

EMG Case No. 86, April 2007

Presenting Symptom(s): Bilateral hand numbness

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Disclosure: S Christensen, None.

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Appropriate Audience: Residents and practicing physicians.

Learning Objectives: After completing this educational activity, participant will be able to: (1) Use a patient's history and clinical examination to give a differential diagnosis of diffuse polyneuropathy; and (2) Recognize when a superimposed process occurs in a neuromuscular disease.

Level of Difficulty: Intermediate

History

A 46 year old right hand dominant male presents with a six month history of numbness in the thumb and first and second digits bilaterally. The symptoms developed insidiously and are gradually getting worse. The right hand is worse than the left. He notes a weakness in his grip bilaterally. His toes also occasionally become numb bilaterally, but his legs do not feel weak. He has had no recent illnesses or infections. He also has pain in multiple joints.

Commentary I

At this point the differential diagnosis includes:

Carpal Tunnel Syndrome

Brachial plexopathy

Polyneuropathy: axonal or demyelinating, distal > proximal

Toxic causes: diabetes, alcohol, medication related

GBS

CIDP

Associated with connective tissue disease

Mononeuritis multiplex

Multifocal motor neuropathy with conduction block

HSMNs

Other acquired polyneuropathies: multifocal motor neuropathy with conduction block, diphtheria, toxic (arsenic, amiodarone, glue sniffing)

Polyradiculopathy

The distribution of numbness suggests carpal tunnel syndrome. However, a sole diagnosis of carpal tunnel syndrome would not explain the numbness in his feet and suggest a more diffuse process with or without superimposed focal mononeuropathies. Brachial plexopathy usually does not present bilaterally but could explain hand numbness.

History, continued

His past medical history includes: seizure disorder, migraine headaches, GERD, cervical arthritis, DJD of various joints, rotator cuff disorder on the left, generalized anxiety disorder, and left ventricular dysfunction. He is status post an unknown left shoulder surgery. He has no documented thyroid disease or diabetes mellitus.

His medications include Flexaryl TID prn, Depakote 750mg BID, Etodolac 400mg BID, Propranolol 40mg BID, Felodipine 10mg QD, Loratadine 10mg QD. He was taking Simvastatin 80mg but stopped this six months prior to presentation as he thought it might be affecting his muscle bulk.

He does not drink or smoke currently. He used cocaine in the past, but has not used since 2001. He stopped marijuana use in 1/06. He never drank alcohol "heavily." His family history is unknown. He works in heavy labor doing odd jobs.

1. If necessary, revise your differential diagnosis based on the additional clinical history.
2. On which details of the physical examination should you focus at this point?

Commentary II

Given his work history, carpal tunnel syndrome is still high on the differential, but still does not explain all of his symptoms. A polyneuropathy is likely given the involvement of the upper and lower extremities. The cause could be toxic, from previous drug use or associated with HIV infection. He could also have a polyneuropathy associated with an undiagnosed connective tissue disorder or other autoimmune process. Although the patient is not currently taking a statin medication, there have been case reports of statin induced polyneuropathy which do not improve after the medication is stopped. His unknown family history does not help to determine if he may have a hereditary polyneuropathy. With a history of cervical arthritis, and likely associated arthritis of the lumbar spine, polyradiculopathy and/or cervical spinal stenosis remains a possibility.

Physical Examination

Strength in the upper extremities is 5/5 in shoulder abduction and elbow flexion; 4/5 in elbow extension, and wrist extension, and 4-/5 in the finger flexors and extensors and hand intrinsics. Some atrophy was noted in the abductor pollicis brevis bilaterally. Strength in the lower extremities was 5/5. He also had atrophy of the foot intrinsics. Reflexes were 1+ at the Biceps, Triceps and Brachioradialis as well as the Patella's. Ankle reflexes could not be obtained. Vibratory sense was diminished at the toes. Pin prick sensation was diminished in a patchy, non-dermatomal distribution. Toes were down going bilaterally. He had a positive Tinel's over the median nerve at the wrist bilaterally. Spurling's test was negative bilaterally.

Commentary III

The physical examination, as well as the history, suggests a symmetric process affecting both the upper and lower extremities, with a distal to proximal gradient. These symptoms would continue to suggest a polyneuropathy. A lack of recent viral illnesses would and the duration of symptoms would argue against an acute diffuse demyelinating disease such as AIDP. Given the atrophy that he has in the foot intrinsic muscles, a chronic condition is more likely. Toxic causes as mentioned above are still possible. Another possibility is a chronic demyelinating polyneuropathy, such as HSMNs, or CIDP. He also has symptoms consistent with carpal tunnel syndrome and a



positive Tinel’s sign at the wrist continues to suggest that this may be super imposed on a polyneuropathy. His weakness in multiple myotomes could still represent a polyradiculopathy, although with a nondermatomal sensory loss pattern, and symmetric reflexes, as well as a negative Spurling’s maneuver, this is less likely.

