



EMG Case No. 57, November 2002

Presenting Symptom: Radiating pain down upper extremity with hand weakness

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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 differential diagnosis for neck pain radiating into the upper extremity, 2) review the electrodiagnostic criteria for chronic demyelinating polyneuropathies, and 3) understand the significance of the presence/absence of temporal dispersion, as related to demyelinating neuropathies.

This case is no longer available for CME credit.

History

The patient is a 36 year old, right-handed man with neck pain and pain radiating down his left arm that started two years after he was involved in a motor vehicle accident (MVA). He also later developed pain radiating down his right upper extremity along with right hand weakness. He has complaints of intermittent numbness in bilateral upper extremities (involving all fingers). He can elicit the paresthesias in the both of the upper extremities by tilting his head toward each extremity. He reports that he was asymptomatic before the MVA.

- *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

The patient presents with radiating pain, diffuse numbness, and weakness of the left upper extremity. He also developed pain radiating down the right upper extremity. Differential diagnosis includes the following:

- Cervical radiculopathy
- Brachial plexopathy
- A diffuse polyneuropathy
- Entrapment neuropathy of the upper extremity
- Early motor neuron disease
- Neuromuscular junction disorder
- Central nervous system process



A cervical radiculopathy is high on the differential diagnosis as it is a very common disorder that may present with radiating pain and/or paresthesias along a specific dermatome. This diagnosis is supported by the fact that the patient's symptoms developed acutely after a traumatic event, has associated neck pain, and can be elicited by movement of his neck. However, because the paresthesias do not localize to a particular dermatome, and in fact are rather diffuse, a diffuse polyneuropathy would also be high on the differential diagnosis.

History, cont.

MRI of the neck had been done previously and demonstrated a left dorsal laterally herniated disc with associated posterior segment degenerative joint changes at C5-6 with minimal deformity of the cervical spinal cord. There was also a minimal right dorsilateral bulging of the C6-7 intervertebral disc. He denies history of diabetes or thyroid disease, and drinks approximately a twelve-pack of beer a month.

On further questioning about his medical status, the patient reports that he has hepatitis B and has been told that he might possibly have Charcot Marie Tooth disease. He feels these diagnoses were given to him inaccurately, and is adamant that his symptoms are a result of the neck injury he received in the motor vehicle accident. In addition, neither his mother nor father ever experienced symptoms of a neuropathy.

- *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 MRI findings are suggestive of a cervical radiculopathy but may be incidental findings as well. The possible history of Hereditary Motor Sensory Neuropathy Type I (HMSN-I), also known as Charcot Marie Tooth type I (CMT I), alters the differential and places a peripheral neuropathy as a leading diagnosis for the current complaints. CMT I is a slowly progressive demyelinating polyneuropathy, and typically affects the distal motor fibers more than the sensory fibers. It most commonly is associated with autosomal dominant inheritance, but may also be a result of autosomal recessive inheritance, as well as X-linked (dominant or recessive) inheritance. Clinical findings such as global areflexia, hypertrophic nerves, and muscle weakness with relative preservation of muscle bulk may suggest a demyelinating neuropathy. Other symptoms associated with Charcot Marie Tooth are pes cavus and hammer toes.

Physical Examination

Physical examination demonstrated enlarged ulnar and superficial radial nerves. He had pes cavus bilaterally. There were mild hammertoe deformities on the left foot. Manual muscle testing demonstrated distal weakness in all extremities with wasting noted in the distal musculature. Strength (right/left) was as follows: deltoid, biceps, triceps, and wrist extensors 5/5; finger extensors 5-/5; interossei, thumb abduction and adduction 4/4; iliopsoas, quadriceps and hamstrings 5/5; anterior tibialis 4/4; extensor hallucis longus 3+/3; ankle invertors, ankle evertors and gastrocnemius 5/5. Muscle stretch reflexes were absent throughout. Sensory examination was remarkable for decreased pin prick sensation in a glove-stocking distribution, decreased proprioception at the great toes, decreased



vibratory sensation to the ankles as well as slightly impaired at the fingers, and impaired temperature sensation to the knees. There were no fasciculations present. Finger-nose-finger and hand tapping were intact. He was unable to heel walk, but was able to demonstrate tandem toe and casual gaits well.

Spurling's is weakly positive on the left and negative on the right.

