



EMG Case No. 76, October 2005

Presenting Symptom(s):

Left Wrist Drop

This case is no longer available for CME credit.

Case prepared by: Edward Babigumira, MD, Stephen Kishner, MD,
Peter A. Zimmerman, MD

Affiliation: Louisiana State University, Section of Physical Medicine and Rehabilitation

Disclosures: E. Babigumira, None; S. Kishner, None; P. Zimmerman, None.

Appropriate Audience: Residents and practicing physicians.

Learning Objectives: After completing this educational activity, participants will be able to: (1) Formulate a differential diagnosis of a wrist drop; (2) Investigate and diagnose multiple pressure nerve palsies and peripheral neuropathy; (3) Use the physical exam and the principles of electrodiagnosis to evaluate conduction block, multifocal motor and hereditary neuropathies.

Level of Difficulty: Advanced.

History

A 25 year old male presented with a three month history of left wrist weakness. The patient stated that about three months ago, he was working as a car mechanic loosening a bolt in a car with a wrench, and apparently the wrench slipped. Since then the patient has been unable to extend his left wrist. The patient denied any numbness or tingling in the left arm.

- Formulate a differential diagnosis for acute wrist drop.
- List the possible areas and causes of radial nerve entrapment.

Commentary I

Patient presents with an acute left wrist drop. The differential diagnosis and common areas of entrapment are as follows;

1. Radial nerve palsy:
 - a. Spiral groove (Saturday night palsy)
 - b. Axillary
 - c. Arcade of Frohse (Posterior interosseous neuropathy)
2. C7 radiculopathy
3. Brachial Plexopathy (posterior cord involvement)
4. Central Nervous System process e.g. Stroke and Multiple Sclerosis.



The common sites of radial nerve compression include the spiral groove due to a humerus fracture or Saturday night palsy, the axillary due to crutches, in the arm at the arcade of Frohse. Superficial radial sensory nerve compression at the wrist is usually due to tight handcuffs, tight wrist ware or local wrist trauma.

History, continued

On review of systems, the patient denied any dizziness, headaches, vertigo, heat intolerance, nausea, skin irritation, and bowel or bladder dysfunction. The patient however reported that he occasionally wakes up at night with numbness and tingling in the first three digits of both hands. The pin and needles are relieved with vigorous hand shaking. The patient denied any weakness in the other extremities including gait abnormalities. The patient denied any drug allergies and was not taking any medications. The patient denied any known family history of neurologic disease. He also denied any history of alcohol, tobacco or drug abuse. The patient was referred to Physical Medicine and Rehabilitation for electrodiagnostic testing.

- What is your new differential diagnosis?
- What kind of investigations or consults would you consider in this case?

Commentary II

The patient's medical history rules out the possibility of a drug induced neuropathy by either alcohol or medication. The patient's family history decreases the chances of hereditary neuropathies but does not rule out spontaneous mutations. The patient also reports a classic "Flick Sign" that increases our suspicion for carpal tunnel syndrome. A young and healthy presenting with probable multiple nerve compressions creates a high suspicion of a generalized or multifocal neuropathy. In addition to electrodiagnostic testing, consider a MRI of the head and neck, blood work for systemic inflammatory markers, serum protein electrophoresis (SPEP), anti GM-1 antibody testing and genetic testing.

Physical Examination

The patient was alert, oriented, in no acute distress. Neurological exam showed no cranial nerve abnormalities. The patient had an absent left upper extremity triceps reflex. The biceps, brachioradialis, patella and ankle reflex were 2+ and symmetric. Manual motor muscle testing (MMT) showed decreased strength (0/5) with wrist and finger extension on the left. There was decreased sensation around the left posterior aspect of the radial hand. The patient had a positive Tinel's and Phalen's sign over the right median nerve at the wrist. He however had 5/5 strength with hip flexion, extension, knee flexion, extension, foot dorsiflexion and plantar flexion in bilateral lower extremities.

The patient had normal pulses, no cyanosis or edema in both extremities.

- In light of the physical findings, what are your diagnostic suspicions?
- What are the electrodiagnostic findings in conduction block?
- What are the common neurological disorders with evidence of conduction block on electrodiagnostic testing?



