



## EMG Case No. 72, April 2005

### Presenting Symptom(s):

Lower extremity weakness and dysesthesias

**This case is no longer available for CME credit.**

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**Disclosures:** J. Gettings, None; B. Weigert, None.

**Appropriate Audience:** Residents and practicing physicians

**Learning Objectives:** After completing this educational activity, participants will be able to: (1) recognize the clinical signs/symptoms of peripheral neuropathy; (2) recognize the EMG findings in peripheral neuropathy; (3) use their knowledge of this unusual and potentially life-threatening cause of polyneuropathy when they evaluate patients in the future.

**Level of Difficulty:** Intermediate.

### History

The patient is a 57 y.o. man who presented with a 4-month history of spontaneous-onset left lower foot and ankle pain and weakness. He noted dysesthesias along the lateral aspect and dorsum of his left foot and reported a foot slap while walking. He was unable to run. He denied low back pain but did report left hip pain. He denied any pain, numbness or weakness in the upper extremities, or the other leg. Past medical history was significant only for benign prostatic hyperplasia (BPH) and asthma. A magnetic resonance imaging (MRI) done prior to this visit showed multilevel degenerative disc disease, most prominent at L5-S1, with some spinal stenosis at this level. His initial physical examination revealed 2/5 motor strength of ankle dorsiflexion, ankle plantar flexion, and extensor hallucis longus (EHL) on the left. Bilateral hip flexion, knee flexion and extension, right ankle dorsiflexion and plantar flexion, and right EHL strength were 5/5. Hip abductors were not tested. He had diminished light touch sensation along the dorsum of his left foot. Lower extremity reflexes were normal except for an absent left achilles reflex.

- 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 which might be helpful in clarifying your differential list or changing its order of priority?

### Commentary I

This gentleman's symptoms, when considered in combination with the MRI results, suggest multi level radiculopathy, potentially involving L4, L5, and S1, related to degenerative disc disease and stenosis. The differential diagnosis includes sciatic neuropathy, lumbosacral plexopathy, myelopathy, polyneuropathy, mononeuritis multiplex, inflammatory demyelinating neuropathy (AIDP, CIDP), hereditary neuropathy, or focal compression



neuropathy. Myopathy, early motor neuron disease and multifocal motor neuropathy are less likely due to the sensory abnormalities and distribution of weakness.

### **History, continued**

The patient underwent fluoroscopically-guided left L5 and S1 transforaminal epidural steroid injections two weeks after presenting to PM&R. None of his symptoms improved following this procedure. While the lower extremity numbness and tingling persisted, he did experience gradual improvement in his left lower extremity strength with physical therapy.

He then presented five weeks after his first visit, reporting right lower extremity symptoms which were similar in character and distribution to those he'd had on the left — predominately numbness and tingling in his toes which had progressed proximally in the foot.

- 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 bilateral nature of his symptoms now makes a focal disorder such as compression neuropathy or sciatic neuropathy less likely. Strength and reflex examination of all extremities as well as sensory examination should be the focus at this point.

### **Physical Examination**

Physical exam revealed slow, wide-based gait, 5/5 bilateral hip flexion, knee flexion, and extension strength; 3+/5 bilateral ankle dorsiflexion, and 2/5 bilateral EHL strength.

Upper extremity strength, sensation, and reflexes remained normal. Hip abductors were not tested.

- 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 bilateral and rapidly-progressive nature of this patient's signs & symptoms suggests a more central or systemic etiology. Bilateral polyradiculopathy related to lumbar stenosis is possible, as is bilateral lumbosacral plexopathy, but less likely.

### **Physical Examination, continued**

Sensory exam of the lower extremities revealed decreased sensation to light touch and pinprick in a stocking distribution of both lower extremities, and decreased joint position sense in both great toes.

- 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

This patient's sensory deficits make etiologies such as AIDP/CIDP polyneuropathy, and mononeuritis multiplex seem more likely. The symmetry of the findings seem to point to a polyneuropathy. The differential still includes polyneuropathy of any etiology, as well as polyradiculopathy and myelopathy.

