

AMSER Case of the Month

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58-year-old male presenting with worsening upper extremity weakness, decreased sensation, tremors, and ataxia

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Patient Presentation

58-year-old male with past medical history of **Type 2 Diabetes Mellitus, Schizophrenia**

Presented to ED with nausea, vomiting, and fever with concern for sepsis related to GI infection

- Later in hospital stay...
 - Develops worsening ataxia and weakness in bilateral upper extremities. Acute on chronic weakness and acute on chronic ataxia noted on prior notes
- **Physical exam:**
 - **Neuro:**
 - **Sensation:** Absent sensation to light touch, pinprick, and proprioception in bilateral lower extremities; proprioception impaired at wrist.
 - **Motor:** Normal bulk; tone decreased in all four extremities. Large amplitude postural tremor. Mild bradykinesia with left > right. Pronator drift absent in bilateral upper extremities. Wrist flexion 4+ in bilateral upper extremities, wrist slips in bilateral upper extremities
 - **Coordination:** Intention tremor present bilaterally, ataxia on heel to shin bilateral

Pertinent Labs

- Basic Metabolic Panel (BMP) has no abnormalities
- Complete Blood Count (CBC) has no abnormalities
- Vitamin B1/B12/E are within normal limits
- Syphilis and HIV Screens are negative
- Copper screen negative

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

- Indication: Acute on chronic ataxia and weakness, prior imaging studies from over 10 years ago

Scenario	Scenario Id	Procedure	Adult RRL	Peds RRL	Appropriateness Category	
Ataxia, nontraumatic, cervical and thoracic spine pathology suspected	3148775	MRI cervical and thoracic spine without IV contrast	0 mSv O	0 mSv [ped] O	Usually appropriate	●
		MRI cervical and thoracic spine without and with IV contrast	0 mSv O	0 mSv [ped] O	Usually appropriate	●
		Arteriography cervical and thoracic spine	10-30 mSv ⊕⊕⊕⊕	Not Assigned	May be appropriate	●
		MRA cervical and thoracic spine with IV contrast	0 mSv O	0 mSv [ped] O	May be appropriate	●
		CT cervical and thoracic spine with IV contrast	10-30 mSv ⊕⊕⊕⊕	Not Assigned	May be appropriate	●
		MRA cervical and thoracic spine without IV contrast	0 mSv O	0 mSv [ped] O	May be appropriate	●
		CT cervical and thoracic spine without IV contrast	10-30 mSv ⊕⊕⊕⊕	Not Assigned	May be appropriate	●
		CT myelography cervical and thoracic spine	10-30 mSv ⊕⊕⊕⊕	Not Assigned	May be appropriate	●
		CTA cervical and thoracic spine with IV contrast	10-30 mSv ⊕⊕⊕⊕	Not Assigned	May be appropriate	●
		Radiography cervical and thoracic spine	1-10 mSv ⊕⊕⊕	Not Assigned	Usually not appropriate	●
		CT cervical and thoracic spine without and with IV contrast	10-30 mSv ⊕⊕⊕⊕	Not Assigned	Usually not appropriate	●

