



April 1999 EMG Case-of-the-Month

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Presenting Symptoms: Lower limb weakness

Learning Objectives: After completing this educational activity, participants will be able to (1) formulate an appropriate differential diagnosis for the lower limb weakness and (2) devise an appropriate treatment plan for the lower limb weakness.

HISTORY

A 21-year-old right hand dominant woman presents with a chief complaint of difficulty ambulating secondary to weakness in her right lower limb. She first noticed weakness approximately two months earlier and her symptoms worsened, causing her to drag her right foot. To compensate, the patient notices picking up her right leg during ambulation. She denies pain or trauma to the affected limb, though she feels her symptoms may be related to the arthritis she was previously diagnosed as having in both knees. There is no weakness in her upper limbs, but she does have occasional weakness in the left lower limb as well. The patient also denies bowel / bladder problems.

- **Prior to continuing, please develop a differential diagnosis and list each possible diagnosis in order of likelihood.**
- **Is there any additional information regarding the clinical history that might be helpful in clarifying your differential list or changing its order of priority?**

COMMENTARY I

From the information presently known, it would appear the patient is describing the typical gait pattern used to compensate for a unilateral "foot drop."

The temporal nature of the patient's symptoms suggests possible neuromuscular causes. An isolated mononeuropathy of the peroneal nerve causing weakness of the ankle dorsiflexors should be strongly considered. Root level pathology, presumably at L5, is possible. However, the absence of radicular pain makes this less likely. Arthritis of the knees would not cause profound weakness, especially in the absence of pain. The differential diagnosis may also include an upper motor neuron syndrome that could produce this pattern of weakness. It is unlikely the patient suffered a CVA, but demyelinating diseases affecting the CNS, such as multiple sclerosis, may be considered. Also to be considered is an unusual presentation of peripheral polyneuropathy.

HISTORY, CONTINUED

The patient denies any previous neurologic symptoms, but now states that her right leg has become "numb." She denies back pain. She is not pregnant. She has no family history of weakness or neurological disease processes. The only medications she is taking are for systemic lupus erythematosus (SLE) which she was diagnosed as having 12 years ago. These include Prednisone (5mg/day for the past 3- years), Plaquenil (the past 6 months)



and coumadin (5mg/day for the past 6 months). The patient states that she was hospitalized 6 months ago because of a DVT in her left lower limb, and that her lupus has occasionally caused rashes and has affected her kidney function.

- **If necessary, revise your differential diagnosis based on the additional clinical history.**
- **On which details of the physical examination should you focus at this point?**

COMMENTARY II

The fact that her SLE has caused renal dysfunction suggests a multi-system pattern of involvement. Connective tissue diseases are known to cause disorders in the central & peripheral nervous systems. Certainly, SLE may explain the patient's history of bilateral knee arthritis. The recent history of DVT, along with her history of SLE, suggests the possibility of a coexistent vasculitis and coagulopathy compromising her PNS. Chronic steroid usage, even in small dosages, has been known to cause myopathy and proximal muscle weakness. The sensory symptoms exclude the sole presence of a myopathic process as well as a disorder affecting the neuromuscular junction.

PHYSICAL EXAMINATION

There is no visible atrophy. The patient is mildly obese with rounding of her face and fading malar rash. Bilateral dorsalis pedis pulses are diminished yet palpable. There is no clubbing or cyanosis. There is 1+ edema in the (L) ankle. She is oriented to person, place and time. Cranial nerves 2-12 are grossly intact. Manual muscle grading of 5/5 throughout both upper limbs. Manual muscle testing of the lower limbs showed: hip flexor, extensor, adductor, abductor, and quadriceps muscle groups are 5/5 bilaterally; (R) hamstrings 4/5; (L) hamstrings 5/5; (R) tibialis anterior, (R) extensor hallucis longus & (R) peronei muscles are 0/5; (L) tibialis anterior, (L) extensor hallucis longus & (L) peronei muscles are 5/5; (R) gastrocnemius 1/5; (L) gastrocnemius 4-/5.

Straight leg raising is negative bilaterally. Muscle stretch reflexes are 2+/4 and symmetric in the biceps, triceps, brachioradialis, and quadriceps muscles, but absent at both ankles. Babinski sign is absent (Toes are flexor bilaterally). There is no ankle clonus bilaterally & Hoffman's sign is negative. Romberg sign is mildly positive. Gait reveals a steppage pattern favoring the right lower limb with visible foot drop. Sensation is diminished to pin prick to light touch over the (R) anterolateral ankle and foot as well as the plantar surface of the (L) foot. Full passive range of motion is present throughout both lower limbs.

- **At this point, review your differential diagnosis and revise as appropriate.**
- **Are there additional observations on physical examination that might be helpful in narrowing your differential list?**

COMMENTARY III

The physical examination shows an asymmetric pattern of weakness in the lower limbs. There is complete paresis of the right peroneal-innervated muscle groups as well as significant weakness of the right gastrocnemius muscles. Mild weakness is noted in the right hamstring and the left gastrocnemius muscle. Radiculopathy is excluded since this pattern of weakness is not in a segmental distribution. Given the asymmetry of the distal motor and sensory findings, a peripheral polyneuropathy seems likely, specifically multiple



mononeuropathies. There is an asymmetric pattern of sensory loss, with right peroneal and left tibial nerve distributions affected. At this point, the possibility of a vasculitic neuropathy associated with SLE should be high on the differential diagnosis list and should warrant further work up including electrodiagnostic testing. The examination has excluded a central nervous system lesion.

