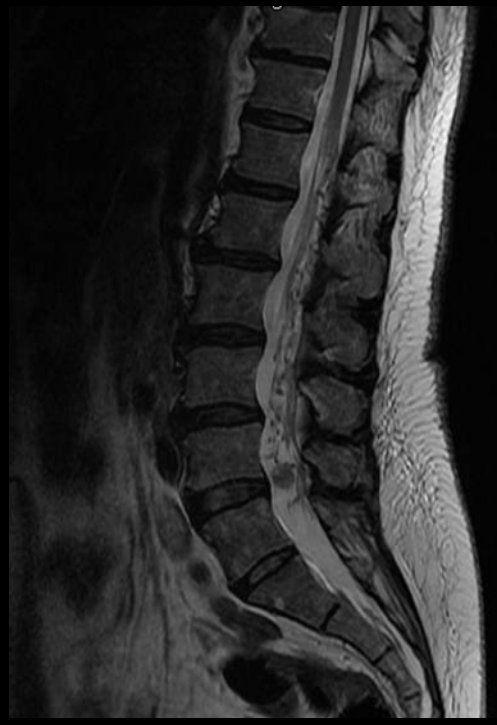
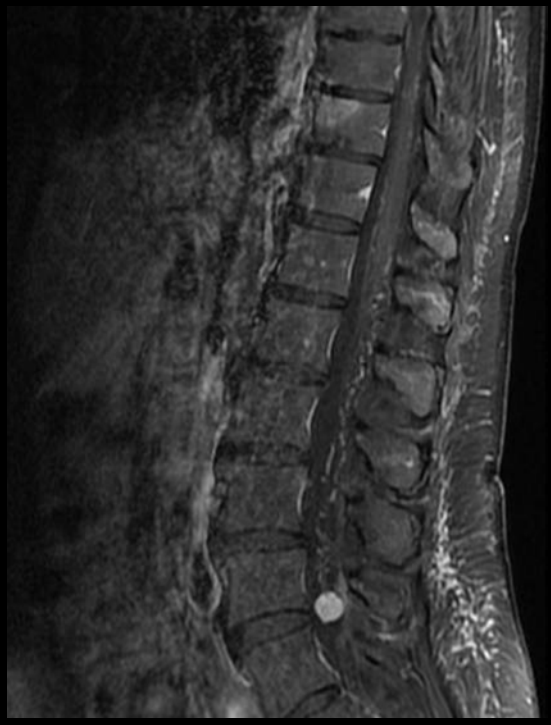
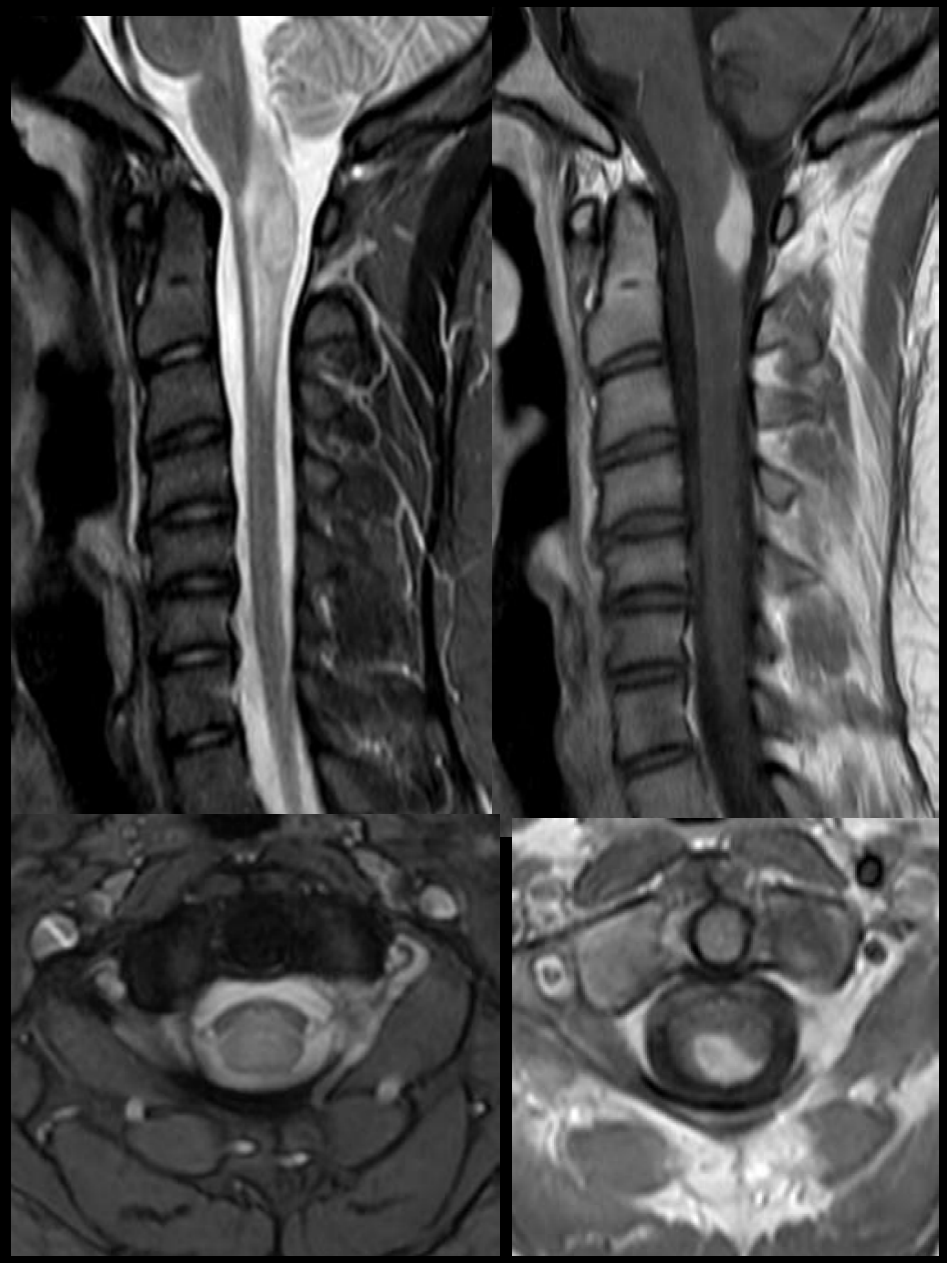




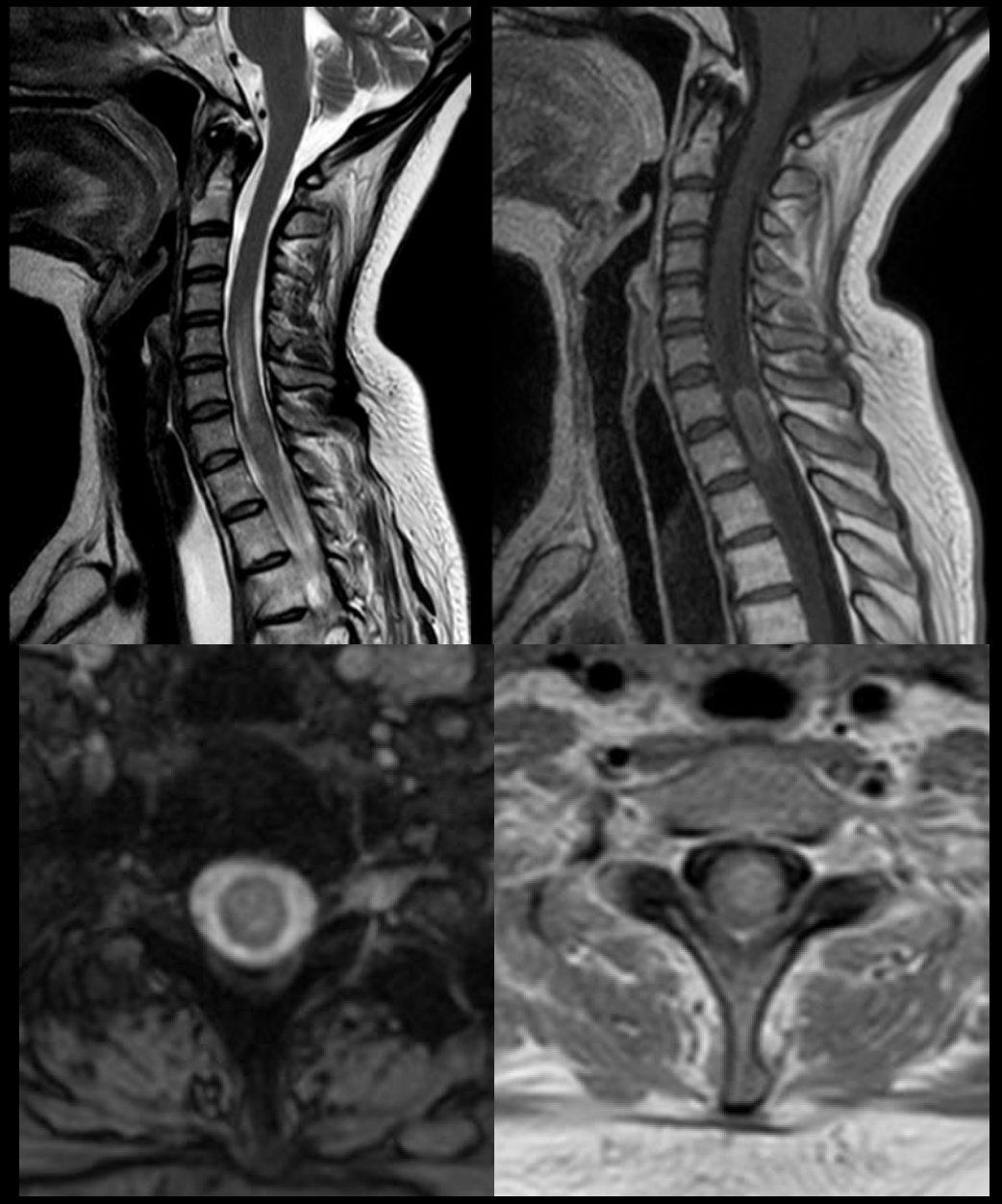
Spinal Cord Tumor

- Ganglioglioma
  - Rare – 1.1% of spinal cord tumors
  - Contains both astrocytic and neural cells
  - Average age at presentation – 23
  - Imaging
    - Nonspecific
      - May show areas of intrinsic T1 shortening



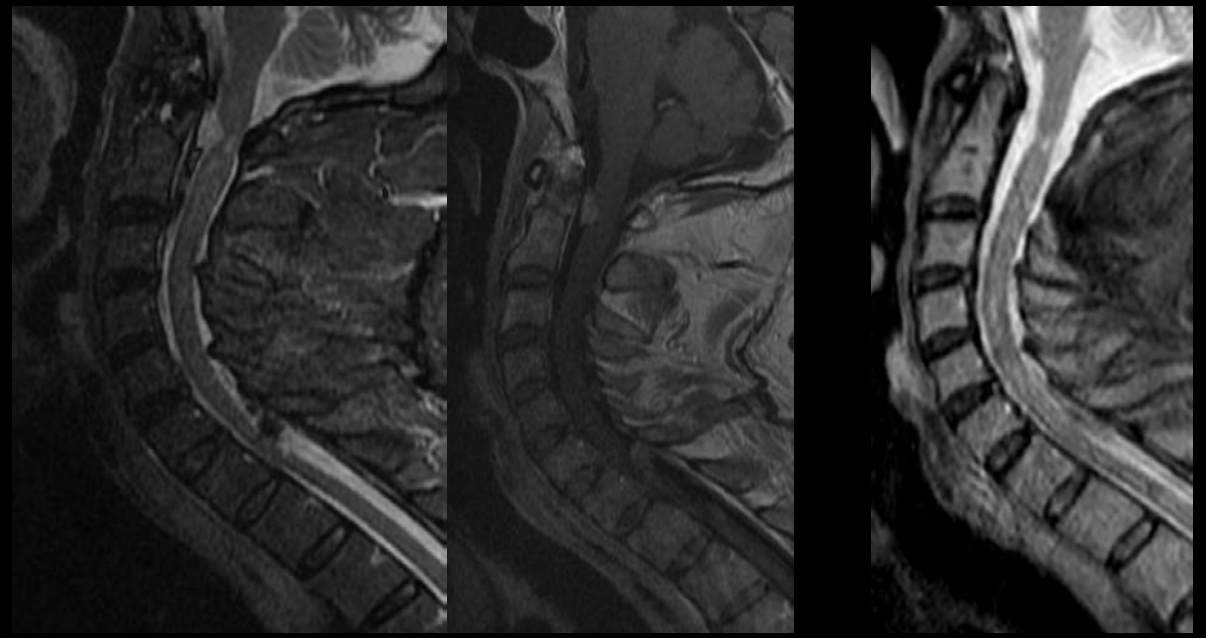
Spinal Cord Tumor

- Spinal cord intramedullary metastatic disease
  - Rare
    - <2% of spinal cord tumors
      - Leptomeningeal metastatic disease much more common
    - Hematogenous spread
    - Most common primary
      - Lung – 50%
    - Spinal cord mass in a patient with a known primary
    - Solitary lesion in 85%
    - 50% of patients will have concomitant brain metastases
    - 25% leptomeningeal disease
  - Poor prognostic indicator
    - Average survival <4 mo following diagnosis



Spinal Cord Atrophy

- End results of any prior spinal cord insult
  - Myelomalacia
  - Wallerian degeneration
- Neurodegenerative process
  - Spinocerebellar ataxia



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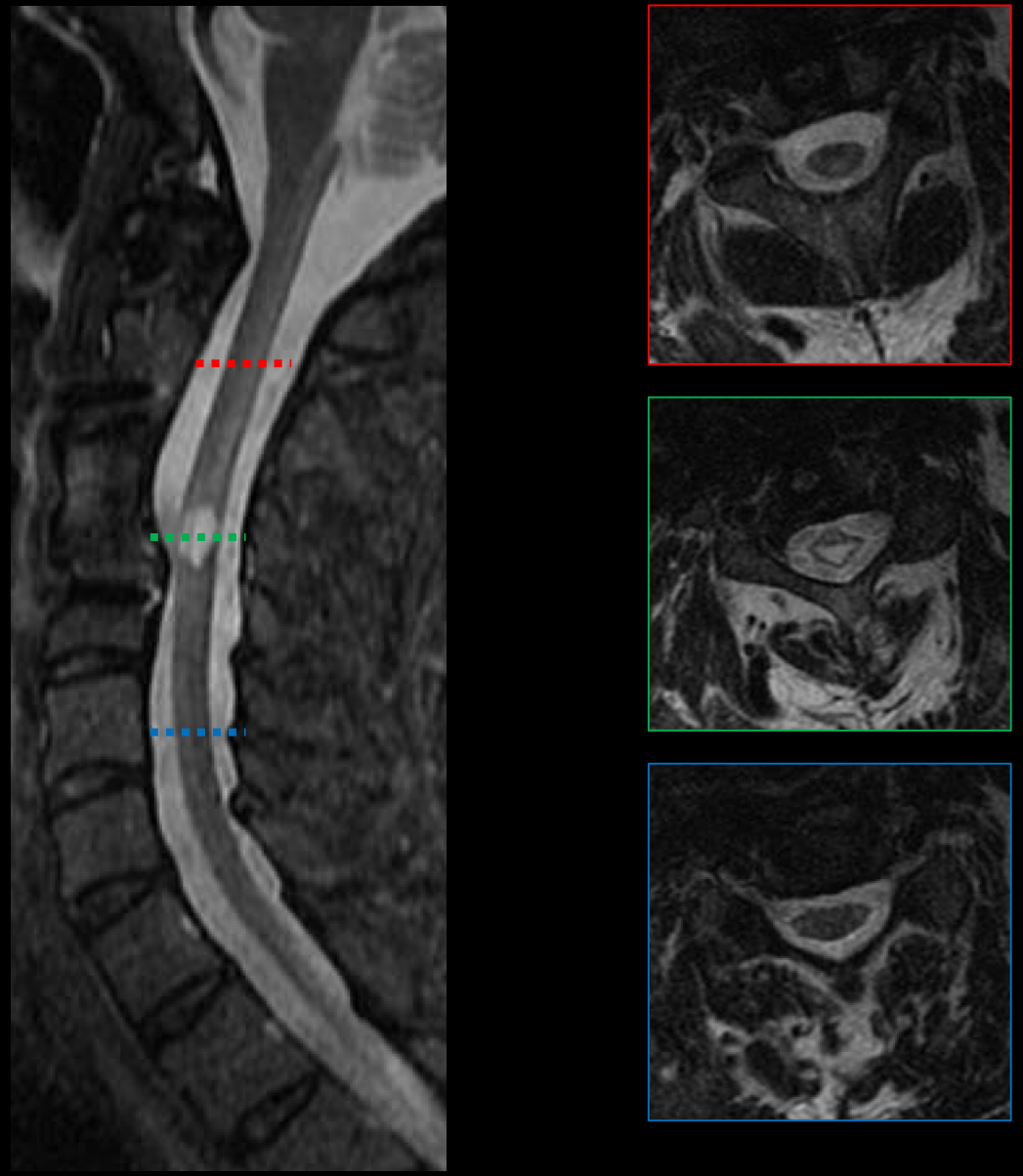

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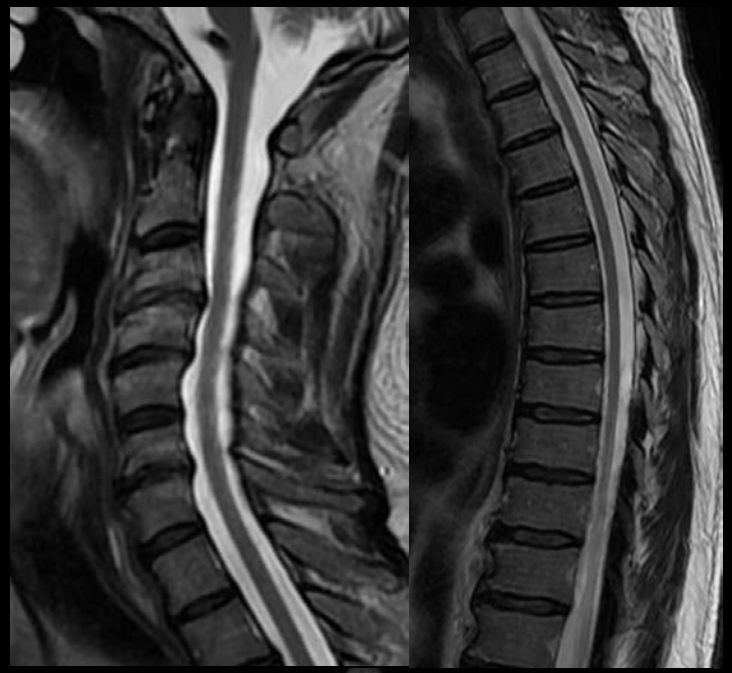
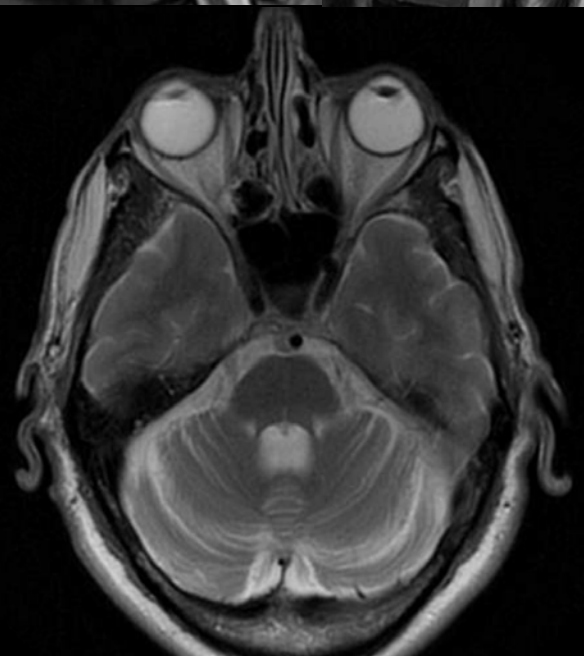
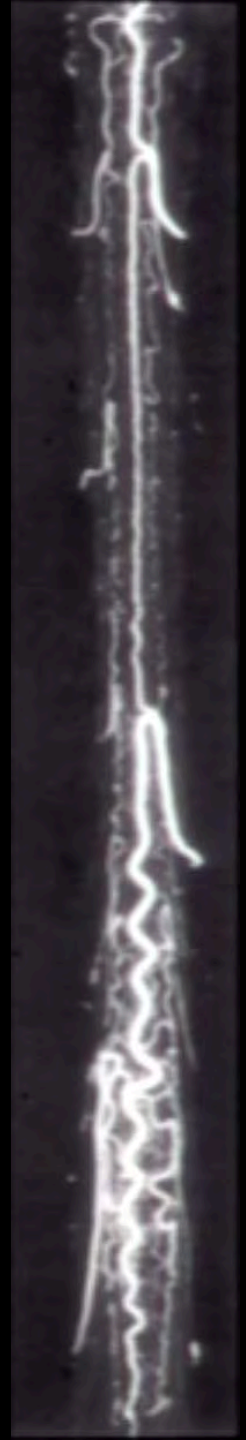




Unique Features – Flow Voids

- Spinal vascular anatomy



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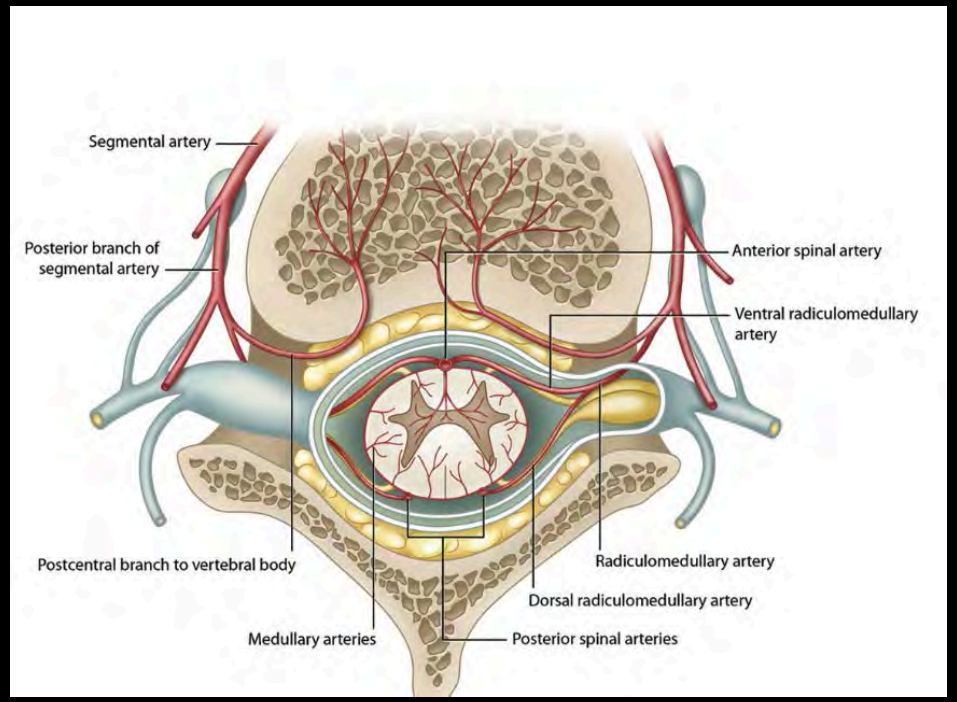