- *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 patient's physical exam findings are as would be expected with Charcot Marie Tooth; the patient has pes cavus, hypertrophied nerves, sensation and motor loss in a distal to proximal gradient. However, it becomes more difficult determining if the patient has a superimposed cervical radiculopathy. The MRI demonstrated a left laterally herniated disc at C5-6, which may possibly correspond with the radiating pain of the left upper extremity. He also had radiating pain down the right upper extremity that may have corresponded with the right C6-7 disc bulge. Symptoms of radiculopathy include pain and paresthesias that radiate along the distribution of a particular nerve root, and may have abnormal muscle stretch reflexes, depending on the affected level. For example, in this case, one of the presumed nerve roots affected is the left C5 or C6 nerve root, the symptoms expected would include sensory symptoms in the lateral arm, forearm and thumb. The biceps and brachioradialis reflexes might be impaired. Unfortunately evaluation of the reflexes does not help in this case as all of his reflexes are impaired. In addition, if the radiculopathy is severe enough, one should expect weakness in the muscles innervated by C5-6, but in this case, he is strong in the proximal C-6 muscles such as the biceps brachii, brachioradialis and deltoid.

Electrophysiologic Data

Sensory Nerve Conduction Studies							
Nerve	Side	Stim Site	Record	Cm	Amp	Lat (peak)	CV (m/s)
Sural	L	Calf	Ankle	14	NR		-
Median	L	Wrist	Index	14	NR		-
Median	R	Wrist	Index	14	NR		-
Ulnar	L	Wrist	5th	14	NR		-



Ulnar	R	Wrist	5th	14	NR		-
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Temperature: R-mid palm = 33.0; L-mid palm = 34.2; L-calf = 32.3

Motor Nerve Conduction Studies							
Nerve	Side	Stim Site	Record	Cm	Amp	Lat (peak)	CV (m/s)
Peroneal	L	Ankle	EDB	9	NR	NR	-
Peroneal	L	BK	AT	10	NR	NR	-
Tibial	L	Ankle	AH	8	0.8	8.0	-
Tibial	L	Knee	AH	38.7	0.6	29.8	17.8
Tibial F-response	L	Ankle	AH	-	-	NR	-
Median	L	Wrist	Thenar	7	4.7	10.5	-
Median	L	Elbow	Thenar	20.9	4.5	221.6	18.8
Median F-response	L	Wrist	Thenar	-	-	83.5	-
Ulnar	L	Wrist	Hypothenar	7	5.2	6.4	-
Ulnar	L	Below Elbow	Hypothenar	21.2	4.7	18.0	18.3
Ulnar	L	Above Elbow	Hypothenar	10	4.6	22.9	20.4
Ulnar F-response	L	Wrist	Hypothenar	-	-	81.0	-

Note: there was no evidence of temporal dispersion on proximal stimulation



Needle Electromyography									
Side	Muscle	Inser	Fib	Oth	Eff	Rec	Amp	Dur	Pol
L	Biceps Brachii	N	0	0	N	N	I+/-	I+/-	I1+
L	Triceps	N	0	0	N	D1+	I1+	N	N
L	Deltoid	N	0	0	N	N	N	N	N
L	Flexor Carpi Radialis	N	0	0	N	N	N	N	N
L	FDI (hand)	I	2+	0	N	D3+	I3+	I3+	I2+
L	Opponens Pollicis	N	0	0	N	D2+	I2+	I1+	I2+
L	Paraspinal-Low Cervical	N	0	0					
R	Biceps Brachii	N	0	0	N	I1+	I1+	I1+	I1+
R	Triceps	N	0	0	N	N	N	N	N
R	Ext Digitorum Communis	N	0	0	N	D1+	I1+	I1+	I2+
R	Opponens Pollicis	N	0	0	N	D+/-	I1+	I1+	I2+
R	FDI (hand)	I	2+	0	N	D2+	I3+	I3+	I3+
R	Paraspinal-Low	N	0	0					



	Cervical								
L	Anterior Tibialis	I	2+	0	N	D2+	I2+	I3+	I3+
L	Vastus Medialis	N		0	N	D2+	I3+	N	N

Nerve conduction studies revealed absent responses in the left peroneal motor response, the left tibial f-response, and all of the sensory nerves tested. The remainder of the nerves studied had decreased amplitude, prolonged distal latencies, and very slowed conduction velocities. The median and ulnar f-responses were significantly delayed. There was no temporal dispersion noted in the CMAP's.