This imaging modality was ordered by the neurology team



Select the applicable ACR Appropriateness Criteria

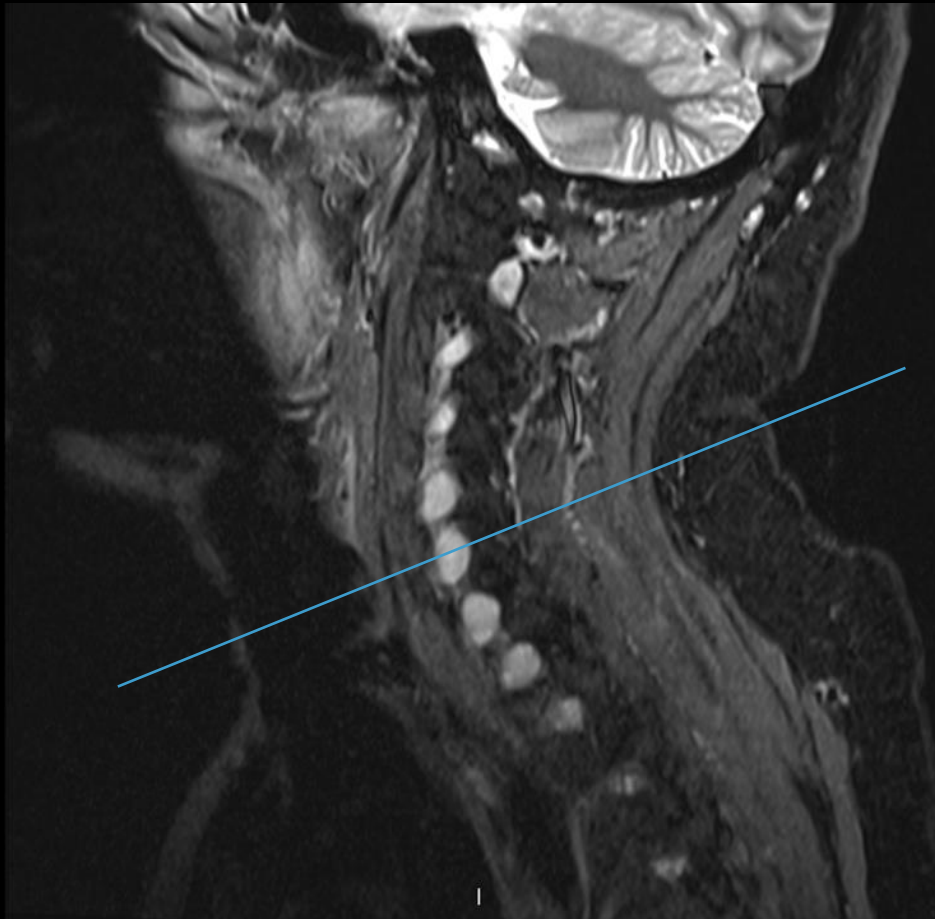
Scenario	Scenario Id	Procedure	Adult RRL	Peds RRL	Appropriateness Category	
Ataxia, nontraumatic, not acute stroke, intracranial process suspected	3149323	MRI head without IV contrast	0 mSv ○	0 mSv [ped] ○	Usually appropriate	●
		MRI head without and with IV contrast	0 mSv ○	0 mSv [ped] ○	Usually appropriate	●
		CT head with IV contrast	1-10 mSv ⊕⊕⊕	0.3-3 mSv [ped]..	May be appropriate	●
		CT head without IV contrast	1-10 mSv ⊕⊕⊕	0.3-3 mSv [ped]..	May be appropriate	●
		CT head without and with IV contrast	1-10 mSv ⊕⊕⊕	3-10 mSv [ped]..	May be appropriate	●
		Arteriography cervicocerebral	1-10 mSv ⊕⊕⊕	3-10 mSv [ped]..	Usually not appropriate	●
		MRA head and neck without IV contrast	0 mSv ○	0 mSv [ped] ○	Usually not appropriate	●
		MRA head and neck without and with IV contrast	0 mSv ○	0 mSv [ped] ○	Usually not appropriate	●
MRV head with IV contrast		0 mSv ○	0 mSv [ped] ○	Usually not appropriate	●	

