



EMG Case No. 79, April 2006

Presenting Symptom(s):

Bilateral leg weakness, right worse than the left, progressively worsening for the past 7-8 years, bilateral arm weakness progressively worsening for the past 2 years

This case is no longer available for CME Credit.

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Disclosures: M.J. Tinney, None; R. Werner, None.

Appropriate Audience: Residents and practicing physicians.

Learning Objectives: After completing this educational activity, participants will be able to: (1) Utilize the patient's history and physical to devise an adequate differential diagnosis of generalized weakness; (2) Describe EMG findings consistent with myopathic disease.

Level of Difficulty: Advanced.

History

A 55 year old man presented for electrodiagnostic evaluation of progressive weakness, starting in his lower extremities (R>L) 7-8 years ago, and in his upper extremities 2 years ago. His symptoms initially started in his calves. He noted weakness was present regardless of his level of activity. He did not report any pain, numbness, tingling, dysphagia, change in voice, or change in vision.

- 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 includes:

- Inflammatory myopathies
- Endocrine associated myopathies
- Drug induced and toxic myopathies
- Metabolic myopathies
- Inclusion Body Myositis
- Muscular dystrophies
- Myotonic dystrophies
- Polyneuropathy
- Myasthenia Gravis
- Lambert-Eaton Myasthenic Syndrome
- CNS lesion
- Cervical Myelopathy
- ALS



- Polyradiculopathies
- Multiple Sclerosis
- Post polio Syndrome

Given his age and gender, the diagnosis of Multiple Sclerosis is unlikely. Without history of headache or cranial nerve symptoms, a CNS lesion is also less likely. Polyradiculopathy is unlikely without a history of pain. The lack of sensory complaints would argue against most peripheral neuropathies.

History, continued

His past medical history was significant for low back pain (he had a negative EMG of lower extremities in 2001), depression, post traumatic stress disorder, and hypertension. His family history was significant for colon cancer, but was negative for muscle disease or similar weakness. His surgical history was significant for normal muscle biopsy of the left quadriceps muscles in 2001 and inguinal hernia repair. Another biopsy of the right hamstring muscles was done on the same day of this EMG, but results were not available. His medications included hydrochlorothiazide, venlafaxine, quetiapine, and Aleve. He had no allergies to drugs. On review of systems, he reported no fasciculations, numbness, tingling, incontinence, dark urine, or rash. His social history reveals he quit smoking cigarettes 14 years ago. He had no other illicit drug use or alcohol use. He was a retired GM factory worker.

- 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

At this point the differential diagnosis is broad and includes:

- Inflammatory myopathies
- Endocrine associated myopathies
- Metabolic myopathies
- Inclusion Body Myositis (IBM)
- Muscular dystrophies
- Myotonic dystrophies
- Cervical Myelopathy
- ALS
- Polyneuropathy
- Myasthenia Gravis
- Lambert-Eaton Myasthenic Syndrome

He has no history of toxin or drug exposure and no history of poliomyelitis therefore drug-induced, toxic myopathy and Post polio syndrome are not possible. The most likely medications that would cause a toxic myopathy would be zidovudine, azidothymidine, cholesterol-lowering agents, and the combination of blocking agents with corticosteroids. Other toxins which can cause myopathy include Pentazocine, chronic alcohol ingestion, gasoline sniffing, and mushroom poisoning. Post polio syndrome can be further ruled out due to the fact that he had a normal EMG back in 2001, which did not show abnormally large amplitude motor unit potentials, typically seen after Polio. The distribution of his



weakness is more consistent with IBM with it being initially distal, but physical examination is required to eliminate other disorders which may account for his symptoms.

Physical Examination

Strength is 5/5 bilaterally at the shoulder abductors, elbow flexors, elbow extensors, wrist extensors, hand intrinsics, finger flexors, hip flexors, and ankle dorsiflexors. Strength is 2/5 for the right hip extensors and 2/5 for the bilateral plantar flexors. Atrophy is seen in the bilateral biceps and quadriceps muscles. Muscle tone is normal.

