Nerve Ultrasound in Motor Conduction Block

Edward C. Smith, MD, Lisa Hobson-Webb, MD, Vern C. Juel, MD, and E.W. Massey, MD

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American Association of Neuromuscular and Electrodiagnostic Medicine

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CME Information
Product: CS26 - Nerve Ultrasound in Motor Conduction Block

Course Description
Intended Audience
This course is intended for Neurologists, Physiatrists, and others who practice neuromuscular, musculoskeletal, and electrodiagnostic medicine with the intent to improve the quality of medical care to patients with muscle and nerve disorders.

Learning Objectives
Upon conclusion of this program, participants should be able to:
1. evaluate patients with MADSAM neuropathy and formulate a differential diagnosis.
2. describe the typical electrodiagnostic findings associated with MADSAM neuropathy.
3. describe the potential role of peripheral nerve ultrasound in the diagnosis of MADSAM neuropathy and in monitoring treatment response.

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Nerve Ultrasound in Motor Conduction Block: Pre- and Post-treatment Findings

January 2009

CME Available from January 2009 through January 2012

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Nerve Ultrasound in Motor Conduction Block: Pre- and Post-treatment Findings

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Presenting Symptom: Right lower limb paresthesias and weakness.

Case prepared by: Edward C. Smith, MD; Lisa Hobson-Webb, MD; Vern C. Juel, MD; E.W. Massey, MD

Affiliations: Duke University Medical Center

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Appropriate Audience: Residents, clinical neurophysiology or neuromuscular fellows, and practicing physicians.

Learning Objectives: After completing this educational activity, participants will be able to:
1) Evaluate patients with MADSAM neuropathy and formulate a differential diagnosis.
2) Describe the typical electrodiagnostic findings associated with MADSAM neuropathy.
3) Describe the potential role of peripheral nerve ultrasound in the diagnosis of MADSAM neuropathy and in monitoring treatment response.

Level of Difficulty: Intermediate/Advanced
Nerve Ultrasound in Motor Conduction Block: Pre- and Post-treatment Findings

History

A 20-year-old right-handed female presented with a 6-week history pain and numbness. Her symptoms began with tenderness in the medial aspect of the right foot, ankle, and calf followed by numbness in the same distribution. She subsequently developed right thigh weakness along with paresthesias in ring and small finger of the right hand. She noted a decline in right grip strength, along with deterioration of her handwriting.

Commentary I

At this time, the differential diagnosis includes:

At this point, the differential diagnosis remains broad and includes multifocal disorders that cause weakness and sensory loss. The history and clinical examination findings should allow one to differentiate between a central and peripheral nervous system disorder. In the absence of long tract findings on examination, the stepwise, multifocal onset suggests a peripheral localization. Pathologic processes affecting sensory pathways in peripheral nerves, plexii, dorsal root ganglia or dorsal roots may explain the sensory symptoms. The weakness could be explained by pathology anywhere in the motor unit, including the anterior horn cell, ventral root, plexus, peripheral nerve, neuromuscular junction or muscle. Given the recent onset of symptoms and patchy distribution of motor and sensory symptoms, an acquired, multifocal polyneuropathy is most likely, although include polyradiculoplexopathy remains a possibility. Further historical details regarding past medical history, family history and occupational history may help focus the differential diagnosis.

History, Continued

The patient is a healthy competitive collegiate athlete with a history of right medial meniscus and right anterior collateral ligament repair two years prior to presentation. Prior to onset of her symptoms, there was no history of recent trauma, illness, travel or toxin exposure. Her appetite has been good without any recent weight loss, rash or fatigue. She recently started taking pregabalin for her pain, but otherwise takes no medications. She has never abused tobacco, alcohol or illicit drugs. There is no family history of unexplained weakness or sensory loss.

