Myopathy and Neuropathy in Internal Medicine

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A series of cases designed to make you feel weak and get on your nerves
OBJECTIVES:
• To present an approach to diagnosing myopathy and neuropathy in internal medicine
• To review and discuss challenging cases of peripheral neuropathy and myopathy due to underlying internal medicine etiologies
Participation is not optional
What is the Motor Unit?

- The motor unit consists of motor neuron, motor nerve fibers, neuromuscular junction, and the activated muscle fibers.
What is the Motor Unit?

- Motor Neuron
- Peripheral Nerve
- NMJ
- Muscle
How can lower motor neuron disorders be separated?

• Sometimes it is difficult, but there are particular examination features that can help to separate these conditions

• First, sensory involvement always tells you it is the peripheral nerve affected
Sensory Loss? → Neuropathy

- (Sensory Loss)

  Is there diplopia, dysarthria, dysphagia, +/- ptosis?
  - (No diplopia, dysarthria, dysphagia, +/- ptosis)

  Are there upper motor neuron signs, fasciculations?
  - (No upper motor neuron signs, fasciculations)

  Is there proximal dominant weakness?
  - (No proximal dominant weakness)

  Consider myasthenia gravis (NMJ Disorder)

+ (Sensory Loss)

  Consider motor neuron disease

  Consider myopathy
Myopathy due to internal medicine causes generally has one clinical phenotype:

- **Proximal Weakness**
When myopathy is due to another cause (genetic, degenerative) there can be differing clinical phenotypes:

Myotonic Dystrophy – Distal dominant weakness

Inclusion Body Myositis – finger flexors, quadriceps predominantly affected
Peripheral neuropathy is a little more heterogeneous in its clinical phenotype

- Most due to internal Medicine causes are length dependent, or stocking and glove
Peripheral Neuropathy

However, other patterns are possible, particularly in cases of inflammatory, toxic or vasculitic causes:

A mononeuritis multiplex pattern results from vasculitis.

Some inflammatory/toxic presentations are proximal.
Documented Approaches to Peripheral Neuropathy

Dyck has proposed steps in the diagnosis of peripheral neuropathy:

- Ten Steps in characterizing patients with peripheral neuropathy (Neurology 47:10-18)
- 10 P’s in characterizing patients with peripheral neuropathy (Neurology 42:14-18)

Schaumberg et al has proposed this as well:

- Diagnostic Algorithm (Disorders of Peripheral Nerve, Edition 2, No. 36)
“Ten Steps”

1) Anatomic-pathologic pattern
2) Confirmation via testing
3) Pathology and Mechanism
4) Onset and Course
5) Inherited or Acquired
6) Association with diseases
7) Appropriate testing
8) Kin evaluation
9) Nerve biopsy
10) Therapeutic Trial
“Ten P’s”

1) Pattern
2) Population of neurons
3) Part of neuron
4) Physiology
5) Pathology
6) Prickling
7) Phenomena
8) Pedigree
9) Plasma
10) Pharmacology
Rate of agreement between final diagnosis and sequential diagnosis after each step of evaluation (Suarez GA et al, Neurology 57:1118-1120)

1) referral history
2) referral exam
3) referral EP
4) Referral pathology (when available)
5) neurologic history at Mayo
6) exam at Mayo
7) EP at Mayo
8) hematology
9) biochemistry
10) serum and urine protein electrophoresis
11) CSF
12) quantitative sensory exam
13) immunology
14) hepatitis, Lyme, HIV, and syphilis
15) radiography
16) biopsied tissue
A Quick and Easy Approach

1) History+Exam

2) Pattern – physical exam phenotype

3) Ax/Demyel – is it axonal or demyelinating?

4) Systemic – what systemic features are present?

5) Confirm – which confirmatory tests are important?
History and Exam

• The history obtained by the neurologist of referral provides the largest increase in diagnostic sensitivity (16%)

• Therefore, this could be considered the most important step

• What are the important historical aspects in the diagnosis of peripheral neuropathy?
## Time Course

<table>
<thead>
<tr>
<th>Acute onset (within days)</th>
<th>Subacute onset (weeks to months)</th>
<th>Chronic course/insidious onset</th>
<th>Relapsing/remitting course</th>
</tr>
</thead>
<tbody>
<tr>
<td>Guillain-Barré syndrome</td>
<td>Maintained exposure to toxic agents/medications</td>
<td>Hereditary motor sensory neuropathies</td>
<td>Guillain-Barré syndrome</td>
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<tr>
<td>Acute intermittent porphyria</td>
<td>Persisting nutritional deficiency</td>
<td>Dominantly inherited sensory neuropathy</td>
<td>CIDP</td>
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<tr>
<td>Critical illness polyneuropathy</td>
<td>Abnormal metabolic state</td>
<td>CIDP</td>
<td>HIV/AIDS</td>
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<tr>
<td>Diphtheric neuropathy</td>
<td>Paraneoplastic syndrome</td>
<td></td>
<td>Toxic</td>
</tr>
<tr>
<td>Thallium toxicity</td>
<td></td>
<td>CIDP</td>
<td>Porphyria</td>
</tr>
</tbody>
</table>

**History+Exam**

Pattern  Ax/Demyel  Systemic  Confirm
History and Exam

• What is the pattern of neuropathy?

Distal and Symmetric

Endocrine diseases
- Diabetes mellitus
- Hypothyroidism
- Acromegaly

Nutritional diseases
- Alcoholism
- Vitamin B₁₂ deficiency
- Folate deficiency
- Whipple's disease
- Postgastrectomy syndrome
- Thiamine deficiency

History+Exam Pattern Ax/Demyel Systemic Confirm
History and Exam

• What is the pattern of neuropathy?

Proximal and Symmetric

- Guillain-Barré syndrome
- CIDP
- Diabetes mellitus
- Porphyria
- Osteosclerotic myeloma
- Waldenstrom's
- Monoclonal gammopathy of undetermined significance
- Acute arsenic polyneuropathy
- Lymphoma

Diphtheria
HIV/AIDS
Lyme disease
Hypothyroidism
Vincristine

History+Exam Pattern Ax/Demyel Systemic Confirm
History and Exam

• What is the pattern of neuropathy?

Polyneuropathy with Cranial Nerve Involvement

Diabetes mellitus
Guillain-Barré syndrome
HIV/AIDS
Lyme disease
Sarcoidosis
Neoplastic invasion of skull base or meninges
Diphtheria
History and Exam

• What is the pattern of neuropathy?

Polyneuropathy with Predominant Arm Involvement

Guillain-Barré syndrome
Diabetes mellitus
Porphyria
Hereditary motor sensory neuropathy
Vitamin $B_{12}$ deficiency
Hereditary amyloid neuropathy type II
Lead neuropathy
Tangier disease
• What is the pattern of neuropathy?

**Mononeuritis Multiplex**

- Vasculitis
- Primary systemic vasculitis:
  - Polyarteritis nodosa
  - Churg-Strauss syndrome
  - Rheumatoid arthritis
  - Sjögren's syndrome
- Primary PNS Vasculitis
- Multiple compression palsies
- Hereditary neuropathy with liability to pressure palsies
- Other
  - Sarcoidosis
  - Lymphoma
  - Carcinoma
  - Amyloid
Why separate into axonal and demyelinating?

Features on exam may be clues to underlying pathology:

- **Axonal**
  - Predominance: Distal
  - Reflexes: May be Diminished
  - Sensory Pattern: Small and Large Fiber

- **Demyelinating**
  - Predominance: May be Proximal
  - Reflexes: Often Absent
  - Sensory Pattern: Large Fiber

History+Exam Pattern Ax/Demyel Systemic Confirm
Why separate into axonal and demyelinating?