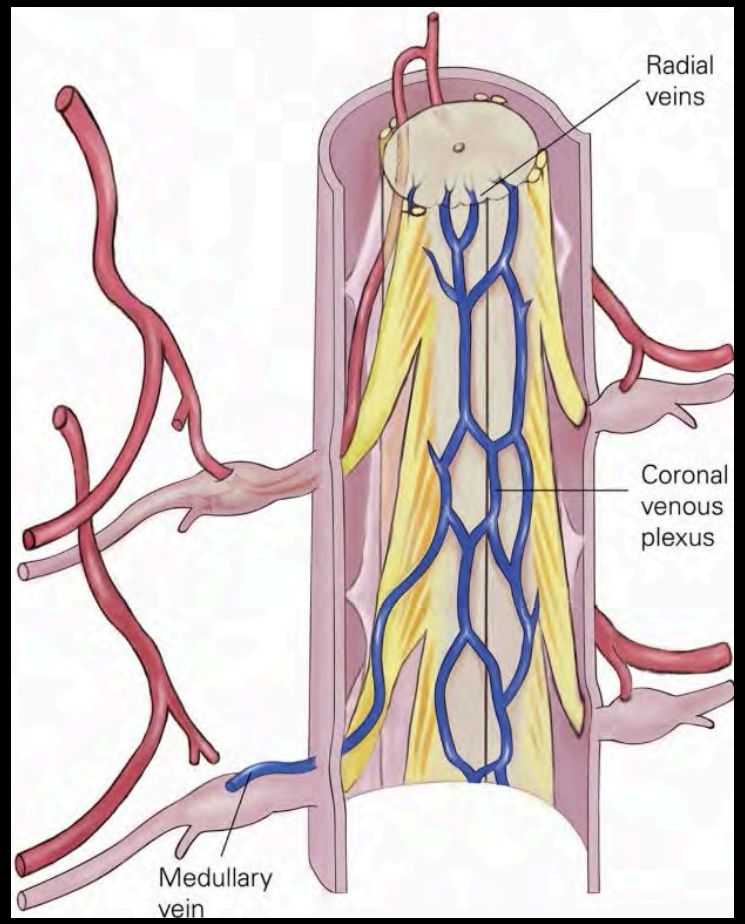

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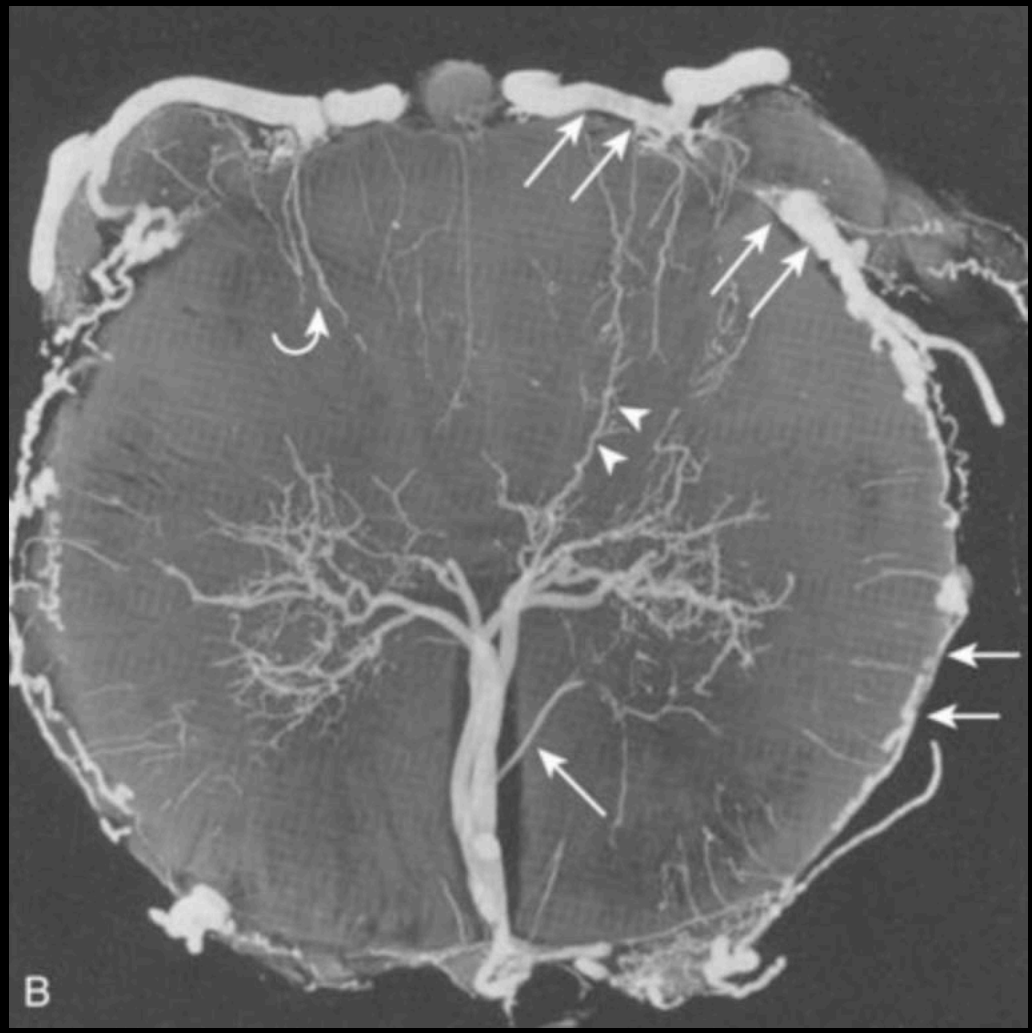
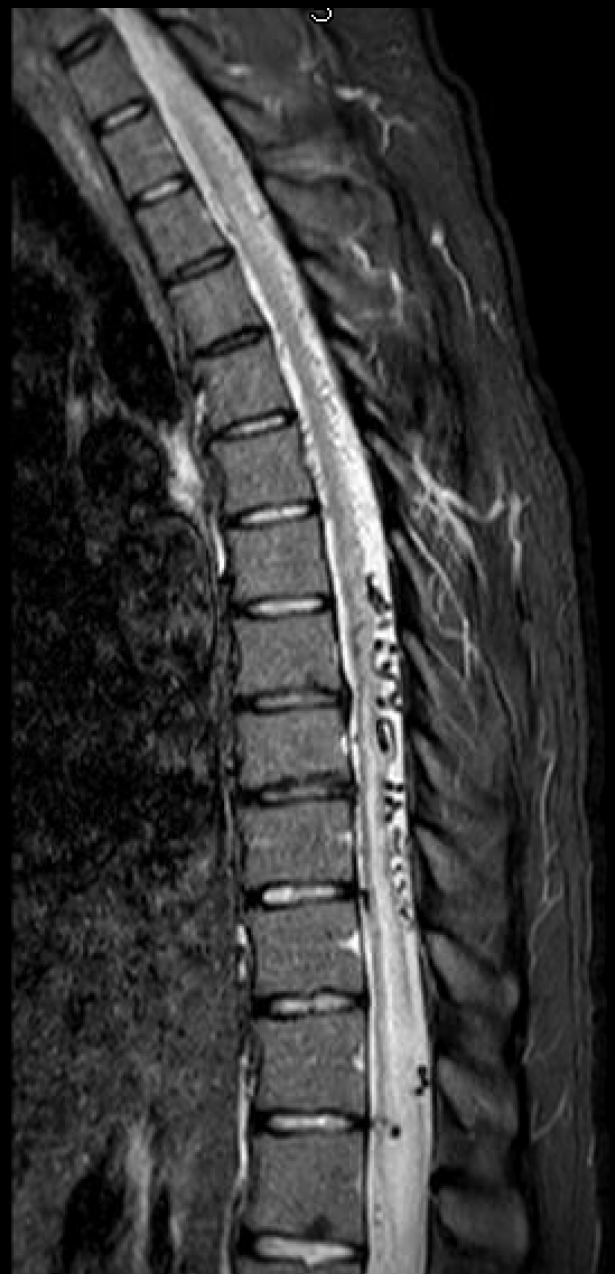


Unique Features – Flow Voids

- Vascular lesions
- Classification



Unique Features – Flow Voids

- Vascular lesions
- Classification

Type I–IV Classification System	Modified Spetzler Classification
Type I (dural AVF or SDAVFs)	Intradural dorsal AVF
Type II (intramedullary or glomus AVM, SCAVMs)	Intramedullary AVM
Type III (metameric or juvenile AVM)	Extradural-intradural AVM
Type IV (perimedullary AVF or SPAVFs)	Intradural ventral AVF (types A, B, and C)
	Extradural AVF
	Conus medullaris AVM

Neurographics, Volume 6, Number 3, 1 May 2016, pp. 159-170(12)

Unique Features – Flow Voids

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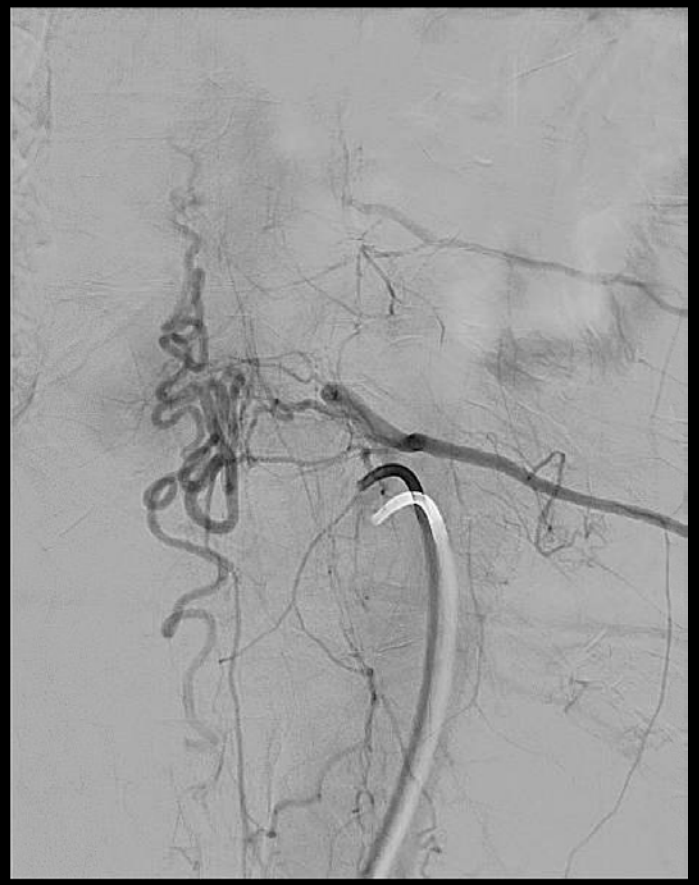
TABLE 2

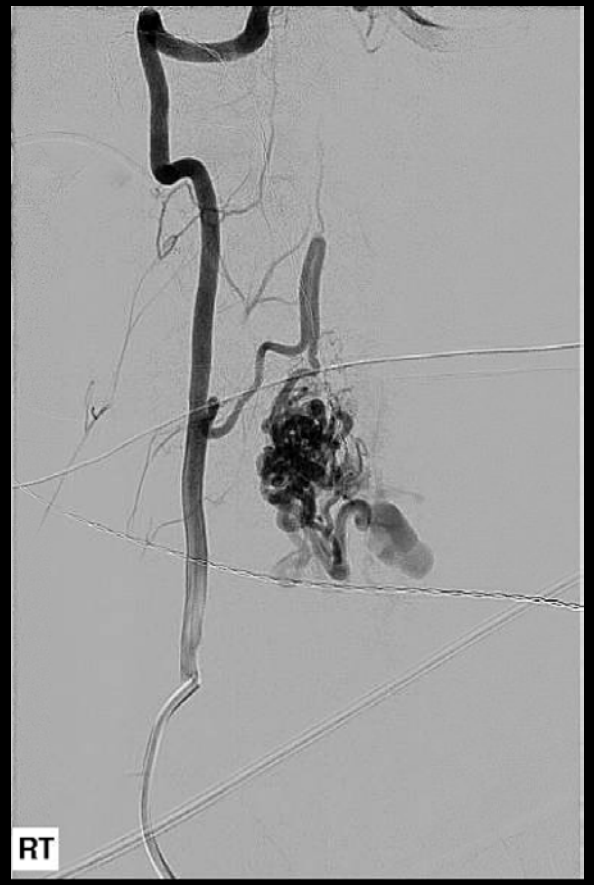
Proposed classification of spinal cord vascular malformations

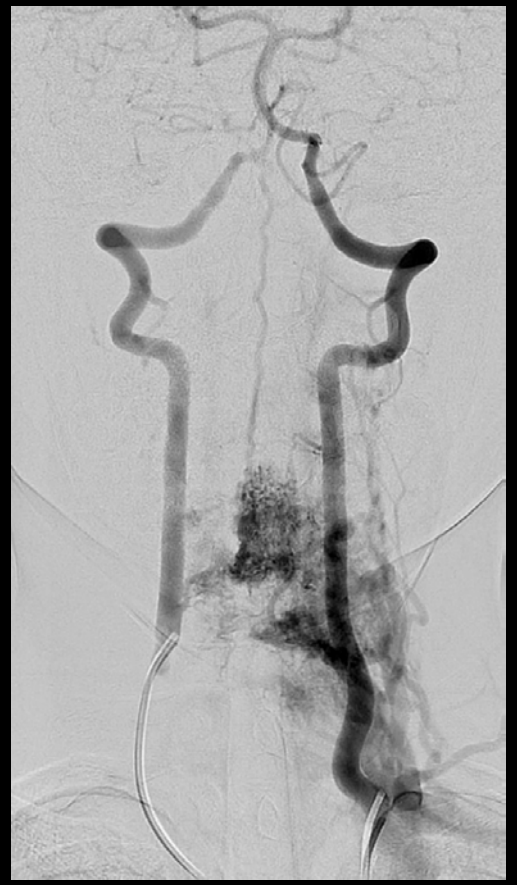
neoplastic vascular lesions
hemangioblastoma
cavernous malformation
spinal aneurysms
arteriovenous fistulas
extradural
intradural
ventral*
dorsal†
AVMs
extradural–intradural
intradural
intramedullary
compact
diffuse
conus medullaris

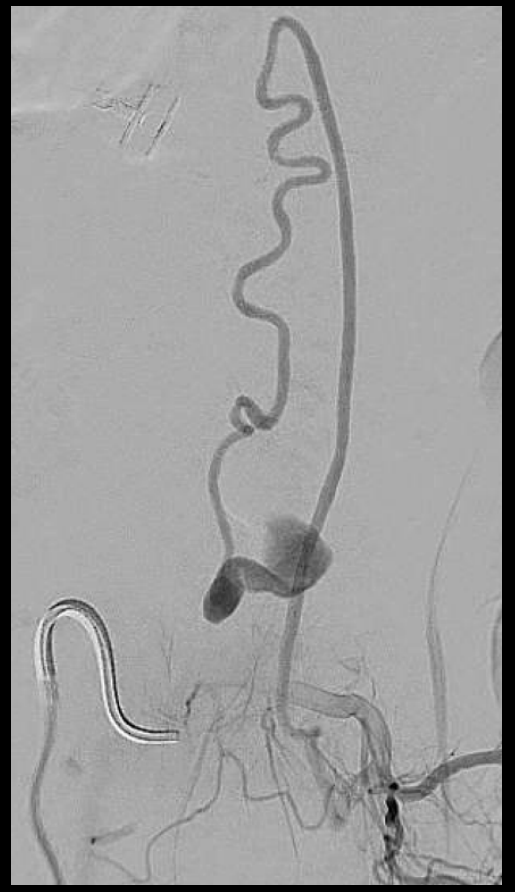
J Neurosurg. 2002 Mar;96:145-56.

