



EMG Case No. 50, April 2001

Presenting Symptoms: Left Thigh Pain and Paresthesias, Left Leg Weakness, Right Hand Weakness, Right Fourth and Fifth Finger Numbness

This case is no longer available for CME credit.

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Appropriate Audience: Residents and practicing physicians

Learning Objectives: After completing this educational activity, participants will be able to (1) formulate a logical algorithm for the electrodiagnostic workup of a patient with multiple neurologic complaints, (2) outline a cogent list of possible systemic medical differential diagnosis for a patient with multiple neurologic complaints, and (3) use electrodiagnostic test results to add to the clinical information needed to make a diagnosis in a patient with multiple neurologic complaints.

History

A 56-year-old woman with systemic lupus erythematosus (SLE) presents for electrodiagnostic evaluation because of a 2-week history of sudden onset of non-radiating left thigh pain and paresthesias. She also complained of significant progressive left leg weakness to the point of being unable to walk for the last 1-2 weeks. In addition, she had slowly worsening right hand weakness and right fourth and fifth finger numbness, causing deterioration of her manual dexterity for the last 6 months. Although she has had a few episodes of lower back pain in the past, she denied having lower back or neck pain at this presentation. She also denied any symptoms in the left upper or right lower limbs. She did not give a history of recent trauma, bowel or bladder symptoms, recent dramatic weight loss, fever or chills, speech or swallowing difficulties.

- *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

At this point the differential diagnosis may include:

- Multiple mononeuropathies
- Peripheral polyneuropathy
- Lumbosacral radiculopathy plus an associated upper extremity problem (radiculopathy/plexopathy/isolated mononeuropathy)
- Lumbosacral plexopathy plus an associated upper extremity problem (radiculopathy/plexopathy/isolated mononeuropathy)
- Mononeuropathy multiplex
- Acute inflammatory demyelinating polyneuropathy (AIDP)
- Chronic inflammatory demyelinating polyneuropathy (CIDP)



- Femoral and ulnar mononeuropathies
- Multiple sclerosis

The diagnosis must take into account the patient's acute and chronic multifocal, asymmetric extremity complaints. Starting in the central nervous system (CNS), it would be very unlikely for one or more lesions in the brain and/or spinal cord to account for this pattern. Although there is not a waxing and waning history, the possibility of multiple sclerosis should be considered. Lesions in the peripheral nervous system (PNS) must then be considered. Although her symptoms could result from more than one lesion, such as lumbosacral and cervical radiculopathy/polyradiculopathy, femoral neuropathy with ulnar neuropathy, or lumbosacral radiculopathy with brachial plexopathy, these would be considered lower on the differential diagnosis, given her neurologic presentation and underlying SLE. A vasculitic neuropathy with multiple, asymmetric mononeuropathies (mononeuropathy multiplex) is more commonly observed in such patients. Other recognized peripheral nerve diseases in patients with SLE are primary sensory or sensorimotor peripheral polyneuropathy, and less commonly AIDP or CIDP.

History, continued

The patient's past medical history includes right lung lobectomy 2 years ago for non-small cell carcinoma. This was thought to be curative and as such she did not undergo any adjunctive treatment. The patient also suffered a right wrist extensor tendon rupture and a left Achilles tendon rupture 8 months ago. These were thought to be the result of attrition due to her rheumatologic disorder and medications. The patient declined surgical intervention for these injuries; thus they were treated non-operatively with casting and splinting.

She smoked 1-3 packs per day for over 40 years but quit after her lung resection 2 years ago.

Her family history is significant for uterine cancer in her mother and prostate cancer in her father.

The patient was sent for a MRI of the lumbosacral spine, with these results:

Diffuse bone marrow heterogeneity of thoracic, lumbar, and sacral vertebral bodies, and pelvic bones. A lytic bone lesion at the greater trochanter of right femur. These bony changes are most likely related to diffuse bone marrow metastases.

Mild degenerative changes in the form of a mild posterior L5-S1 disc bulge, degenerative hypertrophy of L4-L5 and L5-S1 facet joints. No spinal canal stenosis or compression of dural sac or nerve roots. No paravertebral soft tissue mass is seen.

- *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

In view of the past medical history and family history, malignant causes must now be entertained. In the CNS, there may be brain metastases or extradural spinal cord compression from spine metastases. The PNS may be affected by malignancy, typically by tumor invasion into the neural elements or external neural compression. Malignancy related paraneoplastic syndromes commonly manifest with peripheral polyneuropathy, cerebellar



degeneration, or Lambert-Eaton myasthenic syndrome. These are more commonly associated with small cell lung cancer, ovarian cancer, and lymphomas.

