Spinal Neurosarcoidosis: A Case Series

Lucas Marks, DO PGY-3

Yang Tang, MD PHD

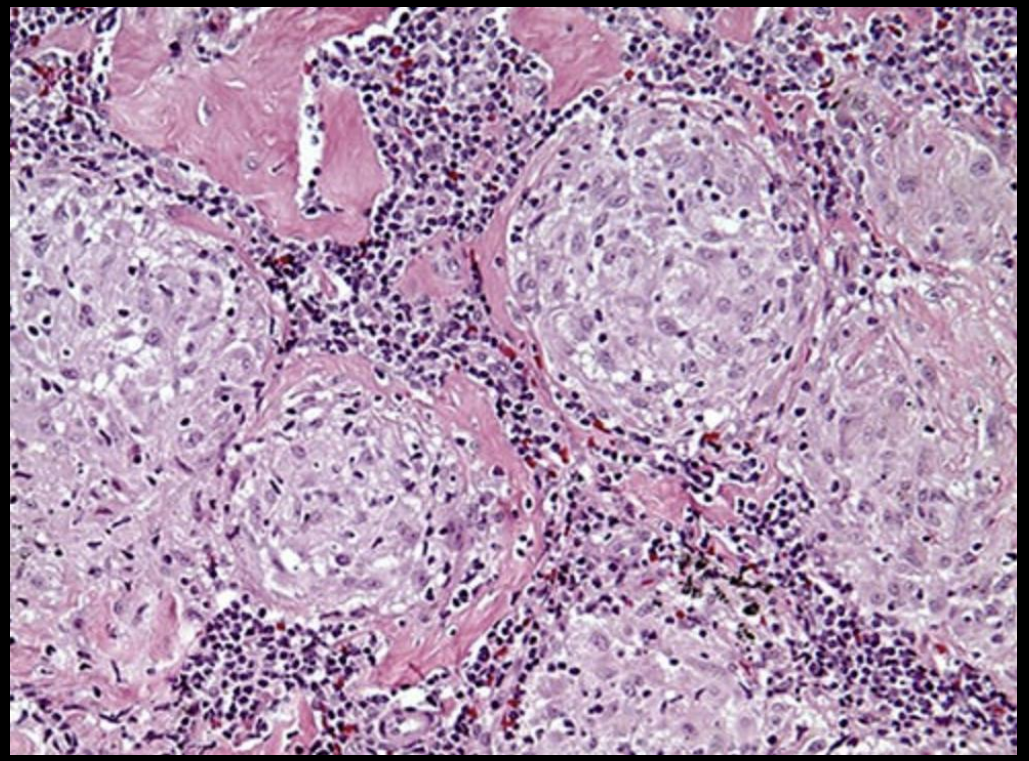
VCU Health

Southeastern Neuroradiological Society

Founded 1977

Intro - Sarcoidosis

- Chronic multisystem disease characterized by the accumulation of noncaseating granulomas.
- Peak incidence individuals 20-39 y/o with highest incidence in African Americans.
- Exact cause still unknown.
- Most commonly involves pulmonary and lymphoid systems.
- Extra-thoracic involvement seen in ~50% of cases.
- Neurologic involvement reported in 5-15% of cases.



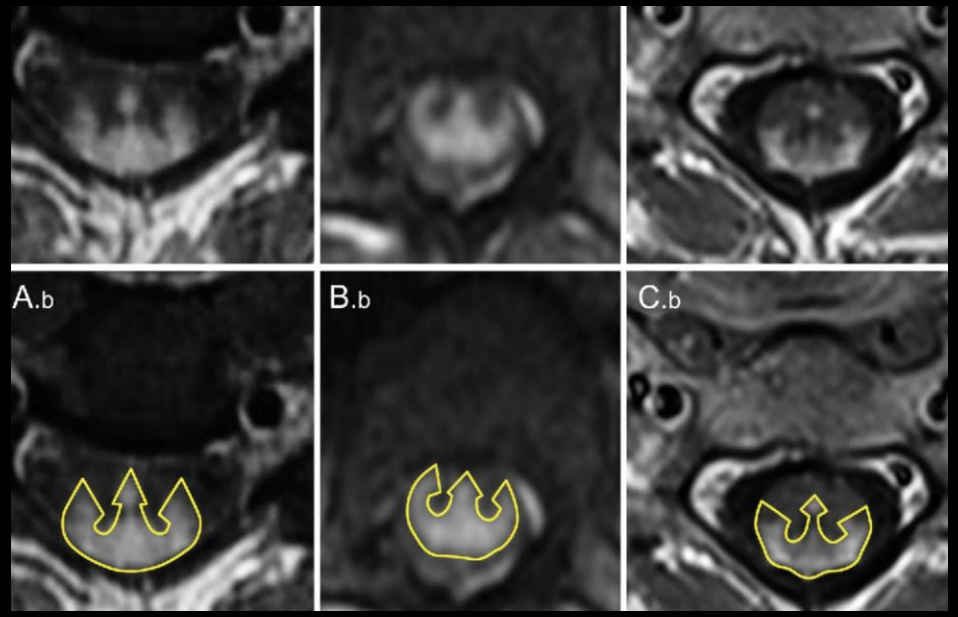
Spinal Neurosarcoidosis

- Only 6-8% of patients with neurosarcoidosis have spinal involvement.
- Spinal involvement may include extradural, intradural-extramedullary, or intramedullary.
- Intramedullary spinal cord involvement seen in <1% of cases.
- Differential dx:
  - Spondylotic myelopathy
  - Demyelinating disease such as Neuromyelitis Optica (NMO)
  - Other inflammatory myelopathies
  - Intradural tumor
  - Dural AVF

Spinal Neurosarcoidosis

- Classic enhancement pattern in intramedullary sarcoidosis:
  - Dorsal-subpial enhancement in combination with central canal enhancement resembling a ’trident’ on axial images.