Type I-IV

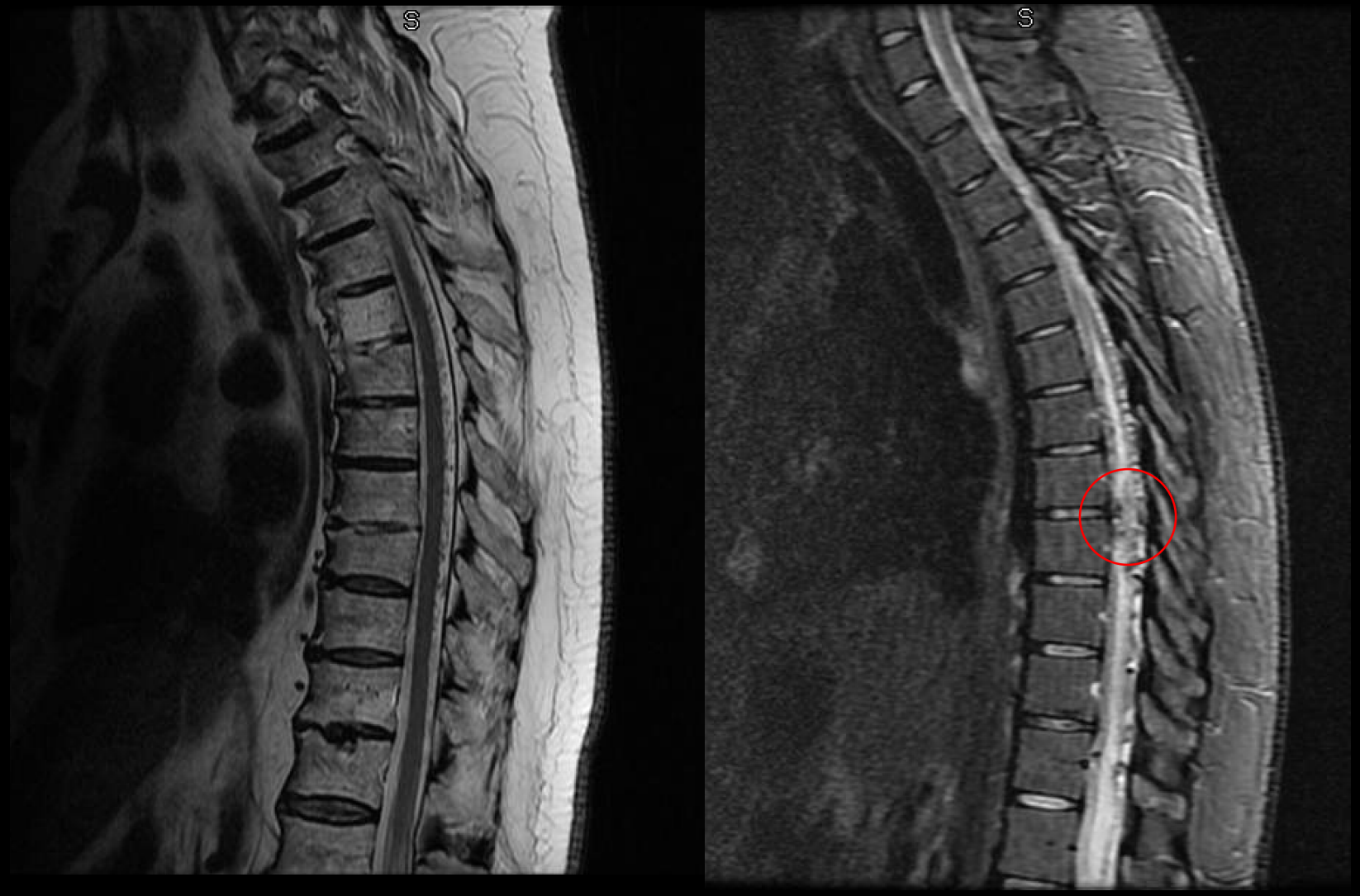






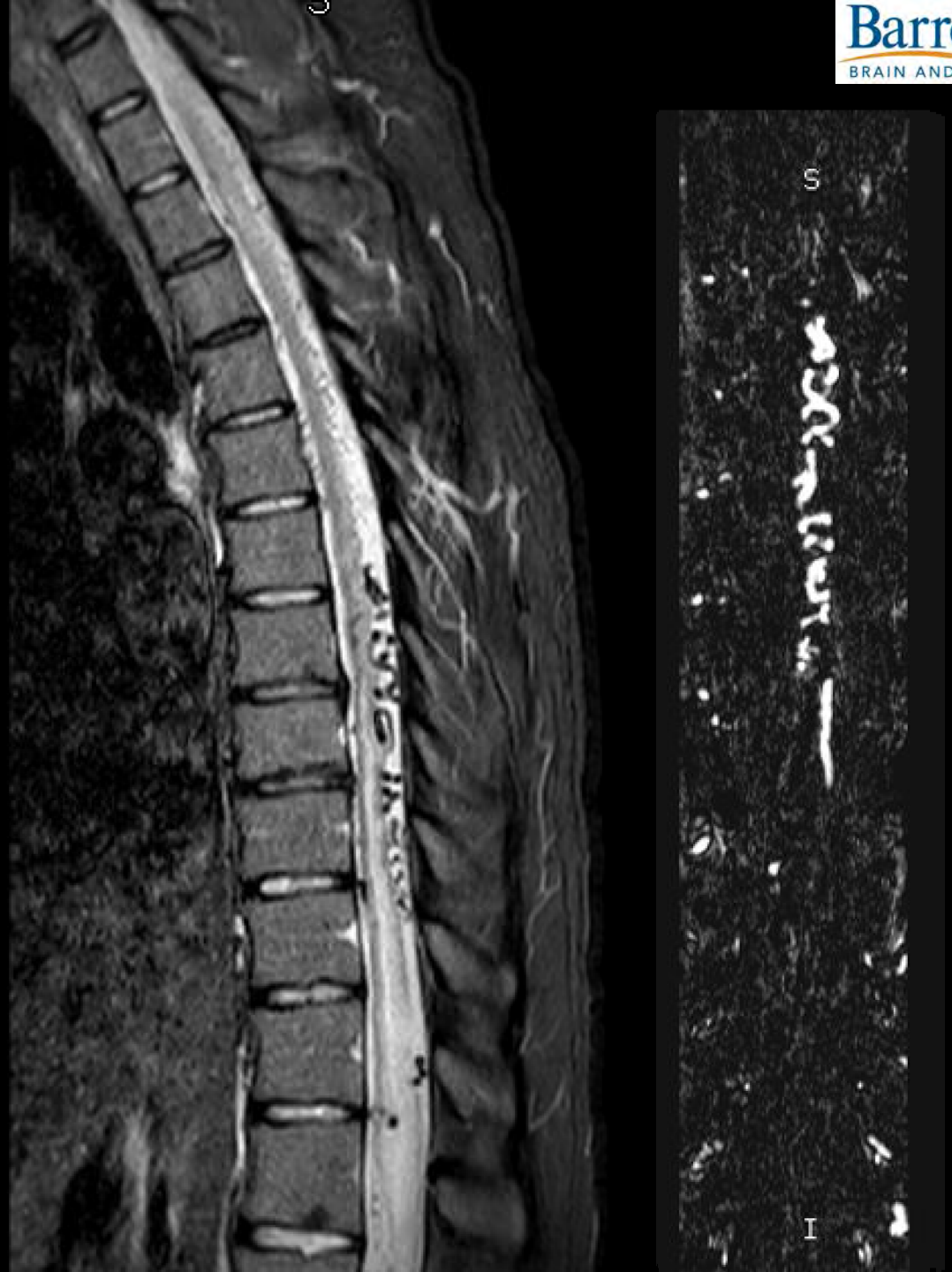


Fistula vs AVM



Spinal Fistulas

- Type I (spinal dural AV fistula, SDAVF)
  - Most common spinal AVF
    - >90%
  - Abnormal shunt between radicular artery and vein
    - Type a – single artery
    - Type B – multiple arteries
  - Level of the dural nerve root sleeve
  - Peak incidence
    - T7-T12
    - 80% between T6-L2
  - Reflux into engorged coronal venous plexus
    - Accounts for typical MR appearance
      - +/- Enlarged veins
      - +/- Cord edema



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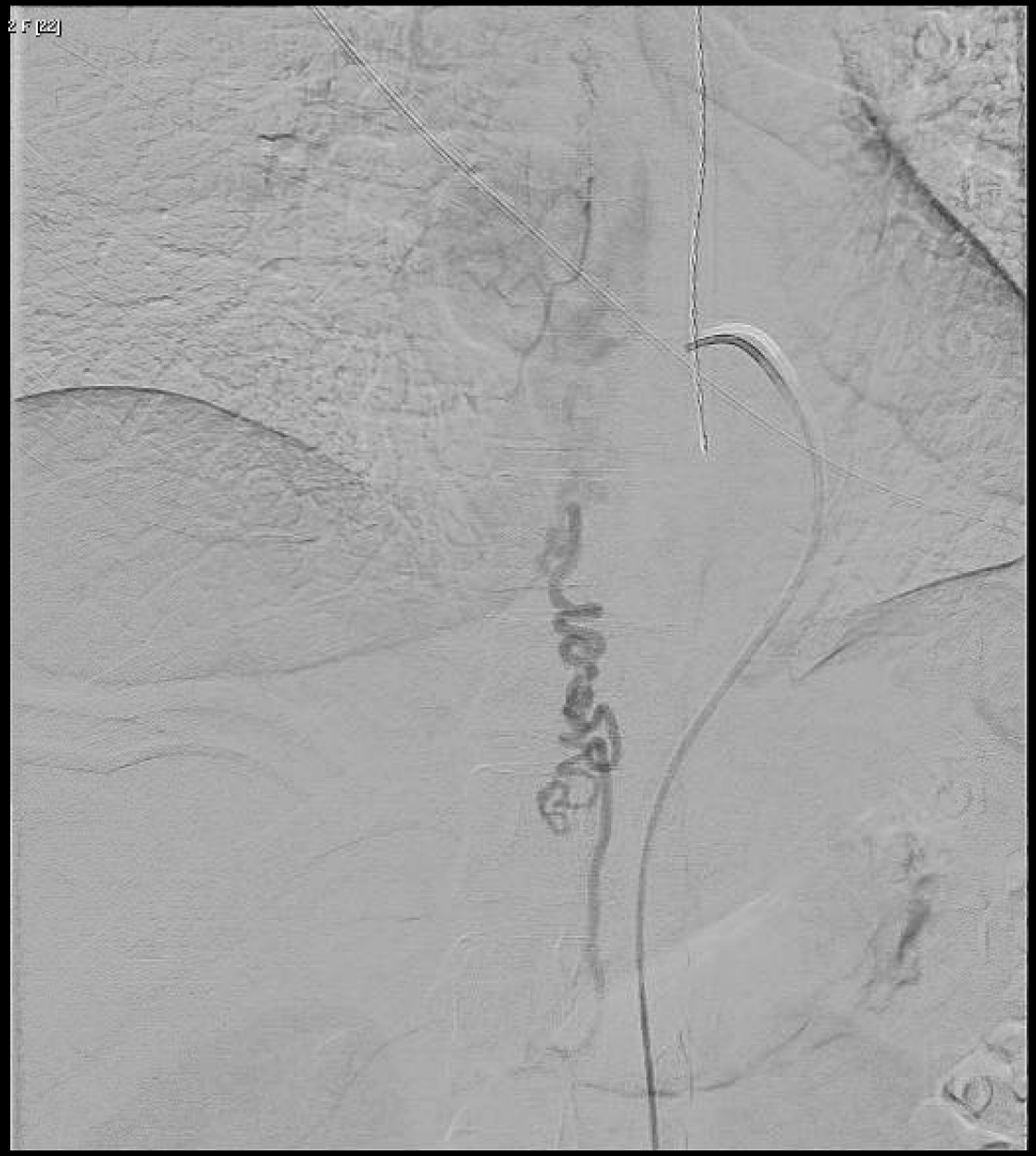
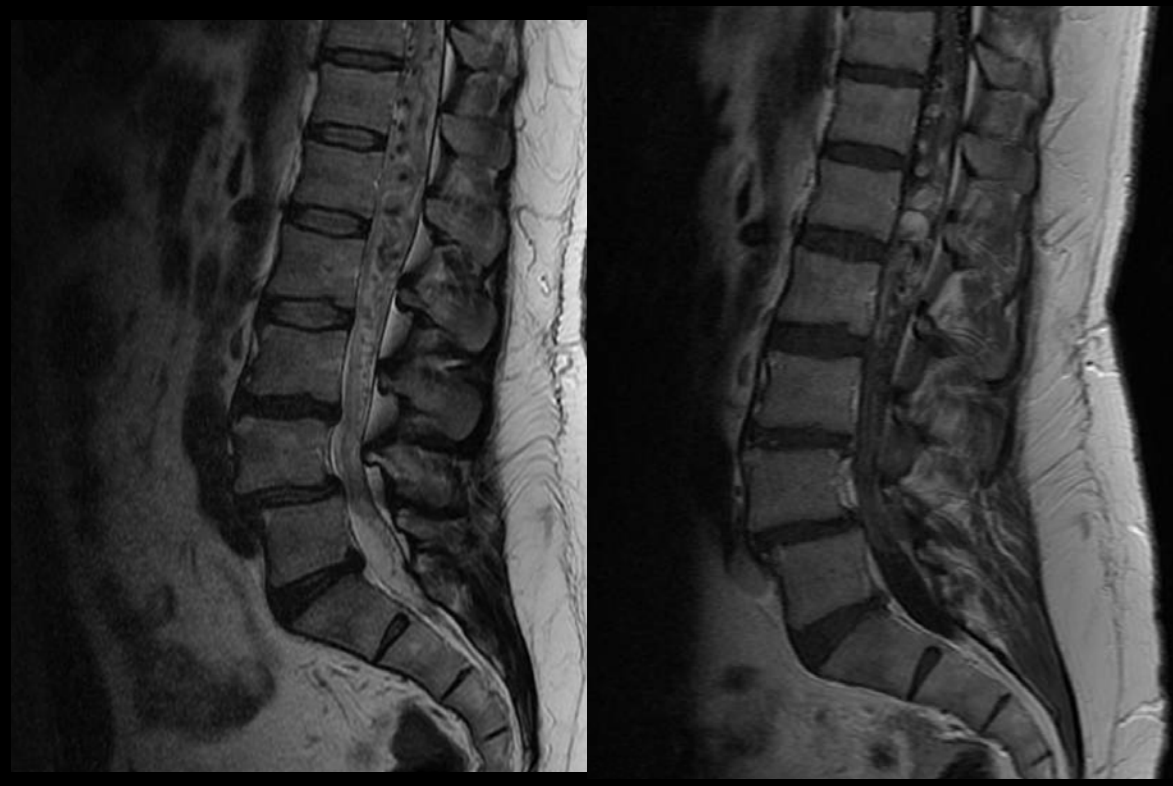
  - Reflux into engorged coronal venous plexus
    - Accounts for typical MR appearance
      - +/- Enlarged veins
      - +/- Cord edema



