AMSER Case of the Month
October 2023

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

<table>
<thead>
<tr>
<th>Scenario Id</th>
<th>Procedure</th>
<th>Adult RRL</th>
<th>Peds RRL [ped]</th>
<th>Appropriateness Category</th>
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<td>MRI cervical and thoracic spine without IV contrast</td>
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This imaging modality was ordered by the neurology team.
Select the applicable ACR Appropriateness Criteria

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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.
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 CIDP\(^1\)
  • 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 paranododal 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 appearance\(^2\)
• 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:


