



## April 1997 EMG Case-of-the-Month

This case is no longer available for CME credit.

Cases prepared by: Ian MacLean, MD; Daniel Dumitru, MD;  
Lawrence R. Robinson, MD; Steve R. Geiringer, MD

### HISTORY

A 32-year-old left hand dominant man has a two month history of progressive left upper limb and neck pain with no known inciting event. He recalls that the pain began insidiously as a dull ache in the posterior left neck and upper shoulder area. Over the course of several weeks the pain progressed to involve his entire left upper limb. There is an associated sensation of numbness affecting the entire left hand. Occasional numbness is noted along the left arm and forearm in no discrete distribution. Weakness is described for the entire left upper limb. The pain is constant in nature but is particularly annoying at night, as it prevents him from sleeping through the night.

He is presently not employed because of his arm pain. The patient had a previous radiculopathy on the right side which was treated with a C5 laminectomy and discectomy in 1994. He is on no medication at this time and is not being treated for any medical conditions.

- **Prior to continuing, please develop a differential diagnosis and list each possible diagnosis in order of likelihood.**
- **On what details of the physical examination do you think you should focus at this point?**

### PHYSICAL EXAMINATION

The patient is alert, oriented, and in no acute distress. Inspection of his torso and arm reveals no evidence of muscle atrophy, skin lesions, or unusual hair loss. His spine is well aligned. Cervical range of motion is unrestricted; however, left cervical pain is described on forward cervical flexion. Manual muscle testing reveals 5/5 strength in the right upper limb as well as both lower limbs. The left upper limb is difficult to accurately examine secondary to pain.

The patient has at least 4-/5 strength in all muscle groups of the left shoulder girdle and limb. Muscle stretch reflexes are symmetrically 2+ for biceps, triceps, brachioradialis, pronator teres, finger flexors, knee extensors, and ankle plantar flexors. Spurling's sign is absent on the left and right.

There are a significant number of trigger points in the left trapezius, sternocleidomastoid, levator scapula, supraspinatus, infraspinatus, and teres major/minor muscles, which appear to replicate a significant portion of the his symptoms. Sensation to pin prick and light touch are intact and symmetric for the left and right upper limbs. No abnormalities in sensation, tone, or strength are noted for the lower limbs.



**ADDITIONAL INFORMATION: LABORATORY DATA**

Serum analysis revealed normal electrolytes and blood sugar. Thyroid studies were unremarkable. Muscle enzymes, in particular serum CK levels, were normal as was the erythrocyte sedimentation rate. Plain X-rays of the cervical region were normal, including flexion/extension and oblique views. An MRI of the cervical region was also noted to be normal.

- **At this point, review your differential diagnosis and determine a final working differential diagnosis from which to design your electrodiagnostic study.**
- **Formulate your approach to the electrodiagnostic study.**

**ELECTROPHYSIOLOGIC DATA:  
 NEEDLE ELECTROMYOGRAPHIC STUDIES**

ELECTROMYOGRAPHY								
N = normal incr = increased decr = decreased 0 = absent 1+ = minimal 4+ = maximal crd = complex repetitive discharge fasc = fasciculation potential myk = myokymic discharge myt = myotonic discharge nmt = neuromyotonic discharge pos wav = positive sharp waves fibrillation = fibrillation potentials recr = recruitment amp = amplitude dur = duration poly = polyphasic potentials								
R/L	MUSCLE	INSERTION SPONTANEOUS			VOLUNTARY			
		activ	p wave	fib	recrt	amp	dur	poly
L	deltoid	incr	1-2+	0	N	N	N	N
L	biceps	incr	1-2+	0	N	N	N	N
L	triceps	incr	1-2+	0	N	N	N	N
L	pronator teres	incr	1-2+	0	N	N	N	N
L	extensor carpi radialis	incr	1-2+	0	N	N	N	N
L	flexor carpi radialis	incr	1-2+	0	N	N	N	N
L	abductor pollicis brevis	incr	1-2+	0	N	N	N	N
L	first dorsal interosseous	incr	1-2+	0	N	N	N	N
L	extensor digitorum communis	incr	1-2+	0	N	N	N	N
R	deltoid	incr	1-2+	0	N	N	N	N
R	biceps	incr	1-2+	0	N	N	N	N
R	triceps	incr	1-2+	0	N	N	N	N
R	pronator teres	incr	1-2+	0	N	N	N	N
R	abductor pollicis brevis	incr	1-2+	0	N	N	N	N
R	first dorsal interosseous	incr	1-2+	0	N	N	N	N
L	vastus medialis	incr	1-2+	0	N	N	N	N
L	tibial anterior	incr	1-2+	0	N	N	N	N



L	gastrocsoleus	incr	1-2+	0	N	N	N	N
R	tibialis anterior	incr	1-2+	0	N	N	N	N
R	gastrocsoleus	incr	1-2+	0	N	N	N	N
R	vastus medialis	incr	1-2+	0	N	N	N	N
L/R	lumbosacral paraspinals	incr	1-2+	0	N	N	N	N
L/R	cervical paraspinals	incr	1-2+	0	N	N	N	N

\* The positive sharp waves observed in all muscles examined were manifest as decrescendo runs. These were not observed to wax and wane. Needle insertion maximally elicited these potentials, but they generally persisted with less intensity than that immediately following needle movement.