Physical Examination, continued

Currently a repeat EMG by neurology is pending. No labs or other studies have been ordered.

1. If necessary, revise your differential diagnosis based on the additional physical findings.
2. 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
Median	R	Wrist	Thenar	7	2.1	6.4	
Median	R	Elbow	Thenar	24	1.8	12.7	27.5
Median	L	Wrist	Thenar	7	5.5	4.7	
Median	L	Elbow	Thenar	23.5	4.8	10.7	27.5
Ulnar	R	Wrist	Hypothenar	7	8.7	3.0	
Ulnar	R	Below elbow	Hypothenar	23	8.6	8.2	31
Ulnar	R	Above elbow	Hypothenar	10	8.5	11.2	33
Ulnar	L	Wrist	Hypothenar	7	5.6	3.3	
Ulnar	L	Below elbow	Hypothenar	23	5.6	9.6	25.5
Ulnar F	R	Wrist	Hypothenar		36 with dispersion		
Peroneal	R	Ankle	EDB	9	3.0	6.3	
Peroneal	R	Below knee	EDB	28	2.7	18.9	15
Peroneal F	R	Ankle	EDB	9	56- with dispersion		

SENSORY NERVE CONDUCTION STUDIES							
NERVE	SIDE	STIM SITE	RECORD	cm	AMPL	LAT	CV
Median	R	Wrist	Index	14	6.1	4.9	35
Median	L	Wrist	Index	14	12	4.3	38.9
Ulnar	R	Wrist	5 th	14	5.5	3.7	46.7
Ulnar	L	Wrist	5 th	14	15.5	3.7	46.7
Radial	L	Forearm	Thenar	10	23	2.5	56
Sural	R	Calf	Ankle	14	5.3	5.5	29.8



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	FDIP	N	0	0	N	N	N	N	N
R	AT	N	0	0	N	N	N	N	N
R	APB	N	0	0	N	N	N	N	N
R	PRT	N	0	0	N	N	N	N	N

Electrophysiologic Data, continued

Nerve conduction studies showed low amplitude responses in the median and ulnar sensory and motor nerves bilaterally as well as sural sensory on the right. The recorded responses in most nerves showed temporal dispersion. Distal latencies were prolonged and conduction velocities were slowed in all nerves tested with the exception of the radial sensory nerve on the right. The motor nerves had a more significant slowing than the sensory nerves tested. The F- wave latencies were prolonged and the responses showed increased chronodispersion. The temperature was initially greater than 32 degrees in all of the extremities tested. In the right ankle, the temperature dropped below 30 degrees and could not be raised despite prolonged warming. The limited needle examination (due to patient tolerance) was normal.

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

Diagnostic Impression

There are electrodiagnostic abnormalities consistent with a moderately severe, age indeterminate, diffuse sensorimotor, primarily demyelinating polyneuropathy, without ongoing denervation. There is also electrodiagnostic evidence of superimposed carpal tunnel syndrome on the right. The patient does not meet electrodiagnostic criteria for carpal tunnel syndrome on the left because the median sensory distal latency is 115% of normal, which is less than the 130% of normal that would be expected for a demyelinating polyneuropathy. Also, the difference between the median and ulnar sensory distal latencies is 0.6 which is within normal limits in most labs.

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

Commentary IV

Polyneuropathies are very common and there are multiple causes, which can be described by the pattern of nerve involvement (proximal vs distal), underlying nerve pathology (axonal, demyelinating or mixed). The patient has a primarily demyelinating polyneuropathy based on the prolonged distal latencies, slowed conduction velocities and low amplitudes in multiple nerves tested. The diagnostic criteria for a demyelinating polyneuropathy include three of the following in motor nerves: Prolonged DLs not at entrapment sites >130% ULN, conduction velocities <75% LLN, Prolonged F or H responses in 2 nerves, conduction block or temporal dispersion in one or more nerves. The criteria for an inherited demyelination polyneuropathy are modified to include 2 of the above since temporal dispersion and conduction block do not occur. EMG finding can include abnormal spontaneous activity in the foot

intrinsic muscles, and sometimes the anterior tibialis. Motor unit changes include decreased recruitment with large amplitude polyphasic motor units can be seen, but motor units are frequently normal.

The differential of demyelinating polyneuropathies can be divided into uniform demyelinating diseases, which include the hereditary polyneuropathies or segmental demyelinating diseases, which include AIDP and CIPD. The temporal dispersion seen in the nerve conduction studies in this patient suggest an acquired etiology. He also has atrophy in the foot intrinsics, which would suggest a chronic condition. The atrophy could be from denervation or disuse. With a mild, purely demyelinating disorder, abnormal spontaneous activity would not be seen as there would be no axonal loss. The low amplitudes noted in the sensory and motor nerves are likely from conduction block. Motor unit changes would also be absent in this case.

