AMSER Case of the Month June 2024

8-year-old presenting with progressive bilateral upper and lower extremity weakness.

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UNIVERSITY of VIRGINIA HEALTH SYSTEM

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Patient Presentation

- **HPI**: Previously healthy 8 YO presents with 4 days of bilateral upper extremity pain and weakness. Also endorsed daily headaches and one episodes of emesis during this time. Recent URI 2-3 weeks ago, which had since resolved.
  - Chart review notable for earlier presentation 2 days prior for bilateral lower extremity weakness but was discharged as symptoms had resolved.
- **Hospital Course**: Imaging initially deferred due to localization favoring peripheral nerve distribution; however, weakness continued to worsen and no longer fit prior characterization.

Patient Presentation

- Physical exam notable for significant bilateral, symmetric weakness of elbow extensors, wrist extensors, finger flexor/extensors that is worse in distal muscle groups. There is also mild bilateral, symmetric weakness of hip and knee flexors. Remainder of neurological exam was otherwise normal.

Pertinent Labs

- BMP and CBC reveal no abnormalities
- Enterovirus/rhinovirus **positive**
- CRP normal, but ESR **mildly elevated at 27** (normal range 0-12)
- B12, Lead, TSH within normal limits
- Autoimmune workup was ultimately negative (NMO, MOG, MGMR)
- Lumbar puncture deferred based on family's wishes

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

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

This imaging modality was ordered by neurology team.

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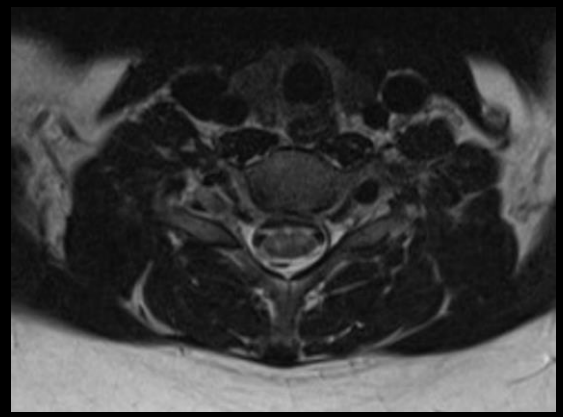
Findings: (unlabeled)