Taking this patient's history of SLE and malignancy into account, one must consider dual etiologies and diagnoses to account for her symptoms. The more pressing issue is malignancy, whether it is a recurrence or a new primary, and which may be manifesting with neurologic symptoms.

Physical Examination

The patient is alert and oriented and in no acute distress. She is afebrile and her vital signs are stable. Cranial nerves II-XII are intact. There is weakness of right wrist extension and right finger flexion, abduction, and adduction. The left lower limb is diffusely weak, with trace hip flexion, adduction, and knee extension. There is also weakness of ankle dorsiflexion, eversion, and plantar flexion, and of great toe extension. All other muscle groups tested in the right upper and left lower limbs are normal. Manual muscle testing of the left upper and right lower limbs is also normal. Light touch and pinprick sensation are diminished in the right ulnar sensory distribution and in the left L2 through L5 dermatomes. Muscle stretch reflexes are diminished in the right upper limb, absent in the left lower limb, but normal in the left upper and right lower extremities. Tests for pathologic reflexes such as Hoffman's and Babinski's are negative. There is also no ankle clonus and toes are down going.

- *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

With the absence of bowel, bladder, or speech and swallowing dysfunctions, no brisk muscle stretch reflexes, negative pathologic reflexes, and no increased muscle tone, it is unlikely that there is a CNS etiology. The right upper extremity neurologic deficits could be explained by C8/T1 radiculopathy, lower trunk brachial plexopathy, or ulnar mononeuropathy. The right wrist extension weakness may be attributed to the patient's history of an old right wrist extensor tendon rupture. The left ankle plantar flexion weakness may be from an old left Achilles' tendon rupture. However, the left lower extremity neurologic deficits may be more difficult to delineate without further investigation.

Physical Examination, continued

Other pertinent positive and negative findings are:

Palpation and active ranges of motion of the neck and back did not elicit any pain or restrictions. Dural tension signs are negative in all four extremities. However, there is pain with range of motion and palpation of the right hip (opposite the side with neurologic deficits).

There are enlarged lymph nodes in both axillae and in the left inguinal region. Multiple subcutaneous nodules are also palpable at the anterior chest and abdominal walls.

- *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

Painless active range of motion of the neck and back, with the absence of dural tension signs, makes the diagnosis of cervical and/or lumbosacral radiculopathy less likely. The right hip pain may suggest an arthritic joint while the multiple enlarged nodes and nodules in various anatomic sites are more suggestive of a diffuse metastatic process. The lytic lesion noted in the right greater trochanter, a presumed metastasis, may be causing her right hip pain.

Electrophysiologic Data

Temperatures: right palm= 32.8 Celsius left ankle=32.6 Celsius

SENSORY NERVE CONDUCTION STUDIES							
NR= no response							
NERVE	SIDE	STIM SITE	RECORD	cm	AMPL (uV)	LAT (ms)	CV (m/sec)
Sural	Left	Calf	Ankle	14	6.7	4.2	40
Sural	Right	Calf	Ankle	14	5.7	4.5	40
Ulnar	Right	Wrist	5 th digit	14	NR		
Ulnar	Left	Wrist	5 th digit	14	14.4	4.1	43.7
Median	Right	Wrist	Index	14	22.8	4.3	43.7

MOTOR NERVE CONDUCTION STUDIES							
NR= no response							
NERVE	SIDE	STIM SITE	RECORD	cm	AMPL (mV)	LAT (ms)	CV (m/sec)
Peroneal	Left	Ankle	EDB	9	2.9	5.7	-
		BK- Ankle	-	30	2.8	14.1	35.7
		AK-BK	-	11.5	2.7	16.8	42.6
Peroneal F-response	Left	Ankle	EDB	-	-	NR	-
Peroneal	Right	Ankle	EDB	9	5.8	5.7	-
		BK- Ankle	-	27.5	5.7	13.0	37.7
		AK-BK	-	11.0	5.3	14.9	57.9



