



April 1998 EMG Case-of-the-Month

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HISTORY

A 68-year-old white man presents for electrodiagnostic medical consultation with the complaint of weakness and numbness limited to the lower limbs. He has recently been admitted to the hospital for inability to ambulate due to progressive bilateral leg weakness that occurred in the two months preceding admission. Four months prior to admission, he fell from a roof and developed low back pain which resolved. For the two weeks prior to admission, he reports numbness in both feet.

- **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?**

COMMENTARY I

The differential diagnosis includes the various causes of acquired polyneuropathy or bilateral lumbosacral polyradiculopathy. Lower limb symptoms of both weakness and numbness make myopathy, neuromuscular junction disorders, and motor neuron disease less likely, as those conditions spare the sensory system.

Other possibilities might be found in the central nervous system. Cerebral parasagittal lesions can cause symptoms limited to the lower limbs. Cervical myelopathy is unlikely without upper limb symptoms, and transverse myelitis causes symptoms at the level of the lesion.

Additional history should assess bowel and bladder function along with upper limb symptoms. Cranial nerve symptoms, e.g. swallowing impairment or visual changes, should be assessed. The nature of the onset makes an inherited disorder very unlikely, but family history should be obtained.

HISTORY, continued

The patient describes progressive difficulty with urination over the previous two months, along with reduced ability to develop and maintain erections. For the last two weeks, he has had pain at night in the lower limbs.

He has no upper limb symptoms nor visual changes, swallowing difficulty, fevers, chills, or night sweats. There have been no recent illnesses, immunizations, or rashes. His family history is negative for neuromuscular disorders. He has not had previous lumbar or cervical spine surgery. He is not diabetic nor does he drink excessively. He is not on coumadin.



- **If necessary, 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?**

COMMENTARY II

The combination of bladder and erectile problems with bilateral weakness of the lower limbs strongly suggests a cauda equina syndrome. The absence of upper limb symptoms argues against a diffuse process, such as polyneuropathy. Physical examination must include assessment of the lower sacral segments.

The patient's nocturnal pain raises the suspicion of malignancy.

PHYSICAL EXAMINATION

The patient is an alert, oriented and slightly overweight man. The examination is performed with the patient in bed. Cranial nerves appear intact. No weakness is detected in the upper limbs and there is no Bevor sign. Strength testing of the lower limbs reveals 3/5 hip flexion, 3/5 knee extension, trace dorsiflexion and trace plantarflexion bilaterally. Anal tone is reduced, but the patient can voluntarily control the anal sphincter. Muscle stretch reflexes in the upper limbs are present and symmetric bilaterally. Knee and ankle reflexes are absent. Hoffman and Babinski signs are not present. The anal wink reflex is absent. There is no sensory deficit in the upper limbs, and no sensory level can be detected on the trunk. There is reduced pinprick sensation over the anterior thighs, medial legs, and soles of the feet.

- **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 demonstrated involvement of the spinal segmental levels from L3-S2 bilaterally with both motor and sensory findings. There was no sensory level on the abdomen to suggest a thoracic lesion. A negative Beever's sign indicates that the lower abdominal muscles are equally as strong as the upper abdominal muscles, further arguing against a mid or low thoracic spinal lesion.

The absent reflexes in the legs suggests lower motor neuron involvement. Upper motor neuron involvement, such as myelopathy, would likely result in extensor plantar responses, clonus, and hyperreflexia. However, with an acute spinal lesion above about T12, flaccid paralysis during the initial period of spinal shock can complicate interpretation of physical exam findings.

The complete sparing of the cranial nerves and the arms argues against a polyneuropathy, although the legs can be much more affected than the arms in an early polyneuropathy.

Motor neuron disease, myopathy, and neuromuscular junction disorders are now very low on the differential, as these disorders spare the sensory system. Sensory testing is a vital part of the electrodiagnostic evaluation. When sensory testing reveals a deficit, attention is focused upon those conditions that affect both the motor and sensory systems.



PHYSICAL EXAMINATION, continued

No further examination data.