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	-	3.7	-	-	11.0	-	-	-	-
ulnar	-	2.9	-	-	10.7	-	-	-	-
median	-	3.2	-	-	15.5	-	-	-	-
median: mid-palm (antidromic)	-	1.8	-	-	23.0	-	-	-	-

MOTOR NERVE CONDUCTION									
nr = no response									
NERVE	LATENCY (ms)			AMPLITUDE (mV)			CONDUCT VEL (m/s)		
	R	L	Norm	R	L	Norm	R	L	Norm
peroneal	-	4.0	-	-	4.5	-	-	50	-
tibial	-	3.9	-	-	5.9	-	-	51	-
median	-	3.1	-	-	9.5	-	-	62	-
ulnar	-	2.6	-	-	7.2	-	-	64	-

- What additional nerve conduction data would be of value given the above findings?
- On the basis of the clinical and electrodiagnostic evaluation, formulate your final impression by determining the most likely diagnosis. List other



**possibilities that are not excluded by the data. Eliminate those diagnoses not supported by the data.**

## **DIAGNOSTIC IMPRESSION**

1. Normal nerve conduction studies.
2. Evidence of diffuse generalized membrane instability in all the muscles examined. It is to be noted that no fibrillation potentials were found and the positive waves were rather profuse and demonstrated decrescendo runs without waxing and waning.

## **ELECTRODIAGNOSTIC MEDICINE IMPRESSION**

1. The electrodiagnostic examination reveals no evidence of a left cervical radiculopathy, left median neuropathy at the wrist, or generalized peripheral neuropathy.
2. There is evidence of generalized muscle membrane instability in all the muscles examined. This finding likely represent a benign muscle membrane disorder possibly related to a genetic abnormality of one of the transmembrane ion channels such as sodium, potassium, or chloride.

## **CLINICAL IMPRESSION**

1. The study does not support the diagnosis of a cervical radiculopathy.
2. The majority of the patient's complaints are related to multiple trigger points in the left shoulder girdle musculature.

## **RECOMMENDATIONS**

1. The patient is to continue follow-up with Neurosurgery.
2. No further electrodiagnostic evaluation in this patient is needed at this time.
3. A comprehensive rehabilitation program will be instituted for his myofascial pain complaint.

## **DIFFERENTIAL DIAGNOSIS**

A patient complaining of neck or back pain associated with corresponding limb pain should be considered to have a possible radiculopathy. In the cervical region, a radicular lesion usually results in pain along the cervical spine region and the medial border of the scapula. The pain radiates into the related upper limb in various distributions depending upon the affected nerve root. Shoulder girdle and upper humeral pain can be described in persons with C5 or C6 radiculopathies. Pain in the above noted regions with accompanying forearm pain may involve the C7 nerve root. An extension of the pain into the hand can be found in C8 and T1 radiculopathies. This pain distribution is somewhat unreliable, as it involves the sclerotomes and myotomes, which have less well defined distributions than dermatomes. Failure to document weakness in a myotomal distribution is not supportive of a radicular lesion. Also, intact muscle reflexes do not suggest a radiculopathy. A vague complaint of



upper limb numbness in no particular dermatome is also not supportive of a radicular injury. Although many patients with radiculopathies do not fulfill all the textbook criteria for a nerve root lesion, they should at least have a few. In this patient the lack of muscle weakness, reflex changes, a poorly defined sensory complaint, and atypical pain do not provide strong evidence for a nerve root insult. A normal MRI of the cervical region in combination with the clinical findings also speak against a radicular lesion. Despite these, radiculopathy should still be kept in mind when performing the needle electromyographic studies.

The clinical finding of numerous trigger points in the shoulder girdle muscles is consistent with a myofascial pain syndrome. This diagnosis is confirmed by the reproduction of symptoms with trigger point palpation. It is important to keep in mind that trigger points may be primary or secondary. Primary trigger points arise in muscles secondary to trauma or some form of dynamic muscle imbalance likely arising from a musculoskeletal injury. Unfortunately, the patient does not recall any inciting events such as heavy lifting, sporting activities, or on-the-job injuries to serve as a cause for the trigger points. Some patients develop primary trigger points for unclear reasons and may have done something as simple as sleeping in a awkward position the night before.