### Electrophysiologic Data

SENSORY NERVE CONDUCTION STUDIES							
NERVE	SIDE	STIM SITE	RECORD	cm	AMPL MicroV	LAT msec	CV
Sural	R	Calf	Lat. Mal.	14	7 (>6)	4.5 (<4.5)	
Sup peroneal	R	Calf	Ankle		Absent	Absent	
Radial	R	Forearm	Thumb		32 (19)	2.6 (<2.8)	

MOTOR NERVE CONDUCTION STUDIES							
NERVE	SIDE	STIM SITE	RECORD	cm	AMPL mV	LAT ms	CV M/sec
Peroneal	R	Ankle	EDB	8	Absent	Absent	
Tibial	R	Ankle	Abd. Hall.	8	Absent	Absent	
Median	R	Wrist	Thenar	6	11.9	3.6 (<4.5)	
		Elbow	Thenar		11.9 (>4)		51 (>48)
Peroneal	L	Ankle	EDB	8	Absent	Absent	
Tibial	L	Ankle	Abd. Hall.	8	Absent	Absent	

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	Inc.	4+	0	N	Dec.	Inc. 2+	Inc. 3+	
R	Medial Gastroc.	Inc.	3+	0	N	Dec.	Inc. 2+	Inc. 3+	
R	Vastus Lat.	N	0	0	N	N	N	N	N
R	Tensor Fascia Lat.	N	0	0	N	N	N	N	N
R	Gluteus Max.	N	0	0	N	N	N	N	N
R	Lumbar Paraspinals	Inc.	2+	0	N	N	N	Inc.1+	
R	Flexor Dig. Ind.	N	0	0	N	N	N	N	N
L	Anterior tibialis	Inc.	3+	0	N	Dec.	Inc. 2+	Inc. 2+	
L	Medial Gastroc.	Inc.	3+	0	N	N	Inc. 3+	Inc. 3+	
L	Vastus Lat.	N	0	0	N	N	N	N	N

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

**Diagnostic Impression**

The NCS/EMG findings reveal a symmetric, predominately axonal polyneuropathy affecting the distal lower extremities. There is spontaneous activity and long duration MUAPs in lumbar paraspinals, which could suggest a superimposed radiculopathy.

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

**Commentary V**

One day after his EMG, this patient presented to the emergency room (ER) with Shortness of breath and hemoptysis. A chest x-ray revealed extensive, right sided infiltrates. Labs showed hypereosinophilia from 1940 up to 4790 and an elevated troponin of 8.3. TTE results included dilated cardiomyopathy with LV EF 30%, moderate MR, and pericardial effusion. A cardiac catheterization showed no significant coronary artery disease. A ventricular endomyocardial biopsy revealed eosinophilic myocarditis. The patient was given a diagnosis of idiopathic hypereosinophilic syndrome. He was started on prednisone and hydroxyurea with clinical improvement.

Idiopathic hypereosinophilic syndrome (HES) was first reported in 1968, when Hardy and Anderson described three individuals who had sustained hypereosinophilia, hepatosplenomegaly, and cardiac or pulmonary involvement. Distinguishing features of HES include eosinophils >1.5 x 10<sup>9</sup>/L lasting longer than 6 months; absence of an identifiable,



underlying cause; and systemic organ involvement. HES is nine times more common in men than in women. Onset is often between 20-50 y.o.

HES is a heterogeneous disorder and has a predilection for the heart, lungs, skin, CNS and PNS. Sensory neuropathy has been reported to be the most common type of peripheral neuropathy in HES, but motor involvement may also occur. The neuropathy is thought to be due to marked demyelination followed by axonal degeneration caused by eosinophilic release of neurotoxins.

Medical treatment involves use of corticosteroids as first-line agents to decrease the number of circulating eosinophils, followed by use of such agents as hydroxyurea or interferon-alpha for recalcitrant cases. The peripheral neuropathy is thought to improve with successful medical treatment but has not been extensively studied. Werner and Wolf reported clinical features such as motor strength and reflexes as well as evoked potential amplitudes, distal latencies, and conduction velocities to be improved 6 months after normalization of the eosinophil count. Cengiz, however, described worsening peripheral neuropathy, which ascended from the lower extremities to involve the upper extremities despite lowered eosinophil counts. Our patient has reported improved sensation and motor strength but has not undergone repeat electrodiagnostic testing.

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