Case presentation 1

- 67 y/o female
- 2011 noticed progressive atrophy in her left FDI muscle
- Excruciating constant 6/10 pain “like when you hit your funny bone” and electrical shocks, some day worse than others
- 11-2016 and 03-2017, two ulnar nerve transposition at the elbow
- First surgery eased the pain little bit, the second did not do anything
- She also has ulnar nerve release at the wrist late 2017
- Bilateral CTS release surgery in 3-2016.

Case presentation 1

- FDI 1/5, severe atrophy of the FDI
- ADM (abductor digiti minimi) 3-4/5, mild to moderate atrophy
- Finger spread 3/5
- No fasciculations
- Sensory exam is normal
- Normal tendon reflexes
Localization

- Ulnar nerve
- Not C8-T1 or brachial plexus because finger extensors, flexor pollicis longus and abductor pollicis brevis muscles are normal
- Not diffuse for motor neuron disease (anterior horn cells), also has pain

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Insertion</th>
<th>Spontaneous Activity</th>
<th>Volitional MUAPs</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st dorsal interosseous</td>
<td>Increased</td>
<td>4+</td>
<td>4+</td>
<td>None</td>
</tr>
<tr>
<td>Extensor indicis proprius</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Abductor digiti minimi</td>
<td>Increased</td>
<td>2+</td>
<td>2+</td>
<td>2+ Red</td>
</tr>
<tr>
<td>Flexor pollicis longus</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>Normal</td>
</tr>
<tr>
<td>Abductor pollicis brevis</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>Normal</td>
</tr>
<tr>
<td>Flexor digitorum profundus</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>Normal</td>
</tr>
</tbody>
</table>

EMG

- Abnormal study. There is evidence for a severe left deep ulnar motor branch neuropathy. MRI of the wrist with contrast is recommended for further evaluation and to rule out compressive lesion of the ulnar motor branch at the wrist. In addition there is evidence for a mild right median neuropathy at the wrist.
MRI

- Heterogeneously enhancing soft tissue mass arising along the ulnar aspect of Guyon's canal, resulting in displacement and mass effect on the ulnar nerve.

Pathology

- Tenosynovial giant cell tumor, localized type

Tenosynovial Giant cell tumor

- Most common in patients after 30 years old
- Second most common benign hand tumor after a ganglion cyst
- Diffuse proliferation of synovial-like cells, giant cells, inflammatory cells and xanthoma cells along tendon sheaths.

Tenosynovial Giant cell tumor

- Group of generally benign intra-articular and soft tissue tumors with common histologic features
- Localized types include giant cell tumors of tendon sheath and localized pigmented villonodular synovitis
- Diffuse types encompass conventional pigmented villonodular synovitis and diffuse-type giant cell tumor
- Localized tumors are generally indolent, whereas diffuse tumors are locally aggressive
Case presentation 1

- Preoperative pain resolved completely
- Marked improvement in hand function

Case 2

- 74 Y/O women
- Six month history of numbness and tingling in the first three fingers of the right hand
- Wrist pain
- Examination: positive Tinel sign, severe atrophy and weakness in the right thenar muscles
Median motor

Median digit II

Median second lumbrical-versus-ulnar interossei distal motor latencies

- Comparing distal latencies of the median motor distal latency recording from the second lumbrical with the ulnar distal latency recording from the interossei
- A difference of more than 0.4 ms is significant

Lumbrical and interossei comparison
Lumbrical and interossei comparison

- Placing an active electrode (G1) slightly lateral to the midpoint of the third metacarpal, with the reference electrode over the proximal interphalangeal joint of the second digit, and stimulating the median and ulnar nerves at the wrist, respectively

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Insertion</th>
<th>Spontaneous activity</th>
<th>Volitional MUAPs</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st dorsal interosseous R</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Extensor indicis proprius R</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Abductor digiti minimi (manus) R</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Flexor pollicis longus R</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Abductor pollicis brevis R</td>
<td>Increased</td>
<td>3+</td>
<td>3+</td>
<td>None</td>
</tr>
<tr>
<td>Pronator teres R</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>None</td>
</tr>
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</table>

**EMG**

<table>
<thead>
<tr>
<th>Muscle</th>
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<th>Volitional MUAPs</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lumbrical-interosseous</td>
<td></td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>
Case 2

• Severe right median neuropathy at the wrist

Case 3

• 33 y/o female
• One month of acute onset of severe pain in the left shoulder and axilla area followed by burning sensation in the first three fingers
• Inability to flex the first three fingers

Examination.