Peroneal F-response	Right	Ankle	EDB	-	-	56.8	-
Tibial	Left	Ankle	AH	8	6.2	4.8	-
		Knee-Ankle	-	41	3.8	14.7	41.4
Tibial F-response	Left	Ankle	AH	-	-	56.7	-
Tibial	Right	Ankle	AH	8	6.0	4.7	-
		Knee-Ankle	-	38	4.8	13.5	43.2
Tibial F-response	Right	Ankle	AH	-	-	56.4	-
Median	Right	Wrist	Thenar	7	4.8	4.4	-
		Elbow-Wrist	-	23	4.1	8.7	53.5
Median F-response	Right	Wrist	Thenar	-	-	30.3	-
Ulnar	Right	Wrist	Hypothenar	7	NR	-	-
Ulnar	Left	Wrist	Hypothenar	7	6.7	3.6	-
		BE-Wrist	-	20	5.3	8.0	45.5
		AE-BE	-	10	5.6	10.0	50.0
Ulnar F-response	Left	Wrist	Hypothenar	-	-	31.2	-
Radial	Right	Forearm	EIP	17	4.0	5.0	-

NEEDLE ELECTROMYOGRAPHY

Insertional activity: N, sust, unsust
 FIB: 0, 1+, 2+, 3+, 4+
 Other: 0 or fascic, myotonia, myokymia
 Effort: N, decr
 Recruitment: N, inc or dec 1+, 2+, 3+, 4+
 Amplitude: N, inc or dec 1+, 2+, 3+, 4+
 Duration: N, inc or dec 1+, 2+, 3+, 4+
 polyphasic: N, inc or dec 1+, 2+, 3+, 4+

R/L	MUSCLE	INSER	PSW/FIB	OTH	EFF	REC	AMP	DUR	POL
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			Fasc						
L	Tibialis Anterior	Sust	2+ 1+	0	N	Dec 2+	N	N	N
L	Peroneus longus	Unsust	1+ 1+	0	N	Dec 1+	Inc 1+	Inc 1+	Inc 1+
L	Short head of Biceps Femoris	N	0 0	0	N	N	N	N	N
L	Medial Gastroc.	N	0 0	0	N	-	N	N	N
L	Rectus femoris	N	0 0	0	N	0	-	-	-
L	Adductor longus	N	0 0	0	N	0	-	-	-
L	External Hamstring	N	0 0	0	N	Dec 1+	N	N	N
L	Tensor Fascia Lata	N	0 0	0	N	Dec 1+	N	N	N
R	FDI (Hand)	Unsust	1+ 1+	0	N	Dec 2+	N	N	Inc 1+
R	FDP	Unsust	1+ 1+	0	N	Dec 1+	N	N	Inc 1+
R	ADQ (Hand)	Unsust	1+ 1+	0	N	Dec 3+	N	N	Inc 1+
R	EDC	N	0 0	0	N	Dec 1+	N	N	N
R	Biceps Brachii	N	0 0	0	N	N	N	N	N
R	Triceps	N	0 0	0	N	N	N	N	N
R	APB	N	0 0	0	N	N	N	N	N
R	Cervical Paraspinal	N	0 0	0	-	-	-	-	-
L	Lumbosacral Paraspinal	N	0 0	0	-	-	-	-	-

- On the basis of both the clinical and electrophysiologic evaluations, formulate your diagnostic impression. List the most likely diagnosis first and follow in order with the other



possibilities that are not excluded by the data. Eliminate those diagnoses not supported by the data.

- *Are there additional electrophysiologic data that you feel would further delineate the diagnosis? (Remember, collecting data that are not needed for the diagnosis is costly and uncomfortable for the patient.)*

Electrophysiologic Data, continued

Summary of previous electrodiagnostic findings:

This patient did have an electrodiagnostic study done for complaints of right upper extremity weakness and numbness 9 months ago. At that time, nerve conduction studies of the right median, ulnar, and radial motor and sensory nerves were normal, as was the right ulnar F-wave response.

Needle EMG study of selected muscles in the right arm (biceps brachii, triceps, flexor carpi radialis, extensor carpi radialis, extensor pollicis longus, extensor indicis, abductor pollicis brevis, and first dorsal interosseous [hand]), and right cervical paraspinal muscles were also normal at that time.

Summary of current electrodiagnostic findings:

Nerve conduction studies of the left peroneal motor nerve show low amplitude with an absent F-wave response, while right peroneal values are normal. Other nerve conduction studies performed in the left and right legs are within normal limits. Nerve conduction studies of the right ulnar motor and sensory nerves show no evoked responses, a change compared to the study of 9 months ago. The other motor nerve conduction studies performed in the right and left arms are within normal limits except for the borderline prolongation of the median, ulnar, and sural sensory evoked responses, which were normal in the previous study. This may be suggestive of a very mild peripheral neuropathy, or simply age related.