Cranial nerves II-XII are intact. Extra ocular movements are normal. No nystagmus is present. Sensation to light touch is intact in all four extremities. Deep tendon reflexes are 2+ bilaterally at the biceps, triceps, brachioradialis, patellar, and Achilles tendons. No Hoffman or Babinski sign is present.

- 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

At this point, the most likely differential diagnosis includes:

- Inflammatory myopathies
- Endocrine associated myopathies
- Metabolic myopathies
- Inclusion Body Myositis
- Muscular dystrophies
- Myotonic dystrophies
- Primarily Motor Polyneuropathy

The physical examination findings are less supportive of myasthenia gravis or LEMS (no pseudobulbar findings, presence of muscle atrophy), cervical myelopathy (no upper motor neuron signs, no hyperreflexia, normal tone), or ALS (less likely due to lack of upper motor neuron signs). The pattern of weakness and quadriceps muscle atrophy found on the physical examination suggest IBM. Other myopathies would likely show more exclusive proximal muscle involvement.

LABS

CPK=3632

Aldolase=23.7

dsDNA=7

RF<20

ANA neg.

TSH=1.0

Basic metabolic panel = normal

Commentary IV

At this point, the most likely differential diagnosis includes:

- Inflammatory myopathy



- Inclusion Body Myositis
- Muscular dystrophies
- Myotonic dystrophies
- Polyneuropathy

The most likely etiology, given the lab findings is a myopathy or myotonic dystrophy. A purely motor polyneuropathy cannot be completely ruled out but is unlikely to cause a CPK over 3,000. The lab findings are less supportive of a metabolic myopathy or endocrine associated myopathy.

- Design your approach to the electrophysiologic examination based on the existing data.

Electrophysiologic Data

MOTOR NERVE CONDUCTION STUDIES							
Nerve	Side	Stim Site	Record	Cm	AMPL	LAT	CV
Ulnar Motor	Right	Wrist	Hypothenar	7.0	15.7	3.7	
No decrement with rep. stim							
Ulnar Motor	Right	Below elbow	Hypothenar	20.0	14.4	7.6	51.3
Ulnar Motor	Right	Above elbow	Hypothenar	10.0	13.9	10.0	41.7
Ulnar F	Right	Wrist	Hypothenar	7.0		28.3	
Peroneal Motor	Right	Ankle	EDB	9.0	11.8	5.1	
Peroneal Motor	Right	Below knee	EDB	27.0	11.0	10.5	50.0
Peroneal F	Right	Ankle	EDB	9.0		47.6	

SENSORY NERVE CONDUCTION STUDIES							
Nerve	Side	Stim Site	Record	Cm	AMPL	LAT	CV
Ulnar	Right	Wrist	5 th digit	14.0	65.3	3.6	46.7
Sural	Right	Calf	Ankle	14.0	28.4	3.9	46.7
Temperature	Right	Midpalm	32.3°C				
Temperature	Right	Calf	32.0°C				



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+ POLyphasia: N, inc or dec 1+, 2+, 3+, 4+									
R/L	MUSCLE	INSER	FIB	OTH	EFF	REC	AMP	DUR	POL
R	Anterior Tibialis	N	0	0	N	N	Inc 1+	N	Inc 1+
R	Vastus Medialis	Sust	2+	0	N	Dec +/-	N	Dec 1+	Inc 2+
R	Biceps Brachii	Sust	2+	0	N	Inc 1+	Dec 1+	Dec 1+	Inc 1+
R	Thoracic Paraspinals	sust	2+	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

The nerve conduction studies were normal as recorded above. The patient cooled quickly despite warming, so the slightly increased amplitude and prolonged latency seen in the right upper extremity may be a result of decreased temperature. There was no decrement seen with repetitive stimulation. There was a slight slowing of the ulnar motor conduction velocity across the elbow but this does not meet criteria for an ulnar mononeuropathy nor does he have associated symptoms. Concentric needle examination showed abnormal spontaneous activity in the proximal muscles tested as recorded above. Insertional activity consistent with fibrotic/fatty change was also seen in all proximal muscles tested. There was increased amplitude, increased recruitment, and polyphasic units in the Biceps Brachii muscle. There were also normal amplitude and polyphasic units in the Vastus Medialis muscle.