• Weakness in the flexion of the first three fingers
• Sensory loss in the first three fingers

Median motor
Is this CTS?

- No
- Low CMAP Median response with normal latency is not CTS
- If median sensory normal, think C8-T1 radiculopathy or anterior horn disease
- If median sensory is affected, think proximal median neuropathy

Diagnosis

- Subacute proximal median neuropathy distal to the branch of the pronator teres
- Idiopathic brachial neuritis or Parsonage-Turner syndrome

MRI forearm
Case 3

- Treated with IV methylprednisolone with complete resolution of her symptoms

Case 4

- 52 y/o male with two year history gradual onset of numbness and tingling in feet

Examination

- Normal strength except for mild weakness of toes extensors and flexors
- Reduced reflexes at the ankles
- Normal vibration and position
- Decreased sensation to pinprick, touch and temperature to mid shin and palm area

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Onset Lat. (ms)</th>
<th>Normal</th>
<th>Amplitude (mV)</th>
<th>Conduction Velocity (m/s)</th>
<th>Norm. CV (ms*mV)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fibular (Peroneal).R Extensor digitorum brevis.R Ankle</td>
<td>5.0</td>
<td>6.0</td>
<td>3.6</td>
<td>2.00</td>
<td>61.0</td>
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<tr>
<td>Fibula (head)</td>
<td>14.2</td>
<td>3.2</td>
<td>37.0</td>
<td>40.0</td>
<td>40.0</td>
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<tr>
<td>Above Knee</td>
<td>16.2</td>
<td>3.1</td>
<td>100</td>
<td>50</td>
<td>50.0</td>
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<tr>
<td>Tibial.R Abductor hallucis.R Ankle</td>
<td>4.9</td>
<td>6.0</td>
<td>7.2</td>
<td>4.00</td>
<td>15.0</td>
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<tr>
<td>Knee</td>
<td>15.5</td>
<td>3.8</td>
<td>44.0</td>
<td>42</td>
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<td>Median.L Abductor pollicis brevis.R Wrist</td>
<td>4.9</td>
<td>3.8</td>
<td>7.5</td>
<td>5.00</td>
<td>19.0</td>
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<td>Elbow</td>
<td>8.6</td>
<td>6.9</td>
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<td>53</td>
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<td>Ulnar.L Abductor digiti minimi (manus).R Wrist</td>
<td>3.0</td>
<td>3.2</td>
<td>7.7</td>
<td>4.00</td>
<td>26.0</td>
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<tr>
<td>Below elbow</td>
<td>7.1</td>
<td>7.6</td>
<td>240</td>
<td>59</td>
<td>240.0</td>
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<tr>
<td>Above elbow</td>
<td>8.6</td>
<td>7.1</td>
<td>100</td>
<td>67</td>
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<td>Nerve</td>
<td>Muscle Insertion</td>
<td>Spontaneous Activity</td>
<td>Volitional MUAPs</td>
<td>Comments</td>
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<td>Normal</td>
<td></td>
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<tr>
<td>Dorsal interossei (pedis) I &amp; II.R</td>
<td>Increased</td>
<td>Increased</td>
<td>Many</td>
<td>-</td>
<td></td>
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<tr>
<td>Peroneus tertius.R</td>
<td>Normal</td>
<td>1+ Red</td>
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<tr>
<td>Vastus medialis.R</td>
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<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
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<tr>
<td>Ulnar.L to Digit V (little finger).R</td>
<td>Normal</td>
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SNCS Nerve

<table>
<thead>
<tr>
<th>Peak Latency</th>
<th>Amplitude</th>
<th>Conduction Velocity</th>
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<tbody>
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<td></td>
<td></td>
<td></td>
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<tr>
<td>Tibialis anterior.R</td>
<td>4.3*</td>
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</tr>
<tr>
<td>Sural.R to Lat Mal.R</td>
<td>4.00</td>
<td>140</td>
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<td>Median.L to Digit II (index finger).R</td>
<td>3.1</td>
<td>3.6</td>
</tr>
<tr>
<td>Ulnar.L to Digit V (little finger).R</td>
<td>3.0</td>
<td>3.1</td>
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</table>