- Cord enhancement in sarcoid usually present at stenotic spinal level and thus difficult to distinguish from spondylotic myelopathy.
  - Caveats – cord edema spanning multiple levels and cord enhancement in the setting of mild/moderate spinal stenosis -> consider spinal neurosarcoid as possibility
  - Consider further evaluation with chest CT/PET-CT to evaluate for pulmonary/systemic sarcoid

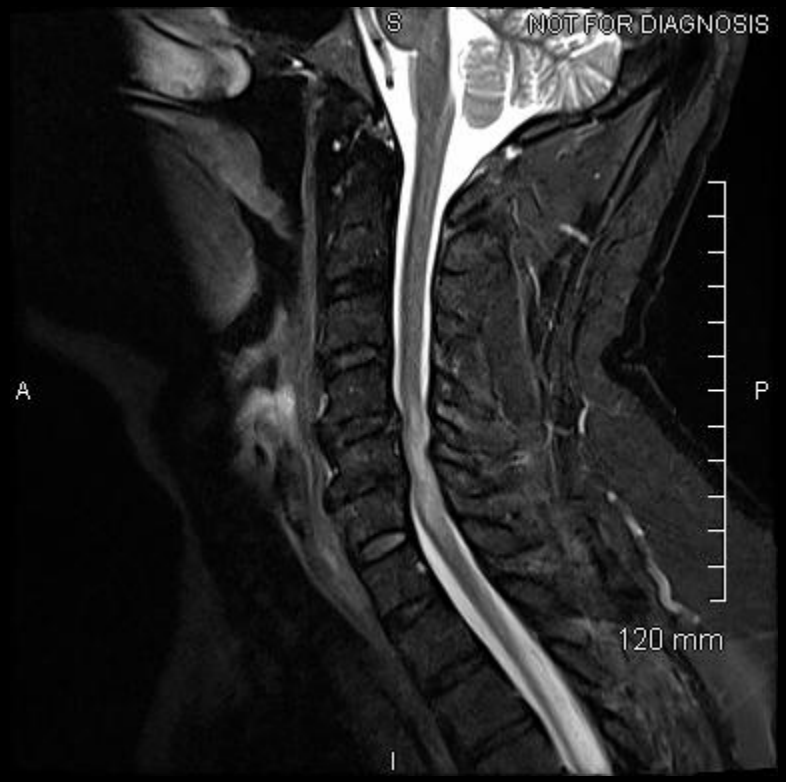


Trident sign

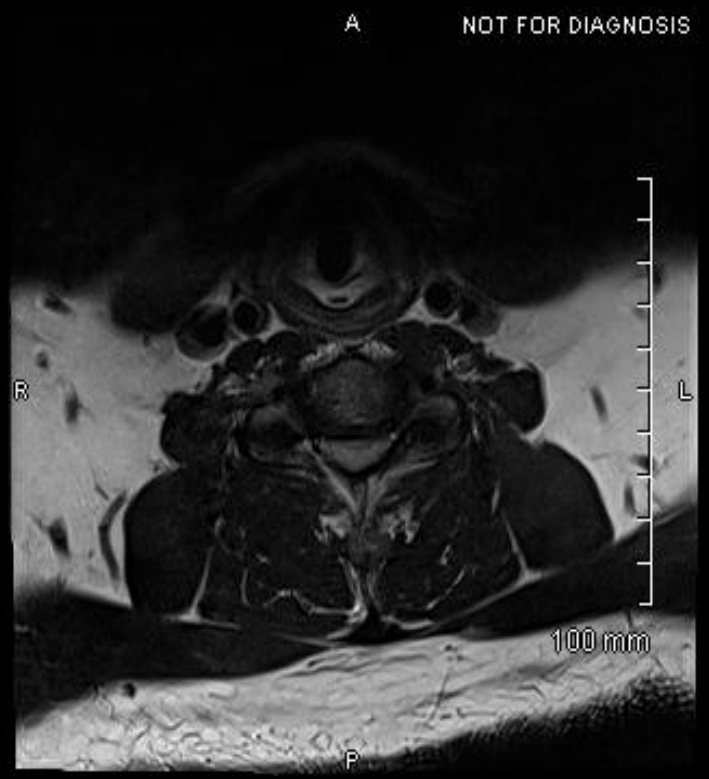
Zalewski, N.L., Krecke, K.N., Weinshenker, B.G., Aksamit, A.J., Conway, B.L., McKeon, A., & Flanagan, E.P. (2016). Central canal enhancement and the trident sign in spinal cord sarcoidosis. *Neurology, 87*, 743 - 744.

Case 1

- 58 y/o M presented with chronic weakness, falls, and upper extremity paresthesias.

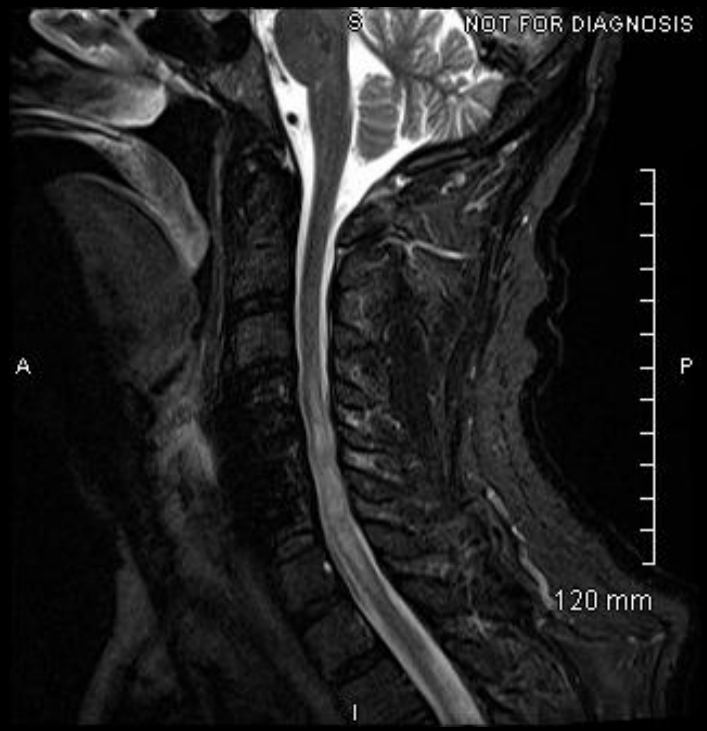


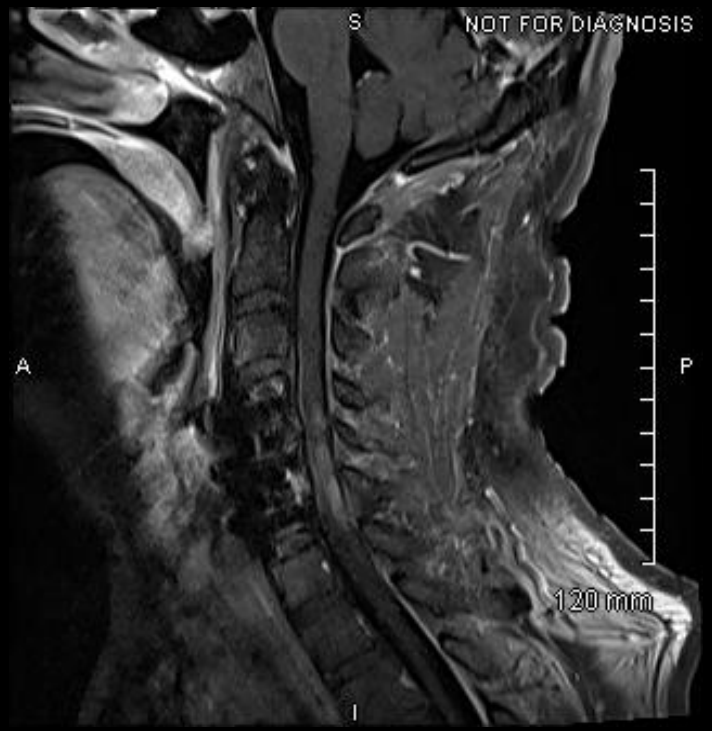
- MRI cervical spine on 5/29/2021
- Disc herniations at C4-5 and C5-6 with moderate to severe spinal stenosis
- Cord edema at C5 and C6
- No contrast was given
- Patient under C4-C6 ACDF 8/2021 without complication