PHYSICAL EXAMINATION, CONTINUED

- **If necessary, revise your differential diagnosis based on the additional physical findings.**
- **Design your approach to the electrophysiologic examination based on the existing data.**

COMMENTARY IV

The MRI of the L/S spine, (ordered by primary care physician one week earlier), is not suggestive of disc disease or root level pathology. Blood tests showed the presence of the lupus anticoagulant & antiphospholipid antibody, suggesting a concomitant *antiphospholipid syndrome* associated with her SLE. Sural nerve biopsy confirmed the presence of inflammatory changes affecting the peripheral nervous system. From all of the information gathered from the history & physical examination, it can be inferred that the patient has a peripheral neuropathic process as the probable cause for her weakness. The pattern, type, and extent of peripheral neuropathy are still unknown. Electrodiagnostic testing can help clarify this issue, prognosticate the likelihood for recovery, and guide appropriate management.

DIAGNOSTIC IMPRESSION

1. Mononeuritis multiplex, primarily axonal, affecting the right sciatic & left tibial nerves.
 2. No evidence of steroid induced myopathy.
- **What other diagnostic procedures (laboratory tests, etc.), if any, are needed?**

MRI of L/S spine for HNP or pathology was normal.

ESR: 110

Antiphospholipid antibody and lupus anticoagulant were both positive (abnormal).

Sural nerve biopsy was positive for inflammatory changes affecting the vaso nervorum.

- **What treatment would you recommend?**

COMMENTARY V

The electrodiagnostic examination was consistent with the history & physical examination in showing a neuropathic process as the cause for the patient's complaints. The sparing of the SNAP amplitude of the right sural nerve was important in showing that a more generalized symmetric peripheral polyneuropathy was not present currently. The biopsy showing inflammatory changes in the vaso nervorum was taken from the patient's left sural nerve.



Sural nerve sparing is reported in many cases of early AIDP, possibly due to its heavy myelination, strong blood supply, and the lack of entrapment sites throughout its distribution. However, the patient's history and clinical course do not support a presentation of AIDP. Conduction block does not appear to be a factor in the patient's electrodiagnostic findings as evidenced by the lack of focal slowing and amplitude reduction across the region of the right fibular head. This also speaks against an isolated entrapment neuropathy of the peroneal nerve across the fibular head, as in peroneal nerve palsy. Abnormalities in motor conduction studies affected the right peroneal and the right & left tibial nerves. The CMAP was absent in the right tibial nerve, and CMAP amplitudes were markedly reduced in the right peroneal & left tibial nerves. Motor nerve conduction velocities were within the range of low/normal for the right tibial nerve and normal for the right & left peroneal nerves.

These findings favor an axonal over a demyelinating process of nerve injury. The needle EMG examination showed membrane instability in muscles supplied by both the right peroneal and tibial nerves as well as increased polyphasicity of motor units in the right long head of the biceps femoris muscle, innervated by the sciatic nerve. Voluntary motor unit recruitment was absent in the right tibialis anterior and peronei muscles and reduced in the right medial gastrocnemius and right long head of the biceps femoris muscles. Membrane instability was also noted in the left gastrocnemius muscle. These findings support the presence of significant axon loss affecting the right sciatic nerve as well as the left tibial nerve to varying degrees. The presence of the left peroneal F-wave with a latency value within normal limits suggests the relative sparing of the proximal conduction pathways of this nerve.

The asymmetry of electrodiagnostic findings supports the diagnosis of a primary axonal *mononeuritis multiplex* affecting the right sciatic, right saphenous and left tibial nerves. There was no evidence of a superimposed steroid myopathy contributing to the patient's symptoms. However, electrodiagnostic detection of a mild steroid myopathy would be difficult since corticosteroids preferentially affect type 2 muscle fibers; motor unit action potential evaluation during needle EMG is primarily of type 1 muscle fibers.

Because of significant axon loss in the involved nerves, a long period of time may be required for recovery of muscle strength. Additionally, the absence of voluntary motor units in the patient's right tibialis anterior muscle along with the duration of her weakness suggest a poor prognosis for functional recovery. Treatment of the right foot drop with an AFO is indicated to improve ambulation. A foot drop splint should also be provided and worn at night to prevent heel cord contracture.

Antiphospholipid Syndrome is an autoimmune disorder of recurrent vascular thrombosis caused by antibody production against phospholipids. Phospholipids on platelets and endothelial cells have an important role in the inhibitory reactions of the clotting cascade. There is a known association between advanced SLE and the presence of antiphospholipid syndrome & vasculitis. SLE patients who have had a long disease duration and early age of onset are the most likely to develop vasculitis, which is confirmed by arteriography or nerve biopsy. The most frequent presentation of neuropathy associated with vasculitis is in the form of an *axonal mononeuritis multiplex* (50-60% of cases). In the *multiple mononeuritis* form, several individual nerves are affected to varying degrees, causing an asymmetric pattern of weakness and sensory loss.



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