Secondary trigger points may develop in response to various disorders. It is incumbent upon the practitioner to investigate the possibility of an underlying condition producing trigger points as one of its associated peripheral markers. The physical examination combined with a few preliminary laboratory studies do not suggest an underlying endocrine reason for the development of trigger points. At the time of the electrodiagnostic medicine examination, it appears as though the patient has primary trigger points as the major presenting reason for his pain complaints.

One must also consider the possibility of an entrapment neuropathy as the underlying cause for the symptoms, i.e. the primary inciting reason for the formation of trigger points. The most common entrapment neuropathy encountered in the upper limb is carpal tunnel syndrome. Patients with carpal tunnel syndrome may indeed complain of not only wrist pain, but forearm, arm, shoulder, and even neck pain. The pain may be rather vague in its distribution, and in advanced cases seem rather similar to that described by the patient in this case. As noted above, however, the patient does not demonstrate any symptoms or signs suggestive of a focal median neuropathy at the wrist. An ulnar or radial neuropathy is even less likely than a median neuropathy at the wrist, particularly in the absence of focal neurologic findings.

A very remote possibility may be some type of occult carcinomatous process, especially since the patient complains of pain worse at night. Fortunately, there are no associated symptoms such as night sweats, weight loss, or other clinical symptoms suggestive of carcinoma. The radiologic imaging studies performed to investigate the possibility of nerve root compromise demonstrate that at least the cervical region appears to be devoid of any mass or lytic lesions suggesting a cancerous process.

## **FORMULATION OF ELECTROPHYSIOLOGIC STUDIES**

The above differential diagnosis discussion permits us to focus on two disorders that only atypically present with a symptom complex as noted for our patient: 1) cervical radiculopathy, and 2) carpal tunnel syndrome. Despite the fact that the cervical MRI did not reveal a structural abnormality, it is certainly still possible for patients to have nerve root dysfunction. A physical compromise of the nerve may be beyond the resolution of the MRI.



Also, not all radicular lesions result from structural nerve root compromise. For example, patients with diabetes mellitus may have multiple radiculopathies resulting from infarction of the vasa nervorum feeding the nerve roots. Despite this information, the patient's physical examination, laboratory studies, and imaging studies do not suggest any type of nerve root lesion. The needle electromyographic examination is primarily performed to investigate the possibility of a radiculopathy on the very remote chance that an atypical disease process is present.

Needle electromyographic examination of this patient began in a rather routine fashion. Positive sharp waves were first noted in the deltoid and biceps brachii muscles. Next, the triceps muscle was examined and when membrane instability was noted, there was some surprise but it was documented. Abnormalities in the pronator teres muscle is to be expected in compromise of the C6 or C7 nerve roots. Detection of abnormalities in the hand intrinsic muscles was rather surprising since the implication is that the patient may have a polyradiculopathy on the left. Additional muscles were examined in the left upper limb to confirm the previous noted abnormalities. All muscles examined showed membrane instability. It was decided to explore the contralateral limb. Positive sharp waves were detected in all muscles studied. The lower limbs were then explored, again showing positive sharp waves to needle insertion with variable degrees of persistence in all locations.

Clearly, the finding of positive sharp waves in every muscle explored is not consistent with a left cervical radiculopathy. It is interesting to note that in no muscle were fibrillation potentials documented. The positive sharp waves were not characteristic of those seen in most lesions of nerve or muscle. Specifically, these occurred in decrescendo trains that decreased in numbers of potentials but continued to persist at all sites. These potentials were somewhat similar to myotonic runs, except that the potentials did not wax and wane. Also, the observed positive sharp waves were relatively short in duration, not exceeding five to ten milliseconds. Positive sharp waves from nerve or muscle disease are usually longer than ten milliseconds and can even reach 100 ms or more, especially when a low frequency filter of 2-3 Hz is used.

Nerve conduction study analysis is very appropriate in this patient. The lower limbs are explored first, as most diffuse peripheral nerve processes preferentially affect the nerves in the lower limbs. Lower limb motor and sensory studies were completely normal, as were those in the left upper limb. There is therefore no suggestion based on nerve conduction studies that the patient has a peripheral polyneuropathy. Also, the motor and sensory studies of the left median nerve are not consistent with a median neuropathy at the wrist. Ulnar nerve motor and sensory studies are also normal.