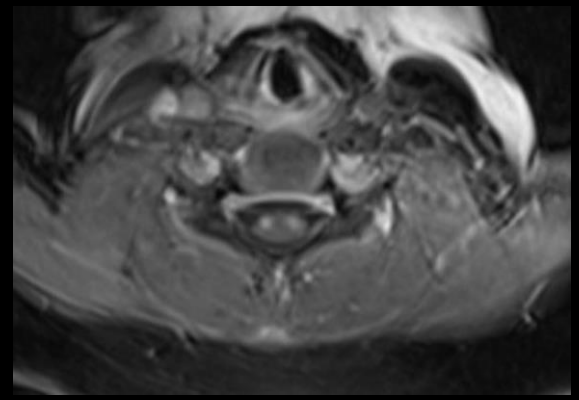
T2-STIR

T1 Post-Contrast

T2

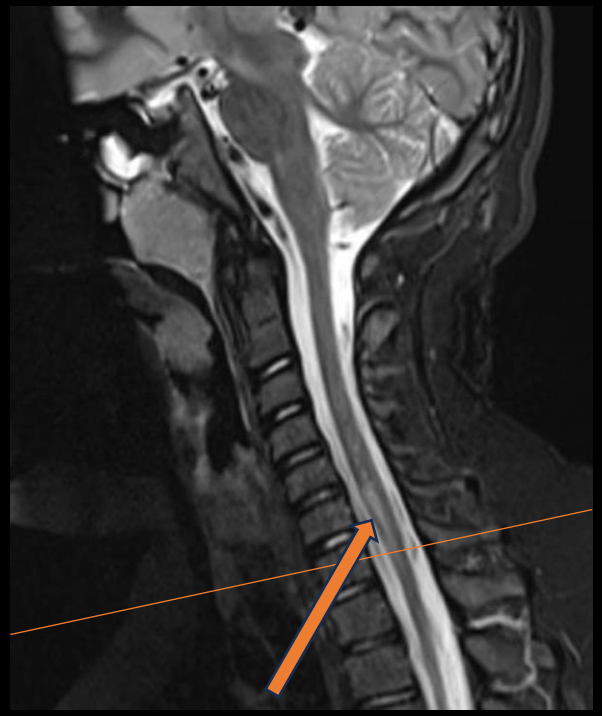


T1 Post-Contrast



Findings: (labeled)

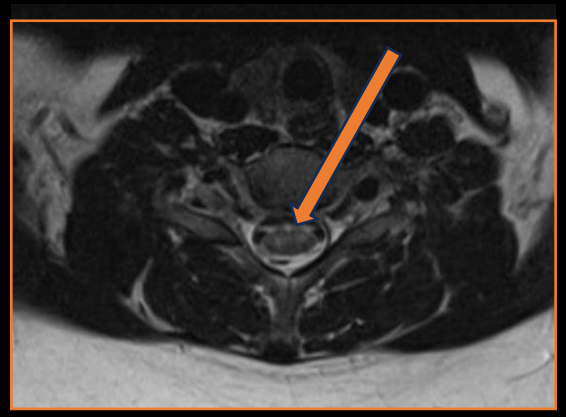
T2-STIR

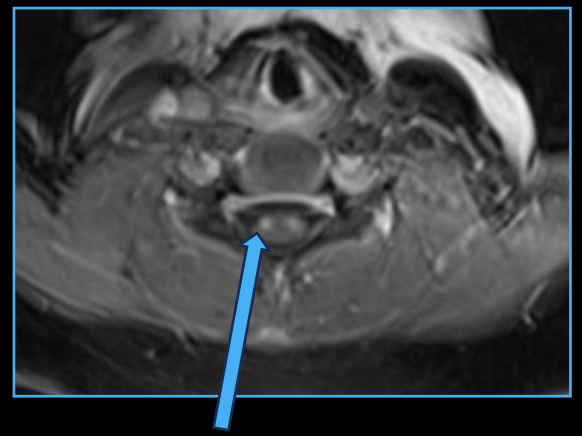


T1 Post-Contrast

Sagittal STIR sequence demonstrates a longitudinal signal hyperintensity in the cervical spinal cord, from the level of C5 to C7.

T2 signal hyperintensity predominately involves spinal cord grey matter ventrally.