CIPD is an acquired demyelinating motor and sensory polyneuropathy. Patients are typically diagnosed in their 40s or 50s, but it can occur at any age. The time course involves symptoms for > 6 weeks and may occur in a stepwise progression, relapsing or remitting course or follow a monophasic progression. They can present with motor or sensory findings. Both the proximal and distal muscles are affected. Reflexes are absent and touch and vibratory sense (large sensory fibers) are affected more than pain and temperature (small sensory fibers). Bulbar and/or respiratory weakness is not common. A wide based gait and ataxia can be present. CSF fluid usually has elevated protein. Nerve biopsy may show segmental demyelination but is often not specific. Nerve root hypertrophy can sometimes be seen on MRI. CIPD is treated with plasma exchange although this is not as successful as in acute idiopathic demyelinating polyneuropathy.

The diagnosis of CIPD is questionable in this patient. He had very minor symptoms, which were mainly sensory, and no appreciable weakness. He also did not note any gait disturbances which are common in CIPD. His course also did not follow any specific progression as his symptoms were mild. However, he does have hyporeflexia, and the slow nerve conduction velocities, as well as prolonged distal latencies which are fairly symmetric do fit the diagnosis of CIPD. He also has a history of polysubstance abuse, which would suggest that CIPD associated with HIV could be a possibility.

Other causes for segmental demyelinating polyneuropathies include multifocal motor neuropathy with conduction block, leprosy, diphtheria, Lyme disease, arsenic, osteosclerotic myeloma, Waldenstrom's macroglobulinemia, monoclonal antibody syndromes, lymphoma, hypothyroid neuropathy, medications, associated with ulcerative colitis, SLE or acromegaly.

Monoclonal antibody syndromes can cause a demyelinating peripheral neuropathy that can mimic CIPD. Patients with MUGA often have a progression of sensory symptoms in a distal to proximal progression. The lower limbs are often more affected, although this patient only had mild nerve conduction slowing in the lower extremities. Benign IgG and IgA monoclonal antibody syndromes often cause a very mild slowing of the nerve conduction in mainly a distal progression. Benign monoclonal antibody syndromes are occasionally associated with CNS lymphoma. Antibodies to myelin-associated glycoprotein or sulfated glucuronyl paragloboside also cause a slowly progressive sensory-motor polyneuropathy. Any of these etiologies are possible in this patient. Lab testing in these patients shows monoclonal antibody spikes on SPEP and CSF protein is elevated. These syndromes often respond to IVIG or plasma exchange. Other immunosuppressive therapies have also been reported to be successful.

The other paraprotein syndromes have systemic features which are lacking in this patient and are not as likely. Amyloidosis also causes distal, symmetric symptoms, mild slowing of conduction velocities and sensory greater than motor symptoms. Osteosclerotic myeloma and Waldenstrom's macroglobulinemia also have distal and symmetric symptoms, but often include more motor findings and have very slowed nerve conduction velocities.

As carpal tunnel syndrome is common with hypothyroidism, and the patient has a superimposed CTS, an undiagnosed thyroid disorder should be entertained. These patients commonly have sensory loss or dysesthesias in a stocking-glove distribution. The lack of systemic symptoms in this patient make SLE, ulcerative colitis, or sarcoidosis. This patient is on no medications that have been associated with demyelinating polyneuropathies.

Multifocal motor neuropathy with conduction block is not as likely, given the fact that there is minimal weakness, there was no evidence of conduction block, and there was no stepwise progression of the symptoms. MMN usually

also has no sensory nerve abnormalities, which are evident in this patient. Leprosy, diphtheria, and Lyme's disease are rare causes, but in a veteran of war, he could have had a mild exposure in the past.

Entrapment neuropathies often co-exist with peripheral neuropathies and are sometimes referred to as a double crush syndrome. This is typically described for carpal tunnel syndrome associated with a cervical radiculopathy. The theory is that the nerve affected by the cervical radiculopathy, or other process affecting the health of the nerve is unable to repair itself as well to minor trauma, thus accelerating damage from an entrapment neuropathy such as CTS. This patient does meet criteria for CTS on the right, which is likely related to his work history.

The patient was seen in the neurology clinic. The neurology team plans to repeat the EMG. No treatment has been started at this time, and no labs have been ordered. Appropriate work-up would include a search for monoclonal antibodies, HIV or other infectious agents as listed above, amyloidosis, or an undiagnosed connective tissue disorder or malignancy. Treatment would be based on the causative agent and was described previously.

Bibliography

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