Spinal Fistulas

- Type IV (Perimedullary AV fistula, SPAVF)
  - Fistula occurs on the pial surface of the cord
    - between the ASA and adjacent coronal venous plexus
  - Type A
    - Small shunts
      - Slow flow
      - Moderate venous hypertension
  - Type B
    - Moderate sized shunt
      - Increased flow and venous hypertension
  - Type C
    - Large shunts ”Giant fistula”
      - Large veins with increased steal and mass effect on the cord



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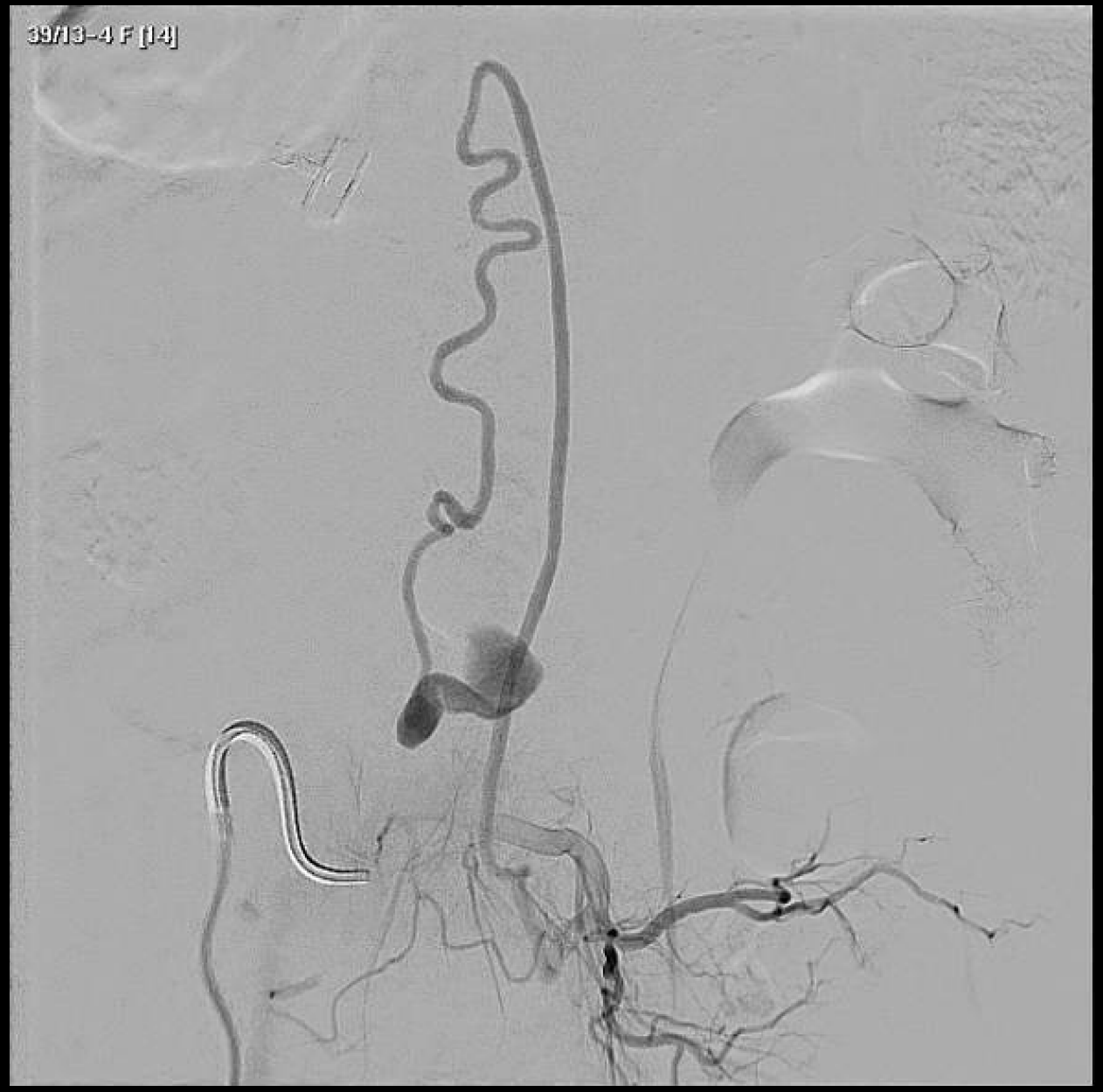
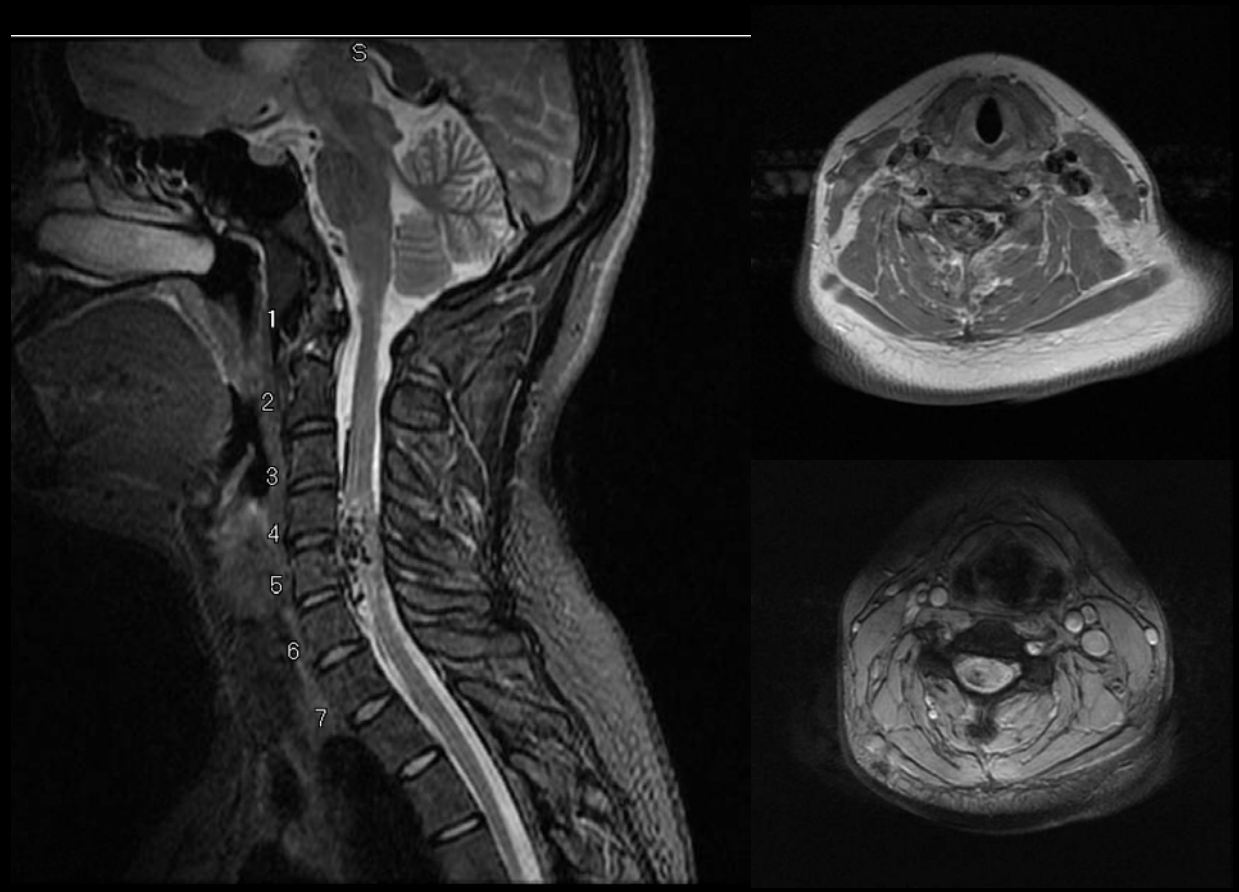


Spinal AVM

- Type II intramedullary or glomus AVM
  - Abnormal connection between artery and vein via a nidus
    - Nidus
      - Compact or diffuse
    - At least 1 feeding vessel from ASA or PSA
      - Characterized by high pressure and high flow
      - May have associated feeding artery aneurysm
  - Risk of intramedullary and subarachnoid hemorrhage
    - AVM >>>> AVF
  - Clinical presentation
    - Acute myelopathy
      - hemorrhage
    - Progressive myelopathy
      - Vascular steal