**Axonal**

- Vincristine (Oncovin, Vincosar PFS)
- Paclitaxel (Taxol)
- Nitrous oxide
- Colchicine
- Isoniazid (Laniazid)
- Hydralazine (Apresoline)
- Metronidazole (Flagyl)
- Pyridoxine (Nestrex, Beesix)
- Didanosine (Videx)
- Lithium
- Alpha interferon (Roferon-A, Intron A, Alferon N)

**Demyelinating**

- Dapsone
- Phenytoin (Dilantin)
- Cimetidine (Tagamet)
- Disulfiram (Antabuse)
- Chloroquine (Aralen)
- Ethambutol (Myambutol)
- Amitriptyline (Elavil, Endep)
- Anti-HIV Meds

**History + Exam Pattern**

Ax/Demyel Systemic Confirm
Is there a toxic cause?

Demyelinating

Amiodarone (Cordarone)
Chloroquine
Suramin (Fourneau 309, Bayer 205, Germanin)
Gold

Neuronopathy

Thalidomide (Synovir)
Cisplatin (Platinol)
Pyridoxine
Systemic Features

- Nerve hypertrophy: leprosy, neurofibromatosis, HMSN 1 and 3, acromegaly, Refsum's disease, and rarely CIDP

- Ocular manifestations include:
  - dry eyes ➔ Sjogren's, amyloidosis, HSAN
  - scleritis ➔ Vasculitis, connective tissue disease
  - uveitis ➔ sarcoid, Behcet's, Rheumatoid
  - corneal opacities ➔ Fabry's, amyloidosis
  - cataracts ➔ Fabry's, Refsum's
  - optic atrophy ➔ Refsum's, HMSN
  - retinitis pigmentosa ➔ Refsum's, HMSN
Systemic Features

• Dermatological exam:
• Alopecia $\rightarrow$ thallium

• tightly curled hair/optic atrophy $\rightarrow$ giant axonal neuropathy

• white transverse nail bands (Mees’ lines) $\rightarrow$ arsenic/thallium

• telangiectasias on abdomen/buttocks $\rightarrow$ Fabry's disease
• Dermatological exam:
  • purpuric skin eruptions of legs → cryoglobulinemia/vasculitides
  • skin pigmentation changes or hypertrichosis → POEMS syndrome
  • Skin lesions – leprosy
  • enlarged yellow-orange tonsils → Tangier disease
  • pes cavus/hammer toes → CMT disease
  • overriding toes/ichthyosis → Refsum's disease
Pattern of Systemic Disease?

- Endocrinological diseases have a high association with relation to peripheral neuropathy
  - Diabetes
  - Thyroid Disease
  - POEMS
  - Acromegaly
  - Adrenoleukodystrophy
  - MEN 2b
Pattern of Systemic Disease?

- Dozens of other diseases may also have relation to peripheral neuropathy in specific cases

**Nutritional diseases**
- Alcoholism
- Vitamin $B_{12}$ deficiency
- Folate deficiency
- Whipple's disease
- Post-gastrectomy syndrome
- Thiamine deficiency
- Vitamin $B_6$ def/excess

**Connective tissue diseases**
- Rheumatoid arthritis
- Polyarteritis nodosa
- Systemic lupus erythematosus
- Churg-Strauss vasculitis
- Cryoglobulinemia

History+Exam Pattern Ax/Demyel Systemic Confirm
### Pattern of Systemic Disease?

<table>
<thead>
<tr>
<th>Hematological</th>
<th>Neoplastic</th>
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<tbody>
<tr>
<td>Uremia</td>
<td>Carcinomatous axonal sensorimotor polyneuropathy</td>
</tr>
<tr>
<td>Hepatic Disease (including hepatities)</td>
<td>Lymphomatous axonal sensorimotor polyneuropathy</td>
</tr>
<tr>
<td>Polycythemia Rubra Vera</td>
<td></td>
</tr>
<tr>
<td>Primary Thrombocytosis</td>
<td></td>
</tr>
<tr>
<td>Leukemias</td>
<td></td>
</tr>
<tr>
<td>Monoclonal gammopathies</td>
<td></td>
</tr>
<tr>
<td>Myelomas</td>
<td></td>
</tr>
<tr>
<td>Lymphomas</td>
<td></td>
</tr>
<tr>
<td>Amyloidoses</td>
<td></td>
</tr>
</tbody>
</table>
Pattern of Systemic Disease?

Infectious and Other

- Hypophosphatemia
- Critical illness polyneuropathy
- Amyloidosis
- Gouty neuropathy
- Infectious diseases
  - AIDS
  - Lyme disease
  - Leprosy
- Sarcoidosis
157 patients suspected of chronic polyneuropathy:

*Polyneuropathy with known cause (n=105)*

*Polyneuropathy cause disclosed by initial investigations (n=11)*

*No neuropathy (n=32)*

*Signs and symptoms of neuropathy, not confirmed by electrophysiology (n=9)*

**Obvious Causes**

**Polyneuropathy with known cause (67%):**

- Diabetes mellitus (n=60)
- HIV (n=21)
- Alcoholism (n=11)
- Drug induced (n=7)
- Renal failure (n=6)
157 patients suspected of chronic polyneuropathy:

Polyneuropathy with cause disclosed by initial investigations (7%): ESR, Hb, MCV, WBCs, platelets, Na+, K+, creatinine, ALP, glucose, BUN, GGT

These conditions were deemed “obvious” and further investigations were not fruitful:

- Diabetes mellitus
- HIV
- Alcoholism
- Known Drug induced
- Renal failure

Rosenberg NR et al. JNNP 71:205-209
Confirmatory investigations would include nerve conduction studies, and specific studies:

- Diabetes mellitus – measures of glycemic control
- HIV – viral loads
- Alcoholism – CBC indices
- Renal failure – renal indices
What are appropriate studies in yet idiopathic cases?

- CBC
- ESR
- 2hGGT
- HbA$_{1c}$
- Electrolytes, BUN, creatinine, Liver enzymes and function tests
- Vitamin B$_{12}$, CK, TSH
- Fasting methylmalonic acid
- Serum protein electrophoresis
Nerve Conduction Studies and EMG

- are particularly useful for classification of neuropathy
- identification of pattern (mononeuritis multiplex, multiple radiculopathies, peripheral neuropathy)
- population of fibers affected (motor, large sensory)
- portion of neuron involved (neuronal vs. axonal)
- pathological pattern (axonal, demyelinating)
- discrimination from mimicking entities
SECOND STAGE DIAGNOSTICS

Additional Blood and Urine Studies for Secondary Diagnostic Testing:

RF ANA ENA ANCA
Cryoglobulins
Syphilis HIV Lyme serology
Urine immunoelectrophoresis
Urine Aminolevulinic acid, porphobilinogen
24-hr urine for heavy metals (Pb, Ti, As)
Hepatitis B and C serology

+ others
When should a nerve biopsy be considered?

- Clinical features
  - *asymmetric* neuropathy
  - sensory loss
  - age > 65 years
  - significant functional disability
- Specific diagnoses are sought

In general, nerve biopsy is of greatest yield when a specific diagnosis is being considered.