Needle EMG study of selected muscles in the right arm and left leg show an acute neurogenic process involving the right ulnar innervated muscles and of the left peroneal innervated muscles below the knee, affecting deep and superficial branches. The lack of voluntarily recruited motor units in the left femoral and obturator innervated muscles may connote abnormality of those nerves as well.

- *Make the final revisions of your diagnostic impression(s).*

Diagnostic Impression

This is an abnormal electrodiagnostic study with findings most consistent with multiple mononeuropathies. Specifically, there is evidence of:

1. a common peroneal mononeuropathy at the left knee associated with mild to moderate denervation;
2. femoral and obturator mononeuropathies (or alternatively a high lumbar plexopathy) on the left that is more recent (likely less than 3 weeks old) than the peroneal mononeuropathy on the same side;



3. ulnar mononeuropathy in the right upper extremity associated with mild to moderate denervation. The location of the ulnar nerve lesion could be anywhere in the upper arm, proximal to the elbow and distal to the brachial plexus.

Taken together, these findings are most consistent with mononeuritis multiplex associated with vasculitis; this is commonly seen in patients with rheumatologic disorders such as SLE.

There may also be a mild generalized peripheral polyneuropathy, reflected in borderline prolongation of the sensory evoked responses in the median, ulnar, and sural nerves. There is, however, no evidence of a lumbosacral radiculopathy in the left lower extremity, a lower trunk brachial plexopathy, a C8/T1 radiculopathy in the right upper extremity, or a myopathic process.

- *What other diagnostic procedures (laboratory tests, etc.), if any, are needed?*
- *What treatment would you recommend?*

Commentary V

Pertinent laboratory studies:

- Sedimentation Rate: 33 (0-20) mm/hr
- C-Reactive Protein: 10.6 (0-. 6) mg/dl
- Calcium: 11.1 (8.6-10.2) mg/dl
- Total Alkaline Phosphatase: 1090 (99-257) U/l
- Liver Fraction: 675 (24-158) U/l
- Bone Fraction: 415 (24-146) U/l
- Intestine Fraction: 0 (0-22) U/l
- ALT: 52 (0-45) IU/l
- AST: 45 (2-35) IU/l
- Anti-dsDNA Antibody: 8.1 (0-7) IU/ml

Pertinent imaging studies:

Head CT: Normal

Bone Scan (Whole Body): Normal

Chest CT:

1. 2 non-calcified pulmonary nodules in the right lung (3 cm x 3 cm , 1 cm x 1 cm);
2. Bilateral axillary lymph node enlargement;
3. Multiple cutaneous non-calcified nodules in the anterior chest wall and abdominal wall, the largest measuring 2 cm x 3 cm.

Abdominal CT:

1. Low attenuation lesion in the left lobe of the liver measuring 1 cm, worrisome for metastases;
2. 2 cm x 3 cm low attenuation lesion is seen posterior to the left psoas muscle, which may also represent metastases.



Pelvic CT:

1. 2 cm x 3.3 cm left inguinal node with a necrotic center, likely representing metastases;
2. 3 cm low attenuation lesion posterior to the right acetabulum, without signs of bony destruction.

Tissue Biopsy:

- Subcutaneous nodules (abdomen):
Degenerated fat and collagen with scattered lymphocytes suggestive of panniculitis
- Left groin lymph node:
Degenerated fat and collagen surrounded with numerous vacuolated cells presumably "histiocytes" and lymphocytes.

Note-definitive tissue diagnosis has yet to be made as to the vasculitic and neoplastic process.

Combining the medical history, physical examination, electrodiagnostic findings, pertinent laboratory studies, imaging studies, and tissue biopsies, we can conclude that two different processes are responsible for this patient's neurologic deficits. Mononeuropathies of the right ulnar and the left common peroneal nerves are probably due to a vasculitic process related to the patient's SLE. The left femoral and obturator nerve compromise is probably due to compression of these neural elements by the left psoas mass either at the origins of the nerves themselves or at the more proximal lumbar plexus level.

Treatment:

There is no definitive treatment for mononeuropathy multiplex itself. Intervention is therefore usually directed at the underlying etiology, which in this case is the vasculitic process related to SLE. Although treating the SLE will not improve the patient's neurologic status, it is hoped that it will halt or slow the deterioration. On the other hand, the medications used to treat or control SLE (corticosteroids and various immunosuppressive agents) may themselves cause neural or neuromuscular damage.

Treatment of nerve compression by an invasive tumor may involve removing or reducing the tumor, via surgical resection, radiation therapy, or both.

Bibliography

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