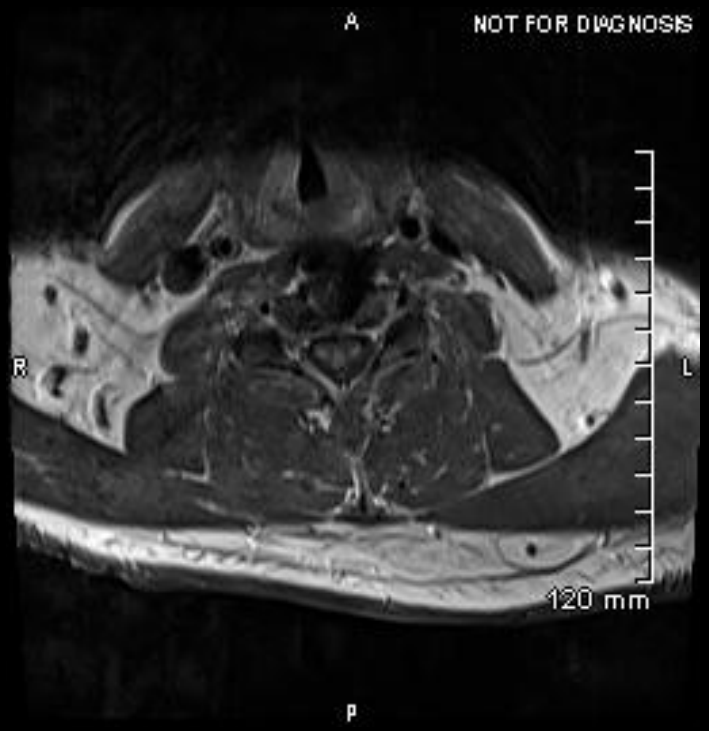


Case 1

- Patient still having progressive upper and lower extremity weakness after ACDF. Underwent cervical and thoracic spine MRI w/w out contrast 12/21/2022