ELECTROPHYSIOLOGIC DATA

ELECTROMYOGRAPHY										
n = normal incr = increased depr = 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 fib = fibrillation potentials recr = recruitment amp = amplitude dur = duration poly = polyphasic potential										
R/L	MUSCLE	INSERTION		SPONTAN		VOLUNTARY				
		activ	p wave	fib	other	recr	amp	dur	poly	effort
L	vastus medialis	n	0	0	-	decr	n	n	n	n
L	rectus femoris	n	0	0	-	decr	n	n	n	n
L	vastus lateralis	n	1+	1+	-	decr	n	n	n	n
L	adductor longus	n	1+	2+	-	no units	-	-	-	-
L	tfl	incr	3+	3+	-	no units	-	-	-	-
L	anterior tibialis	incr	3+	3+	-	no units	-	-	-	-
L	medial gastroc	incr	1+	1+	-	decr	n	n	n	n
L	lateral gastroc	n	2+	2+	-	decr	n	n	n	n
L	biceps femoris (sh)	n	1+	1+	-	decr	n	n	n	n
L/R	lumbar paraspinals	incr	3+	3+	-	-	-	-	-	-
L	orbicularis oris	n	0	0	-	n	n	n	n	n
R	vastus medialis	n	1+	1+	-	decr	n	n	n	n
R	anterior tibialis	incr	2+	3+	-	decr	n	n	incr	n
R	medial gastroc	incr	3+	3+	-	decr	n	n	n	n
L	thoracic paraspinal	n	0	0	-	-	-	-	-	-
L	cervical paraspinal	n	0	0	-	-	-	-	-	-
L	first dorsal interos	n	0	0	-	n	n	n	n	n
L	biceps	n	0	0	-	n	n	n	n	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
sural (14 cm)	3.7	4.1	<4.0	9	4	>5			-
superficial radial (14 cm)	-	2.9	<3.3	-	23	>13			-

MOTOR NERVE CONDUCTION									
nr = no response									
NERVE	LATENCY (ms)			AMPLITUDE (mV)			CONDUCT VEL		
	R	L	Norm	R	L	Norm	R	L	Norm
peroneal	4.1	5.6	<6.2	7	1.5	>2	41	42	>39
ulnar	3.0	-	<4.2	8	-	>2.5	52	-	>48

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	-	30	-	-	-	-	-
R	peroneal	-	62	-	-	-	-	-
L	tibial h-reflex	-	nr	-	-	-	-	-



REPETITIVE STIMULATION

Repetitive stimulation of the right common peroneal nerve and recording at the EDB showed no decrement to 3 hz stimulation, no facilitation immediately post-exercise, and no post-tetanic decrement (exhaustion).

- **On the basis of both the clinical and electrodiagnostic evaluations, 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.**
- **What other diagnostic procedure are needed?**

DIAGNOSTIC IMPRESSION

The findings are most suggestive of bilateral lumbosacral polyradiculopathies affecting L3-S2 root levels. There was marked acute denervation with minimal reinnervation. These findings can be seen in lumbar spinal stenosis and with intraspinal lesions.

DISCUSSION

The EMG examination showed severe bilateral recent denervation in the lower limb muscles innervated by L3 to S2 nerve roots, with no denervation in the upper limb, thoracic paraspinals, or in a facial nerve supplied muscle. The marked denervation in the lumbar paraspinal muscles bilaterally localizes the lesion/lesions to the lumbosacral root level. Recruitment was neuropathic, not myopathic, and argues against a myopathy. Because of spinal cord and cauda equina anatomy within the lower thoracic and lumbar spinal canal, it is important to realize that EMG findings cannot precisely localize the site of a lesion within the spinal canal. A lesion of the conus that destroys anterior horn cells can yield the same EMG and nerve conduction findings as an L2-3 herniated nucleus pulposus.

Nerve conduction studies were normal in the upper limbs, but showed a low amplitude peroneal response, absent H reflex, and prolonged peroneal F-wave latency even corrected for height and age. Of particular importance is the preservation of the sural sensory responses bilaterally. This suggests that the lesion or lesions are proximal to the dorsal root ganglia.

Repetitive stimulation of a weak muscle did not demonstrate a decrement, arguing against myasthenia gravis or other neuromuscular junction disorder. No facilitation after a short contraction excludes Lambert-Eaton syndrome. The suspicion for a neuromuscular junction disorder was very low, so other proximal nerves were not evaluated with repetitive stimulation. If the findings on EMG were not so compelling, neuromuscular junction testing of the femoral and spinal accessory nerves would be warranted.

Polyneuropathy is not likely as the sensory system and the arms were normal. Motor nerve conduction velocity was normal, even in the left peroneal nerve where the amplitude was reduced, indicating axon loss as the cause of the reduced amplitude.



Motor neuron disease can present with involvement of a single limb or just the lower limbs. If the symptoms were to generalize, a repeat study of the arms, cranial nerve supplied muscles, and thoracic paraspinal muscles would be indicated.