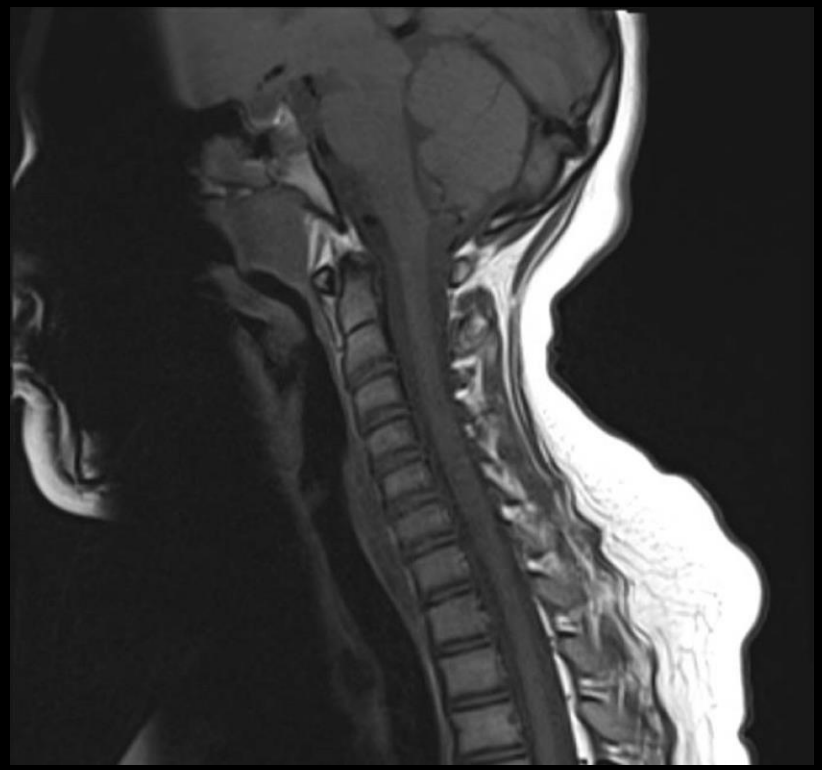




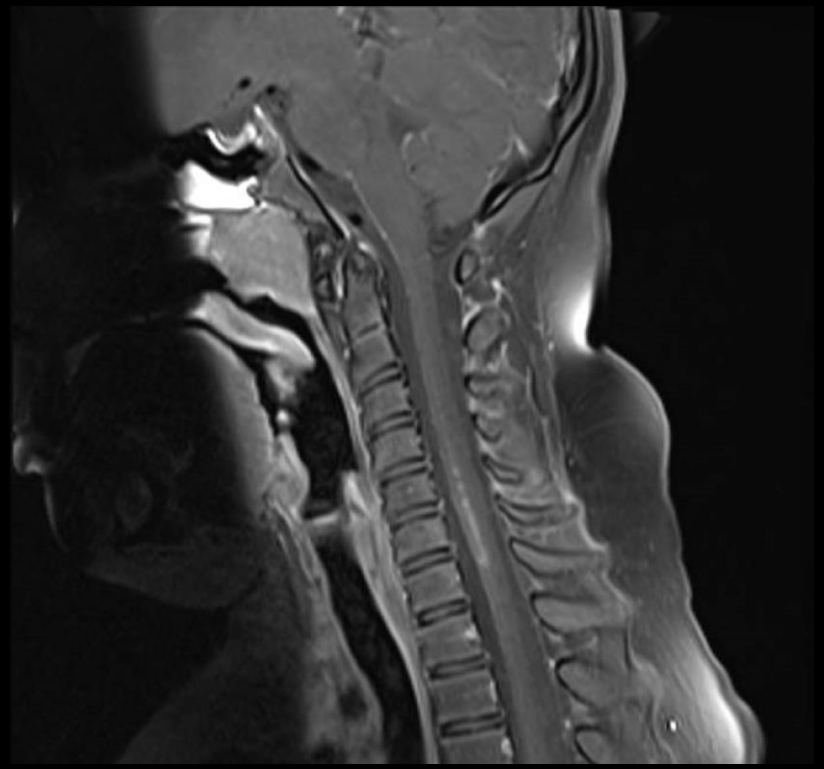
Axial T1 image at the level of lesion shows focal contrast enhancement at bilateral anterior horns.

Findings (unlabeled)