This imaging modality was ordered by the neurology team

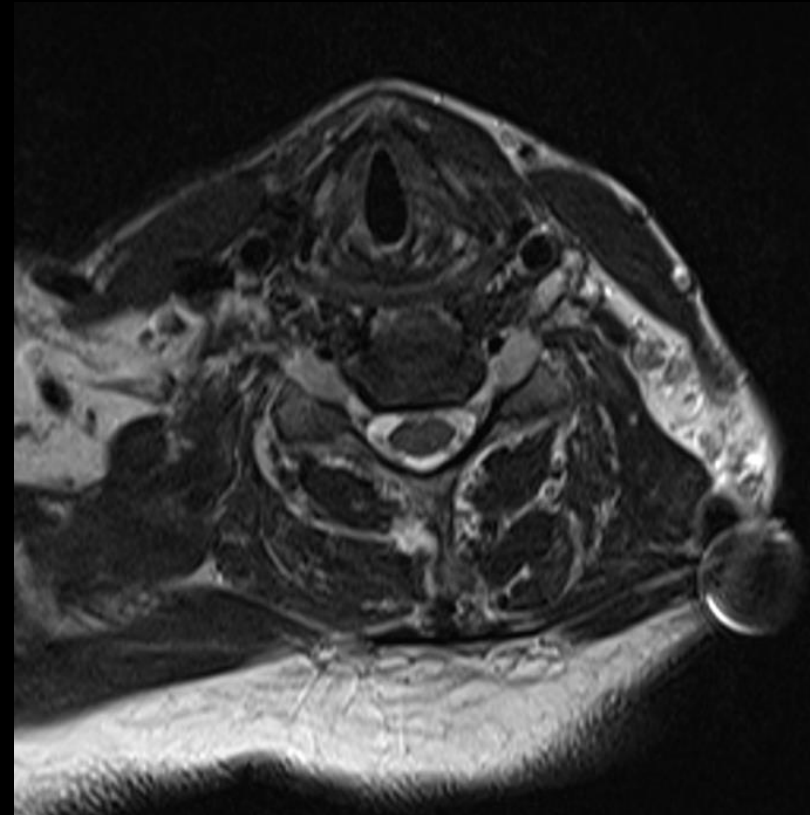


Findings (unlabeled)

- STIR Sagittal



- T2 Axial



Findings (unlabeled)