*Don’t do it just because of a lack of diagnosis – it will leave pain, numbness and infection risk*
CASE 1

• 51F presenting with paraplegia of subacute onset in March 2012

• History of DM2*2y without complications

• Started to lose weight on purpose with change in diet (2011)

• PMHx also of possible rheumatoid arthritis diagnosed in 2008
CASE 1

• In July 2010, she was diagnosed with squamous cell Ca of the cervix

• She travelled to Mexico in August 2010 to receive chelation therapy. Believed to be in remission during 2011
CASE 1

• Received Chemotherapy consisting of carboplatin and taxol in late 2010 and early 2011
• Doing well during early 2011
• Walking on own during January 2012
• Reported having numbness and tingling affecting her toes in mid-2011
CASE 1

• On February 10, 2012, she awoke having difficulty moving her legs
• Unable to walk on her own
• Believes that she went to bed able to walk normally the night before and had walked out of the hospital after an appt the day before
CASE 1

• Intensified bilateral leg pain developed over next few days
• Pain was radiating into the anteromedial thighs bilaterally
• There was change in numbness or tingling based upon history
• There was some background of mild mechanical back pain, without change
CASE 1

- Bowel and bladder function reported normal
- No other history of note except for an incisional hernia repair
- Medications: metformin, advil
- Allergies: nil
- Family history: unremarkable
- Social history: prior caretaker, on disability. No alcohol or smoking
Initial Neuro Exam (2 days after onset of bilateral leg problem)

Normal cranial nerve function
Normal neck strength
Normal strength to left and right arm

Leg strength:

<table>
<thead>
<tr>
<th>Left</th>
<th>Right</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hip Flexor</td>
<td>Hip Flexor</td>
</tr>
<tr>
<td>Hip Extensor</td>
<td>Hip Extensor</td>
</tr>
<tr>
<td>Hip Abductors</td>
<td>Hip Abductors</td>
</tr>
<tr>
<td>Hip Adductors</td>
<td>Hip Adductors</td>
</tr>
<tr>
<td>Quads</td>
<td>Quads</td>
</tr>
<tr>
<td>Hamstrings0</td>
<td>Hamstrings0</td>
</tr>
<tr>
<td>Tibialis Anterior</td>
<td>Tibialis Anterior</td>
</tr>
<tr>
<td>Tibialis Posterior</td>
<td>Tibialis Posterior</td>
</tr>
<tr>
<td>Peronei</td>
<td>Peronei</td>
</tr>
<tr>
<td>Gastrocnemius</td>
<td>Gastrocnemius</td>
</tr>
<tr>
<td>EHL</td>
<td>EHL</td>
</tr>
<tr>
<td>EDB</td>
<td>EDB</td>
</tr>
<tr>
<td>Toe Flexors</td>
<td>Toe Flexors</td>
</tr>
</tbody>
</table>

- Hip Flexor: 0
- Hip Extensor: 0
- Hip Abductors: 0
- Hip Adductors: 0
- Quads: 0
- Hamstrings0: 0
- Tibialis Anterior: 0
- Tibialis Posterior: 0
- Peronei: 0
- Gastrocnemius: 0
- EHL: 1
- EDB: 1
- Toe Flexors: 1
Initial Neuro Exam (2 days after onset of bilateral leg problem)

No atrophy nor fasciculations could be detected, but she was emaciated
Reflexes hyporeflexic to brachioradialis, biceps, normal to triceps, absent to the knees and ankles
Plantars bilaterally mute
Normal tone to arms; total flaccidity to both legs
Subjective decreased sensation (pin, $T^\circ$) over the entirety of both legs up to the abdomen at the level of just below the umbilicus ($T_{11}$) bilaterally; similar loss of pinprick over all sacrolumbar dermatomes and up to $T_{11}$ over the back bilaterally
Normal sensation to the upper limbs
Initial Neuro Exam (2 days after onset of bilateral leg problem)

Impaired vibration threshold with no vibration detection to left ASIS and right knee, with normal detection to fingertips and xiphoid process

No proprioception to level of knees bilaterally

Coordination normal to upper extremities

Gait could not be tested
Differential Diagnosis and Plan

What are you thinking?
# Initial Nerve Conduction Studies

## Nerve Conduction Studies

### Anti Sensory Summary Table

<table>
<thead>
<tr>
<th>Site</th>
<th>NR</th>
<th>Onset (ms)</th>
<th>Peak (ms)</th>
<th>O-P Amp (μV)</th>
<th>Neg Dur (ms)</th>
<th>Site1</th>
<th>Site2</th>
<th>Dist (cm)</th>
<th>Vel (m/s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left Sup Peron Anti Sensory (Ant Lat Mall) Calf</td>
<td>NR</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>10.0</td>
<td></td>
</tr>
<tr>
<td>Right Sup Peron Anti Sensory (Ant Lat Mall) Calf</td>
<td>1.7</td>
<td>2.0</td>
<td>5.0</td>
<td>1.06</td>
<td>Calf</td>
<td>Ant Lat Mall</td>
<td></td>
<td>10.0</td>
<td>59</td>
</tr>
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</table>

## Motor Summary Table

<table>
<thead>
<tr>
<th>Site</th>
<th>NR</th>
<th>Onset (ms)</th>
<th>O-P Amp (mV)</th>
<th>Neg Area (mV·ms)</th>
<th>Amp% (Prev)</th>
<th>Neg Dur (ms)</th>
<th>Site1</th>
<th>Site2</th>
<th>Dist (cm)</th>
<th>Vel (m/s)</th>
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</thead>
<tbody>
<tr>
<td>Left Peroneal Motor (Ext Dig Brev) Ankle</td>
<td>4.8</td>
<td>3.2</td>
<td>9.19</td>
<td>100.0</td>
<td>4.77</td>
<td></td>
<td>Ankle</td>
<td>Ext Dig Brev</td>
<td>8.0</td>
<td>42</td>
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<tr>
<td>B Fib</td>
<td>11.4</td>
<td>2.7</td>
<td>8.08</td>
<td>84.4</td>
<td>5.23</td>
<td></td>
<td>B Fib</td>
<td>Ankle</td>
<td>28.0</td>
<td>42</td>
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<tr>
<td>Poplt</td>
<td>12.9</td>
<td>3.2</td>
<td>6.29</td>
<td>81.5</td>
<td>6.25</td>
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<td>Poplt</td>
<td>B Fib</td>
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<tr>
<td>Right Peroneal Motor (Ext Dig Brev) Ankle</td>
<td>4.2</td>
<td>1.3</td>
<td>4.13</td>
<td>100.0</td>
<td>5.16</td>
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<td>Ankle</td>
<td>Ext Dig Brev</td>
<td>8.0</td>
<td>41</td>
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<tr>
<td>B Fib</td>
<td>10.8</td>
<td>1.1</td>
<td>3.46</td>
<td>84.6</td>
<td>5.70</td>
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<tr>
<td>Poplt</td>
<td>13.2</td>
<td>0.9</td>
<td>3.02</td>
<td>81.8</td>
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<td>Poplt</td>
<td>B Fib</td>
<td>10.0</td>
<td>42</td>
</tr>
<tr>
<td>Left Tibial Motor (Abd Hall Brev) Ankle</td>
<td>3.2</td>
<td>5.4</td>
<td>12.43</td>
<td>100.0</td>
<td>5.16</td>
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<td>Abd Hall Brev</td>
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<td>Knee</td>
<td>13.0</td>
<td>4.0</td>
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<td>Knee</td>
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<td>Right Tibial Motor (Abd Hall Brev) Ankle</td>
<td>3.4</td>
<td>3.1</td>
<td>9.73</td>
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<td>Knee</td>
<td>Ankle</td>
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</tbody>
</table>
Initial Nerve Conduction Studies

### F Wave Studies

<table>
<thead>
<tr>
<th>Side</th>
<th>Nerve (Mrkrs) (EDB)</th>
<th>F-Lat (ms)</th>
<th>Lat Norm (ms)</th>
<th>L-R F-Lat (ms)</th>
<th>L-R Lat Norm</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left</td>
<td>Peroneal</td>
<td>&lt;60</td>
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<td></td>
<td>&lt;5.1</td>
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<tr>
<td>Right</td>
<td>Peroneal</td>
<td>&lt;60</td>
<td></td>
<td></td>
<td>&lt;5.1</td>
</tr>
<tr>
<td>Left</td>
<td>Tibial (Mrkrs) (Ahd Hallucis)</td>
<td>&lt;61</td>
<td></td>
<td></td>
<td>&lt;5.7</td>
</tr>
<tr>
<td>Right</td>
<td>Tibial (Mrkrs) (Ahd Hallucis)</td>
<td>&lt;61</td>
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<td></td>
<td>&lt;5.7</td>
</tr>
</tbody>
</table>