Commentary II

The absence of a family history and the multifocal, asymmetric distribution of symptoms make an inherited polyneuropathy such as Charcot-Marie-Tooth disease unlikely. At this point, the differential diagnosis includes acquired causes of sensorimotor polyneuropathy due to (1) toxic/metabolic, (2) autoimmune, and (3) infectious etiologies:

(1) Toxic/Metabolic
- Diabetes mellitus
- Vitamin deficiency (B1, B12)
- Neurotoxins
- Arsenic and other heavy metals
- Colchicine
- N-hexane

(2) Immune-mediated
- Acute inflammatory demyelinating polyneuropathy (AIDP or Guillain-Barre syndrome)
- Acute motor and sensory axonal polyneuropathy (AMAN)
Chronic inflammatory demyelinating polyneuropathy (CIDP)
Multifocal motor neuropathy (MMN)
Multifocal acquired demyelinating sensory and motor neuropathy (MADSAM)
Distal acquired demyelinating sensorimotor neuropathy (DADS)
Multifocal acquired motor axonopathy (MAMA)
Connective tissue disorders (Sjögren’s, systemic lupus erythematosus)
Cryoglobulinemia
Isolated peripheral nervous system vasculitis
Vasculitis associated with systemic disease
Sarcoidosis

(3) Neoplastic
Secondary to infiltrative tumors
Paraneoplasia
Associated with lymphoproliferative disorders and plasmacytomas (multiple myeloma, MGUS, Waldenstrom macroglobulinemia, osteosclerotic myeloma)

(4) Infectious
Diphtheria
Hepatitis C
HIV
Leprosy
Lyme disease
West Nile virus

Given the patient’s age, lack of systemic symptoms, pain and asymmetric distribution, an acquired, immune-mediated etiology seems most likely.

Physical Examination

General Examination: Healthy-appearing female in no distress. No rashes. No muscular atrophy. Cranial Nerves: Normal. No optic disc edema. No sensory loss. No weakness or facial asymmetry. The tongue appeared normal. Motor: Normal muscle tone and bulk. In the upper limbs, examination revealed 4/5 weakness of the interossei bilaterally, and milder weakness of right hand pronation and wrist extension. In the lower limbs, there was 4/5 weakness of right knee extension and right foot dorsiflexion, eversion and inversion were graded at 4+/5.

Commentary III

The physical examination findings suggest a more widespread process than initially suspected based on the patient’s complaints. There is weakness in a bilateral ulnar distribution, along with weakness of right radial and median-innervated muscles. There is also weakness in the right leg and thigh that does not correspond to a single nerve or nerve root level. This process is truly multifocal and asymmetric.

While the patient’s symptoms suggest sensory and motor involvement, further examination is needed. At this time, the differential diagnosis remains unchanged.
PHYSICAL EXAMINATION, CONTINUED

Reflexes: Trace at the bilateral Achilles and triceps, otherwise absent. Plantar responses were flexor.
Sensation: Absent pinprick sensation over the medial aspect of the right calf in a saphenous distribution.
Pinprick and vibratory sensation were otherwise normal.

COMMENTARY IV

There is now evidence of sensory and motor involvement on examination with loss of pinprick sensation in a right saphenous distribution. This further demonstrates the multifocal nature of this process and eliminates exclusively motor neuropathic processes (e.g., multifocal motor neuropathy) from the differential diagnosis.
Laboratory evaluation revealed no evidence of diabetes mellitus, hypothyroidism, vitamin deficiencies (B1 or B12). Anti-neuronal (paraneoplastic) autoantibodies and ganglioside antibodies were absent. Hepatitis serology was negative. Lumbar puncture was normal with a protein of 43 mg/dL. Testing for connective tissue disorders revealed a low positive ANA titer of 1:160.