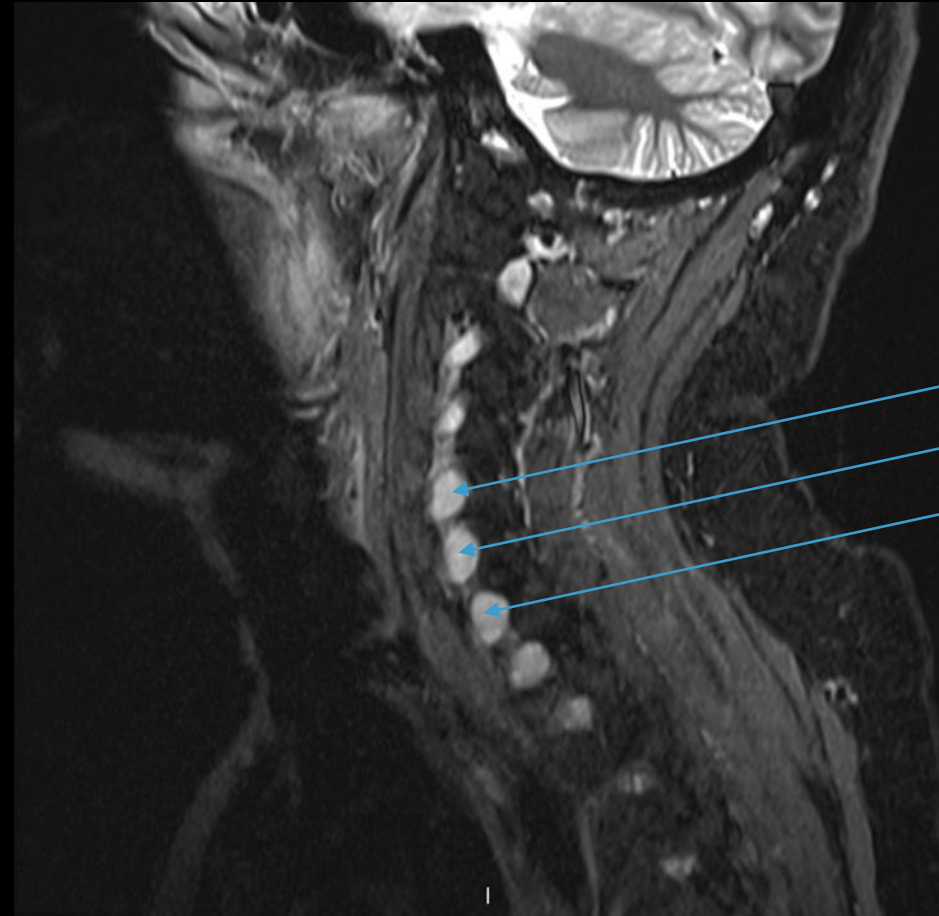
- T2 Coronal



- T2 Axial

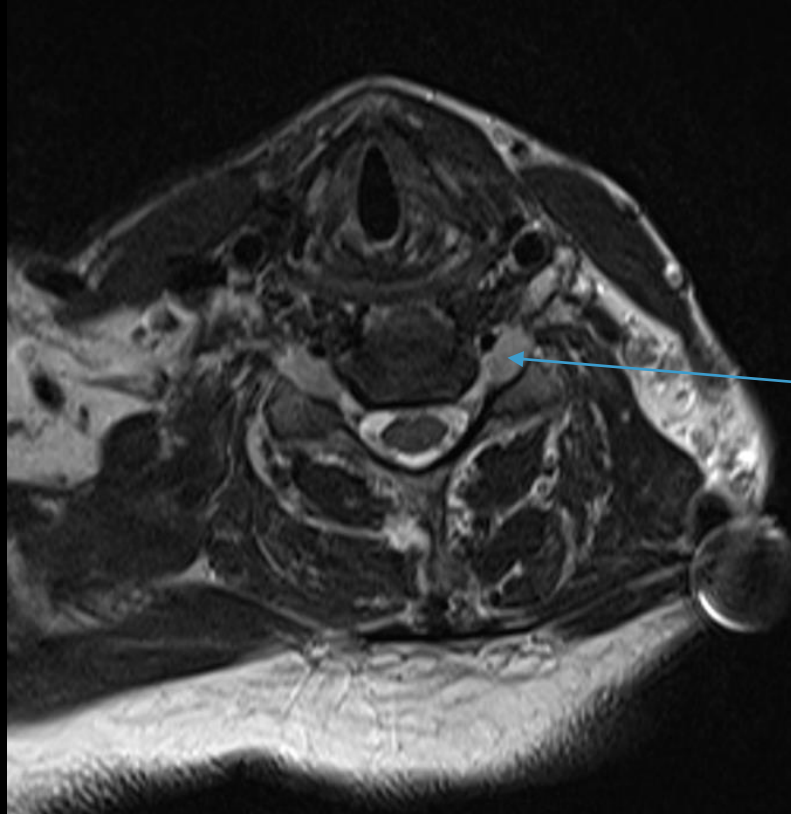


Findings: (labeled)



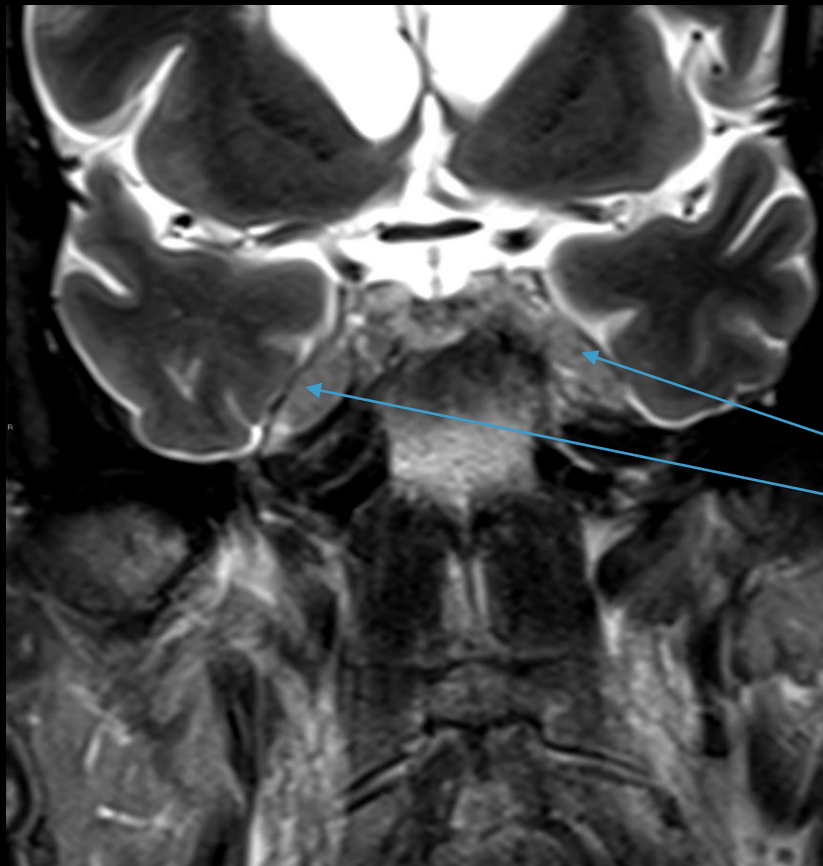
Diffuse thickening of the cervical nerve roots (C4-C6 highlighted)