### EMG

<table>
<thead>
<tr>
<th>Side</th>
<th>Muscle</th>
<th>Nerve</th>
<th>Root</th>
<th>Ins Act</th>
<th>Fib s</th>
<th>Ps w</th>
<th>Fasie</th>
<th>Spont Act</th>
<th>Amp</th>
<th>Dur</th>
<th>Poly</th>
<th>Recr</th>
<th>Int Pat</th>
<th>Firing rate</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left</td>
<td>1stDorlar</td>
<td>Ulnar</td>
<td>C8-T1</td>
<td>Nm</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>Nml</td>
<td>Nml</td>
<td>Nm</td>
<td></td>
<td></td>
<td>Nml</td>
<td></td>
<td>Nml</td>
</tr>
<tr>
<td>Left</td>
<td>AntTibialis</td>
<td>Dp Br Peron</td>
<td>L4-5</td>
<td>Nm</td>
<td>2+</td>
<td>2+</td>
<td>0</td>
<td>Nml</td>
<td>Nml</td>
<td>Nm</td>
<td></td>
<td></td>
<td>Nml</td>
<td></td>
<td>Nml</td>
</tr>
<tr>
<td>Left</td>
<td>VastusLat</td>
<td>Femoral</td>
<td>L2-4</td>
<td>Nm</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>Nml</td>
<td>Nml</td>
<td>Nm</td>
<td></td>
<td></td>
<td>Nml</td>
<td></td>
<td>Nml</td>
</tr>
<tr>
<td>Left</td>
<td>Iliopsoas</td>
<td>Femoral</td>
<td>L2-3</td>
<td>Nm</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>Nml</td>
<td>Nml</td>
<td>Nm</td>
<td></td>
<td></td>
<td>Nml</td>
<td></td>
<td>Nml</td>
</tr>
</tbody>
</table>

Reduce d Activation: Nm
Reduce d Activation: Nm
Reduce d Activation: Nm
## Laboratory Testing

<table>
<thead>
<tr>
<th>Test</th>
<th>Value</th>
<th>Reference Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>CSF protein</td>
<td>0.62</td>
<td>(&gt;0.65)</td>
</tr>
<tr>
<td>CSF WBC</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>CSF culture</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>CSF viral PCR</td>
<td>–</td>
<td></td>
</tr>
</tbody>
</table>

### Blood Work

<table>
<thead>
<tr>
<th>Test</th>
<th>Value</th>
<th>Reference Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mg</td>
<td>0.64</td>
<td>(&gt;0.65)</td>
</tr>
<tr>
<td>Creatinine</td>
<td>130</td>
<td>(&lt;100)</td>
</tr>
<tr>
<td>Serum folate</td>
<td>12.0</td>
<td>(&gt;12.1)</td>
</tr>
<tr>
<td>Vit. E, vit B12</td>
<td>N</td>
<td></td>
</tr>
</tbody>
</table>

### CBC Differential

<table>
<thead>
<tr>
<th>Test</th>
<th>Value</th>
<th>Reference Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hb</td>
<td>104</td>
<td>(&gt;4.0)</td>
</tr>
<tr>
<td>RBC</td>
<td>3.3</td>
<td>(&gt;4.0)</td>
</tr>
<tr>
<td>RDW</td>
<td>19.1</td>
<td>(&lt;16)</td>
</tr>
<tr>
<td>Platelets</td>
<td>298</td>
<td>(150-400)</td>
</tr>
<tr>
<td>WBC</td>
<td>14.5</td>
<td>(&lt;11.0)</td>
</tr>
<tr>
<td>Myelocytes</td>
<td>1.0</td>
<td>(&lt;0.1)</td>
</tr>
<tr>
<td>Neutrophils</td>
<td>9.4</td>
<td>(&lt;9.0)</td>
</tr>
<tr>
<td>Bands</td>
<td>1.2</td>
<td>(&lt;1.3)</td>
</tr>
<tr>
<td>Lymphocytes</td>
<td>0.1</td>
<td>(&gt;0.5)</td>
</tr>
<tr>
<td>Metamyelocytes</td>
<td>1.1</td>
<td>(&lt;0.1)</td>
</tr>
<tr>
<td>Blasts</td>
<td>0.1</td>
<td>(0.0)</td>
</tr>
<tr>
<td>(toxic changes)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Repeat Imaging

DWI
Repeat Imaging

CT of brain - normal
What is this?

I can't stop thinking!!
Laboratory Testing

REPEAT CSF testing

CSF protein 0.60
CSF WBC None
CSF culture –
CSF viral PCR –

CSF cytology – atypical cells
CASE 1

• She developed incontinence of bowel and bladder function about 1 week after presentation

• There was no improvement or decline in her neuro exam of the legs
## Follow-up Nerve Conduction

### Nerve Conduction Studies
**Anti Sensory Summary Table**

<table>
<thead>
<tr>
<th>Site</th>
<th>NR</th>
<th>Onset (ms)</th>
<th>Peak (ms)</th>
<th>O-P Amp (μV)</th>
<th>Neg Dur (ms)</th>
<th>Site1</th>
<th>Site2</th>
<th>Dist (cm)</th>
<th>Vel (m/s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left Sup Peron Anti Sensory (Ant Lat Mall)</td>
<td>Calf</td>
<td>NR</td>
<td></td>
<td></td>
<td></td>
<td>Calf</td>
<td>Ant Lat Mall</td>
<td>14.0</td>
<td></td>
</tr>
<tr>
<td>Right Sup Peron Anti Sensory (Ant Lat Mall)</td>
<td>Calf</td>
<td>NR</td>
<td></td>
<td></td>
<td></td>
<td>Calf</td>
<td>Ant Lat Mall</td>
<td>14.0</td>
<td></td>
</tr>
<tr>
<td>Left Sural Anti Sensory (Lat Mall)</td>
<td>Calf</td>
<td>NR</td>
<td></td>
<td></td>
<td></td>
<td>Calf</td>
<td>Lat Mall</td>
<td>14.0</td>
<td></td>
</tr>
<tr>
<td>Right Sural Anti Sensory (Lat Mall)</td>
<td>Calf</td>
<td>NR</td>
<td></td>
<td></td>
<td></td>
<td>Calf</td>
<td>Lat Mall</td>
<td>14.0</td>
<td></td>
</tr>
</tbody>
</table>

### Motor Summary Table

<table>
<thead>
<tr>
<th>Site</th>
<th>NR</th>
<th>Onset (ms)</th>
<th>O-P Amp (mV)</th>
<th>Neg Area (mV·ms)</th>
<th>Amp% (Prev)</th>
<th>Neg Dur (ms)</th>
<th>Site1</th>
<th>Site2</th>
<th>Dist (cm)</th>
<th>Vel (m/s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left Peroneal Motor (Ext Dig Brev)</td>
<td>Ankle</td>
<td>NR</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Ankle</td>
<td>Ext Dig Brev</td>
<td>8.0</td>
<td></td>
</tr>
<tr>
<td>Right Peroneal Motor (Ext Dig Brev)</td>
<td>Ankle</td>
<td>NR</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Ankle</td>
<td>Ext Dig Brev</td>
<td>8.0</td>
<td></td>
</tr>
<tr>
<td>Left Tibial Motor (Abd Hall Brev)</td>
<td>Ankle</td>
<td>NR</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Ankle</td>
<td>Abd Hall Brev</td>
<td>8.0</td>
<td></td>
</tr>
<tr>
<td>Knee</td>
<td>13.8</td>
<td>0.0</td>
<td>0.02</td>
<td></td>
<td>1.88</td>
<td></td>
<td>Ankle</td>
<td>Abd Hall Brev</td>
<td>0.0</td>
<td></td>
</tr>
<tr>
<td>Right Tibial Motor (Abd Hall Brev)</td>
<td>Ankle</td>
<td>13.8</td>
<td>0.0</td>
<td>0.02</td>
<td>100.0</td>
<td>1.88</td>
<td>Ankle</td>
<td>Abd Hall Brev</td>
<td>8.0</td>
<td></td>
</tr>
</tbody>
</table>