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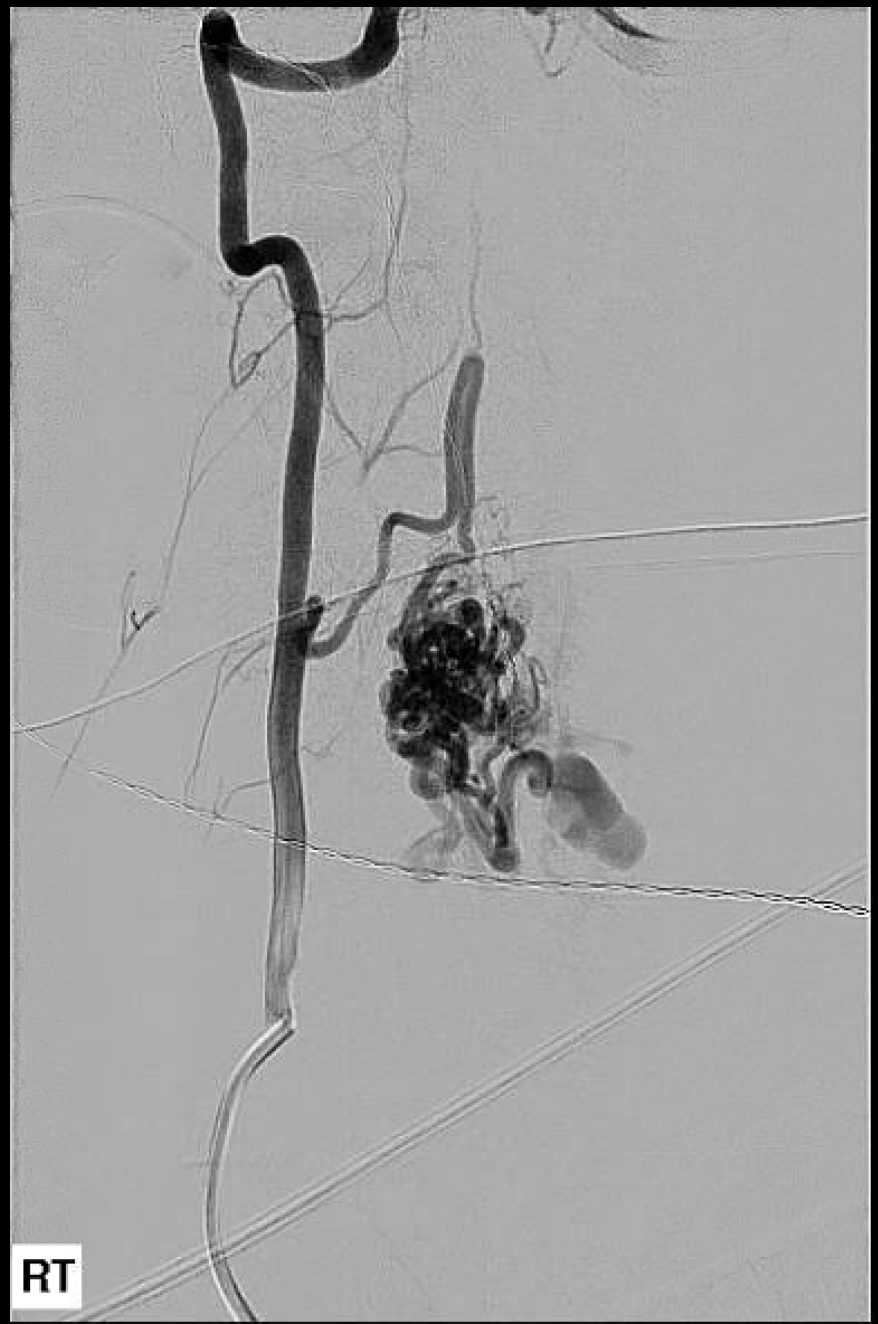
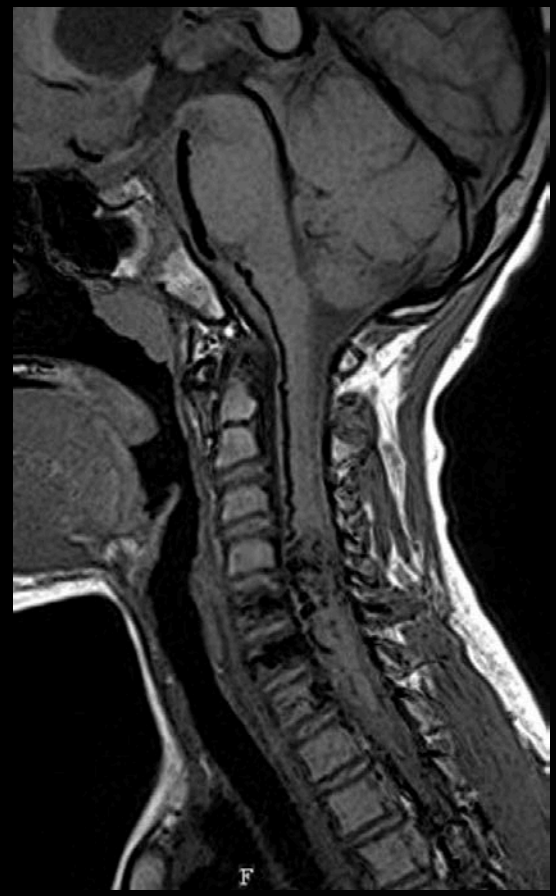
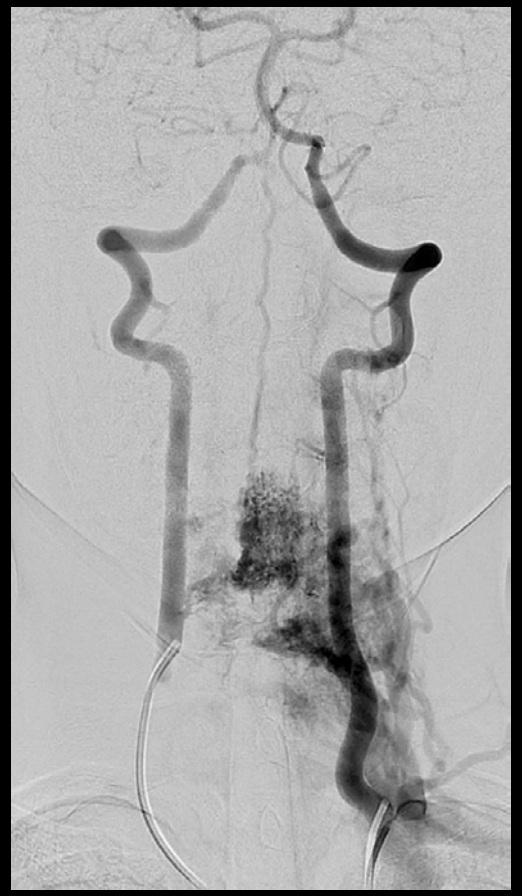


Spinal AVM

- Type III (Metameric or Juvenile AVM)
  - Also called intradural-extradural AVM
  - Can involve the spinal cord and any component of the metamere at a given level
  - Extremely uncommon
  - Cobb syndrome
  - Requires multidisciplinary approach to treatment

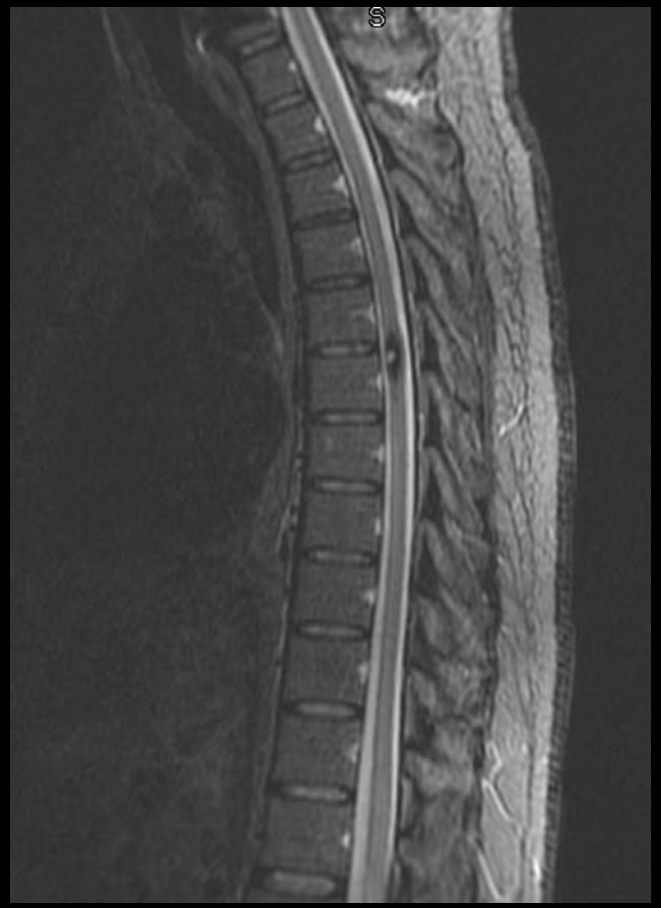


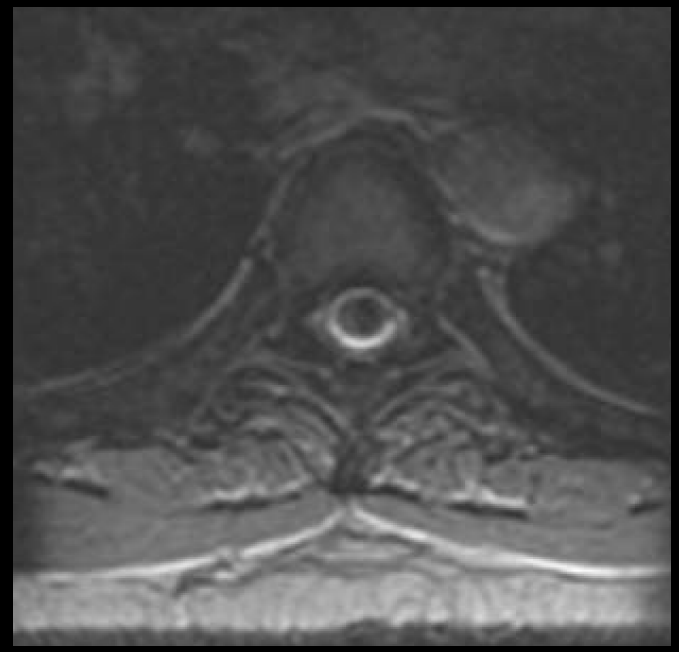


AJNR, Case of the week, Katsunari Namba, MD

Cavernous Malformation

- Vascular lesion with risk of hemorrhage
  - If hemorrhagic present acutely similar to spinal cord AVM
- Vascular channels formed by endothelial cells
- associated with DVA
  - Delayed venous infarct associated with resection of DVA
- Characteristic MR appearance
  - Circumscribed lesion with T1/T2 hypointense signal
    - Due to hemosiderin rim
  - Absence of surrounding edema unless hemorrhagic
  - Not visible on spinal angiography
    - Angiographically occult





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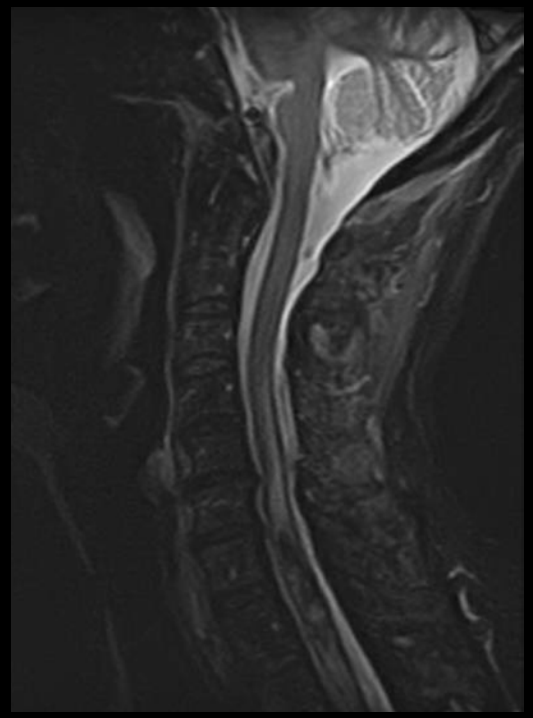
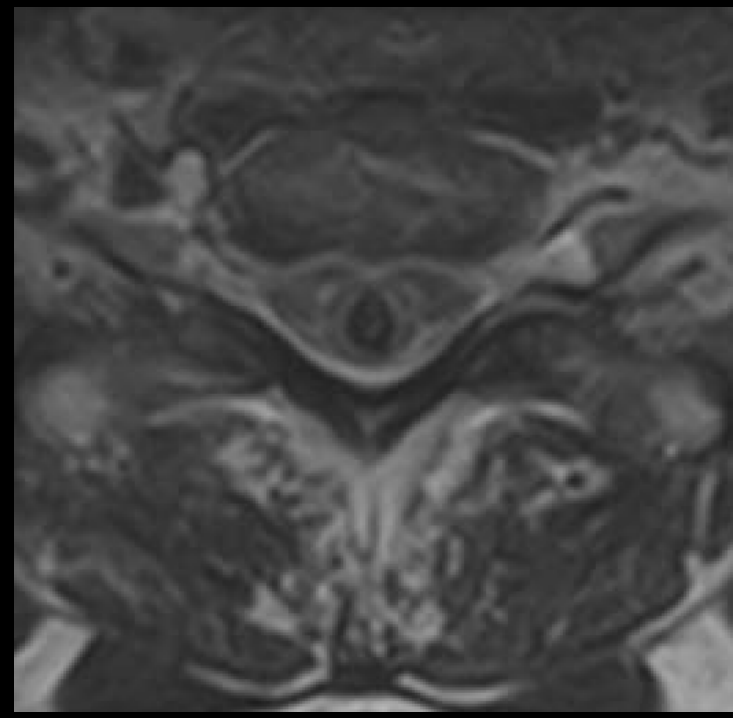
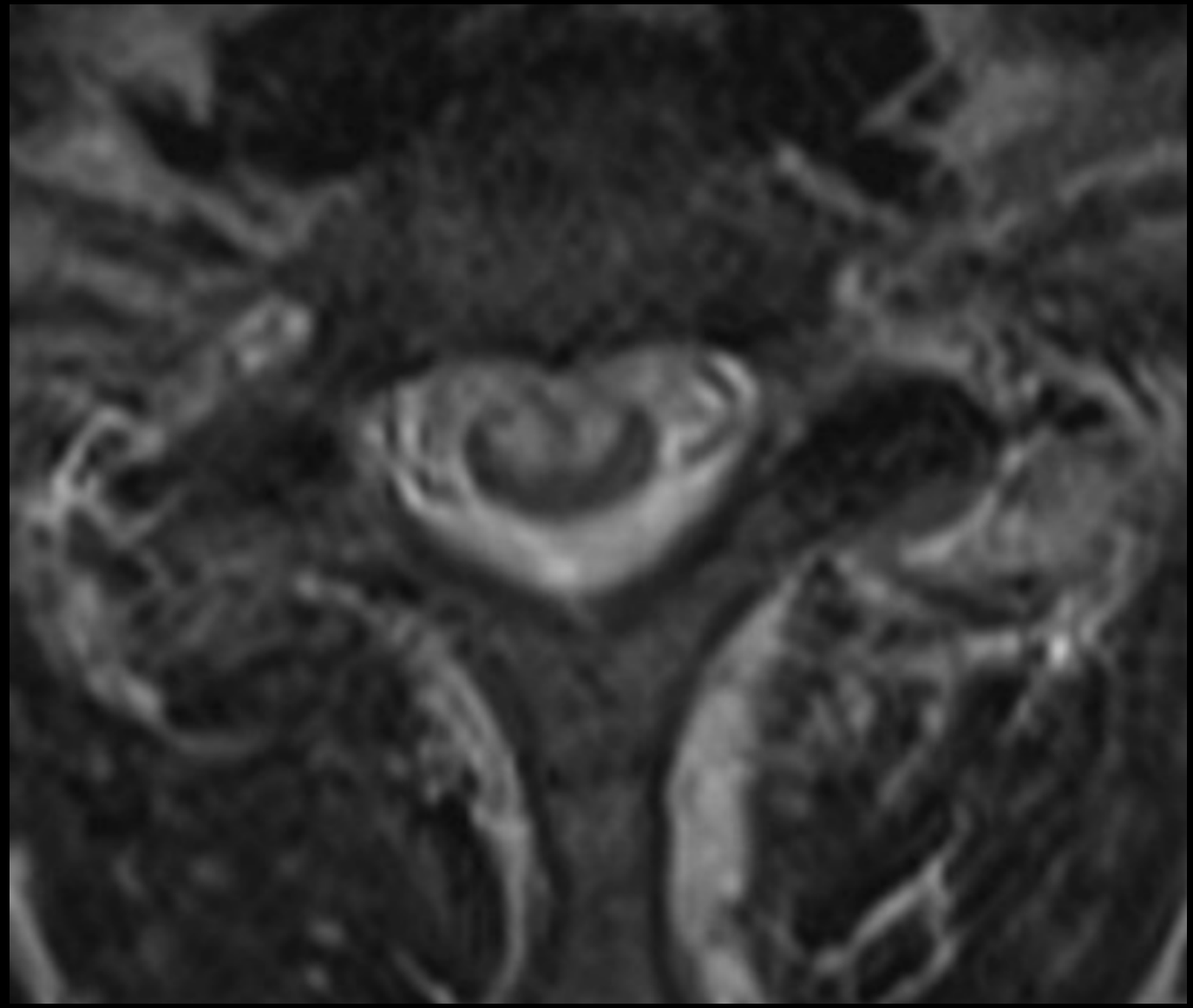