Electrophysiologic Data (Abnormal Values in Blue)

### SENSORY NERVE CONDUCTION STUDIES
Nerve Conduction Studies performed at 34° C

<table>
<thead>
<tr>
<th>NERVE</th>
<th>SIDE</th>
<th>STIM SITE</th>
<th>RECORD</th>
<th>Distance cm</th>
<th>AMPL microvolts</th>
<th>LAT msec</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median Right</td>
<td>D2</td>
<td>Wrist</td>
<td>13</td>
<td>16</td>
<td>2.9</td>
<td></td>
</tr>
<tr>
<td>Median Right</td>
<td>Palm</td>
<td>Wrist</td>
<td>8</td>
<td>50</td>
<td>1.9</td>
<td></td>
</tr>
<tr>
<td>Median Left</td>
<td>D2</td>
<td>Wrist</td>
<td>13</td>
<td>26</td>
<td>4.5</td>
<td></td>
</tr>
<tr>
<td>Median Left</td>
<td>Palm</td>
<td>Wrist</td>
<td>8</td>
<td>50</td>
<td>2.1</td>
<td></td>
</tr>
<tr>
<td>Ulnar Left</td>
<td>D5</td>
<td>Wrist</td>
<td>11</td>
<td>9</td>
<td>2.8</td>
<td></td>
</tr>
<tr>
<td>Ulnar Left</td>
<td>Palm</td>
<td>Wrist</td>
<td>8</td>
<td>15</td>
<td>2.1</td>
<td></td>
</tr>
<tr>
<td>Ulnar Right</td>
<td>D5</td>
<td>Wrist</td>
<td>11</td>
<td>7</td>
<td>2.8</td>
<td></td>
</tr>
<tr>
<td>Ulnar Right</td>
<td>Palm</td>
<td>Wrist</td>
<td>8</td>
<td>9</td>
<td>2.3</td>
<td></td>
</tr>
<tr>
<td>Radial Left</td>
<td>Forearm</td>
<td>Wrist</td>
<td>10</td>
<td>27</td>
<td>2.1</td>
<td></td>
</tr>
<tr>
<td>Sural Right</td>
<td>Calf</td>
<td>Ankle</td>
<td>14</td>
<td>14</td>
<td>3.8</td>
<td></td>
</tr>
</tbody>
</table>

### MOTOR NERVE CONDUCTION STUDIES
Nerve Conduction Studies performed at 34° C

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Side</th>
<th>Stim Site</th>
<th>Record</th>
<th>Cm</th>
<th>Amplitude millivolts</th>
<th>Lat msec</th>
<th>CV M/sec</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median Left</td>
<td>Wrist</td>
<td>APB</td>
<td>6.5</td>
<td>7.0</td>
<td>5.3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Median Right</td>
<td>Wrist</td>
<td>APB</td>
<td>6.5</td>
<td>8.8</td>
<td>4.3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Median Left</td>
<td>Elbow</td>
<td>APB</td>
<td>26</td>
<td>4.5</td>
<td>10.6</td>
<td>49.0</td>
<td></td>
</tr>
<tr>
<td>Median Right</td>
<td>Elbow</td>
<td>APB</td>
<td>27</td>
<td>8.4</td>
<td>9.6</td>
<td>52.0</td>
<td></td>
</tr>
<tr>
<td>Median F-wave Left</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>32.1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Median F-wave Right</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>30.5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ulnar</td>
<td>Left</td>
<td>Wrist ADQ</td>
<td>6.5</td>
<td>9.9</td>
<td>3.5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ulnar</td>
<td>Right</td>
<td>Wrist ADQ</td>
<td>6.5</td>
<td>8.0</td>
<td>3.3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ulnar</td>
<td>Left</td>
<td>Below Elbow ADQ</td>
<td>22.5</td>
<td>9.3</td>
<td>7.7</td>
<td>54.0</td>
<td></td>
</tr>
<tr>
<td>Ulnar</td>
<td>Right</td>
<td>Below Elbow ADQ</td>
<td>22</td>
<td>7.0</td>
<td>8.2</td>
<td>45.0</td>
<td></td>
</tr>
<tr>
<td>Ulnar</td>
<td>Left</td>
<td>Above Elbow ADQ</td>
<td>34</td>
<td>6.0</td>
<td>11.6</td>
<td>42.0</td>
<td></td>
</tr>
<tr>
<td>Ulnar</td>
<td>Right</td>
<td>Above Elbow ADQ</td>
<td>34</td>
<td>3.2</td>
<td>12.5</td>
<td>37.0</td>
<td></td>
</tr>
<tr>
<td>Ulnar F-wave</td>
<td>Left</td>
<td></td>
<td></td>
<td></td>
<td>36.2</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ulnar F-wave</td>
<td>Right</td>
<td></td>
<td></td>
<td></td>
<td>40.7</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Peroneal</td>
<td>Left</td>
<td>Ankle EDB</td>
<td>7.5</td>
<td>3.1</td>
<td>9.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Peroneal</td>
<td>Right</td>
<td>Ankle EDB</td>
<td>7.5</td>
<td>4.6</td>
<td>8.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Peroneal</td>
<td>Left</td>
<td>Knee EDB</td>
<td>41</td>
<td>2.7</td>
<td>17.9</td>
<td>46.0</td>
<td></td>
</tr>
<tr>
<td>Peroneal</td>
<td>Right</td>
<td>Knee EDB</td>
<td>43.5</td>
<td>4.3</td>
<td>18.2</td>
<td>43.0</td>
<td></td>
</tr>
<tr>
<td>Peroneal F-wave</td>
<td>Left</td>
<td></td>
<td></td>
<td></td>
<td>58.3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Peroneal F-wave</td>
<td>Right</td>
<td></td>
<td></td>
<td></td>
<td>NR</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Note: The ulnar motor to the thenar recording is a volume conducted compound muscle action potential from the ulnar muscles under the thenar eminence. It depends on the muscles innervated by the ulnar nerve (flexor pollicis brevis, adductor pollicis, intrinsics, or even opponens and abductor pollicis brevis if there is Riche-Cannieu or Martin-Gruber anastomosis).