### EMG

<table>
<thead>
<tr>
<th>Side</th>
<th>Muscle</th>
<th>Nerve</th>
<th>Root</th>
<th>Inst Act</th>
<th>Fib s</th>
<th>Ps w</th>
<th>Fast c</th>
<th>Spon t Act</th>
<th>Amp</th>
<th>Dur</th>
<th>Pol y</th>
<th>Recrt</th>
<th>Int Pat</th>
<th>Firin g rate</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Righ t</td>
<td>AntTibiali</td>
<td>Dp Br</td>
<td>L4-5</td>
<td>5</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
<td>Nml</td>
<td>Nml</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Nml</td>
</tr>
<tr>
<td>Righ t</td>
<td>VastusMe</td>
<td>Femoral</td>
<td>L2-4</td>
<td>1</td>
<td>1</td>
<td>1+</td>
<td>0</td>
<td></td>
<td>Nml</td>
<td>Nml</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Nml</td>
</tr>
<tr>
<td>Righ t</td>
<td>MedGastro</td>
<td>Tibial</td>
<td>S1-2</td>
<td>2</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
<td>Nml</td>
<td>Nml</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Nml</td>
</tr>
</tbody>
</table>

**Modifier:**
- **Nm:** Normal
- **Recur:** Recurrent
- **Firing rate:**
- **Activation:** Positive
- **Reduce:** Negative
Cauda Equina Syndrome

The caudal end of the spinal cord is the conus medullaris and is attached to the coccyx by a thin non-neural filament, the filum terminale.

Cauda equina = Latin for “horse’s tail”

Cauda equina syndrome (CES) is a compression of some or all of these nerve roots, resulting in symptoms that include:

- Bowel and bladder dysfunction,
- Saddle anesthesia,
- Varying degrees of loss of lower extremity sensory and motor function.
Cauda Equina Syndrome
Saddle Anesthesia: Dense sensory loss involving the perineum, buttocks
- If found, may indicate poor potential for recovery of normal bladder function.

Sensation to pinprick in the perianal region (S2-S4 dermatomes), perineum, and posterior thigh is altered/diminished.
- there may be preserved sensation to pressure and light touch so discrimination between pinprick and light touch sensation should be sought

BLADDER DYSFUNCTION
• Considered a required element in CES
• May begin with urinary hesitancy, with progression to urinary retention
• Eventually overflow incontinence can develop

A rectal examination is critical to assess the tone and voluntary contracture of the external anal sphincter. Decreased rectal tone is often found early in CES.
Case 2

- 61 y/o RHD M
  - Progressive bilateral lower extremity weakness over 1 week. Thought to have cord compression on MR in Cranbrook; sent to Calgary

- PMHx:

<table>
<thead>
<tr>
<th>COPD</th>
<th>OSA</th>
<th>DM2: 20 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>HTN</td>
<td>Dyslipidemia</td>
<td>CAD</td>
</tr>
<tr>
<td>Cardiomyopathy</td>
<td>GERD</td>
<td>Low Testosterone</td>
</tr>
<tr>
<td>L2-L3 Stenosis</td>
<td>B12 Def</td>
<td></td>
</tr>
</tbody>
</table>
History of presenting illness:
- presents to his GP with bilateral leg swelling, bilateral circumferential leg paresthesias and corresponding leg weakness.
- Paresthesias: “numbness” with pins and needles starting in the feet ascending to the level of the hips over ~4-6 weeks.
- Gait: Substantial loss of balance and stability owed primarily to loss of leg strength. Pt was still able to ambulate with assistance and a cane.
- Lumbar back: long-standing history of “mild” lumbar back pain with no radiation below the leg.

ROS: +Fevers, +Night sweats, Wt loss of 265 ->220 over 9/12 without intention. Head tremor (began 3 years ago)
- Pertinent Negatives: No SOB, no CP, no palpitations, no recent travel, no sick contacts, no bladder/bowel incontinence.
Presentation to hospital

- **Medications:**
  - Pantoprazole 40mg OD
  - Metoprolol 25 mg bid
  - Atorvastatin 20mg OD
  - Spironolactone 25mg OD
  - Enalapril 10 mg bid
  - Novomix insulin 30/70 bid
  - Advair 250 bid
  - ASA 81 mg OD

- **Social Hx:**
  - Living independently
  - Smoke: 30 PY Hx
  - EtOH: Nil
  - Rec Drugs: Nil

- **Family Hx:**
  - Father: Alzheimer’s
  - Mother: Nil
  - No neurological problems in the siblings
Physical Examination

- T=36 HR=103 BP= 107/63 RR=16 O2=92% on R/A
- CV: S1S2, no EHS, no murmurs; leg edema to above knees bilaterally.
- Resp: AE to the bases bilaterally, no crackles or wheeze.
- Abdo: Soft, nontender, abdomen. Unable to assess for splenomegaly due to body habitus.
- Derm: blanchable, erythematous rash on the lower extremities with a hypermelanotic appearance involving limbs.
Physical Examination

- Neurological Examination
  - Oriented x 3
  - CN2 – 12 = Normal
  - Cerebellar:
    - N RAM
    - N finger to nose on the R but unable to perform on the L because of weakness.
    - Gait: Unable to be assessed due to weakness.
  - Sensory:
    - Decreased light touch, pin, and temperature from the feet to the hips
    - Decreased vibration bilaterally
    - N proprioception bilaterally
## Physical Examination

### Motor

<table>
<thead>
<tr>
<th>Muscle Group</th>
<th>Left</th>
<th>Right</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shoulder Abduction</td>
<td>2/5</td>
<td>4/5</td>
</tr>
<tr>
<td>Elbow Flexor</td>
<td>4/5</td>
<td>4/5</td>
</tr>
<tr>
<td>&quot; Extensor</td>
<td>4/5</td>
<td>4/5</td>
</tr>
<tr>
<td>Wrist Flexor</td>
<td>4/5</td>
<td>4/5</td>
</tr>
<tr>
<td>&quot; Extensor</td>
<td>4/5</td>
<td>4/5</td>
</tr>
<tr>
<td>Hand</td>
<td>4/5</td>
<td>4/5</td>
</tr>
<tr>
<td>Hip Flexor</td>
<td>2/5</td>
<td>2/5</td>
</tr>
<tr>
<td>&quot; Extensor</td>
<td>2/5</td>
<td>2/5</td>
</tr>
<tr>
<td>Knee Flexor</td>
<td>2/5</td>
<td>2/5</td>
</tr>
<tr>
<td>&quot; Extensor</td>
<td>2/5</td>
<td>2/5</td>
</tr>
</tbody>
</table>
Thoughts?
INVESTIGATIONS SUMMARY

- EMG confirmed: Distal Sensory and Motor Neuropathy
- L1 Lytic/Sclerotic Lesion seen on CT/MRI
- Mild Hepatomegaly seen on CT
- Mild hepatomegaly and Lymphadenopathy seen on PET
- Low Testosterone
- MGUS: see below