Notes

<table>
<thead>
<tr>
<th>Muscle Insertion</th>
<th>Spontaneous Activity</th>
<th>Volitional MUAPs</th>
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<tbody>
<tr>
<td>Tibialis anterior.R</td>
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<td>Normal</td>
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<tr>
<td>Gastrocnemius (Medial head).R</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
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<td>Normal</td>
<td>Normal</td>
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<tr>
<td>Dorsal interossei (pedis) I &amp; II.R</td>
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<td>Increased</td>
<td>Many</td>
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<tr>
<td>Peroneus tertius.R</td>
<td>Normal</td>
<td>1+ Red</td>
<td>1+</td>
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<tr>
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<td>Normal</td>
<td>Normal</td>
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<td>Ulnar.L toDigit V (little finger).R</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
</tbody>
</table>
Sensory conduction in medial plantar nerve

Normal values, clinical applications, and a comparison with the sural and upper limb sensory nerve action potentials in peripheral neuropathy

R. J. GUILLOFF AND R. M. SHERRATT
From the National Hospital for Nervous Diseases, Queen Square, London

SUMMARY A method for recording the medial plantar sensory nerve action potential at the ankle with surface electrodes is described. Normal values in 69 control subjects are given and compared with the sural sensory nerve action potential in the same limb in the same subjects. Clinical applications were studied in 57 patients. The procedure may be applied in the diagnosis of L4-5 nerve plexus or root lesions, lesions of the sciatic, posterior tibial, and medial plantar nerves, and is a more sensitive test than other sensory nerve action potentials in the diagnosis of peripheral neuropathy.

SHORT REPORT

ABSTRACT. The objective of this study was to compare the sensitivity of needle electromyography of the abductor hallucis and peroneus tertius muscles in the diagnosis of medium-length-dependent peroneal neuropathy (MLDPN) in 50 patients with clinical evidence of MLDPN. Results demonstrated that the peroneus tertius is as sensitive and more specific than the abductor hallucis. It is particularly useful when more proximal muscles, such as the tibialis anterior and medial gastrocnemius, are not yet involved.

Electromyographic Sensitivity of Peroneus Tertius Relative to Abductor Hallucis in Assessment of Peripheral Neuropathy

ANDREW J. BUCH, M.D. and C. MICHEL HARPER, M.D.*

*Division of Clinical Neurophysiology, Mayo Clinic and Foundation, 200 First Street SW, Rochester, Minnesota 55905, USA
Division of Physical Medicine and Rehabilitation, Mayo Clinic and Foundation, Rochester, Minnesota, USA
Department of Neurology, Mayo Clinic and Foundation, Rochester, Minnesota, USA

August 28, 2009

PERONEUS TERTIUS

Illustration: Deep Peroneal Nerve, Common Peroneal Nerve, Sural Nerve, Posterior Division Sural Nerve, K.S.
Distal large neuropathy

- Always check for medial planate response if younger than 55 year-old
- Always examine distal muscles, including foot muscles

Case 4

- 35 y/o male with medically refractory localization related epilepsy presented with status epilepticus
- Now s/p trach/PEG
- Mechanical ventilation for respiratory failure
- Hypotension, pneumonia and UTI
- Prolonged ICU stay 2 months

Examination

- Unresponsive for 6 weeks
- Cough reflex is absent and gag reflex is absent. Pupils reactive. No movement to peripheral or central stimulation.
Examination

- Flaccid weakness
- Does not withdraw to noxious stimuli
- Absent tendon reflexes
- Diffuse muscle atrophy

• CK 188 (49 - 397 U/L)
SHORT REPORT

ABSTRACT: Critical illness myopathy (CIM) is a frequent cause of generalized weakness in the intensive care unit. Prolonged compound muscle action potential (CMAP) durations have been described in this setting previously, and the CMAP duration further serves as an indicator of poor recovery. In this report, we describe a patient with a prolonged CMAP duration on initial presentation who had no other clinical evidence of muscle weakness. The patient subsequently developed multiple new muscle weaknesses over time, including the quadriceps, abdominal, and iliopsoas muscles. Recognition of the pattern, which has not been widely described, can facilitate the diagnosis of CIM.

Wu et al., Annals of Intensive Care, 2019

PROLONGED COMPOUND MUSCLE ACTION POTENTIAL DURATION IN CRITICAL ILLNESS MYOPATHY

MELANIE S. GODFREY, ET AL.