Needle electromyography showed abnormal insertional and spontaneous activity in the first dorsal interossei of the hand bilaterally, and the left anterior tibialis. There were motor unit changes noted in three different extremities, including the following muscles: the biceps brachii bilaterally, the left triceps, the first dorsal interossei of the hand bilaterally, the opponens pollicis bilaterally, the right extensor digitorum communis, the left anterior tibialis, and the left vastus medialis.

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

Diagnostic Impression

The electrodiagnostic abnormalities noted on the nerve conduction studies and needle examination are consistent with that of a diffuse, hereditary, sensory-motor demyelinating peripheral polyneuropathy. The diagnosis of a superimposed radiculopathy of the upper extremity is difficult to confirm or exclude on this study due to the diffuse changes related to the demyelinating neuropathy. It is difficult to determine if the abnormal findings in the proximal muscles are related to a radiculopathy or the demyelinating neuropathy. The proximal findings, for the most part, are symmetrical indicating that their abnormalities are most likely related to the polyneuropathy. In addition, the lack of abnormal EMG findings in the cervical paraspinals, deltoid, triceps, and flexor carpi ulnaris make C5, C6, C7 radiculopathy less likely. A superimposed C8-T1 radiculopathy is even more difficult to rule out on this study, however it is unlikely, as the paraspinals are not involved and it does not correlate with the patient's symptoms or MRI findings.

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



- *What treatment would you recommend?*

Commentary V

The electrodiagnostic criteria required to make a diagnosis of a chronic demyelinating neuropathy include:

1. Prolonged distal motor latencies symmetrically in both upper and lower extremities (in at least two nerves not at entrapment sites) greater than or equal to 130% of the upper limit of normal
2. Slowed conduction velocity of less than or equal to 75% of the lower limit of normal
3. F-responses and H-reflex greater than 130% of the upper limit of normal
4. Conduction block and temporal dispersion do not occur in inherited demyelinating neuropathies such as it does in acquired demyelinating neuropathies.

Sensory nerve conduction studies are abnormal in both lower and upper extremities. The sensory nerve responses are commonly absent and are commonly less than 60% of normal. Motor nerve conduction studies may be difficult to obtain when recording from the extensor digitorum brevis or the abductor hallucis, thus requiring the anterior tibialis muscle to be used, as was attempted in this case. If there is a response recorded from the EDB or AH (such as the response recorded from the AH in this case), the CMAP amplitude is decreased and the distal latency is significantly prolonged. Conduction velocities of 20-25m/s are common. The characteristic findings in HMSN I is a dramatically reduced nerve conduction velocities, usually in the 10-15 m/s range (18m/s in this particular case) with good preservation of waveform morphology.

Temporal dispersion or conduction block is generally not seen in hereditary demyelinating neuropathies, but is seen in acquired demyelinating neuropathies such as AIDP or CIDP. This is a key feature to help differentiate between inherited and acquired demyelinating polyneuropathies. With inherited demyelinating neuropathies, all the myelin is effected equally causing uniform symmetric slowing of conduction velocity (thus no temporal dispersion). In acquired demyelinating neuropathies, temporal dispersion and conduction block occurs due to patchy, multifocal demyelination causing asymmetry in nerve conduction studies.

Proximal nerve conduction using f-waves is generally considerably prolonged and may be absent.

The EMG findings usually show evidence of re-inervation with little spontaneous activity. Spontaneous activity may be seen in demyelinating neuropathies, as there may be secondary axonal loss. Abnormal spontaneous activity may be present, and when found, is mostly commonly in the distal muscles. The tibialis anterior is the muscle most likely to have spontaneous activity. Motor unit changes include decreased recruitment, long duration, high amplitude and polyphasia. The motor unit findings tend to be more prominent in the distal limb muscles, but may also be seen in the proximal and limb girdle muscles. The degree of abnormalities on the nerve conduction studies and electromyography do not necessarily correlate with the severity of the disease.



The diagnosis of HMSN-I may be confirmed by genetic testing, as was done and was positive in this patient. There is no effective treatment for HMSN-I other than supportive care with physical therapy, occupational therapy and assistive orthotic devices.

Bibliography

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