**Serum Protein Electrophoresis**

<table>
<thead>
<tr>
<th>Protein</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Protein</td>
<td>62 L</td>
</tr>
<tr>
<td>Albumin</td>
<td>31.8 L</td>
</tr>
<tr>
<td>Monoclonal #1</td>
<td>3.9</td>
</tr>
<tr>
<td>Monoclonal protein identified as IgG lambda.</td>
<td></td>
</tr>
</tbody>
</table>

**Hematologic Work Up**

<table>
<thead>
<tr>
<th>Test</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Beta 2 Microglobulin</td>
<td>2.65 H</td>
</tr>
<tr>
<td>Immunoglobulin Free Light Chains</td>
<td></td>
</tr>
<tr>
<td>-Kappa</td>
<td>25.10 H</td>
</tr>
<tr>
<td>-Lambda</td>
<td>31.20 H</td>
</tr>
<tr>
<td>-Kappa/Lambda Ratio</td>
<td>0.80</td>
</tr>
</tbody>
</table>
Diagnosis?
POEMS

- P - Polyneuropathy – Motor/Sensory
- O - Organomegaly – Hepato, Lympha
- E - Endocrinopathy – Low Test., DM2
- M - Monoclonal Protein - MGUS
- S - Skin Changes – New onset Rash
Pathophysiology

PLASMA CELLS

CYTOKINE PRODUCTION

ENDOTHELIAL CELLS

PROLIFERATION

NEURONAL DAMAGE

Axonal loss

Demyelination

NEUROLOGY 2006;66:10–12
### TABLE I. Criteria for the Diagnosis of POEMS Syndrome

<table>
<thead>
<tr>
<th>Mandatory major criteria</th>
<th>1. Polyneuropathy (typically demyelinating)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>2. Monoclonal plasma cell-proliferative disorder (almost always λ)</td>
</tr>
<tr>
<td>Other major criteria (one required)</td>
<td>3. Castleman disease*</td>
</tr>
<tr>
<td></td>
<td>4. Sclerotic bone lesions</td>
</tr>
<tr>
<td></td>
<td>5. Vascular endothelial growth factor elevation</td>
</tr>
<tr>
<td>Minor criteria</td>
<td>6. Organomegaly (splenomegaly, hepatomegaly, or lymphadenopathy)</td>
</tr>
<tr>
<td></td>
<td>7. Extravascular volume overload (edema, pleural effusion, or ascites)</td>
</tr>
<tr>
<td></td>
<td>8. Endocrinopathy (adrenal, thyroid, pituitary, gonadal, parathyroid, pancreatic)</td>
</tr>
<tr>
<td></td>
<td>9. Skin changes (hyperpigmentation, hypertrichosis, glomeruloid hemangioma, plethora, acrocyanosis, flushing, white nails)</td>
</tr>
<tr>
<td>Other symptoms and signs</td>
<td>10. Papilledema</td>
</tr>
<tr>
<td></td>
<td>11. Thrombocytosis/polycythemia</td>
</tr>
<tr>
<td></td>
<td>Clubbing, weight loss, hyperhidrosis, pulmonary hypertension/restrictive lung disease, thrombotic diatheses, diarrhea, low vitamin B&lt;sub&gt;12&lt;/sub&gt; values</td>
</tr>
</tbody>
</table>
POEMS, polyneuropathy, organomegaly, endocrinopathy, M protein, skin changes.

The diagnosis of POEMS syndrome is confirmed when both of the mandatory major criteria, one of the three other major criteria, and one of the six minor criteria are present.

\( ^a \) There is a Castleman disease variant of POEMS syndrome that occurs without evidence of a clonal plasma cell disorder that is not accounted for in this table. This entity should be considered separately.

\( ^b \) Because of the high prevalence of diabetes mellitus and thyroid abnormalities, this diagnosis alone is not sufficient to meet this minor criterion.

\( ^c \) Approximately 50% of patients will have bone marrow changes that distinguish it from a typical MGUS or myeloma bone marrow. [39] Anemia and/or thrombocytopenia are distinctively unusual in this syndrome unless Castleman disease is present.
CASE 3

- 48F presenting with inability to rise from chairs and dyspnea upon exertion
- Progressive over 3 months
- Difficulty raising her hands above head
- Can’t get out of chair without using arms on armrests
CASE 3

• There is a chronic dry cough
• No chest pain, no limb pain
• No numbness or tingling
• PMHx: nil      Meds: nil
• Rest of history: nil
Mild facial weakness
Neck strength weak for neck flexion/extension
Limb strength:

<table>
<thead>
<tr>
<th></th>
<th>Left</th>
<th>Right</th>
</tr>
</thead>
<tbody>
<tr>
<td>Deltoid</td>
<td>4-</td>
<td>Deltoid</td>
</tr>
<tr>
<td>Triceps</td>
<td>4</td>
<td>Triceps</td>
</tr>
<tr>
<td>Biceps</td>
<td>4</td>
<td>Biceps</td>
</tr>
<tr>
<td>Wrist Extensors</td>
<td>5</td>
<td>Wrist Extensors</td>
</tr>
<tr>
<td>Finger Extensors</td>
<td>5</td>
<td>Finger Extensors</td>
</tr>
<tr>
<td>Hand Intrinsics</td>
<td>5</td>
<td>Hand Intrinsics</td>
</tr>
<tr>
<td>Hip Flexor</td>
<td>3</td>
<td>Hip Flexor</td>
</tr>
<tr>
<td>Hip Extensor</td>
<td>4-</td>
<td>Hip Extensor</td>
</tr>
<tr>
<td>Hip Abductors</td>
<td>4-</td>
<td>Hip Abductors</td>
</tr>
<tr>
<td>Hip Adductors</td>
<td>4</td>
<td>Hip Adductors</td>
</tr>
<tr>
<td>Quads</td>
<td>4+</td>
<td>Quads</td>
</tr>
<tr>
<td>Hamstrings4+</td>
<td></td>
<td>Hamstrings4+</td>
</tr>
<tr>
<td>Tibialis Anterior</td>
<td>5</td>
<td>Tibialis Anterior</td>
</tr>
<tr>
<td>Tibialis Posterior</td>
<td>5</td>
<td>Tibialis Posterior</td>
</tr>
<tr>
<td>Gastrocnemius</td>
<td>5</td>
<td>Gastrocnemius</td>
</tr>
<tr>
<td>Toe Flexors</td>
<td>5</td>
<td>Toe Flexors</td>
</tr>
</tbody>
</table>
CASE 3

Reflexes normal
Normal responses to pin and temperature sensation
Normal vibration, proprioception to great toes
Waddling, Trendelenberg gait
Can’t get up from chair without using arms
Not able to rise from crouch w/o assistance

Diffuse velcro-sounding crackles to bilateral lung fields
No skin changes visible
CASE 3

Her chest XR showed diffuse interstitial changes:

CT scan showed ground glass changes:
CASE 3

Blood Work

CBC – slight normocytic anemia

Electrolytes N

Liver, Renal Functions N

TSH, vitamin D, PTH N

SPEP N

CK 12200

C-RP 77
CASE 3

EMG

Muscles had irritative changes, with fibrillations and positive sharp waves

Early recruitment of small appearing motor unit potentials

Consistent with an inflammatory myositis, no evidence for neuropathic changes
CASE 3

Muscle Biopsy

- Variation in fiber size
- Small rounded fibers

- Mononuclear cells
Diagnosis?
Polymyositis with IPF

Immune myositis – Dermatomyositis/Polymyositis

There are certain Antibody mediated conditions to be considered

This patient has an anti-Jo-1 Antibody
CASE 3

Dermatomyositis

• Retinopathy, Cardiovascular Disease, Interstitial lung disease, tentative association with Rheumatoid arthritis; Scleroderma; CREST

• Important to remember association with malignancy
Polymyositis

• Although dermatological changes are absent, other systemic conditions are frequently observed

• Cardiac Arrhythmias, Myocarditis

• Respiratory muscle weakness; Interstitial lung disease

• Esophageal paresis

• Autoimmune: Lupus, Sjögren's, Anti-phospholipid antibodies & syndrome: 5-8%

• Thyrotoxicosis, Rare
Polymyositis

- There are antibodies associated with immune myopathy syndromes:
  - Jo-1 & t-RNA synthetase antibody
  - Mi-2 antibody
  - PM-Scl antibody
  - Malignancy-associated
  - Graft-versus-host disorders
Jo-1 & t-RNA synthetase antibody

- Proximal Weakness
- Raynaud’s phenomenon
- Mechanic’s hands
- Arthritis

- Interstitial lung disease – Dyspnea, Cough
  - Pathology shows non-specific interstitial pneumonia (NSIP) or Bronchiolitis obliterans organizing pneumonia (BOOP)
  - Inflammation without granulomas

- High Mortality due to lung involvement
Mi-2 antibody syndrome

- Proximal Weakness in child or adult
- Dermatomyositis changes
- Nailfold involvement common
- Arthritis and other immune conditions uncommon, but +ANA is ubiquitous
CASE 3

PM-Scl antibody syndrome

- Polymyositis picture
- Scleroderma changes or Systemic Sclerosis
- Mechanic’s hands common, along with calcinosis and telangiectasia
- +ANA very common
CASE 4

- 52F presenting with dimpling of her right buttock and paresthesiasa of the right leg in March 2007 (w/o radiation)
- History of DM2*1y without complications
- Numbness and tingling over posterior right calf and dorsal right foot
• PMHx of Crohn`s Disease diagnosed in 2006
Initial Neuro Exam (2007)

Normal cranial nerve function
Normal neck strength
Normal strength to left and right arm

Leg strength:

<table>
<thead>
<tr>
<th>Left</th>
<th>Right</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hip Flexor</td>
<td>Hip Flexor</td>
</tr>
<tr>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Hip Extensor</td>
<td>Hip Extensor</td>
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<td>Tibialis Anterior</td>
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<td>5</td>
<td>4+</td>
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<td>Tibialis Posterior</td>
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<tr>
<td>5</td>
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<td>Peronei</td>
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<tr>
<td>Gastrocnemius</td>
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<tr>
<td>EHL</td>
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<td>5</td>
<td>4+</td>
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<tr>
<td>EDB</td>
<td>EDB</td>
</tr>
<tr>
<td>5</td>
<td>4+</td>
</tr>
<tr>
<td>Toe Flexors</td>
<td>Toe Flexors</td>
</tr>
<tr>
<td>5</td>
<td>4+</td>
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</table>
No atrophy nor fasciculations could be detected, but the right buttock was dimpled. Refluxes hyporeflexic to right ankle, otherwise normal. Plantars bilaterally down. Normal tone to arms and both legs. Subjective decreased sensation (pin, T°) over the dorsal right foot and lateral right lower leg. Normal sensation to the upper limbs. Vibration, proprioception normal to great toes. Antalgic gait without difficulties on Romberg, tandem gait, or walking on heels or toes (no steppage gait).
Impaired vibration threshold with no vibration detection to left ASIS and right knee, with normal detection to fingertips and xiphoid process
No proprioception to level of knees bilaterally
Coordination normal to upper extremities
Gait could not be tested
Differential Diagnosis and Plan
Neuroimaging
Neuroimaging
Neuroimaging
# Initial Nerve Conduction Studies

## Nerve Conduction Studies

### Anti Sensory Summary Table

<table>
<thead>
<tr>
<th>Site</th>
<th>NR</th>
<th>Onset (ms)</th>
<th>Peak (ms)</th>
<th>O-P Amp (μV)</th>
<th>Neg Dur (ms)</th>
<th>Site1</th>
<th>Site2</th>
<th>Dist (cm)</th>
<th>Vel (m/s)</th>
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<tbody>
<tr>
<td>Left Sup Peron Anti Sensory (Ant Lat Mall)</td>
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<tr>
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## Motor Summary Table

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<th>Site2</th>
<th>Dist (cm)</th>
<th>Vel (m/s)</th>
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<td>Left Tibial Motor (Abd Hall Brev)</td>
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## Initial Nerve Conduction Studies

### Right Tibial Motor (Abd Hall Brev) 29.6

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### F Wave Studies

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<th>Lat Norm (ms)</th>
<th>L-R F-Lat (ms)</th>
<th>L-R Lat Norm</th>
<th>M-Lat (ms)</th>
<th>HLat-MLat (ms)</th>
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<tr>
<td>Left Peroneal (Mrkrs) (EDB)</td>
<td>50.35</td>
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<td>Left Tibial (Mrkrs) (Abd Hallucis)</td>
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<td>&lt;5.7</td>
<td>5.73</td>
<td>48.81</td>
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<tr>
<td>Right Tibial (Mrkrs) (Abd Hallucis)</td>
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<td>&lt;61</td>
<td>&lt;5.7</td>
<td>7.05</td>
<td>52.99</td>
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<th>L-R H-Lat (ms)</th>
<th>L-R Lat Norm</th>
<th>M-Lat (ms)</th>
<th>HLat-MLat (ms)</th>
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<tbody>
<tr>
<td>Left Tibial (Gastroc)</td>
<td>30.64</td>
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<td>NR</td>
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Initial Nerve Conduction Studies

<table>
<thead>
<tr>
<th>Side</th>
<th>Muscle</th>
<th>Nerve</th>
<th>Root</th>
<th>Ins Act</th>
<th>Fibs</th>
<th>Ps w</th>
<th>Fasic</th>
<th>Spon Act</th>
<th>Amp</th>
<th>Dur</th>
<th>Poly</th>
<th>Recert</th>
<th>Int Pat</th>
<th>Firin g Rate</th>
<th>Comment</th>
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<tr>
<td>Left</td>
<td>AntTibialis</td>
<td>Dp Br</td>
<td>L4-5</td>
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<td>0</td>
<td>1+</td>
<td>Nml</td>
<td>Nml</td>
<td>Nm 1</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
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<td>Left</td>
<td>MedGastroc</td>
<td>Peron</td>
<td>S1-2</td>
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<td>0</td>
<td>0</td>
<td>Nml</td>
<td>Nml</td>
<td>Nm 1</td>
<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
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<tr>
<td>Left</td>
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<td>Tibial</td>
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<td>Nm 1</td>
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<td>0</td>
<td>Nml</td>
<td>Nml</td>
<td>Nm 1</td>
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<td>Nml</td>
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<td>Nml</td>
<td>Nml</td>
<td>Nml</td>
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</tbody>
</table>

**EMG**

**NCV (Left Peroneal Motor)**

**NCV (Right Peroneal Motor)**

**NCV (Left Tibial Motor)**

**NCV (Right Tibial Motor)**

**H-Reflex (Left Tibial)**

**H-Reflex (Right Tibial)**
Laboratory Testing

CSF protein 0.38
CSF WBC None
CSF culture –
CSF viral PCR –

Blood Work

CBC N
Electrolytes N
Liver, Renal Functions N
Serum folate, Vit. E, vit B12 N
SPEP N
Pestronk Ab testing negative
# Laboratory Testing