## **FORMULATION OF AN IMPRESSION**

The formulation of an impression is not particularly straightforward. Clinically, the patient has a myofascial pain syndrome with multiple trigger points in the left shoulder girdle region. This likely accounts for the patient's major complaint of pain. Neither the clinical examination nor electrophysiologic findings suggest a cervical nerve root lesion, median neuropathy at the wrist, or generalized peripheral neuropathy. In short, the clinical impression in this patient is multiple trigger points which are to be considered primary in nature, treated with an aggressive rehabilitation program.

Diffuse membrane instability consisting of only positive sharp waves which manifest in numerous trains of decrescendo runs is consistent with a poorly defined entity commonly



referred to as "EMG Disease". Other terms applied to this disorder are "Diffuse or Widespread insertional activity" and "Forme-fruste of myotonia congenita". Unfortunately, no one has characterized this disorder from a genetic standpoint with respect to a particular chromosome or particular membrane defect. It is known that this disorder occurs in an autosomal dominant form because several families have been extensively studied. Laboratory analysis of serum enzymes and electrolytes are typically normal, as is the muscle biopsy.

It is typical for persons with widespread membrane instability from this entity to be discovered only when an electromyographic examination is performed for possible focal disease such as median mononeuropathy or radiculopathy. This disorder is benign when present in isolation. That is, when family members of the patient are examined, there is nothing that can be found on clinical examination to suggest a generalized muscle disorder. The benign nature of the disorder is likely why it has been poorly characterized genetically.

The fact that widespread membrane instability is noted on needle examination with no associated weakness or myotonia suggests that the primary abnormality is a very mild defect in one of the transmembrane ion channels such as sodium, potassium, or chloride. This disorder should most likely be classified as a channelopathy, along with myotonia congenita, paramyotonia, hyperkalemic periodic paralysis, and related diseases. It is unclear why fibrillation potentials are not observed. This discrepancy clearly points out our deficient knowledge regarding the generation of positive sharp waves and fibrillation potentials. Further work is required to more completely elucidate the generation of these decrescendo runs of positive waves in this disorder.

It is necessary to be aware of this disorder so as to not confuse it with some other disorder that carries a much more ominous prognosis. Detecting widespread membrane instability may cause one to consider a disorder such as motor neuron disease; the lack of associated EDx findings speak against a motor neuron process. A diffuse peripheral neuropathy may also be entertained when "positive sharp waves" are detected in all muscles. Again, the lack of fibrillation potentials and the normal motor unit potentials and conduction study values mitigate against a diffuse peripheral nerve problem. Inflammatory myopathy might also be considered, however, normal motor units and the lack of fibrillation potentials are inconsistent with a myogenic disorder. Therefore, the observation of short duration positive sharp waves appearing in decrescendo runs (without waxing and waning), unaccompanied by fibrillation potentials, with normal motor unit potentials and nerve conduction studies should raise the suspicion of this benign disorder.

One may ask how it is possible to diagnose an axon loss lesion in a person with an underlying primary muscle membrane abnormality. Simply put, it is not easy. First, there has not been a patient described who has been carefully studied with this disorder who has had fibrillation potentials accompanying the positive sharp waves. This author has studied three families, each with multiple affected members. In no person with this disorder were fibrillation potentials found. Therefore, if fibrillation potentials are detected, one may cautiously conclude that an axonal lesion is present, particularly if they are documented in a myotomal or peripheral nerve distribution. Second, the MUAP morphology and recruitment in these patients is normal. There does not appear to be intrinsic loss of muscle fibers either from axon loss or muscle disease. If MUAPs are abnormal, then a disease process is likely present producing motor unit remodeling, whether myopathy or neuropathy. Third, the nerve conduction studies in asymptomatic persons with this disorder are normal. Clinically, these patients do not have the focal or generalized weakness of a disease process. In short,



the only abnormality noted is the diffuse muscle membrane instability. Therefore, any abnormalities of the type noted above suggest a disease process other than the benign channelopathy, and the patient should be treated accordingly.

## **BIBLIOGRAPHY**

1. Dumitru D: *Electrodiagnostic Medicine*. Philadelphia, Hanley & Belfus, 1995.
2. Nutter P, Collins K: Diffuse positive waves: case report. *Arch Phys Med Rehabil* 1988; 69: 295-296.
3. Wiechers DO, Johnson EW: Diffuse abnormal electromyographic insertional activity: a preliminary report. *Arch Phys Med Rehabil* 1979; 60: 419-422.
4. Wiechers DO, Johnson EW: Syndrome of diffuse abnormal insertional activity. *Arch Phys Med Rehabil* 1982; 63: 538.
5. Wright KC, Ramsey-Goldman R, Nielson VK, et al: Syndrome of diffuse abnormal insertional activity: case report and family study. *Arch Phys Med Rehabil* 1988; 69: 534-536.