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

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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 episode 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.

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<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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<td>MRI spine area of interest without and with IV contrast</td>
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<td>MRI spine area of interest without IV contrast</td>
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This imaging modality was ordered by neurology team.
Findings: (unlabeled)
Findings: (labeled)

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

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

T2 signal hyperintensity predominately involves spinal cord grey matter ventrally.

Axial T1 Post-Contrast
Findings (labeled)

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

T1 Pre-Contrast

T1 FS Post-Contrast
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:


