



February 1997 EMG Case-of-the-Month

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HISTORY

A 51-year-old woman presents with progressive distal upper limb weakness over the last 3 months, characterized by frequently dropping objects. She has also noted twitching in the lower limbs and occasionally in the trunk, and reports tripping while walking.

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

HISTORY CONTINUED

She has a burning sensation in the forearms and intermittent urinary incontinence. She denies dysphagia or dysarthria.

No contributory family history was elicited. Her only medication is Xanax, taken intermittently for panic attacks.

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

PHYSICAL EXAMINATION

Mental status is intact. Cranial nerves II-XII are normal on examination; specifically there is no dysarthria, facial weakness, or tongue fasciculation. Muscle bulk in the upper limbs is normal proximally, but there is wasting of thenar and hypothenar eminences bilaterally. Strength is 4+/5 in the shoulder abductors, shoulder flexors, and elbow flexors, 3+/5 in forearm pronation, wrist extension and flexion, and elbow extension, and 3/5 for intrinsic hand muscles. Lower limb strength is 5/5. Sensation to pinprick and light touch is intact except for an area of questionable hypesthesia along the medial volar forearm on the left. Muscle stretch reflexes in the upper limbs are 3+ at the biceps, brachioradialis, and triceps. Lower limb reflexes are 3+ at the knees and 4+ at the ankles, with extensor plantar responses bilaterally.

- **At this point, review your differential diagnosis and revise as appropriate.**



- Determine a final working differential diagnosis from which to design your electrodiagnostic study.
- Formulate your approach to the electrodiagnostic study.
- On the basis of the clinical and electrodiagnostic evaluation, formulate your final impression. List the most likely diagnosis followed by other possibilities that are not excluded by the data. Eliminate those diagnoses not supported by the data.

ELECTROPHYSIOLOGIC DATA

ELECTROMYOGRAPHY										
N = normal incr = increased decr = decreased 0 = absent 1+ = minimal 4+ = maximal crd = complex repetitive discharge fasc = fasciculation potential myk = myokymic discharge myt = myotonic discharge nmt = neuromyotonic discharge p wave = positive sharp waves fibrillation = fibrillation potentials recr = recruitment amp = amplitude dur = duration poly = polyphasic potentials										
R/L	MUSCLE	INSERTION SPONTAN				VOLUNTARY				
		activ	p wave	fib	other	recr	amp	dur	poly	effort
L	deltoid	N	0	0	0	N	N	N	N	full
L	biceps brachii	N	0	0	0	N	N	N	N	full
L	triceps brachii	incr	3+	3+	0	decr	N	N	N	full
L	flexor carpi radialis	incr	3+	2+	fasc	decr	N	N	incr	full
L	abductor pollicis brevis	incr	3+	2+	0	discrete	N	N	incr	full
L	1 st dorsal interosseous	incr	3+	2+	fasc	discrete	incr	incr	incr	full
R	biceps brachii	N	0	0	0	N	N	N	N	full
R	triceps brachii	incr	1+	1+	0	central	N	N	N	full
R	1 st dorsal interosseous	incr	2+	2+	fasc	decr	N	N	N	full
L	vastus medialis	N	0	0	0	N	N	N	N	full
L	tibialis anterior	N	0	0	0	N	N	N	N	full
L	medial gastroc	N	0	0	0	N	N	N	N	full
L	mid-thoracic paraspinal	N	0	0	0	-	-	-	-	-
R	mid-thoracic paraspinal	N	0	0	0	-	-	-	-	-
-	tongue	N	0	0	0	N	-	-	-	-



SENSORY NERVE CONDUCTION									
nr = no response									
NERVE	LATENCY (ms)			AMPLITUDE (µV)			CONDUCT VEL(m/s)		
	R	L	Norm	R	L	Norm	R	L	Norm
ulnar	-	3.3	-	-	69	-	-	-	-
med. antebrachial cut.	-	2.6	-	-	22	-	-	-	-

MOTOR NERVE CONDUCTION									
nr = no response									
NERVE	LATENCY (ms)			AMPLITUDE (mV)			CONDUCT VEL (m/s)		
	R	L	Norm	R	L	Norm	R	L	Norm
ulnar	-	-	-	-	-	-	-	-	-
wrist	-	3.5	-	-	1.8	-	-	-	-
below elbow	-	7.2	-	-	1.8	-	-	51	-
above elbow	-	9.1	-	-	1.8	-	-	62	-
axilla	-	12.0	-	-	1.8	-	-	51	-

F-WAVE								
# = number of stimuli P = persistence CD = chronodispersion F:M = ratio of average F-wave amplitude to M-wave amplitude								
R/L	NERVE	#	LATENCY (ms)			CD (ms)	P (%)	F:M (%)
			min	mean	max			
L	ulnar to ADM	10	32.6	-	-	-	-	-



SOMATOSENSORY EVOKED POTENTIALS							
* = abnormally small potentials							
NERVE	RECORDING SITE	LATENCY (ms)			AMPLITUDE (µV)		
		R	L	NORM	R	L	NORM
ulnar	axilla (N1)	7.5	7.4	<8.5	7.3	5.4	>1.6
	C7 spine - Fz (N1)	13.9	13.8	<15.8	0.4*	1.6	>0.5
	Fz - mastoid (P1)	14.5	14.5	<17.8	0.5	0.9	>0.3
	C3'/C4' - Fz (N1)	19.6	19.5	<21.8	5.2	6.3	>0.9
	-	23.2	24.6	<26.2	-	-	-

DIAGNOSTIC IMPRESSION

1. Probable cervical myelopathy.
2. The diagnosis of motor neuron disease is less likely given the absence of findings in the bulbar, thoracic or lumbosacral regions. However, one cannot completely exclude the possibility of motor neuron disease initially affecting only the upper limbs.