Department of Neurology, Mayo Clinic, Rochester, Minnesota, USA

12.3.12.2019

Table 1. Maximal compound muscle action potential-duration from healthy controls and critical illness myopathy cohorts.

<table>
<thead>
<tr>
<th>CMAP Group</th>
<th>Healthy Controls</th>
<th>Critical Illness Myopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maximal CMAP Durations in ms</td>
<td>Maximal CMAP Durations in ms</td>
<td></td>
</tr>
<tr>
<td>Healthy Controls</td>
<td>13.2 ± 1.6</td>
<td>28.8 ± 8.6</td>
</tr>
<tr>
<td>Critical Illness</td>
<td>13.2 ± 1.6</td>
<td>28.8 ± 8.6</td>
</tr>
</tbody>
</table>

Note: Values are mean ± standard deviation, all values from 20 healthy controls.

Graphs showing two different muscle waveforms.
<table>
<thead>
<tr>
<th>Muscle</th>
<th>Activity</th>
<th>Insertion</th>
<th>Spontaneous Activity</th>
<th>Volitional MUAPs</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tibialis anterior (L)</td>
<td>Increased</td>
<td>1+</td>
<td>None</td>
<td>None</td>
<td>-</td>
</tr>
<tr>
<td>Gastrocnemius (Medial head) (L)</td>
<td>Increased</td>
<td>1+</td>
<td>None</td>
<td>None</td>
<td>-</td>
</tr>
<tr>
<td>Vastus medialis (L)</td>
<td>Increased</td>
<td>2+</td>
<td>None</td>
<td>None</td>
<td>-</td>
</tr>
<tr>
<td>Iliopsoas (L)</td>
<td>Increased</td>
<td>2+</td>
<td>None</td>
<td>None</td>
<td>-</td>
</tr>
<tr>
<td>1st dorsal interosseous (L)</td>
<td>Increased</td>
<td>1+</td>
<td>None</td>
<td>None</td>
<td>-</td>
</tr>
<tr>
<td>Deltoid (L)</td>
<td>Normal</td>
<td>2+</td>
<td>None</td>
<td>None</td>
<td>-</td>
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</table>

**Direct muscle stimulation**

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Activity</th>
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</tr>
</thead>
<tbody>
<tr>
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<td>Increased</td>
<td>1+</td>
<td>None</td>
<td>None</td>
<td>-</td>
</tr>
<tr>
<td>Gastrocnemius (Medial head) (L)</td>
<td>Increased</td>
<td>1+</td>
<td>None</td>
<td>None</td>
<td>-</td>
</tr>
<tr>
<td>Vastus medialis (L)</td>
<td>Increased</td>
<td>2+</td>
<td>None</td>
<td>None</td>
<td>-</td>
</tr>
<tr>
<td>Iliopsoas (L)</td>
<td>Increased</td>
<td>2+</td>
<td>None</td>
<td>None</td>
<td>-</td>
</tr>
<tr>
<td>1st dorsal interosseous (L)</td>
<td>Increased</td>
<td>1+</td>
<td>None</td>
<td>None</td>
<td>-</td>
</tr>
<tr>
<td>Deltoid (L)</td>
<td>Normal</td>
<td>2+</td>
<td>None</td>
<td>None</td>
<td>-</td>
</tr>
</tbody>
</table>
Conclusion

Abnormal study. The findings are most consistent with severe critical illness myopathy. No evidence for critical illness polyneuropathy.

Direct muscle stimulation

For stimulation, a stainless steel subdermal electrode (12 mm in length, 0.4 mm in diameter, Nicolet Biomedical, Madison, WI) is placed in the distal third of the muscle, away from the end-plate region. Another subdermal needle was placed 5 mm laterally as an anode.

The muscle is stimulated with gradually increasing strength until a clear twitch is palpable (10-100 mA, 0.1 msec, 0.5 Hz).

Guided by the twitch, a recording needle is placed (stainless steel subdermal electrode, 12 mm in length, 0.4 mm in diameter, Nicolet Biomedical) 1 to 3 cm away from the stimulation electrode and moved it until the amplitude of the CMAP is maximized.

A surface electrode was used as a reference.