Unique Feature - Anatomic Distribution

- Anterior horn cell
  - Spinal cord infarct
    - Complication of aortic aneurysm surgery
    - Dissection
    - Catheterization



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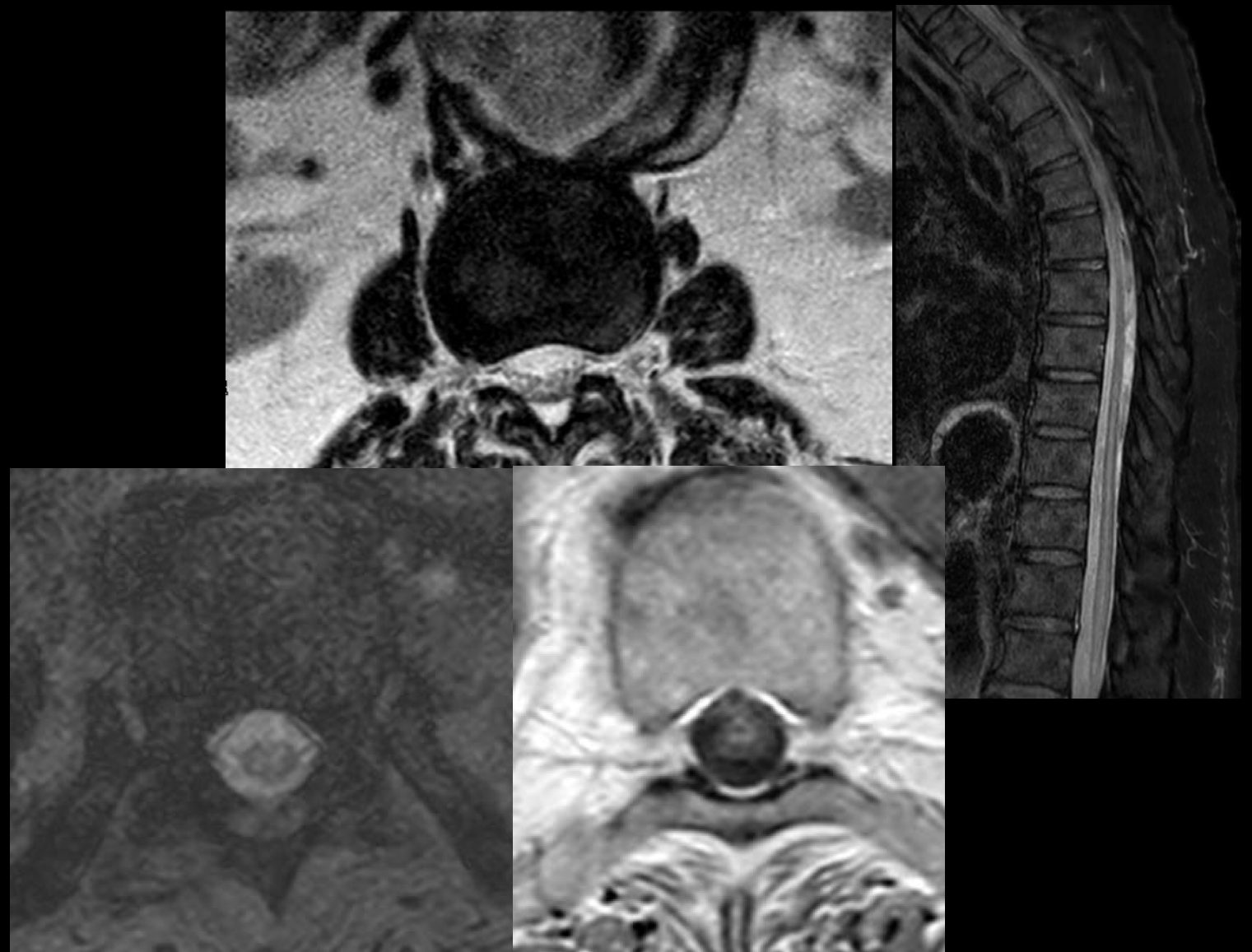
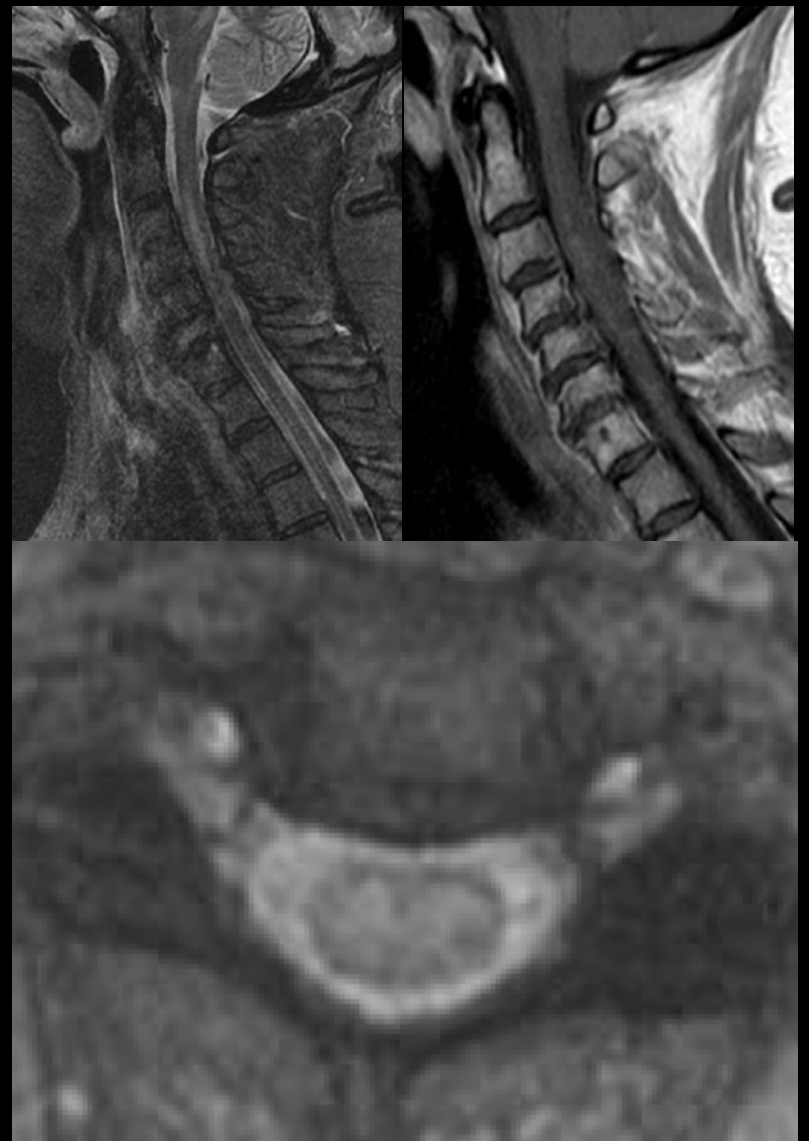


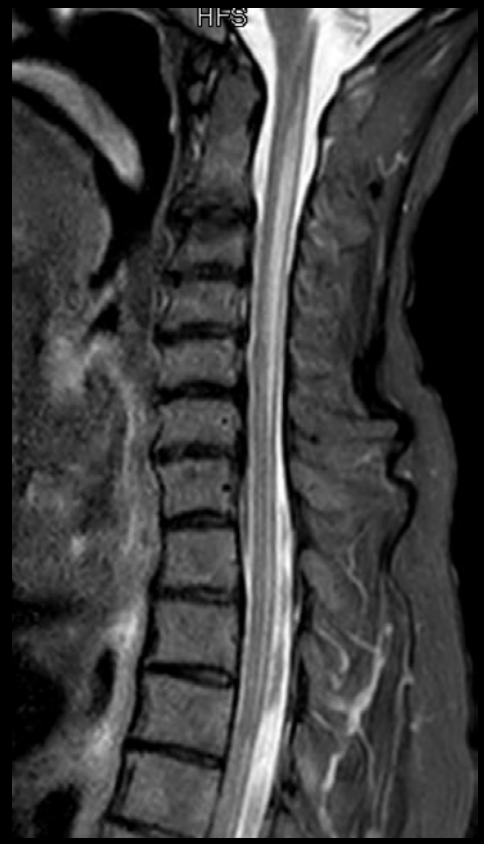
Unique Feature - Anatomic distribution

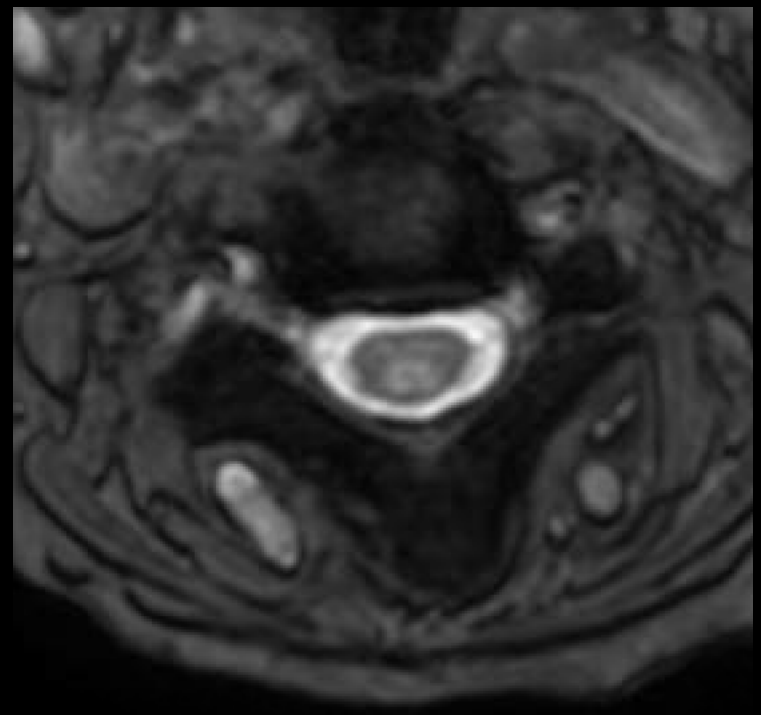
- Anterior horn cell
  - Poliomyelitis
  - Acute flaccid myelitis
    - Unknown etiology
      - Epidiomologic evidence suggests an association with enterovirus D68
    - Predominantly children
    - Increased incidence 2012-2015
    - 2018 – 201 cases
      - 4 confirmed cases in AZ