T1 Pre-Contrast

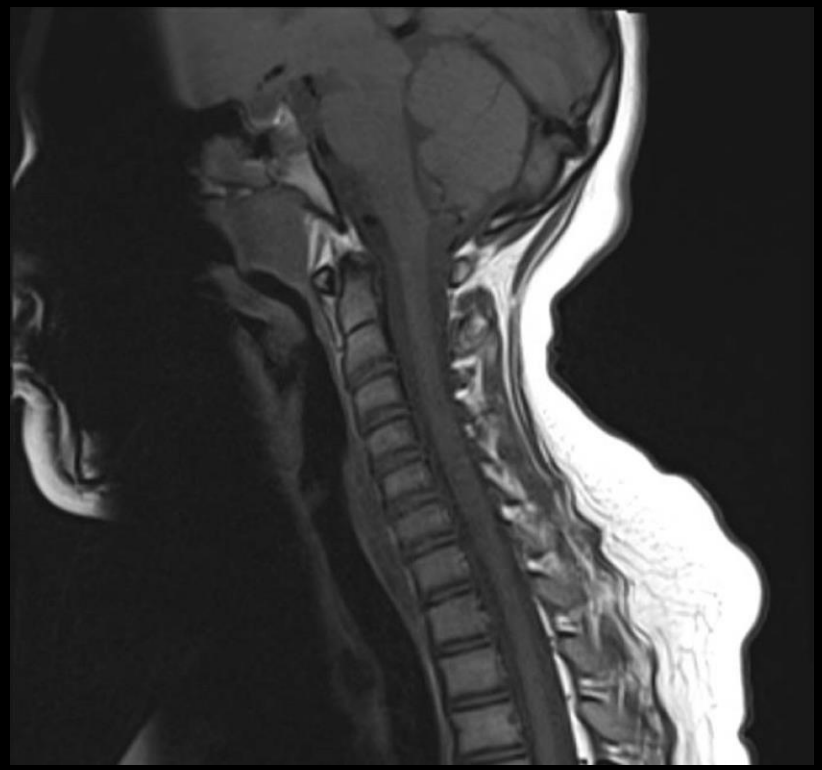


T1 FS Post-Contrast

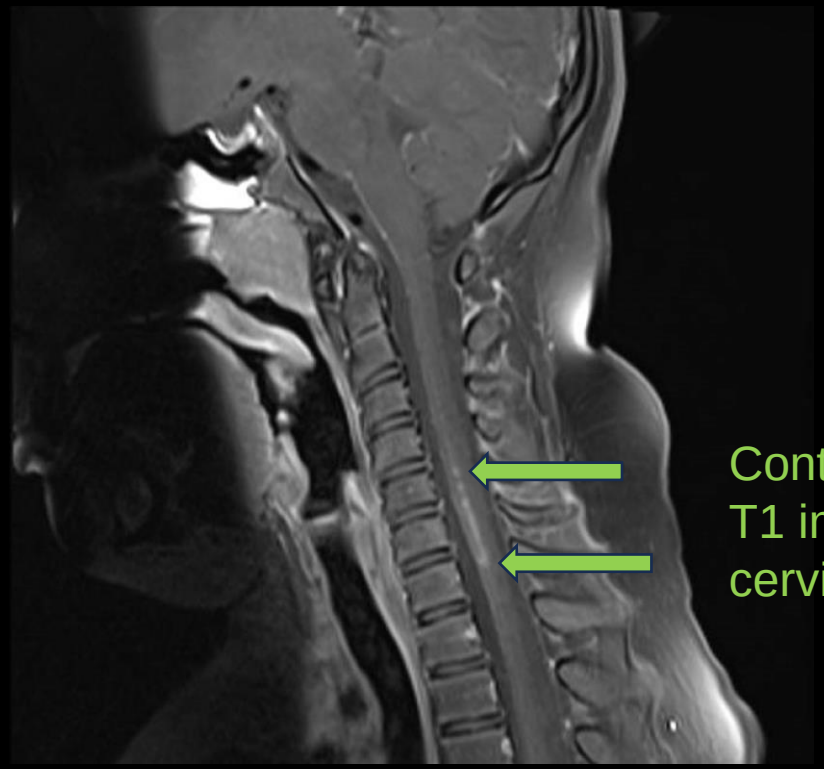


Findings (labeled)

T1 Pre-Contrast



T1 FS Post-Contrast



Contrast enhancement on sagittal T1 images which corresponds to cervical spinal cord lesion.

Final Dx:

Acute Flaccid Myelitis

Case Discussion

- Acute Flaccid Myelitis (AFM) is a rare cause of acute flaccid paralysis in young children that typically follows a viral respiratory illness (i.e.: enteroviruses).
- There have been 750 confirmed cases in the U.S. since the CDC began tracking AFM in 2014.
- Clinical presentation may mimic several other causes of acute flaccid paralysis, including:
  - Guillain-Barre Syndrome
  - Acute transverse myelitis
  - Spinal cord infarction
  - Poliomyelitis (or other infectious myelitis)

Diagnosis and Treatment

- It is ultimately a diagnosis of exclusion that depends on fulfilling clinical criteria of acute flaccid paralysis, ruling out alternative diagnoses, and collecting supportive imaging/laboratory studies.
- Current treatment guidelines for AFM focus on supportive care.
  - 20-35% of patients develop respiratory failure due to respiratory muscle and/or bulbar weakness, requiring ventilatory support.
  - IVIG, glucocorticoids, plasma exchange, and immunomodulators have been used in published case reports with varying degrees of success.
  - There are currently no prospective, randomized trials that demonstrate efficacy of these agents.

Imaging Findings and Recommendations

- MRI is the modality of choice for an acute myelopathy per ACR guidelines.
  - Imaging findings may be too subtle in the first 72 hours of symptom onset (acute phase), but they typically become more pronounced as disease progresses.
  - Key imaging finding is a longitudinal, extensive T2 hyperintense signal in the grey matter of the spinal cord, typically restricted to the anterior horns.
    - T2 signal hyperintensity extends to the brainstem in up to 35-75% of cases.
  - Contrast enhancement of the lesion on T1 sequences when present is typically a feature of the subacute phase of AFM, as was demonstrated in this case.

Case Resolution

- Our patient also underwent an MRI brain which was negative for any acute intracranial abnormality. The clinical presentation and imaging findings were restricted to AFM of the cervical spinal cord.
- There were no signs of respiratory compromise or further progression of weakness. Patient remained stable for discharge the following week and has since had an incomplete neurologic recovery with persistent weakness of his distal BUE – which is typical of most patients with AFM.

References:

1. Caceres JA, Saucier L, Murphy OC, Gordon-Lipkin EM, Santoro JD, Van Haren K, Pardo CA, Hopkins S. Brain Magnetic Resonance Imaging Abnormalities in Acute Flaccid Myelitis. Pediatr Neurol. 2023 Dec;149:56-62. doi: 10.1016/j.pediatrneurol.2023.08.021. Epub 2023 Aug 19. PMID: 37797356.
2. Murphy OC, Messacar K, Benson L, Bove R, Carpenter JL, Crawford T, Dean J, DeBiasi R, Desai J, Elrick MJ, Farias-Moeller R, Gombolay GY, Greenberg B, Harmelink M, Hong S, Hopkins SE, Oleszek J, Otten C, Sadowsky CL, Schreiner TL, Thakur KT, Van Haren K, Carballo CM, Chong PF, Fall A, Gowda VK, Helfferich J, Kira R, Lim M, Lopez EL, Wells EM, Yeh EA, Pardo CA; AFM working group. Acute flaccid myelitis: cause, diagnosis, and management. Lancet. 2021 Jan 23;397(10271):334-346. doi: 10.1016/S0140-6736(20)32723-9. Epub 2020 Dec 23. PMID: 33357469; PMCID: PMC7909727.
3. Okumura A, Mori H, Fee Chong P, Kira R, Torisu H, Yasumoto S, Shimizu H, Fujimoto T, Tanaka-Taya K; Acute Flaccid Myelitis Collaborative Study Investigators. Serial MRI findings of acute flaccid myelitis during an outbreak of enterovirus D68 infection in Japan. Brain Dev. 2019 May;41(5):443-451. doi: 10.1016/j.braindev.2018.12.001. Epub 2018 Dec 26. PMID: 30594353.
4. J.A. Maloney, D.M. Mirsky, K. Messacar, S.R. Dominguez, T. Schreiner, N.V. Stence. MRI Findings in Children with Acute Flaccid Paralysis and Cranial Nerve Dysfunction Occurring during the 2014 Enterovirus D68 Outbreak. American Journal of Neuroradiology Feb 2015, 36 (2) 245-250; DOI: 10.3174/ajnr.A4188

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