Rich MM, Muscle and Nerve, 1997;20:665–75
Lefaucheur J, JNNS 2006, 77: 500–506

Direct muscle stimulation

A: Normal pattern
B: Neuromyopathic
C: Myopathic
D: Neurogenic
Case 5

- 52 y/o male with 6 years history of progressive distal weakness in the feet more than hands, burning pain and sensory loss.

Examination

- Exam showed weakness in distal muscles, ADM, FDI, finger spread, finger extensors and APB 4/4
- Foot dorsiflexion, inversion and eversion, toe flexors and extensors 0/5, foot plantar flexion 3/5.
- Tendon reflexes are absent

- Decreased pinprick, touch and temperature to the wrist and knees
- Absent vibration and position in the toes and ankles, decreased vibration and position in the fingers

Case 5

- Treated with multiple courses of IVIG and prednisone without any significant improvement in his symptoms
- Outside EMG with demyelinating features
- VEGF level is normal.
- IgA lambda on serum immunofixation
- Bone survey is negative
- CSF protein 1.448 (0.15–0.45 G/L), WBC 0
MNC

Nerve Latency

Amplitude

Conduction Velocity

Neg Area

Onset Lat.

ms

Normal ≤ mV

Distance mm

Normal CV ≥ ms*mV

Fibular (Peroneal).R Extensor digitorum brevis.R

Ankle NR

NR

60 NR

Tibial.R Abductor hallucis.R

Ankle NR

NR

60 NR

Fibular (Peroneal).R Tibialis anterior.R

Fibula (head) 4.9

0.7*

2.60

100

4.5

Popliteal fossa 9.3

0.6

95

22*

40.0

3.2

Median.R Abductor pollicis brevis.R

Wrist 5.2*

3.8

5.4

5.00

60

19.6

Elbow 16.0

3.3

280

26*

50.0

14.4

Ulnar.R Abductor digiti minimi (manus).R

Wrist 5.4*

3.2

2.8*

4.00

60

10.3

Below elbow 15.9

2.4

235

22*

50.0

10.4

Above elbow 19.9

2.3

100

25*

50.0

9.8

Median elbow NR

NR

NR

Median wrist NR

NR

NR

Notes

Muscle Insertion Spontaneous Activity Volitional MUAPs Comments

Activity Fibs PSW Fasc Other Activation Recruit Dur Amp Poly

Tibialis anterior.R Increased 2+ 2+ None Normal 2+ Red 2+ 2+ 100%

Gastrocnemius (Medial head).R Increased 2+ 2+ None Normal 2+ Red 2+ 2+ 25%

Vastus medialis.R Normal 0 0 None Normal Normal Normal Normal Normal 50%

Tensor fasciae latae.R Normal 0 0 None Normal Normal Normal Normal Normal None

Deltoid.R Normal 0 0 None Normal Normal Normal Normal Normal None

1st dorsal interosseous.R Increased 2+ 2+ None Normal 2+ Red 2+ 2+ 50%
Conclusion

- Abnormal study. There is evidence for a moderate to severe, diffuse demyelinating sensorimotor peripheral neuropathy with secondary axonal loss distally. The findings are more in keeping with an acquired process. The uniform slowing of conduction velocities along with reported finding of IgA lambda are concerning for POEMS syndrome.
PET scan

- Bone marrow biopsy showed 5% lambda-predominant plasma cells

POEMS syndrome

- CT: Showed moderate moderate hepatosplenomegaly
- Thrombocytosis
- Darkening of his skin color

Research Paper

Uniform demyelination and more severe axonal loss distinguish POEMS syndrome from CIDP

Michelle L Maslennikova,1 Eric J Drennan,1 Angela Depoorter2, Jay Mantravadi,3 Galleria A Guerci,1 Peter J Dyck,1 P. James E Dyck1

PATIENTS OF NERVE CONDUCTION ABNORMALITIES IN POEMS SYNDROME

J-B. YAO, MD, BS, SATISH KUMAR, MD, KAZUE OSIYAMA, MD, KAZUHIRO KIMU, MD, and
TAE-MIN PATTN, MD
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Abstract