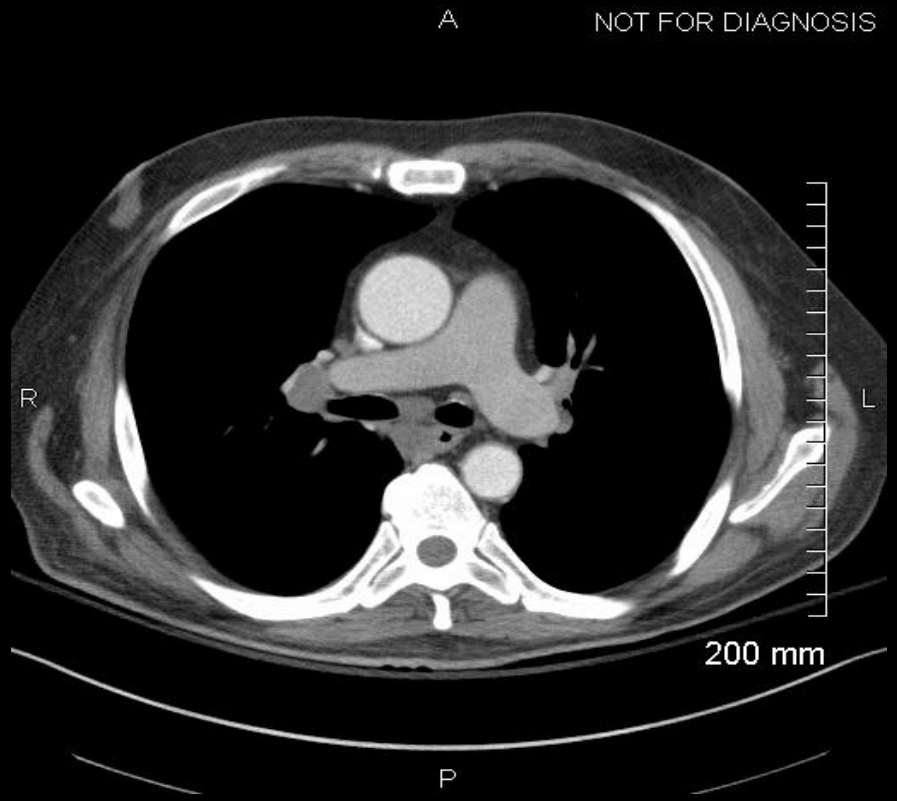


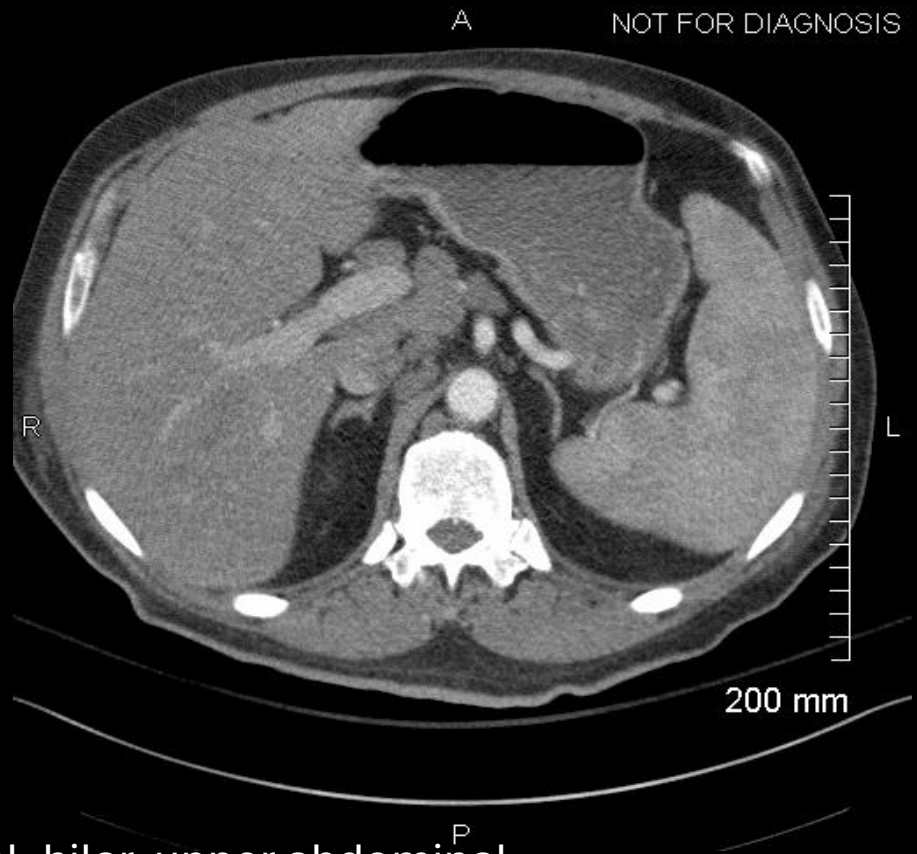


- Resolution of spinal stenosis post ACDF, however cord edema has progressed now extending from C4 to T1.
- Postcontrast sag and axial T1 show subpial enhancement along the lateral and dorsal cord with “trident” configuration.

Case 1

- With findings concerning for spinal neurosarcoidosis, a chest CT with contrast was performed on 12/17/2022.





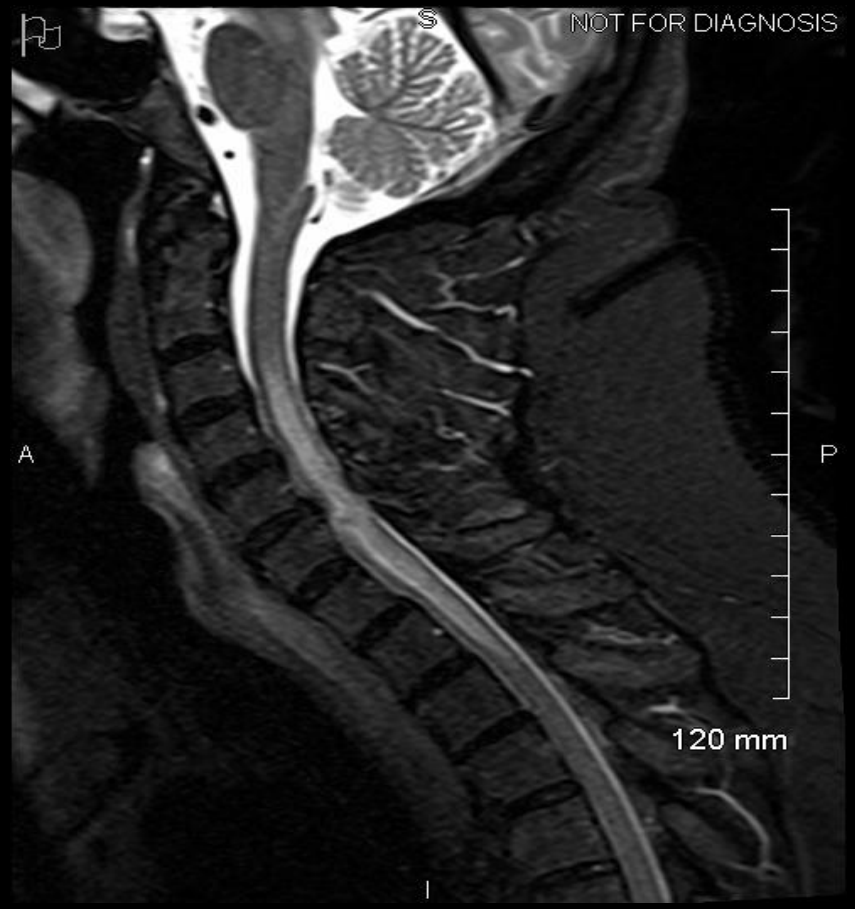
Findings notable for mediastinal, hilar, upper abdominal adenopathy, and splenomegaly.

Case 1

- Brain MRI w/w out IV contrast 12/17/2022 with no significant findings.
- HIV/ANCA/MOG/NMO antibodies all negative
- Lumbar puncture 12/20/2022 with elevated CSF protein
- Trial of high dose IV steroids with significant improvement in neurologic symptoms.
- Underwent endobronchial mediastinal lymph node biopsy 12/23/2022 with pathology showing non-necrotizing granulomas confirming diagnosis of sarcoidosis.

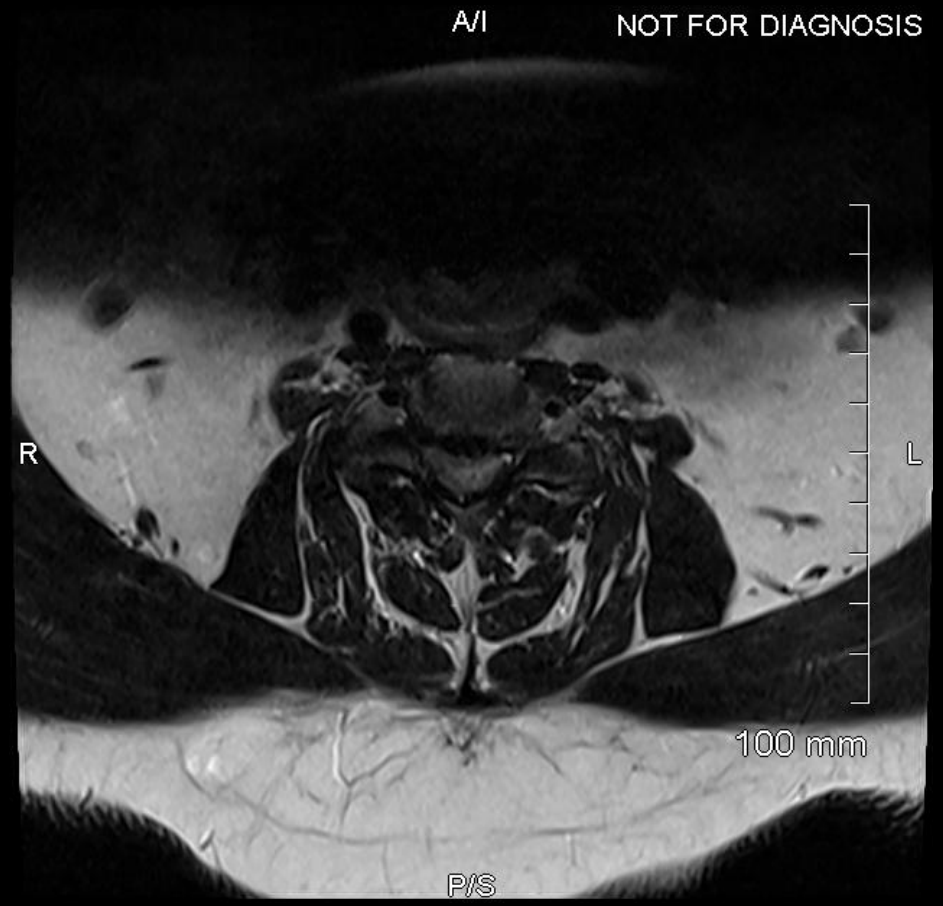
Case 2

- 66 y/o F presented with multiple months of progressive L>R upper extremity weakness as well as difficulty walking.