### NEEDLE ELECTROMYOGRAPHY

**INSertional activity**: N, sust, unsust  
**FIB**: 0, 1+, 2+, 3+, 4+  
**OTH**er: 0 or fascic, myotonia, myokymia  
**EFFort**: N, decr  
**RECruitment**: N, inc or dec 1+, 2+, 3+, 4+  
**AMPlitude**: N, inc or dec 1+, 2+, 3+, 4+  
**DURation**: N, inc or dec 1+, 2+, 3+, 4+  
**POLyphasia**: N, inc or dec 1+, 2+, 3+, 4+  

<table>
<thead>
<tr>
<th>R/L</th>
<th>MUSCLE</th>
<th>INSER</th>
<th>FIB</th>
<th>OTH</th>
<th>EFF</th>
<th>REC</th>
<th>AMP</th>
<th>DUR</th>
<th>POL</th>
</tr>
</thead>
<tbody>
<tr>
<td>R</td>
<td>First Dorsal Interosseous (hand)</td>
<td>N</td>
<td>0</td>
<td>0</td>
<td>N</td>
<td>Dec 1</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>R</td>
<td>Deltoid</td>
<td>N</td>
<td>0</td>
<td>0</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
</tbody>
</table>
Electrophysiologic Data, Continued

Ultrasonography was performed on both upper limbs, focusing on the bilateral median and right ulnar nerves. A 15-MHz linear array probe was used for imaging and the examiner was blinded to the electrodiagnostic data. The cross-sectional areas provided in the table are for the median nerve. All distances are listed as proximal to the distal wrist crease.

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Wrist</th>
<th>8cm</th>
<th>14cm</th>
<th>18cm</th>
</tr>
</thead>
<tbody>
<tr>
<td>R Median</td>
<td>12mm²</td>
<td>9mm²</td>
<td>8mm²</td>
<td>8mm²</td>
</tr>
<tr>
<td>L Median</td>
<td>14mm²</td>
<td>7mm²</td>
<td>14mm²</td>
<td>7mm²</td>
</tr>
</tbody>
</table>

The values for the right ulnar nerve are listed below:

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Wrist</th>
<th>Forearm</th>
<th>Ulnar Grove</th>
<th>4cm Above Elbow</th>
<th>11cm Below Elbow</th>
</tr>
</thead>
<tbody>
<tr>
<td>R Ulnar</td>
<td>7mm²</td>
<td>7mm²</td>
<td>12mm²</td>
<td>10mm²</td>
<td>7mm²</td>
</tr>
</tbody>
</table>

An inching study of the left median nerve was performed. The values are listed below:

<table>
<thead>
<tr>
<th>Stimulation Site</th>
<th>Recording Site</th>
<th>Latency (ms)</th>
<th>Amplitude (mV)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wrist Crease (WC)</td>
<td>APB</td>
<td>7.0</td>
<td>9.3</td>
</tr>
<tr>
<td>4 cm prox to WC</td>
<td>APB</td>
<td>7.4</td>
<td>8.7</td>
</tr>
<tr>
<td>8 cm prox to WC</td>
<td>APB</td>
<td>7.9</td>
<td>9.4</td>
</tr>
<tr>
<td>12 cm prox to WC</td>
<td>APB</td>
<td>8.0</td>
<td>9.9</td>
</tr>
<tr>
<td>14 cm prox to WC</td>
<td>APB</td>
<td>8.7</td>
<td>2.5</td>
</tr>
</tbody>
</table>

Diagnostic Impression

The electrophysiologic data reveal a multifocal, motor greater than sensory, demyelinating neuropathy with conduction blocks. The conduction blocks in the ulnar nerves localize to the elbow and could be secondary to compression as opposed to an inflammatory process. The same can be said for the ultrasonographic enlargement of the median and ulnar nerves at the wrist crease and ulnar groove, as nerves are known to enlarge at compression sites. However, the left median nerve conduction block is located in the forearm and correlates with an area of ultrasonographic enlargement. The lack of spontaneous activity on electromyography also supports the case for a demyelinating process. In summary, this appears to be a multifocal, motor > sensory neuropathy with segmental demyelination.
The most likely diagnosis is multifocal acquired demyelinating sensory and motor neuropathy (MADSAM), also known as Lewis-Sumner syndrome. However, other related disorders should be excluded, particularly the paraproteinemias.

Commentary V

MADSAM was first described in 1982 and is an important condition to consider in patients where mononeuritis multiplex and multifocal motor neuropathy (MMN) are considerations. It is defined as a chronic, asymmetric sensory and motor demyelinating polyneuropathy. The age of onset is widely variable, although onset in middle age is most frequent. The asymmetric distribution of findings is characteristic. Although the motor symptoms dominate, the presence of sensory loss and abnormalities in sensory nerve conduction studies help differentiate it from MMN. This distinction is important, as MADSAM is often responsive to steroids.

The patient in this case was treated with intravenous immunoglobulin and returned for repeat studies exactly 4 weeks later. She reported improvement in right grip strength and her motor examination was normal with the exception of 4+/5 weakness of the bilateral first dorsal interosseus. Sensory loss persisted in a right saphenous distribution. On electrodiagnostic testing, there was resolution of the conduction block in the left median nerve. On ultrasonography, the left median nerve measured 8mm throughout the forearm.

In this particular case, peripheral nerve ultrasonography (US) played a significant role in the diagnostic process. Over the last decade, US has proven to be a useful tool in the diagnosis of compression neuropathies. At sites of compression, nerves tend to enlarge and become hypoechochogenic in appearance. More recently, it has been suggested that this phenomenon also occurs in inflammatory neuropathies, including CIDP and MMN. In this particular case, an area of focal enlargement was identified in the left median nerve 14cm proximal to the distal wrist crease. This correlated with the site of conduction block seen on nerve conduction studies and also correlated with improvement. Although this may have been coincidental, it emphasizes the value that US can add to electrodiagnostic studies. With US, it may be possible to increase the yield of nerve conduction studies in demonstrating conduction block by focusing on areas of focal enlargement. US may also prove to be a valuable tool in monitoring response to treatment.

Bibliography