AANEM Case Study:
Critical Illness Myopathy

Author Information

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<thead>
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</tbody>
</table>

No one involved in the planning of this CME activity had any relevant financial relationships to disclose. Authors/Faculty had nothing to disclose.

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ELECTRODIAGNOSTIC MEDICINE
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Rochester, MN 55901
Critical Illness Myopathy

**Educational Objectives:** Upon completion of this case study, the participants will acquire skills to 1) identify uncommon presentation of critical illness myopathy, (2) differentiate among various causes of brachial diplegia (man in the barrel syndrome), and (3) outline the electrodiagnostic (EDX) changes in critical illness myopathy.

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EMG CASE: Critical Illness Myopathy

Raghav Govindarajan, MD; Danita Jones, DO; Nestor Galvez, MD, MS, FAAN

Case Information

<table>
<thead>
<tr>
<th>Presenting Symptom(s):</th>
<th>Weakness of bilateral upper limbs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case-specific Diagnosis:</td>
<td>Critical illness myopathy</td>
</tr>
<tr>
<td>Appropriate Audience:</td>
<td>Residents and/or practicing neurologists and physiatrists</td>
</tr>
<tr>
<td>Level of Difficulty:</td>
<td>Intermediate</td>
</tr>
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1. HISTORY

Chief complaint: Weakness of both upper limbs.

A 52-year-old male with no significant past medical history drowned in his swimming pool after an alcoholic binge. He was brought intubated to the intensive care unit (ICU). His ICU course was complicated by aspiration pneumonia, sepsis, and acute respiratory distress syndrome, requiring multiple antibiotics, intravenous steroids, nondepolarizing neuromuscular paralytics, and high positive end-expiratory ventilator pressure. Forty days post-cardiac arrest he was following simple commands but was still connected to the ventilator with tracheostomy. His nurse noticed he was not moving his upper limbs although he was moving his lower limbs. His face appeared symmetric.

2. COMMENTARY I

This brief history describes a patient who was critically ill and now appears to have disproportionate weakness of the upper limbs as compared to the lower limbs and face. Such a clinical presentation fits the description of man-in-the-barrel syndrome (MIBS) or brachial diplegia.¹ Below is the differential diagnosis of MIBS:

1. Central
   A. Cerebrum: Bilateral frontal lobe lesions (precentral gyrus) or bilateral anterior watershed infarction. The causes of these can be global hypoperfusion, tumors, or emboli.
   B. Brainstem: Pontine or extrapontine myelinolysis.
2. Cervical spinal cord (cruciate paralysis)
   A. Vascular: Anterior spinal artery infarction.
   C. Variant of amyotrophic lateral sclerosis.

3. Peripheral
   A. Degenerative disc disease affecting multiple cervical roots.
   B. Bilateral brachial plexopathy due to trauma, postsurgical inflammation.
   C. Multifocal motor neuropathies.
   D. Myasthenia gravis.
   E. Myopathies: Polymyositis, dermatomyositis, critical illness myopathy (CIM).

Based on the history and presentation, this broad differential diagnosis can be narrowed down to the following:

1. Watershed infarctions due to global hypoperfusion as a result of drowning.
2. Traumatic cervical spinal cord injury during drowning or emergent intubation.
3. Traumatic vertebral artery dissection.
4. Bilateral traumatic brachial plexopathy or brachial plexopathy developed in the setting of surgeries/medical procedures (e.g., Parsonage–Turner’s syndrome).
5. CIM due to the history of use of steroids and neuromuscular paralytics.
6. Myasthenia gravis unmasked in the setting of a critically ill patient.
7. Inflammatory myopathy precipitated in this critically ill setting.

Reader question: Is the pattern of weakness in MIBS proximal or distal?

3. HISTORY, CONTINUED

The patient could not lift his upper limbs off the bed, although he could wiggle his fingers. He could lift his legs off the bed and wiggle his toes. He could lift his neck off the bed. He denied any pain, numbness, or tingling in his upper or lower limbs. He denied double vision or blurry vision.

4. COMMENTARY II

The history provides some clues to the differential diagnosis as one would typically expect some sensory involvement in a brachial plexopathy. Although the patient has yet to be examined, the pattern of weakness appears to suggest proximal more than distal involvement. To narrow down the differential, there is no preferential involvement of antigravity muscles which would be expected with an upper motor neuron lesion. This can be further confirmed by examination.

At this point, the differential diagnosis includes the following:

1. Watershed infarctions due to global hypoperfusion.
2. Traumatic cervical spinal cord injury.
3. Vertebral artery dissection.
4. CIM.
5. Myasthenia gravis.
6. Inflammatory myopathy.
5. PHYSICAL EXAMINATION

The patient was examined on the bed. He was connected to the ventilator though the tracheostomy, needing minimal ventilator support. He had a percutaneous endoscopic gastrostomy (PEG) tube in place through which he was receiving nutrition. The Foley catheter was draining clear urine. He could follow most commands. He verbalized by nodding his head or blinking his eyes. He had been off sedation for 10 days and off of paralytics for 15 days.