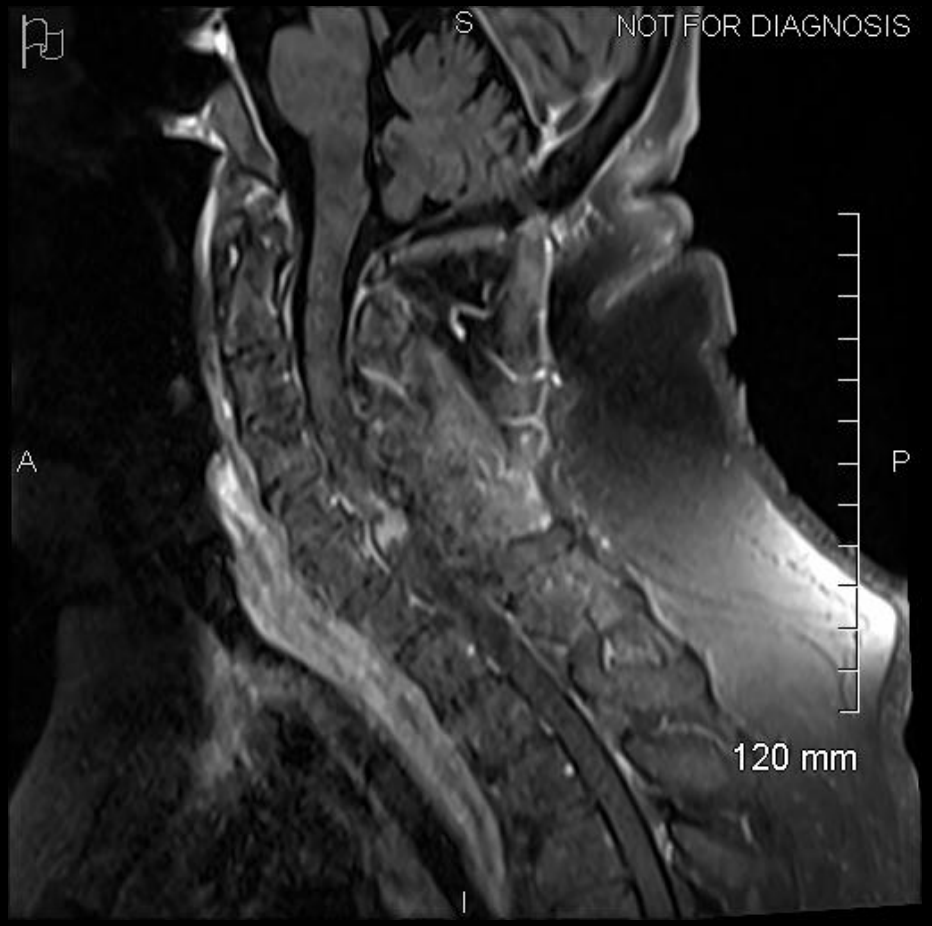


MRI cervical spine w & wo 11/8/2020 demonstrating:

- Extensive cord edema C3-T2
- Mild spinal stenosis C5-C6

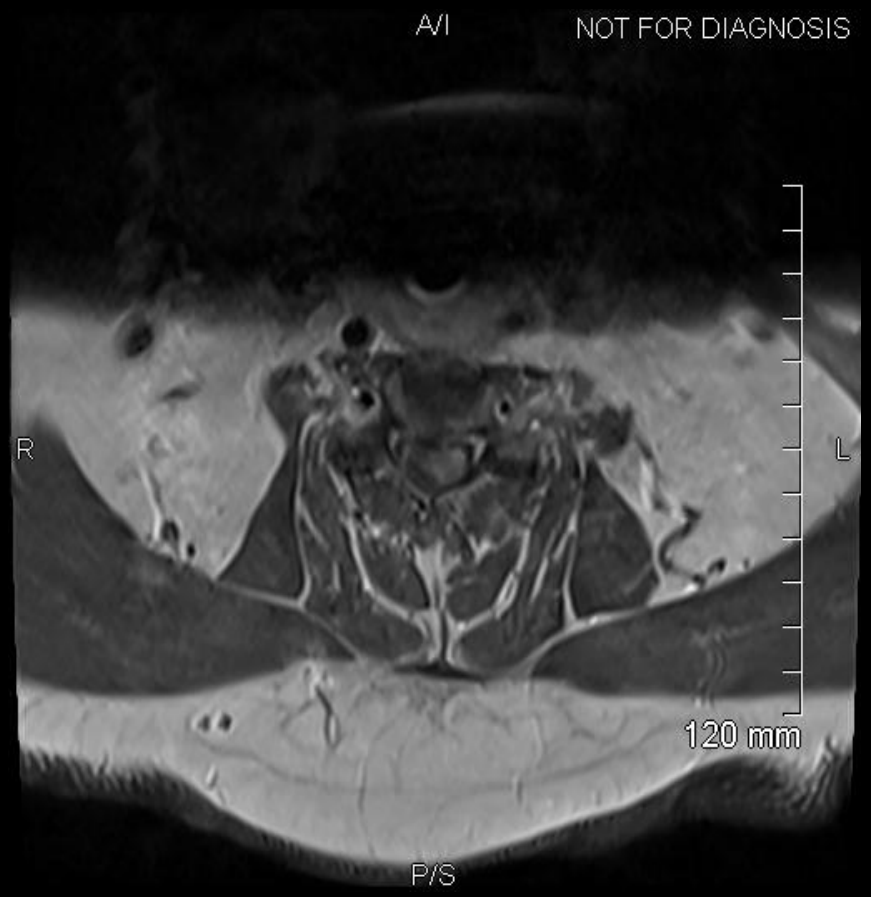


Case 2



MRI cervical Spine w & wo 11/8/2020, post contrast T1 sequence demonstrating:

- Focal heterogenous subpial enhancement primarily at C5-C6

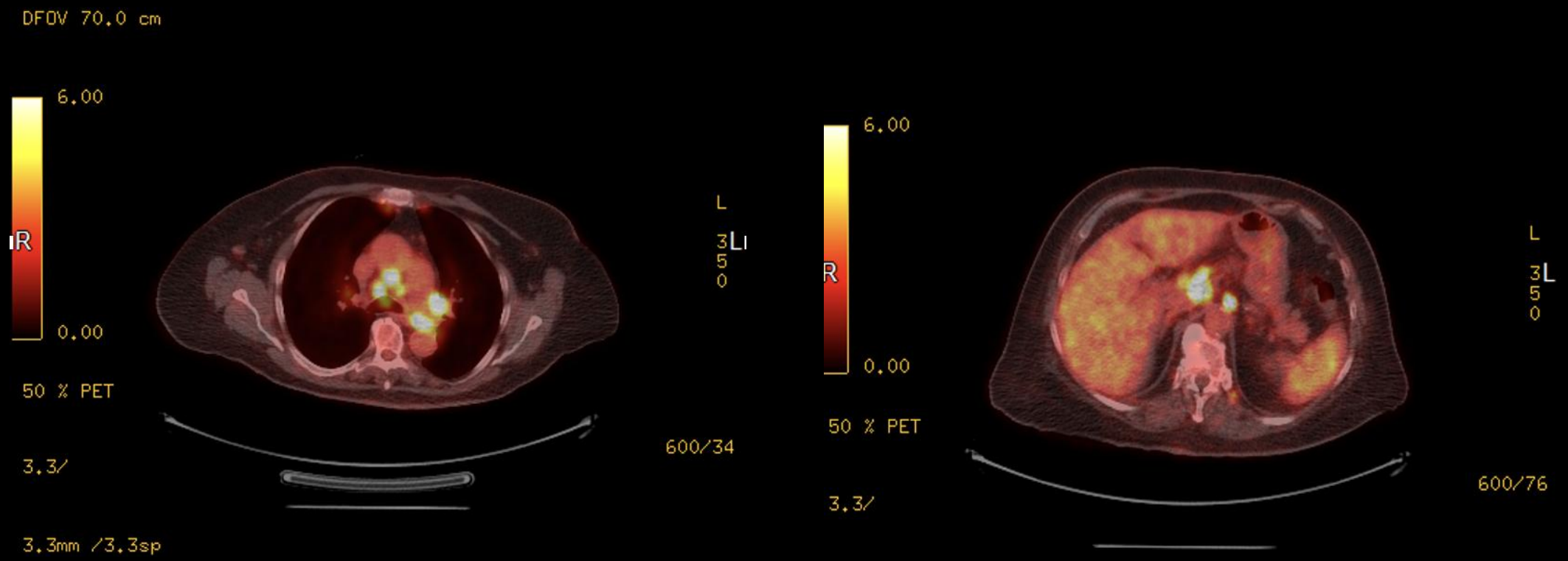


Case 2

- MRI brain, T-spine, and L-spine were negative.
- Underwent lumbar puncture with elevated CSF protein and lymphocytic pleocytosis.
- ANA, NMO antibodies were negative
- Started on high dose steroids with symptoms improvement.
- Patient underwent C5-C6 ACDF on 11/10/2020. Biopsy at C5-C6 with pathology demonstrating neuroglial tissue with granulomatous inflammation confirming diagnosis of intramedullary sarcoidosis.
- Symptoms returned several months later with subsequent imaging showing enlargement of the cord lesion at C5-C6.

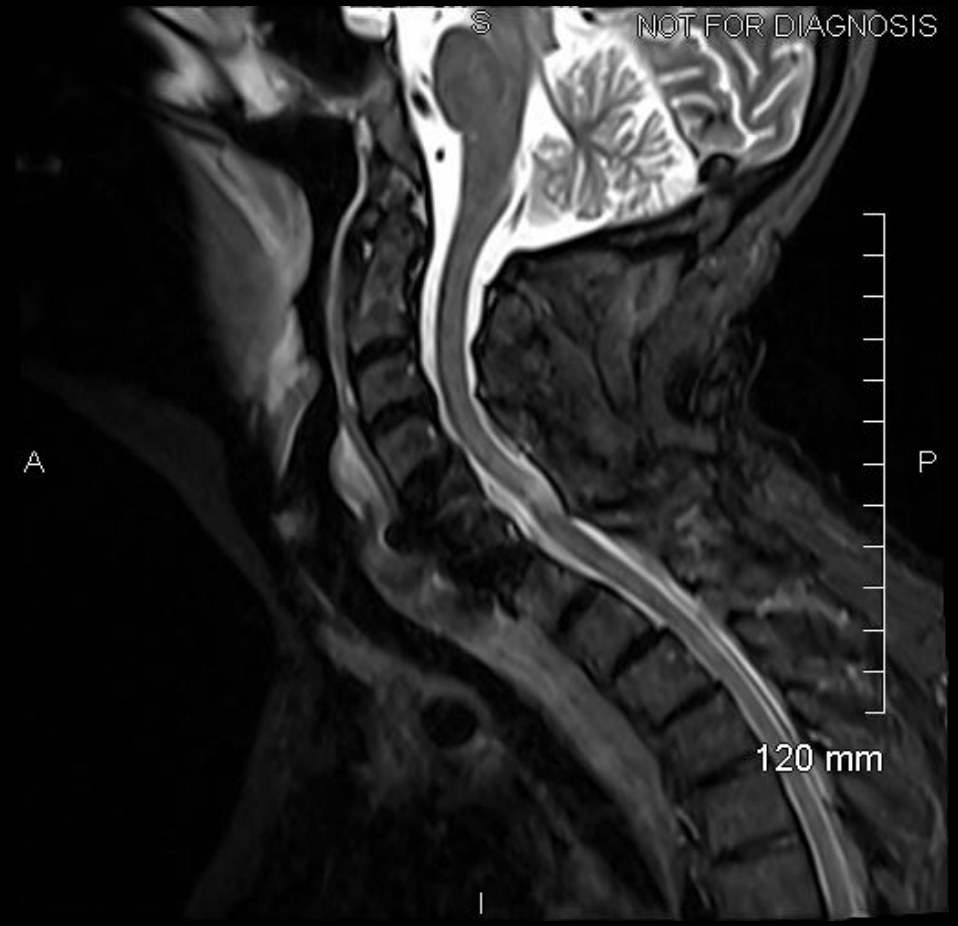
Case 2

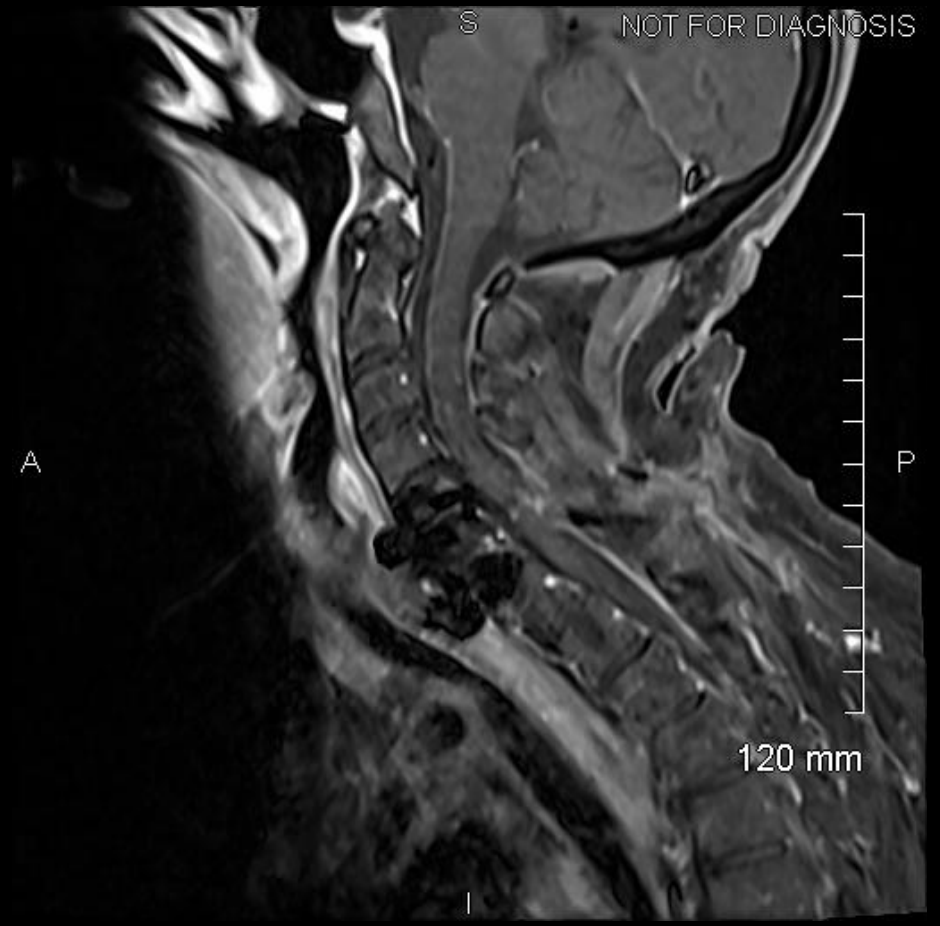
- Cardiac MRI on 11/08/2021 showed findings concerning for cardiac sarcoid involvement.
- PET/CT 6/10/2022 demonstrating multiple hypermetabolic mediastinal, hilar, and upper abdominal lymph nodes further confirming diagnosis.



Case 2

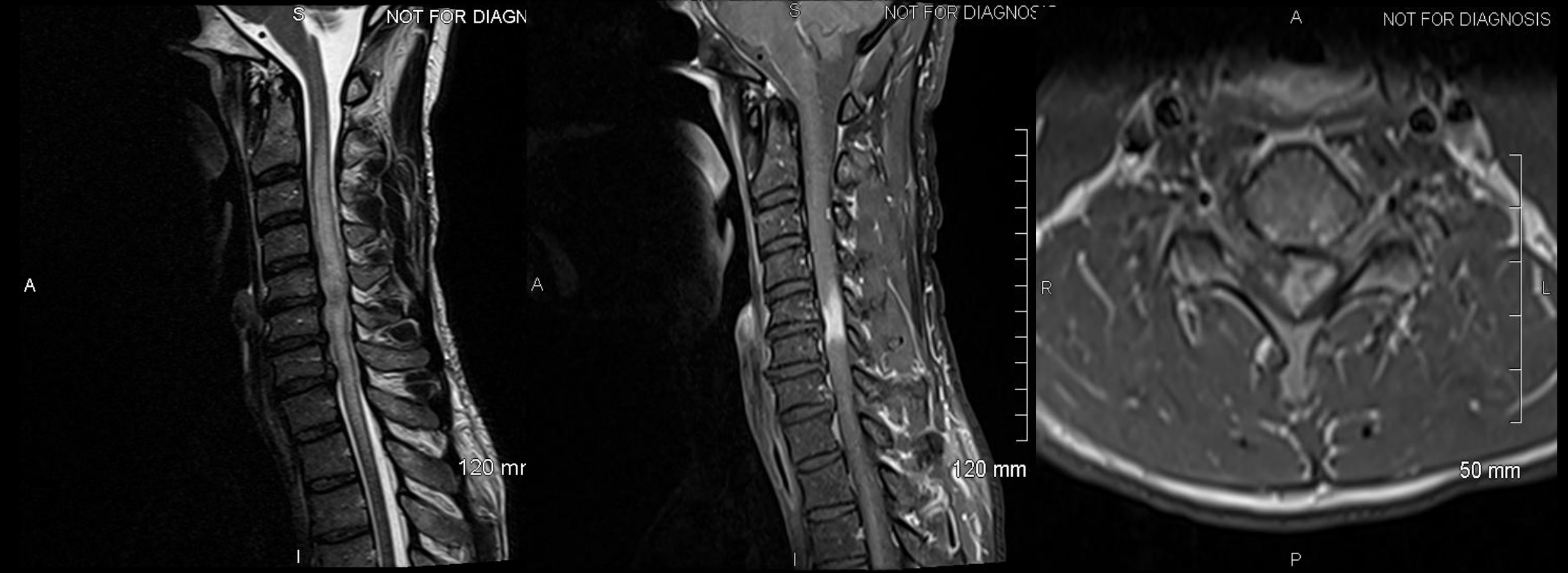
- Patient continued on oral steroids. Recent follow up c-spine MRI 5/28/2025 showing ACDF with resolution of cord edema and minimal residual enhancement at C5-C6.





