GENERALIZED WEAKNESS CLINICAL CASE FINDINGS

Case 1 – 67-year-old woman with leg weakness

8-year history of slowly progressive, painless weakness

- 8 years ago - trouble arising from floor
- 4 years ago - trouble arising from chair
- 2 years ago - falls, give way of left leg
- Denies atrophy, fasciculations, muscle pain, sensory symptoms, or any upper extremity or trunk symptoms
- Previous needle electromyography (EMG) and nerve conduction studies (NCSs) normal

Clinical Examination: Uses upper extremities to arise from seated

- -2 weakness quadriceps with mild, bilateral atrophy
- -1 to -2 weakness left-right finger flexors and wrist flexors
- Remainder of neurologic examination is normal, including reflexes and sensory examination

NCS – normal peroneal, tibial, and sural

COMMENT

Scattered, mytonic discharges in multiple muscles

REPORT

Summary - NCSs are normal. Needle EMG shows fibrillation potentials and myotonic discharges with a mixture of short and long duration, polyphasic, rapidly recruited motor unit action potentials (MUAPs) in proximal and distal muscles.

Interpretation – The findings are those of a chronic, severe, diffuse myopathy. Fibrillation potentials in myopathic disorders may indicate the presence of necrosis, splitting, or vacuolization of muscle fibers. Mixed MUAPs can be seen in disorders that affect both nerve and muscle or in very chronic myopathies such as inclusion body myositis (IBM), chronic polymyositis or some muscular dystrophies. IBM is most likely with this combination of EMG and clinical findings.

MUSCLE BIOPSY – IBM

COMMENT

The distribution of weakness is an important clue to IBM. Mixed MUAP changes are also typical, but can be seen in very chronic myositis.

<table>
<thead>
<tr>
<th>EMG</th>
<th>Fibrillation</th>
<th>Recruit</th>
<th>Duration</th>
<th>Amplitude</th>
<th>Phases</th>
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<tr>
<td>Biceps brachii</td>
<td>0</td>
<td>reduced</td>
<td>+ long +short</td>
<td>+low</td>
<td>25%</td>
</tr>
<tr>
<td>FDI</td>
<td>0</td>
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<td>+long +short</td>
<td>+low</td>
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<tr>
<td>FCR</td>
<td>++</td>
<td>reduced</td>
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<td>25%</td>
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<tr>
<td>Medial gastrocnemius</td>
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<td>+long +short</td>
<td>+high +low</td>
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<tr>
<td>TA</td>
<td>+</td>
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<td>+long</td>
<td>+high</td>
<td>15%</td>
</tr>
<tr>
<td>Vastus medialis</td>
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<tr>
<td>TFL</td>
<td>+</td>
<td></td>
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</tbody>
</table>

EMG = electromyography; FCR = flexor carpi radialis; FDI = first dorsal interosseous; TA = tendon achilles; TFL = tensor fasciae latae
Case 2: 20-year-old college student 2 weeks progressive generalized weakness

Day 1
1. Myalgia, headache, sore throat, fever
10. Student Health: penicillin for “strep throat”, persistent emesis
11. Emergency Room - Urinary retention, lethargy, unsteady
12. Diplopia, mild proximal weakness, brisk deep tendon reflex (DTR), bilateral Babinski
13. Reduced reflexes, progressive weakness, shortness of breath (SOB), tachycardia
14. Hospitalized: Head CT and MRI normal; Cerebrospinal fluid (CSF) cells & protein increased

Diagnosis – Guillain–Barré with myelopathy, polyradiculoneuropathy, and autonomic neuropathy

Treatment Plan: Start 5 days intravenous immunoglobulin (IVIg)

Day 14

NCS

All motor amplitudes were low with no dispersion
Median motor and peroneal responses absent
No F waves
Conduction velocities and blink reflexes normal
Repetitive stimulation normal

EMG

Poor or no activation
Two muscles with reduced recruitment
No fibrillation potentials

REPORT

Acute, severe, diffuse, axonal polyradiculopathy. A severe motor neuropathy with distal conduction block cannot be excluded, but is less likely. Repeat study in 7 - 10 days for more definitive information.

Day 21

Total paralysis. Pulse dose methylprednisolone; tracheostomy; percutaneous endoscopic gastrostomy (PEG); C. difficile colitis; GM1 < 250; viral t/c (-)

NCS

Motor and sensory responses all showed marked reduction from the study of 9 days ago, despite high-intensity stimulation. The remaining, very low-amplitude responses had markedly long distal latencies and reduction in amplitude with proximal stimulation. No blink reflexes could be recorded.

EMG

No voluntary MUAPs could be activated in any muscle tested. Minimal fibrillation potentials were recorded only in hand and cervical paraspinal muscles.

REPORT

There has been marked progression of the disorder. There is now much clearer evidence of a widespread, demyelinating neuropathy with distal involvement that could account for the paralysis.

Day 23

IVIg 5 days - slow improvement; antibiotics continued

Day 60

NCS

NCSs reveal markedly reduced or absent compound muscle action potentials throughout with markedly slowed conduction velocities and prolonged distal latencies. There is an ulnar conduction block in the forearm on the left. The blink reflex was present, but markedly prolonged.