Unique Feature – Anatomic Distribution

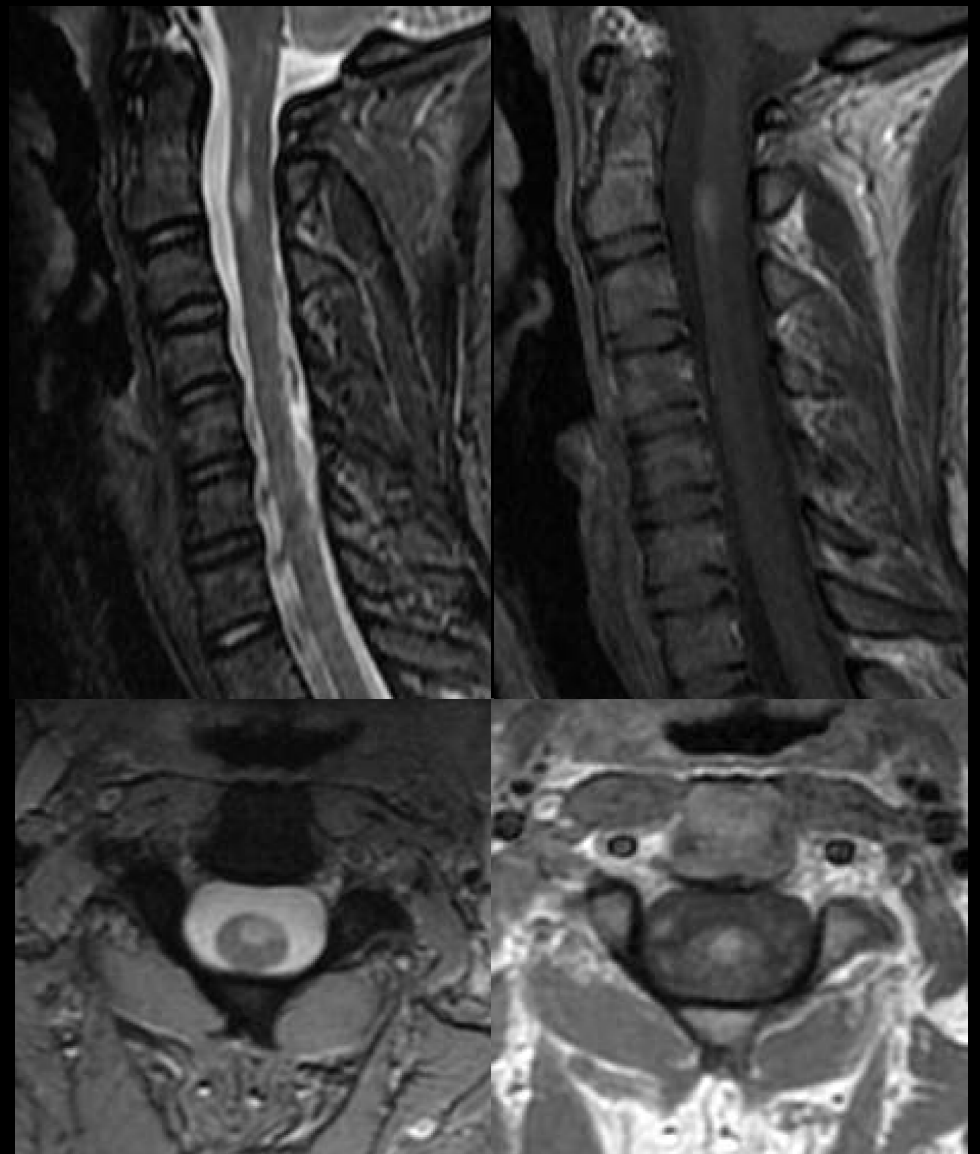
- Dorsal column system
  - Subacute combined degeneration
    - Vitamin B12 deficiency
    - Copper deficiency
  - Nitrous oxide toxicity
    - Recreational drug use
      - “Whippets”
      - Age, history
  - Imaging
    - Dorsal column involvement
    - Cervical/Thoracic cord
    - Inverted “V”, Rabbit ears





Unique Features – Enhancement Pattern

- Enhancement reflect breakdown of the blood-spinal cord barrier
  - Many etiologies
  - Importance
    - Must entertain tumor
- Parenchymal only
  - MS
  - Tumor
  - ADEM
  - Tranverse myelitis
- Leptomeningeal only
  - Meningitis
  - Leptomeningeal tumor spread
- Parenchymal and leptomeningeal enhancement
  - Neurosarcoidosis
  - Hemangioblastoma
  - Vasculitis



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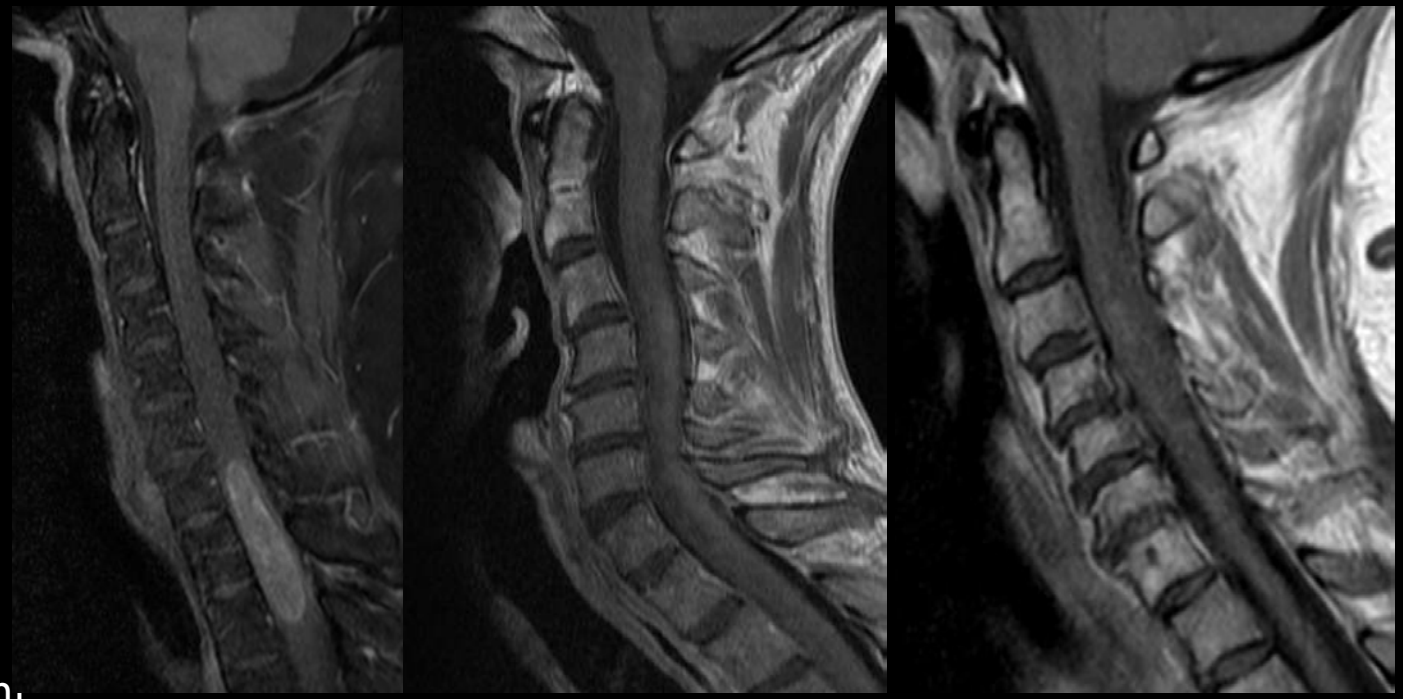

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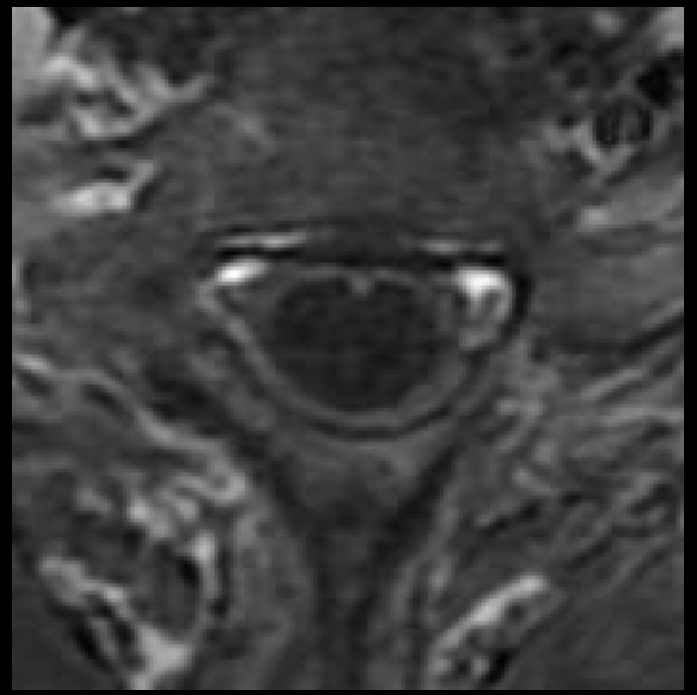

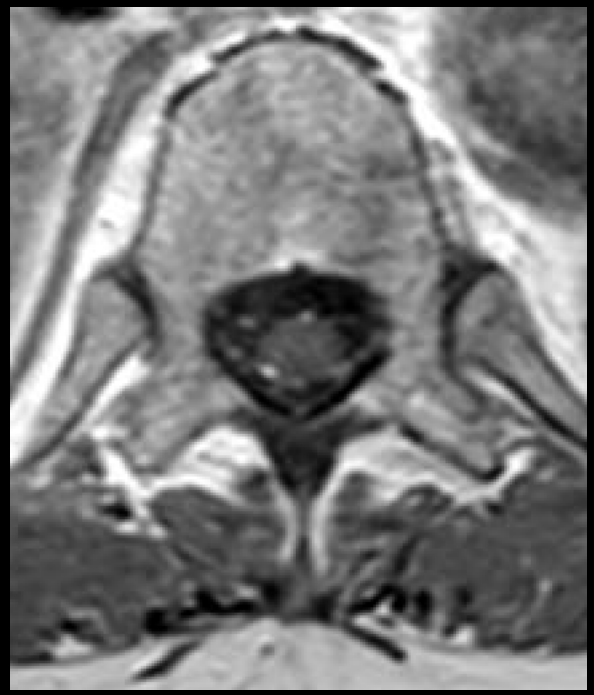

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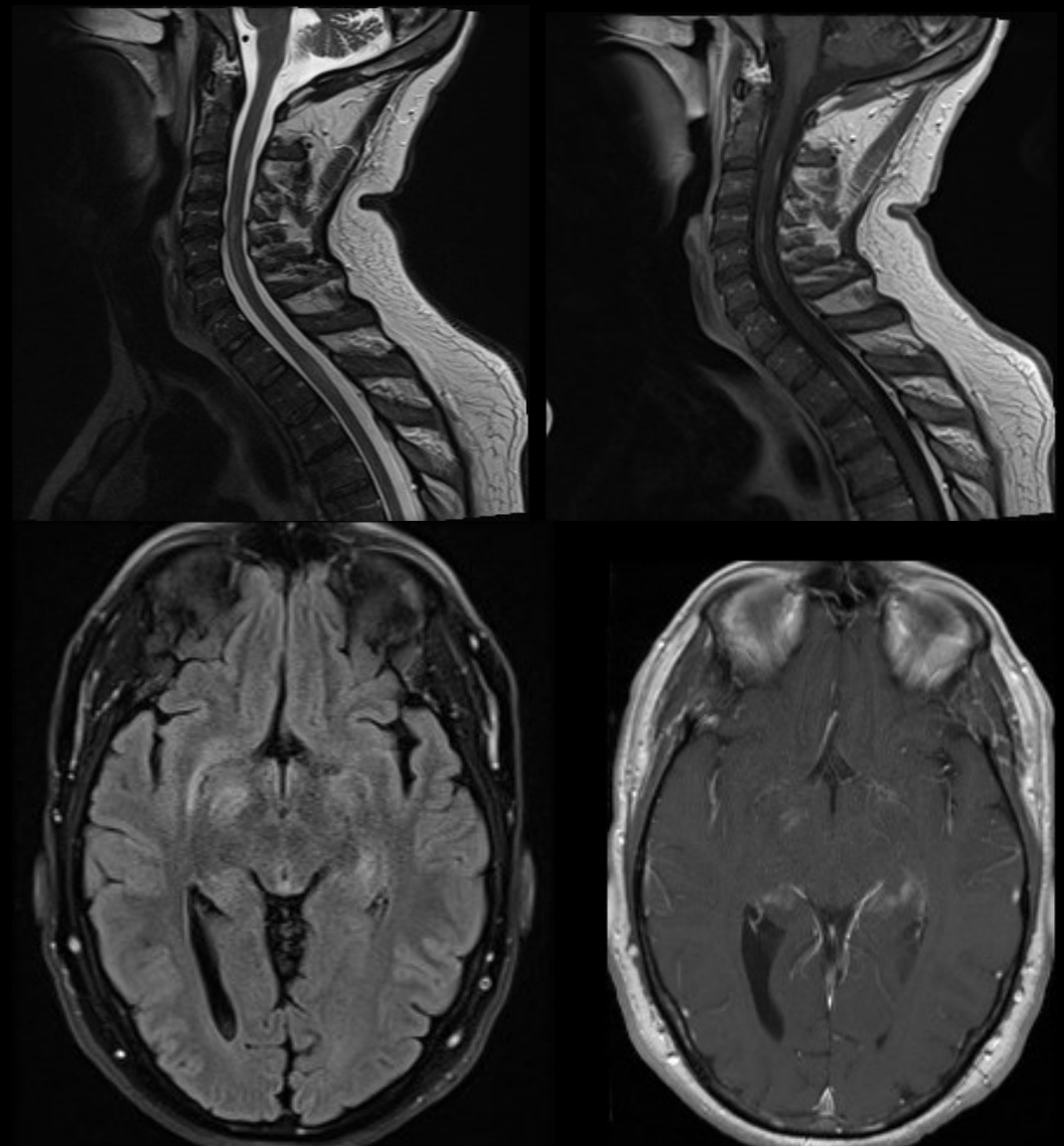

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