Vital signs: BP: 138/80 mmHg; Pulse: 80/min; RR: 20/min; Ventilator setting: Continuous positive airway pressure (CPAP) with pressure of 10 mmHg.
Respiratory: Breath sounds were coarse with air entry equal bilaterally.
Cardiovascular: Heart sounds normal, no murmurs.
Gastrointestinal: Abdomen soft, non-tender with normal bowel sounds.
Cutaneous: No rashes or other neurocutaneous stigmata.
Extremities: 1+ pitting pedal edema.

6. COMMENTARY III

The general physical examination provides a few clues. It is imperative to note that although still connected to a ventilator, the patient’s breathing capacity has significantly improved, thus reducing the likelihood of a high cervical lesion. Clear urine rules out an underlying rhabdomyolysis. Further, there are no suspicious cutaneous rashes as can be seen in dermatomyositis. Finally, since he is off sedation, he would be alert to be able to participate in the physical examination and needle electromyography (EMG) which will result in more reliable studies.

7. PHYSICAL EXAMINATION, CONTINUED

Neurological examination:

- The patient is alert, appropriate, and oriented to place and person.

- Cranial nerve examination:
  - 2nd cranial nerve: Pupils are round, reactive, symmetric, and 3 mm in size. Fundus is normal.
  - 3rd, 4th, and 6th cranial nerves: Extraocular eye movements are intact in all directions of gaze.
  - 7th cranial nerve: Bilateral face strength is normal.
  - 8th cranial nerve: Hearing is intact to finger rub.
  - 9th and 10th cranial nerves: Uvula is midline and palate elevates symmetrically, gag reflex is intact.
  - 11th cranial nerve: Shoulder shrug is 5/5 on the Medical Research Council (MRC) scale.
  - 12th cranial nerve: Tongue is midline and strength is normal.

- Motor system:
  - Tone: Mildly reduced in upper limbs and normal in lower limbs.
• Bulk: Normal throughout.

<table>
<thead>
<tr>
<th>Strength</th>
<th>Right</th>
<th>Left</th>
</tr>
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<tbody>
<tr>
<td>Deltoid</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Biceps</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Triceps</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Wrist extensors</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Grip</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>First dorsal interosseous</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Abductor pollicis brevis</td>
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<td>2</td>
</tr>
<tr>
<td>Hip flexors</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Quadriceps</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Hamstrings</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Ankle flexors</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Ankle extensors</td>
<td>5</td>
<td>5</td>
</tr>
</tbody>
</table>

There was no scapular winging

<table>
<thead>
<tr>
<th>Reflexes</th>
<th>Right</th>
<th>Left</th>
</tr>
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<tbody>
<tr>
<td>Biceps</td>
<td>2+</td>
<td>2+</td>
</tr>
<tr>
<td>Triceps</td>
<td>1+</td>
<td>1+</td>
</tr>
<tr>
<td>Brachioradialis</td>
<td>2+</td>
<td>2+</td>
</tr>
<tr>
<td>Knee</td>
<td>2+</td>
<td>2+</td>
</tr>
<tr>
<td>Ankle</td>
<td>2+</td>
<td>2+</td>
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</table>

Hoffman’s sign: Negative
Plantar response: Flexor

• Sensory system: Sensation intact to light touch, pin prick, vibration, and proprioception bilaterally in both upper and lower extremities.

• Cerebellum and gait were not assessed.

8. COMMENTARY IV

The physical examination confirms the clinical suspicion. The pattern of weakness is proximal predominant affecting the upper limbs while sparing the lower limbs and face. Further sensory examination is normal and there are no upper motor neuron signs. These findings taken together put myopathy high on the differential. Thus, the differential diagnosis can be revised to the following:

1. CIM.
2. Inflammatory myopathy.
3. Watershed infarctions due to global hypoperfusion.
5. Vertebral artery dissection.

The signs of an upper motor neuron lesion may not be obvious within the first week of injury due to neurogenic shock. Although it is unclear as to when exactly the patient became weak, the lack
of other clinical findings such as alteration in mentation and absent deep tendon reflexes argues against an upper motor neuron lesion.

9. ELECTROPHYSIOLOGIC DATA

### SENSORY NERVE CONDUCTION STUDIES

<table>
<thead>
<tr>
<th>NERVE</th>
<th>SIDE</th>
<th>STIM SITE</th>
<th>RECORD</th>
<th>cm</th>
<th>AMPL (µV)</th>
<th>LAT (onset, ms)</th>
<th>CV (m/s)</th>
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</thead>
<tbody>
<tr>
<td>Median</td>
<td>R</td>
<td>Mid wrist</td>
<td>Index finger</td>
<td>14</td>
<td>14 (&gt;7)</td>
<td>2.4 (&lt;3.4)</td>
<td>58 (&gt;44)</td>
</tr>
<tr>
<td>Ulnar</td>
<td>R</td>
<td>Mid wrist</td>
<td>Little finger</td>
<td>14</td>
<td>16 (&gt;7)</td>
<td>2.2 (&lt;3.1)</td>
<td>63 (&gt;44)</td>
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<tr>
<td>Sural</td>
<td>R</td>
<td>Lateral leg</td>
<td>Lateral malleolus</td>
<td>14</td>
<td>14 (&gt;5)</td>
<td>2.8 (&lt;3.7)</td>
<td>51 (&gt;38)</td>
</tr>
<tr>
<td>Sural</td>
<td>L</td>
<td>Lateral leg</td>
<td>Lateral malleolus</td>
<td>14</td>
<td>12</td>
<td>2.7</td>
<td>67</td>
</tr>
</tbody>
</table>