Findings: (labeled)

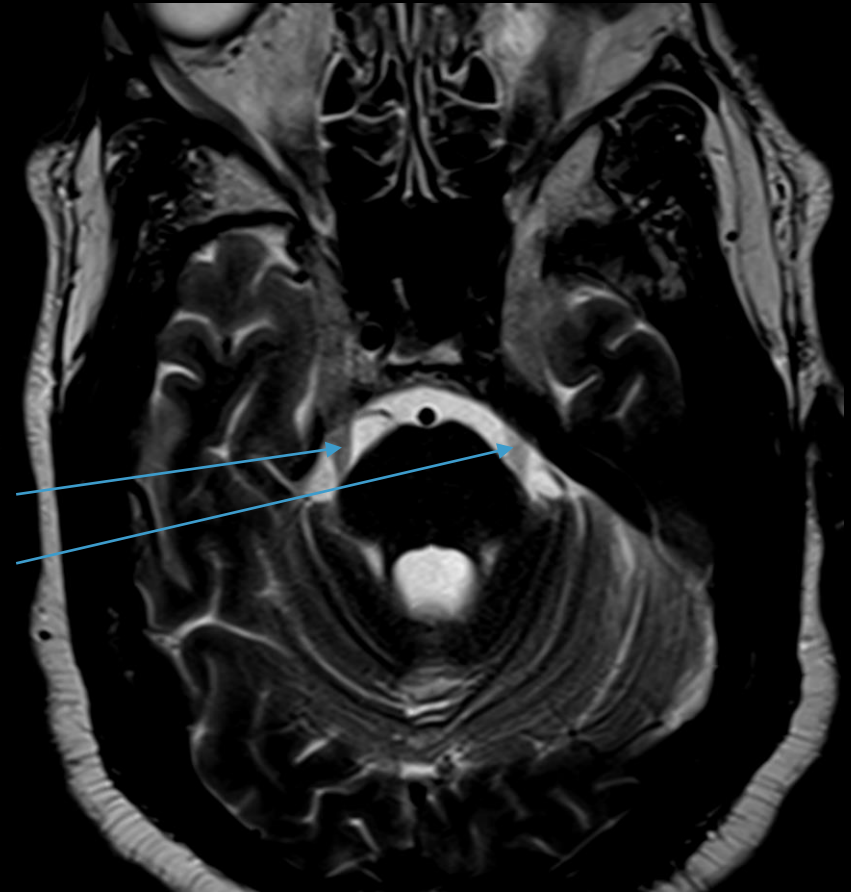


Redemonstrated thickening of bilateral cervical nerve roots, with normal spinal cord signal

Findings: (labeled)



Diffuse thickening of the trigeminal nerve divisions, especially V2 and V3. Other cranial nerves show no apparent thickening



Final Dx:

Chronic inflammatory demyelinating
polyneuropathy (CIDP)

Case Discussion

- Further history clarification:
 - Patient did not report initial history on first neuro team consult – only after further discussion and outside chart review it was discovered that he did carry this diagnosis from the 1990s and had even received prior IVIG treatment 20 years ago!

Case Discussion (2)

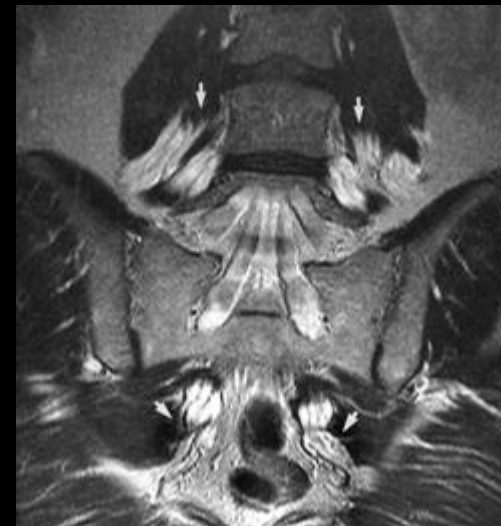
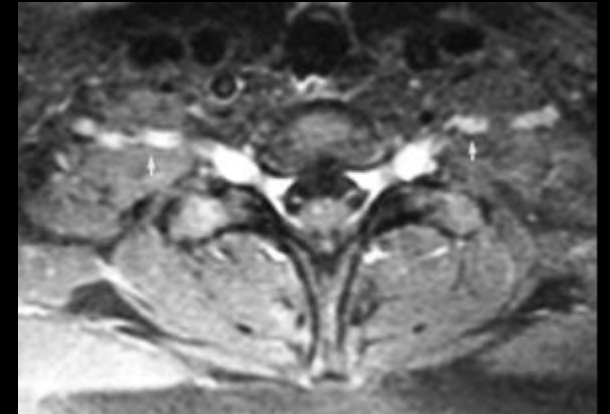
- Pathogenesis of CIDP¹
 - Two distinct mechanisms
 - Macrophage-induced demyelination via phagocytosis, due to deposition of autoantibody
 - IgG4 antibodies, such as anti-neurofascin 155 and anti-contactin 1 antibodies, against components at nodal and paranodal junctions implicated; **patients with these antibodies show characteristic clinical features, such as sensory ataxia and tremor [such as seen in our patient]**
 - Pathology: repeated inflammation results in proliferation of Schwann cells, deposition of collagen leading to thickening, and development of onion bulb appearance²
 - Types
 - Typical CIDP
 - Atypical CIDP
 - Multifocal acquired demyelinating sensory and motor (MADSAM), distal acquired demyelinating symmetric (DADS), pure sensory, pure motor, and focal subtypes

Case Discussion (3)

- Imaging Findings

- MRI: STIR/T1/T2 will show marked thickening of spinal nerve roots and peripheral nerves, lumbar and brachial plexuses, and can sometimes involve cranial nerves (particularly the trigeminal nerves)³
- Lesions can also enhance in the acute to subacute phase, with increase in peak signal intensity⁴
- Supplied muscles can also demonstrate atrophy

T1 (Gd) study shows enhancement of brachial plexus



T2 sequence demonstrates thickened spinal roots in the lower spinal canal

References:

1. Koike H, Katsuno M. Pathophysiology of Chronic Inflammatory Demyelinating Polyneuropathy: Insights into Classification and Therapeutic Strategy. *Neurol Ther.* 2020;9(2):213-227. doi:10.1007/s40120-020-00190-8
2. Louis E, Mayer S, Noble J. Merritt's Neurology. Lippincott Williams & Wilkins. (2010) ISBN:0781791863.
3. Abe Y, Terashima H, Hoshino H, et al. Characteristic MRI features of chronic inflammatory demyelinating polyradiculoneuropathy. *Brain Dev.* 2015;37(9):894-896. doi:10.1016/j.braindev.2015.01.006
4. Tanaka K, Mori N, Yokota Y, et al. MRI of the cervical nerve roots in the diagnosis of chronic inflammatory demyelinating polyradiculoneuropathy: a single-institution, retrospective case–control study. *BMJ Open* 2013;3:e003443. doi: 10.1136/bmjopen-2013-003443