## Table of Antibody Results

<table>
<thead>
<tr>
<th>Antibody</th>
<th>Patient Values</th>
<th>Normal Values</th>
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<tr>
<td>IgM vs GM1</td>
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<tr>
<td>NP-9</td>
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<tr>
<td>IgM vs Histone H3</td>
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<tr>
<td>IgM vs GalNAc-GD1a</td>
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<td></td>
</tr>
<tr>
<td>IgM vs asialo-GM1</td>
<td>&lt;1000</td>
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</tr>
<tr>
<td>IgM vs GD1b</td>
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<td>&lt;1000</td>
</tr>
<tr>
<td>IgG vs GM1</td>
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<tr>
<td>IgG vs GD1b</td>
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<tr>
<td>IgG vs GT1a</td>
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<td>&lt;2000</td>
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<tr>
<td>IgM vs Sulfatide</td>
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<tr>
<td>IgG vs Sulfatide</td>
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<tr>
<td>IgM vs MAG</td>
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<td>IgG vs Yo</td>
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<tr>
<td>IgM vs Decorin</td>
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**Report Date:** 11-14-2008
**Sample Number:** 06-08-305-0323
What’s Going on?
Case 4 Follow-up

• Crohn`s Disease reactivated in 2010. Her GI specialist started her on remicade without Neuro consultation
• Remained neurologically stable
• Neurosurg consultation re: sciatic nerve lesion – no biopsy
• Patient declined further therapy
## Follow-up Nerve Conduction Studies

<table>
<thead>
<tr>
<th>Site</th>
<th>NR</th>
<th>Onset (ms)</th>
<th>Peak (ms)</th>
<th>O-P Amp (µV)</th>
<th>Neg Dur (ms)</th>
<th>Site1</th>
<th>Site2</th>
<th>Dist (cm)</th>
<th>Vel (m/s)</th>
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<tbody>
<tr>
<td>Left Sup Peron Anti Sensory (Ant Lat Mall)</td>
<td>Calf</td>
<td>3.2</td>
<td>3.8</td>
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<td>1.34</td>
<td>Calf</td>
<td>Ant Lat Mall</td>
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<td>3.1</td>
<td>3.7</td>
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### Motor Summary Table

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<th>Site</th>
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<th>Neg Area (mV/µs)</th>
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<th>Site2</th>
<th>Dist (cm)</th>
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<tr>
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<td>10.0</td>
<td>40</td>
</tr>
<tr>
<td>Left Tibial Motor (Abd Hall Brev)</td>
<td>Ankle</td>
<td>6.3</td>
<td>8.3</td>
<td>23.75</td>
<td>100.0</td>
<td>5.23</td>
<td>Ankle</td>
<td>Abd Hall Brev</td>
<td>8.0</td>
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<tr>
<td></td>
<td>Knee</td>
<td>15.4</td>
<td>4.8</td>
<td>12.07</td>
<td>57.8</td>
<td>5.63</td>
<td>Knee</td>
<td>Ankle</td>
<td>39.0</td>
<td>40</td>
</tr>
<tr>
<td>Right Tibial Motor (Abd Hall Brev)</td>
<td>Ankle</td>
<td>5.9</td>
<td>6.2</td>
<td>15.21</td>
<td>100.0</td>
<td>7.27</td>
<td>Ankle</td>
<td>Abd Hall Brev</td>
<td>8.0</td>
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<tr>
<td></td>
<td>Knee</td>
<td>15.7</td>
<td>1.1</td>
<td>2.62</td>
<td>17.7</td>
<td>6.02</td>
<td>Knee</td>
<td>Ankle</td>
<td>39.0</td>
<td>40</td>
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</table>
Follow-up Nerve Conduction Studies

F Wave Studies

<table>
<thead>
<tr>
<th>NR</th>
<th>F Lat (ns)</th>
<th>Lat Norm (ns)</th>
<th>L-R F Lat (ms)</th>
<th>L-R Lat Norm</th>
<th>M Lat (ms)</th>
<th>F Lat-M Lat (ms)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left Peroneal (Mkrs) (EDB)</td>
<td>50.16</td>
<td>&lt;60</td>
<td>4.55</td>
<td>&lt;3.1</td>
<td>3.95</td>
<td>44.21</td>
</tr>
<tr>
<td>Right Peroneal (Mkrs) (EDB)</td>
<td>54.71</td>
<td>&lt;60</td>
<td>4.55</td>
<td>&lt;3.1</td>
<td>5.95</td>
<td>48.76</td>
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<tr>
<td>Left Tibial (Mkrs) (Abd Hallucis)</td>
<td>53.84</td>
<td>&lt;61</td>
<td>8.05</td>
<td>&lt;5.1</td>
<td>6.48</td>
<td>47.36</td>
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<tr>
<td>Right Tibial (Mkrs) (Abd Hallucis)</td>
<td>61.89</td>
<td>&lt;61</td>
<td>8.05</td>
<td>&lt;5.7</td>
<td>6.83</td>
<td>55.06</td>
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</table>
Laboratory Testing

Pestronk Ab testing repeated

<table>
<thead>
<tr>
<th>Antibody</th>
<th>Patient Values</th>
<th>Normal Values</th>
</tr>
</thead>
<tbody>
<tr>
<td>IgM vs GM1</td>
<td>0</td>
<td>&lt;2000</td>
</tr>
<tr>
<td>NP-9</td>
<td>Negative</td>
<td>Negative</td>
</tr>
<tr>
<td>IgM vs NS6S</td>
<td>22,000</td>
<td>&lt;15,000</td>
</tr>
<tr>
<td>IgM vs GalNAc-GD1a</td>
<td>0</td>
<td>&lt;8,000</td>
</tr>
<tr>
<td>IgM vs asialo-GM1</td>
<td>2100</td>
<td>&lt;1,000</td>
</tr>
<tr>
<td>IgM vs GD1b</td>
<td>0</td>
<td>&lt;3,000</td>
</tr>
<tr>
<td>IgG vs GM1</td>
<td>0</td>
<td>&lt;1,000</td>
</tr>
<tr>
<td>IgG vs GQ1b</td>
<td>0</td>
<td>&lt;2,000</td>
</tr>
<tr>
<td>IgM vs Sulfaide</td>
<td>&lt;1,500</td>
<td>&lt;1,500</td>
</tr>
<tr>
<td>IgG vs Sulfaide</td>
<td>&lt;2,000</td>
<td>&lt;2,000</td>
</tr>
<tr>
<td>IgM vs MAG</td>
<td>0</td>
<td>Negative</td>
</tr>
<tr>
<td>MAG Western blot</td>
<td></td>
<td>Negative</td>
</tr>
<tr>
<td>IgM vs TS-HDS</td>
<td>&lt;10,000</td>
<td></td>
</tr>
<tr>
<td>IgM vs Tubulin</td>
<td>&lt;2,500</td>
<td>&lt;2,500</td>
</tr>
<tr>
<td>IgG vs Tubulin</td>
<td>&lt;2,500</td>
<td>&lt;2,500</td>
</tr>
<tr>
<td>Tubulin Western blot</td>
<td>Negative</td>
<td></td>
</tr>
<tr>
<td>IgG vs Hu (Western &amp; IHC)</td>
<td>Negative</td>
<td></td>
</tr>
<tr>
<td>IgG vs Yo (Western &amp; IHC)</td>
<td>Negative</td>
<td></td>
</tr>
<tr>
<td>IgM vs Decorin</td>
<td>&lt;2,500</td>
<td></td>
</tr>
</tbody>
</table>

Control antigens: (Histone H3, GD1a, GM1 & Sulfatide)

**INTERPRETATION:** IgM in this serum shows selective binding to NS6S disaccharide using ELISA. IgG shows no reactivity.
Case 4 Follow-up

- Repeat MRI – negative – sciatic nerve lesion disappeared
- Discussed d/c of remicade – reluctant due to Crohn`s doing well
- IVIG trial – developed hemolytic anemia
- Imuran – idiosyncatic reaction
- Now, managing conservatively only
• Muscle and Nerve diseases aren’t hard to figure out

• Always make first step of differentiating type of motor unit dysfunction

• Remember systemic associations!
Muscle and Nerve