### MOTOR NERVE CONDUCTION STUDIES

<table>
<thead>
<tr>
<th>NERVE</th>
<th>SIDE</th>
<th>STIM SITE</th>
<th>RECORD</th>
<th>cm</th>
<th>AMPL (mV)</th>
<th>LAT (ms)</th>
<th>CV (m/s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median</td>
<td>R</td>
<td>Wrist</td>
<td>Abductor pollicis brevis</td>
<td>7</td>
<td>4.7 (&gt;4)</td>
<td>3.3 (&lt;4.4)</td>
<td>53 (&gt;49)</td>
</tr>
<tr>
<td>Ulnar</td>
<td>R</td>
<td>Medial wrist Below elbow</td>
<td>Abductor digitorum brevis</td>
<td>7</td>
<td>10.5 (&gt;6)</td>
<td>10 (3.5)</td>
<td>51 (&gt;49)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Above elbow</td>
<td></td>
<td>17.2</td>
<td>10.1</td>
<td>6.4</td>
<td>59</td>
</tr>
<tr>
<td></td>
<td>R</td>
<td>Mid ankle</td>
<td>Extensor digitorum brevis</td>
<td>10</td>
<td>3.1 (&gt;2)</td>
<td>4.0 (&lt;6.1)</td>
<td>49 (&gt;41)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Fibular head Popliteal fossa</td>
<td></td>
<td>26.5</td>
<td>3.0</td>
<td>9.4</td>
<td>50</td>
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<tr>
<td></td>
<td></td>
<td>Abductor hallucis</td>
<td></td>
<td>8.5</td>
<td>2.9</td>
<td>11.1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>R</td>
<td>Medial ankle Popliteal fossa</td>
<td></td>
<td>8.5</td>
<td>14.3 (&gt;3)</td>
<td>3.7 (&lt;6.1)</td>
<td>57 (&gt;41)</td>
</tr>
<tr>
<td>Median</td>
<td>L</td>
<td>Mid wrist</td>
<td>Abductor pollicis brevis</td>
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<tr>
<td>Ulnar</td>
<td>L</td>
<td>Medial wrist Below elbow</td>
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<td>7</td>
<td>11.5</td>
<td>3.1</td>
<td>55</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Above elbow</td>
<td></td>
<td>20.2</td>
<td>10.9</td>
<td>6.8</td>
<td>56</td>
</tr>
</tbody>
</table>
2-Hz repetitive nerve stimulation of the right spinal accessory nerve was normal.

10. DIAGNOSTIC IMPRESSION

The final diagnostic impression for this patient is irritable myopathy, probably CIM.

The central causes of MIBS are less likely with this clinical and EDX picture.
Nerve conduction studies showed a small symmetric compound motor action potential amplitude from the median nerve when recorded from the abductor pollicis brevis. There was no conduction block or other signs of demyelination. The sensory studies were spared as were the lower limb studies. Needle EMG showed a proximal predominant irritable myopathy involving the upper limbs. It is imperative to rule out a neuromuscular junction (NMJ) disorder with myopathic changes on needle EMG. Thus, a slow repetitive nerve stimulation of the right spinal accessory nerve was performed. This was normal, thereby reducing the likelihood of an NMJ disorder.

Although inflammatory myopathies can produce similar changes on EDX studies, the clinical picture of prolonged intubation, use of steroids, and use of nondepolarizing neuromuscular paralytics is more consistent with CIM. A biopsy would have been helpful to differentiate these two, but it was not pursued. At the last followup at about 6 months since the initial evaluation, the patient had recovered strength in his upper limbs with a MRC grade of 3/5 proximally in all muscles and 4/5 distally.

CIM is an underrecognized clinical entity. The predisposing factors are acute respiratory disorder (e.g., acute respiratory distress syndrome, pneumonia, or severe asthma) in conjunction with the use of high-dose intravenous steroids, nondepolarizing blocking agents, and aminoglycosides. Other predisposing conditions include liver and lung transplantation, hepatic failure, and acidosis.

Classically, CIM presents with proximal weakness affecting all the limbs and thus has been referred to in the past as acute quadriplegic myopathy. However, patchy or selective involvement of the limbs can occur, as seen in this case, thus confusing the clinical picture. A high degree of clinical suspicion is needed to initiate a proper workup in cases such as the one presented here.

The prognosis in CIM appears to be fairly good in terms of recovery. In a recent study comparing 8 patients with only CIM and 11 patients with CIM and critical illness polyneuropathy, 8/11 CIM patients had completely recovered in 1 year as compared to 6/11 of those with both.

11. COMMENTARY V

12. BIBLIOGRAPHY