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

Diagnostic Impression

There are electrodiagnostic abnormalities consistent with a mixed myopathic and neuropathic process. A proximal to distal gradient and changes seen in the biceps brachii muscles (short duration, low amplitude, and polyphasia) are more characteristic of a classic



inflammatory myopathy. The differential diagnosis includes inflammatory myopathies, inclusion body myositis, and adult-onset muscular dystrophies.

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

Commentary V

Muscular dystrophy usually presents at an earlier age, but cannot be entirely ruled out for adult-onset dystrophies such as limb-girdle muscular dystrophy. A pure motor neuropathy can still be a possible diagnosis.

The only way to obtain a more definitive diagnosis is to perform a muscle biopsy along with genetic testing. Laboratory tests which can help in making the diagnosis including CK levels and inflammatory markers.

The inflammatory myopathies include polymyositis and dermatomyositis. It is usually characterized by progressive weakness, but a rash can occur with dermatomyositis. 20% of these patients can also have an associated autoimmune disease or connective tissue disease. The pattern of weakness is usually proximal and symmetric. Symptoms are usually subacute, but may be chronic. Nerve conduction studies are usually normal. Fibrillation potentials and positive sharp waves can indicate inflammation, although denervation potentials can also be seen in other myopathies.

Inclusion body myositis (IBM) is the most common inflammatory myopathy in individuals older than 50 years. It usually presents as slowly progressive weakness and is more common in men than women (3:1). The age of presentation is commonly in the 6th decade. Distal muscles may be more involved along with proximal weakness and the distribution is commonly symmetric but may be asymmetric. Prominent muscle atrophy, especially of the quadriceps, occurs. A subset of patients demonstrates mild sensory or sensorimotor polyneuropathy, which complicates the diagnosis. The combination of myopathic and neuropathic findings on EMG may suggest a chronic myopathy such as IBM.

The patient's muscle biopsy performed the day of this EMG evaluation ultimately showed necrotizing myopathy, characterized by small groups of muscle fibers undergoing regeneration, occasional degenerating muscle fiber with no inflammatory response, and enhanced alkaline and acid phosphatases. Possible etiologies outlined by the pathologist included a myopathy from a toxin, drug, a paraneoplastic syndrome, or a connective tissue disorder. No inclusion bodies were noted. Clinical correlation was advised.

There was also focal type I fiber predominance in the sections reacted for ATPase, rare denervated muscle fibers in the section reacted for nonspecific esterase, and probably two or three muscle fibers low or negative for cytochrome C oxidase. Myophosphorylase, phosphofructokinase, and the amount of lipids and glycogen were not unusual. Myoadenylate deaminase was lower than in the normal muscle control, which could be either secondary to the necrotizing myopathy or a primary partial deficiency.

Recent labs have been normal including lactic acid = 3.7, C-ANCA = negative, P-ANCA = neg., Anti-Jo = 3, CRP = 0.644, HIV-1/2 ab = negative, SPEP = 6.8, C4 complement = 29, C3 complement = 36.6. Genetic testing for Fascioscapulohumeral dystrophy is currently pending, although age onset is usually at the end of the first decade of life.



A repeat EMG was performed which again showed a mixed myopathic and neurogenic process but it was felt the myopathic process predominated. There was less spontaneous activity noted on the follow-up study and more fibrotic muscle noted. There was also a repeat muscle biopsy from the deltoid but there were only non-specific changes noted consistent with a non-inflammatory myopathic process. The treating neurologist felt that the patient had either limb girdle or facioscapulohumeral (FSH) muscular dystrophy and no treatment was initiated. (If it had been determined that this patient had an inflammatory myopathy or IBM, treatment would entail corticosteroid or immunosuppressive therapy. In refractory cases of IBM, intravenous immunoglobulin and low-dose whole body or lymphoid radiation are options.)

Bibliography

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