Case 3

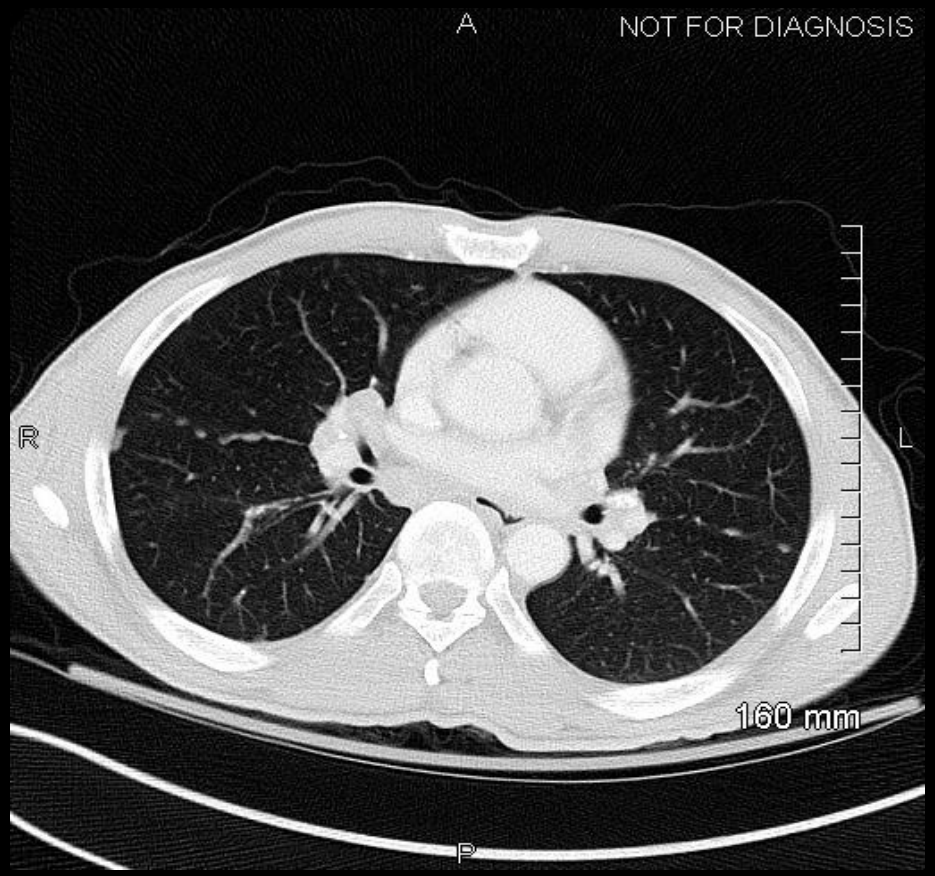
- 53 y/o M presented with 2 weeks of rapidly progressive lower extremity weakness

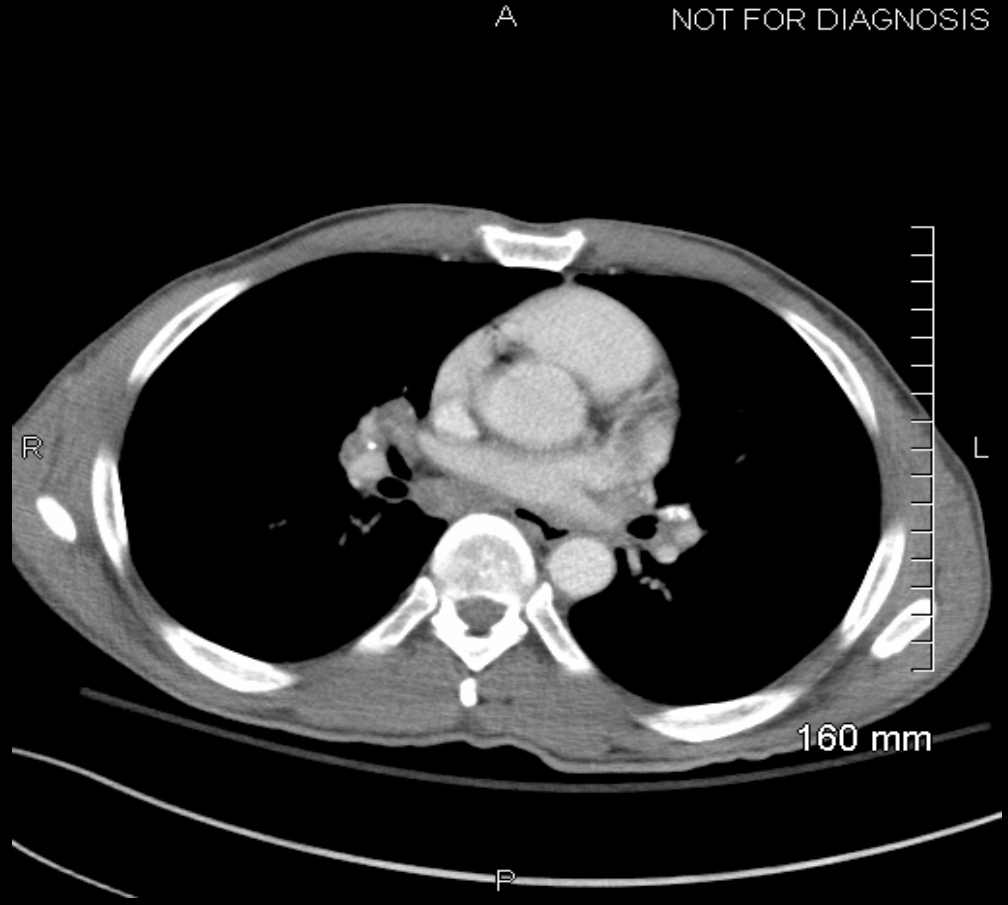


MRI C-spine w & w/out contrast 4/4/2021 demonstrates longitudinally extensive cord edema and cord expansion from C2-T2, mild spinal stenosis with intense subpial enhancement of the cord at C5-C6.

Case 3

- Chest CT 4/5/2021 was ordered to evaluate for possible infection vs malignancy.





Chest CT reveals perilymphatic nodules along the fissures as well as calcified mediastinal lymph nodes. Subsequent PET/CT demonstrated low grade FDG uptake within these mediastinal nodes.

Case 3

- The patient underwent LP with CSF studies demonstrating elevated protein with a lymphocytic predominant pleocytosis. All CSF infectious markers and cultures negative. CSF and serum NMO, serum MOG, and CSF oligoclonal bands were all negative.
- CSF angiotensin converting enzyme (ACE) elevated, in combination with imaging findings in the cervical spine and chest, suggestive of sarcoidosis
- Patient treated with high dose steroids with rapid improvement in symptoms and cord edema.

Take home points

- Intramedullary spinal neurosarcoidosis is a relatively rare disease process, which can be hard to make the diagnosis based on imaging findings alone.
- If longitudinal extensive cord edema and focal enhancement is centered as the level of mild/moderate spinal stenosis, consider neurosarcoidosis as a differential diagnosis.
- The ”trident” sign is a relatively nonspecific MRI finding, however can suggest spinal neurosarcoidosis if seen.

References

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