POEMS syndrome is a rare cause of demyelinating and axonal neuropathy. Electrodiagnostic findings may be ordered, but the two diseases are distinct. To evaluate the neuro-phenotypic features of POEMS syndrome, 16 POEMS syndrome cases were examined. Our results with 13 of 16 patients and CIDP. The patients with POEMS syndrome showed a more severe degree of axonal nerve conduction slowing than with CIDP. Conduction slowing was more prominent in the intermediate nerve segments (forearm or leg segment) than in the distal nerve segments. Less conduction block and temporal dispersion were observed in POEMS syndrome compared with CIDP. Lower limbs were more affected than upper limbs. More axonal loss was observed in POEMS syndrome. The slowing of conduction velocity was more uniform in POEMS syndrome than in CIDP.
46 y/o male with 3 months of progressive proximal and distal muscle weakness and sensory loss with absent reflexes. Negative monoclonal protein and MAG antibody, CSF protein 299 (15-45 mg/dl)

<table>
<thead>
<tr>
<th>Muscles</th>
<th>Needle EMG</th>
<th>Spontaneous Activity</th>
<th>Voluntary MUPs</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tibialis anterior L</td>
<td>Normal</td>
<td>None</td>
<td>Normal</td>
<td>Normal</td>
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<tr>
<td>Vastus lateralis L</td>
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<td>Normal</td>
<td>Normal</td>
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<tr>
<td>Iliopsoas L</td>
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</table>

![Table of nerve conduction studies](image)

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Conduction Velocity</th>
<th>Normal CV</th>
<th>Mean Amplitude</th>
<th>Normal Amplitude</th>
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<tbody>
<tr>
<td>Sural L to Lat Mal L</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Median L to Digit II L</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ulnar L to Digit V L</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

![Recordings](image)
58 y/o male with 15 years of proximal and distal muscle weakness, sensory loss, absent reflexes, IgG lambda. Negative MAG antibody.

Diagnosis

- CIDP
### Acknowledgment

- Margaret Rolle
- Daniella Marks
- Sarah Main
- Allison Owen

### Case presentation

- 69 y/o female
- 2008 Dx with lymphoplasmacytic lymphoma (Waldenstrom macroglobulinemia)
- Elevated monoclonal gammopathy IgM lambda: 2511 (40-230 mg/dl) 12/2014 and bone marrow biopsy
- S/P rituximab treatment 2010, 2012 and 2015, felt to be in remission

### Diagnosis

- CIDP

### Table

<table>
<thead>
<tr>
<th>Model</th>
<th>Spinal level</th>
<th>Spontaneous activity</th>
<th>Normal MUAP</th>
<th>Comment</th>
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<tbody>
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<td>R</td>
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<td>Gastrocnemius (Medial head)</td>
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<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Vastus medialis</td>
<td>R</td>
<td>Normal</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Tensor fasciae latae</td>
<td>R</td>
<td>Normal</td>
<td>0</td>
<td>None</td>
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</tbody>
</table>
Case presentation

- 02-2015: 3 weeks history of left arm pain and numbness
- The pain is 8/10 worse at night, burning type pain, the skin hurt to touch, is hypersensitive
- MRI C-spine 2015 unremarkable, ACDF C5, C6
- 09-2016: Muscle wasting and weakness since 09-2016, repeat MRI c-spine again unremarkable
- 09-2016: underwent C6-C7 foraminotomy. Pain improved after surgery, but recurs after 7-10 days, pain got worse in the last 3 months.
- 01-2017: Pain in the left side of the face
- 03-2017 presented to OHSU

Case presentation

- Decreased sensation left side of face
- Diffuse weakness in left arm
- Mild in left shoulder rotators, deltoid, triceps, biceps 4/5
- Severe in wrist extensors, finger extensors, finger flexors and hand muscles 1-2/5

Case presentation

- Absent reflexes left biceps, triceps, brachioradialis
- Sensory loss in left medial antebrachial nerve distribution