RECOMMENDATIONS

Workup should include:

- Imaging studies of the cervical spine to look for causes of myelopathy.
- Consideration of repeat EDx studies in 3 - 6 months, if the diagnosis of motor neuron disease remains a clinical possibility.

DIFFERENTIAL DIAGNOSIS

Based upon the initial clinical data, differential diagnosis primarily includes motor neuron disease (amyotrophic lateral sclerosis) and cervical myelopathy. Motor neuron disease typically presents with mixed upper and lower motor neuron findings; this patient has atrophy in hand muscles and hyperreflexia in upper and lower limbs, as well as Babinski signs. There is minimal, if any, sensory loss.

Cervical spondylosis may also present with hand weakness and atrophy, though patients often have neck pain and prominent sensory symptoms. Cervical myelopathy could account for upper motor neuron signs as well as intermittent urinary incontinence.

Given her marked upper motor neuron signs, a strictly lower motor neuron lesion, such as peripheral polyneuropathy or multifocal motor neuropathy, is unlikely.



FORMULATION OF ELECTROPHYSIOLOGIC STUDIES

The electrophysiologic studies were initially formulated to answer the following questions:

1. Does the EMG confirm motor axon loss in the atrophic upper limbs?
2. Is there evidence of denervation outside the cervical myotomes (i.e. suggesting a more diffuse process)?
3. Is there involvement of peripheral or central sensory pathways?

This patient's needle EMG findings are most striking for evidence of denervation in the lower cervical myotomes (at least the C7-C8 levels, possibly also including a level higher or lower). There is, however, no evidence for denervation outside these cervical levels, arguing against motor neuron disease. Fasciculations can be seen in cervical myelopathy as well as motor neuron disease and hence are non-specific.

Nerve conduction studies do not show evidence of peripheral nerve involvement. There is no slowing of motor or sensory conduction. Sensory amplitudes are normal, arguing against a post-ganglionic lesion. Low ulnar compound muscle action potential amplitudes are consistent with lower motor neuron loss, which can occur anywhere from the anterior horn cell distally.

Somatosensory evoked potentials are helpful in this case. They demonstrate a small amplitude cervical spine potential with right ulnar nerve stimulation. This is one of the most common findings in cervical myelopathy, often preceding changes in the cortically generated scalp potentials. Motor neuron disease is not expected to produce similar findings.

FORMULATION OF AN IMPRESSION

Combining the EMG, NCS and SEP data, evidence appears strongest for a cervical myelopathy affecting both upper and lower motor neurons, the latter most severely at the C7/C8 segmental levels.

Patients with motor neuron disease may present initially with hand weakness and denervation most apparent in distal upper limb muscles. However, in this patient, denervation was limited to muscles supplied by a focal area in the cervical cord and did not affect bulbar, thoracic, or lumbosacral regions. Thus, by any criteria, the data are insufficient to permit the diagnosis of motor neuron disease. However, one cannot completely exclude the possibility that this represents an initial presentation of motor neuron disease.

FOLLOW-UP

Cervical spine films demonstrated multilevel degenerative disc disease from C3-C7 with osteophytes at C5-6 and C6-7.

MRI revealed spondylotic ridging with thecal sac compression at C4-5 and C5-6 as well as an area of high signal in the spinal cord at C6-7.

Repeat studies three months later, when the patient had clinically improved, showed less evidence of denervation in the upper limbs and a normal examination elsewhere.



CLINICAL AND ELECTRODIAGNOSTIC FEATURES OF CERVICAL SPONDYLITIC MYELOPATHY

Patients with cervical myelopathy may present with clinical findings similar to those of motor neuron disease. These include progressive upper limb weakness, fasciculations, and minimal or transient sensory symptoms. Important symptoms that should alert one to the possibility of cervical myelopathy include: sensory symptoms, bowel or bladder dysfunction, and neck pain. Physical examination is usually remarkable for features of an upper motor neuron lesion with hyperreflexia and upgoing toes. Fasciculations may be present in the upper limbs, but are not usually seen in the lower limbs or tongue. Sensation is often reduced, but this sign is not always marked.

Electrophysiologic studies may demonstrate evidence of denervation in both upper limbs when compression affects either the roots or grey matter (anterior horn cells) of the spinal cord. Unlike motor neuron disease, the thoracic paraspinal muscles (below the area of innervation from cervical roots), lumbosacral innervated muscles, and bulbar muscles do not show evidence of denervation.

Nerve conduction studies may demonstrate small amplitude compound muscle action potentials (CMAPs) reflecting the motor neuron or axon loss, though peripheral conduction is usually normal or only slightly slowed. Sensory nerve action potentials (SNAPs) are usually normal since the lesion is proximal to the dorsal root ganglia.

SEPs, with upper or lower limb stimulation, may be abnormal in cervical myelopathy. When stimulating the upper limb, cortical responses are often normal since the lesion may spare dorsal column tracts. The cervical spine response, however, is more commonly reduced in amplitude since this reflects synaptic activity at the cervical cord level (i.e. not just dorsal column pathways). Lower limb SEP abnormalities may reflect myelopathy affecting ascending dorsal column function through the cervical cord.

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