This case illustrates several important issues surrounding electrodiagnosis of lower limb weakness and evaluation for spinal conditions. Spinal metastases causing a cauda equina syndrome in patients with cancer occur in about 5-10% (Rodriguez and Dinapoli; 1980). Intramedullary metastases are considerably rarer with incidences of 1-3% (Rodriguez and Dinapoli; 1980). Perrin, Livingston and Aarabi (1982) in their review of spinal metastases at their institution reported that of 200 patients, only one had an intramedullary lesion. Intradural extramedullary tumors occurred in 10 of the 200 patients. Metastases to the spine are usually hematogenously spread and most commonly involve the cancellous portion of the vertebral body with spread into the epidural space (Errico and Kostuik; 1986). Patients who present with acute cauda equina symptoms represent a neurosurgical emergency, and correctable structural causes must be sought. The electrodiagnostic evaluation can direct clinicians to the correct region of the spine for evaluation. However, the EMG examination takes about three weeks to become fully manifest. For this reason, spine MRI is the optimal diagnostic evaluation for acute symptoms. In this patient's case, the progression was subtle, and other diagnostic possibilities were raised.

Patients with cauda equina syndrome due to malignancy generally have a poor prognosis. In one study, median survival was 50 days after leptomeningeal metastases were detected in patients with oat cell carcinoma (Aisner, et al; 1981). Clinical findings of cauda equina syndrome associated with malignancy often include fairly rapid progression of leg weakness and bladder dysfunction. Pain at night is a worrisome symptom for cancer of the spine and warrants imaging (Errico and Kostuik; 1986).

The spinal cord can be involved in a variety of ways by malignancy elsewhere in the body, without direct metastases. Paraneoplastic encephalomyelitis is an inflammatory disorder of the brain and spinal cord thought to be immune mediated, primarily affecting the gray matter. It is most often associated with oat cell cancer of the lung (Palma; 1985). When the conus is involved, there is lower motor neuron involvement on EMG (Palma; 1985). Necrotizing myelopathy, an acute necrosis of the spinal cord occurring in patients with cancer, often spans many cord segments (Palma; 1985). In one autopsy study of necrotizing myelopathy associated with multiple myeloma, the spinal cord was necrotic without any direct tumor involvement (Story and McKelvie; 1991).

The precise electrodiagnostic findings in many of these conditions are not well described. In one report, spinal cord infarction due to a venous malformation at T9 demonstrated fibrillations in all myotomes between L4 and S2 bilaterally (Levin and Daube; 1984). In this case, like the one presented above, the sural sensory responses were spared. Sparing of the sural sensory responses, particularly in the face of severe sensory loss and profound weakness and denervation, suggests intraspinal causes proximal to the dorsal root ganglia and not polyneuropathy. In a large series of patients referred for presumed polyneuropathy, Bourque and Dyck (1990) reported normal sural nerve responses in four cases which were found to have intraspinal causes of their lower limb weakness. In patients with a laterally herniated disk or with extensive bony destruction of spinal elements, the dorsal root ganglion may be involved and the presence of abnormal sensory responses must be integrated into the overall clinical and electrodiagnostic presentation (Ball; 1989). Again, for emphasis, if the sural responses are spared in the face of profound clinical sensory loss and



muscle denervation localized to the lower limbs, intraspinal causes rather than polyneuropathy should be considered.

Imaging plays a vital and complementary role in spinal diagnosis. In order to assess the spine for bony metastases, epidural lesions, and intramedullary tumors, MRI must be performed before and after gadolinium (Chamberlain, Sandy, and Press; 1991).

CLINICAL RECOMMENDATIONS TO THE REFERRING CLINICIAN

Suggest imaging of the spine with MRI (with and without gadolinium) and plain x-rays with lateral flexion and extension views to assess for structural causes of this polyradiculopathy.

FOLLOW UP

CSF analysis was normal; ESR, RPR, HIV, and SPEP were normal. CPK was elevated at 564. Chest x-ray revealed a pulmonary nodule. Bronchoscopic biopsy demonstrated that this tumor was oat cell carcinoma of the lung.

Initial lumbar spine MRI (without gadolinium) showed only degenerative changes consistent with this patient's age. Plain x-rays of the lumbar spine with flexion and extension views did not reveal spondylolisthesis, tumor invasion, or instability.

Given the profound and compelling electrodiagnostic and clinical findings pointing to a cauda equina syndrome, the MRI was repeated with views pre- and post- administration of gadolinium. This showed an enhancing intramedullary lesion in the conus medullaris at the T11 level. MRI of the brain revealed several enhancing masses consistent with metastases.

The patient progressed to paraparesis. He required a catheter for bladder management. After a short stay on the rehabilitation service, he was discharged home and underwent chemotherapy and radiation therapy to the brain and spine.

At one year followup, he is alive and nearly paraplegic, using a wheelchair and managing his bladder with a Foley catheter. His mental status is still preserved. Repeat MRI of the brain and spinal cord revealed that those lesions had increased in size, consistent with metastases.

FINAL DIAGNOSIS

Metastatic oat cell carcinoma of the lung involving the conus medullaris, resulting in a cauda equina syndrome.

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