Localization
**Localization**

- Brachial plexus
- Trigeminal nerve

**Differential diagnosis**

- Involvement by Waldenstrom macroglobulinemia
- Vasculitic process

### Motor Nerve Conduction Studies

<table>
<thead>
<tr>
<th>Location</th>
<th>Data</th>
<th>Normal</th>
<th>Data</th>
<th>Normal</th>
<th>Data</th>
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<tbody>
<tr>
<td>Sural L to Lat Mal L Post Calf</td>
<td>Latency ms</td>
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<td>Amplitude 4.0</td>
<td>85</td>
<td>Conduction Velocity 4.0</td>
<td>100</td>
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<tr>
<td>Sural R to Lat Mal R Post Calf</td>
<td>Latency ms</td>
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<td>Amplitude 4.0</td>
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<td>Conduction Velocity 5.0</td>
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</tr>
<tr>
<td>Median L to Digit II (Index finger) L Wrist</td>
<td>Latency ms</td>
<td>3.7*</td>
<td>Amplitude 3.6</td>
<td>10</td>
<td>Conduction Velocity 16</td>
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</tr>
<tr>
<td>Ulnar L to Digit V (Little finger) L Wrist</td>
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<td>Amplitude 3.1</td>
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<td>Amplitude 2.9</td>
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<td>Conduction Velocity 7</td>
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<tr>
<td>Medial Ante Brachial Cutaneous L to Forearm L Elbow</td>
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<td>Amplitude 2.8</td>
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<td>Conduction Velocity 8</td>
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<td>Amplitude 2.6</td>
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<td>Ulnar R to Digit V (Little finger) R Wrist</td>
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<td>Amplitude 3.1</td>
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<td>Conduction Velocity 21</td>
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<td>Radial R to Anatomical Snuff Box R Forearm</td>
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<td>Amplitude 2.9</td>
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<td>Conduction Velocity 27</td>
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<td>Medial Ante Brachial Cutaneous R to Forearm R Elbow</td>
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<td>Amplitude 2.6</td>
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<td>Conduction Velocity 33</td>
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### Sensory Nerve Conduction Studies

<table>
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<tr>
<th>Location</th>
<th>Data</th>
<th>Normal</th>
<th>Data</th>
<th>Normal</th>
<th>Data</th>
<th>Normal</th>
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<tbody>
<tr>
<td>Sural L</td>
<td>Latency ms</td>
<td>0.0</td>
<td>Amplitude 0.4</td>
<td>85</td>
<td>Conduction Velocity 0.4</td>
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<td>Sural R</td>
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<td>Amplitude 0.4</td>
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<td>Conduction Velocity 0.4</td>
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<tr>
<td>Median L</td>
<td>Latency ms</td>
<td>0.0</td>
<td>Amplitude 0.4</td>
<td>10</td>
<td>Conduction Velocity 0.4</td>
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</tr>
<tr>
<td>Ulnar L</td>
<td>Latency ms</td>
<td>0.0</td>
<td>Amplitude 0.4</td>
<td>14</td>
<td>Conduction Velocity 0.4</td>
<td>110</td>
</tr>
<tr>
<td>Radial L</td>
<td>Latency ms</td>
<td>0.0</td>
<td>Amplitude 0.4</td>
<td>7</td>
<td>Conduction Velocity 0.4</td>
<td>150</td>
</tr>
<tr>
<td>Medial Ante Brachial Cutaneous L</td>
<td>Latency ms</td>
<td>0.0</td>
<td>Amplitude 0.4</td>
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<td>Conduction Velocity 0.4</td>
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<td>0.0</td>
<td>Amplitude 0.4</td>
<td>18</td>
<td>Conduction Velocity 0.4</td>
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<td>Median R</td>
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<td>Amplitude 0.4</td>
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<td>Conduction Velocity 0.4</td>
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<td>Ulnar R</td>
<td>Latency ms</td>
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<td>Radial R</td>
<td>Latency ms</td>
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<td>Amplitude 0.4</td>
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<td>Conduction Velocity 0.4</td>
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</table>

***Empty data fields denote absent responses.***
Needle EMG Examination

<table>
<thead>
<tr>
<th>Muscles</th>
<th>Activity</th>
<th>Spontaneous Activity</th>
<th>Volitional MUAPs</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st dorsal interosseous L</td>
<td>Increased</td>
<td>1+</td>
<td>1+</td>
<td>Many</td>
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<tr>
<td>Extensor indicis proprius L</td>
<td>Increased</td>
<td>2+</td>
<td>2+</td>
<td>None</td>
</tr>
<tr>
<td>Flexor pollicis longus L</td>
<td>Increased</td>
<td>1+</td>
<td>1+</td>
<td>Rare</td>
</tr>
<tr>
<td>Pronator teres L</td>
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<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Biceps brachii L</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Deltoid</td>
<td>Increased</td>
<td>1+</td>
<td>1+</td>
<td>None</td>
</tr>
<tr>
<td>Triceps brachii L</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Infraspinatus L</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Tibialis anterior L</td>
<td>Normal</td>
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<td>0</td>
<td>None</td>
</tr>
<tr>
<td>Gastrocnemius (Medial head) L</td>
<td>Normal</td>
<td>0</td>
<td>0</td>
<td>None</td>
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<tr>
<td>Vastus medialis L</td>
<td>Normal</td>
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<td>0</td>
<td>None</td>
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<tr>
<td>Biceps femoris (long head) L</td>
<td>Normal</td>
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</table>