EMG

There are fibrillation potentials throughout, but with clear MUAP activation of normal or mildly long duration in the proximal upper limb.

REPORT

The findings are now those of a severe polyradiculopathy with prominent demyelination, as well as moderate axon loss. There has been improvement since the previous study. Fibrillation potentials are more prominent, but this reflects the Wallerian degeneration that has occurred, and does not indicate a worsening.

3 months – Reflexes return; tracheostomy and PEG out
6 months – Return to school with good limb strength

COMMENT

The distinction between an axonal motor neuropathy (AMAN) and an acute demyelinating neuropathy is not always easy. Every attempt should be made to define the diagnosis and stop specific therapy too soon.

Case 3: 65 –year-old woman with fatigue

• 2 years of difficulty with household chores, “tired”
• Difficulty squatting at “Curves”
• EMG 1 year ago – normal NCS and EMG
• Limited improvement on sertraline hydrochloride
• Examination - mild proximal weakness including difficulty swallowing. Normal reflexes, cranial nerves, sensation, and gait

NCS

Motor and sensory NCSs are normal in the arm and leg, including F-wave latencies. Slow repetitive stimulation of ulnar/hypothenar,
accessory/trapezius and peroneal/anterior tibial muscles before and after exercise is normal.

**EMG**

Needle EMG showed short duration MUAPs in several proximal muscles, including the genioglossus. A few fibrillation potentials were seen in the thoracic paraspinals.

**REPORT**

The findings are those of a very proximal myopathy. The absence of many fibrillation potentials argues against, but does not exclude, an active inflammatory myopathy.

**BIOPSY**

Sarcoidosis

**COMMENT**

While the clinical and EMG picture make an inflammatory myopathy most likely, a muscle biopsy should always be considered to obtain a more definitive diagnosis. Sarcoidosis may present initially with muscle involvement

**Case 4: 29-year-old woman with muscle aching**

- Healthy - 5 years muscle aching
- Mild elevations of creatine kinase (CK) (300 - 550)
- Examination normal
- Mild weakness, limited to left triceps

**NCS**

Motor and sensory NCSs normal in arm and leg. Normal repetitive stimulation.

**EMG**

Short duration MUAPs with mild, scattered fibrillation potentials in proximal muscles.

**REPORT**

The findings are mild, but consistent with a proximal myopathy, possibly inflammatory.

**BIOPSY**

Scattered, small, atrophic fibers with rare necrotic fibers. Consider myotonic dystrophy, Type 2.

**GENE TEST**

Myotonic dystrophy type 2 (DM2) showed CCTG repeat expansions (>12,100 bp). Confirmed in relatives.

**COMMENT**

Myotonic discharges in DM2 are more prominent proximal, predominantly wane, and may be few and far between.

**Case 5: 45-year-old interior designer with 3 months generalized weakness**

- Brain stem astrocytoma-stable 2 yrs after radiation therapy
- Temporal lobe herpes simplex virus (HSV) encephalitis-better year after acyclovir
- Bulbar dysfunction from tumor-dexamethasone and temazolamide
- 2 mo progressive weakness with no other symptoms or signs
- CSF and EMG performed at home - axonal and demyelinating neuropathy
- Hospital transfer – flaccid quadriplegia with mild facial weakness and dyspnear
- Other cranial nerves, reflexes and sensation normal
- MRI 10 mm mass and residuals of HSV; with deep vein thrombosis (DVT)

**EMG**

Short duration, polyphasic, stable MUAPs with rapid recruitment in all muscles. Scattered fibrillation potentials

**NCS**

CMAPs are low amplitude and markedly long duration without dispersion. F-wave latencies are normal, but F waves are small. Normal sensory NCSs. Normal repetitive stimulation.

**ADDITIONAL STUDIES**

Low amplitude MUAP with direct muscle stimulation. Normal motor unit number estimates.

**REPORT**

The findings are those of a severe, generalized myopathy of the type seen with critical illness. This pattern can occur with high dose steroid medication.

No evidence of an additional, neurogenic process or defect of neuromuscular transmission was found.

**FOLLOW-UP**

Full return of strength after 4 months

**COMMENT**

Critical illness myopathy can be difficult to distinguish from neuropathy. Prolonged CMAP without dispersion and small MUAPs are important distinguishing features.
Case 6: 84-year-old woman with weakness

- Diabetes mellitus and hypothyroidism with 2 months of painless arm weakness
- Examination: Proximal, symmetric arm weakness; normal sensation and reflexes

NCS
Normal

EMG

Proximal and distal muscles of the arm and leg show rapid recruitment of short duration MUAPs primarily in the proximal, muscles, a bit more in the arm than leg. A few scattered myotonic discharges were seen, but no fibrillation potentials were found.

REPORT

The findings are those of a proximal myopathy. The findings do not clearly suggest an inflammatory or other specific muscle disorder. This pattern could be seen with a hypothyroid myopathy.

COMMENT

Severe hypothyroid myopathy often shows brief runs of myotonic like activity. Biopsy is nonspecific.