EMG

- Abnormal study. There is evidence for a moderate to severe, left lower trunk brachial plexopathy with milder involvement of the upper trunk. Those findings given the patient’s history are concerning for infiltration of the nerves by the lymphoproliferative disorder.
- There is also evidence for superimposed bilateral ulnar neuropathy at the elbow.

Labs

- IgM level: 485 (40-230 mg/dl)
- KAPPA LT CHAIN, SERUM: 3.3 - 19.4 mg/L, 12.0
- LAMBDAtA CHAIN, SERUM: 5.7 - 26.3 mg/L, 16.7
- KAPPA/LAMBDA RATIO: 0.26 - 1.65 ratio, 0.25
- IgM Lambda on immunofixation: 0.3 g/dl

Case presentation 2

- C-REACTIVE PROTEIN: <10.0 mg/L, 2.5
- SEDIMENTATION RATE: 0 - 30 mm/hr, 60
- ANA, ENA, ANCA, cryoglobulin negative
Case presentation 2

- CSF WBC 0 - 5 / cu mm 0
  - NEUTROPHIL (CSF) 0 - 6 % 0
  - LYMPHOCYTES (CSF) 68 - 80 % 17
  - MONOCYTES (CSF) 15 - 45 % 83
- TOTAL PROTEIN CSF 15 - 45 mg/dL 150
- LEUKEMIA/LYMPHOMA MARKER - CSF/BODY FLUID
- Cerebrospinal fluid:
  - Involvement by B cell lymphoma

MRI brain W/Wo contrast

- Right nasopharyngeal mass with abnormal enhancement of the bilateral V3 segments of the trigeminal nerves at the level of the foramina ovale and just below the skull base, with involvement of the left Meckel cave. Findings are favored to represent lymphoma/lymphoproliferative disease. Nasopharyngeal carcinoma with perineural spread is considered less likely.

MRI of total spine

- Abnormal marrow signal and enhancement throughout the C5-T1 vertebral bodies with thickening of the bilateral brachial plexus cysts nerve roots, left greater than right. Findings overall are concerning for lymphoma or other malignant marrow replacing process.

STIR coronal
Contrast coronal

Contrast axial

Bing-Neel syndrome

- Infiltration of the central nervous system or meninges by plasmacytoid lymphocytes
Bing-Neel syndrome

- The median time interval between the appearance of neurological symptoms and the diagnosis of BNS was 4 months, with an upper limit of 36 months.

Case presentation

- High dose MTX/leukovorin rescue and Rituximab in 4/1/17
- She developed elevated LFT's, mental status changes, and marrow suppression

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Case presentation

- Treated with ibrutinib 420 mg once a day
- Remarkable improvement in her symptoms
- Pain resolved and weakness improved
- In remission again
IMBRUVICA® (ibrutinib)

- Ibrutinib is a small molecule inhibitor of BTK (Bruton's tyrosine kinase).
- BTK is a signaling molecule of the B cell antigen receptor (BCR) and cytokine receptor pathways.
- BTK’s role in signaling through the B cell surface receptors results in activation of pathways necessary for B cell trafficking, chemotaxis, and adhesion.
- Nonclinical studies show that ibrutinib inhibits malignant B cell proliferation and survival *in vivo* as well as cell migration and substrate adhesion *in vitro*.

IMBRUVICA® (ibrutinib)

- Mantle cell lymphoma (MCL) who have received at least one prior treatment.
- Chronic lymphocytic leukemia (CLL)/Small lymphocytic lymphoma (SLL).
- Chronic lymphocytic leukemia (CLL)/Small lymphocytic lymphoma (SLL) with 17p deletion.
- Waldenström's macroglobulinemia (WM).
- Marginal zone lymphoma (MZL) who require a medicine by mouth or injection (systemic therapy) and have received a certain type of prior treatment.
- Chronic graft versus host disease (cGVHD) after failure of 1 or more lines of systemic therapy.