Commentary III

The physical exam widens our differential diagnosis, we now suspect probable carpal tunnel syndrome in addition to radial nerve palsy. The decreased sensation in the left upper extremity clinically decreases the possibility of a posterior interosseous neuropathy. Radial nerve compression at the spiral groove of the humerus, axillary, posterior cord brachial plexopathy and the possibility of a multifocal motor neuropathy are still high on our list. Conduction block is defined as a failure of an action potential to be conducted past a particular point in the nervous system, whereas conduction is possible below the point of the block. Mechanisms include demyelination, sodium channel blockade, depolarization and hyperpolarization. Findings include decreased amplitude, temporal dispersion and prolonged F-waves. Common disorders with conduction block are Chronic Inflammatory Demyelinating Polyneuropathy (CIDP), Guillian Barre Syndrome (GBS) and Multifocal Motor Neuropathy.

Electrophysiologic Data

NR = no response

SENSORY NERVE CONDUCTION STUDIES							
NERVE	SIDE	STIM SITE	RECORD	cm	AMPL	LAT	CV
Sural	R	Calf	Lateral malleolus	14	NR		
Ulnar	R	Wrist	Little finger	14	23	4.02	
Ulnar	L	Wrist	Little finger	14	21	4.08	
Median	R	Wrist	Long finger	14	10	5.51	
Median	L	Wrist	Long finger	14	NR	0	
Radial	L	Wrist	1st web space	14	NR	0	

MOTOR NERVE CONDUCTION STUDIES							
NERVE	SIDE	STIM SITE	RECORD	cm	AMPL	LAT	CV
Median	L	Wrist	Abductor Pollicis Brevis	8	15.1	5.54	
Median	L	Elbow	Abductor Pollicis Brevis	22	14.7	10.12	49.3
Ulnar	L	Wrist	Abductor Digiti Minimi	8	15.8	3.68	
Ulnar	L	Elbow	Abductor Digiti Minimi	24	14.2	7.64	60.6
Median	R	Wrist	Abductor Pollicis Brevis	8	2.5	5.62	
Median	R	Elbow	Abductor Pollicis Brevis	20	2.4	10.14	44.2
Ulnar	R	Wrist	Abductor Digiti Minimi	8	18.2	4.02	
Ulnar	R	Elbow	Abductor Digiti Minimi	22	16.5	8.14	53.4
Peroneal	R	Ankle	Extensor Digitorum Brevis	8	2.9	5.95	
Peroneal	R	Head of fibula	Extensor Digitorum Brevis	32	2.8	12.25	50.8
Peroneal	R	Popliteal	Extensor Digitorum Brevis	10	1.1	14.95	37.0



Radial	L	Forearm	Extensor Indicis Proprius	14	3.1	6.74	
Radial	L	Spiral groove	Extensor Indicis Proprius		NR	0	

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
L	Deltoid	Normal	0	0	N	Full	0	normal	N
L	Biceps Brachii	Normal	0	0	N	Full	0	normal	N
L	Triceps	Normal	0	0	N	Full	0	normal	N
L	Brachioradialis	Normal	3+	0	N	Full	0	normal	N
L	Extensor Indicis Proprius	Normal	1+	0	N	Dec	0	normal	N
L	1st Dorsal Interosseous	Normal	0	0	N	Full	0	normal	N
L	Abductor Pollicis Brevis	Normal	0	0	N	Full	0	normal	N
L	Paraspinal, Cervical	Normal	0	0	N	Full	0	normal	N
L	Pronator Teres	Normal	0	0	N	Full	0	normal	N
L	Pronator Quadratus	Normal	0	0	N	Full	0	normal	N

The patient declined further needle EMG testing in the lower extremity.

Diagnostic Impression

The electrodiagnostic exam showed multiple findings. Needle EMG showed positive sharp waves (PSW's) and fibrillations (Fibs) in the left brachioradialis and extensor indicis proprius. Nerve conduction studies in the upper extremities showed prolonged motor distal latencies in the median nerves bilaterally. The left median and right radial sensory responses were absent. The left radial nerve showed no motor response with stimulation at the spiral groove. The ulnar sensory latencies were mildly prolonged bilaterally while the right median sensory response was significantly prolonged. There was an absent right sural sensory response. The EMG and nerve conduction studies showed evidence of left radial neuropathy at the spiral groove with partial axon loss and evidence of continued conduction



block. There is a right median neuropathy at the wrist. There was also evidence of right peroneal nerve slowing across the knee with conduction block. Based upon the clinical exam and electrodiagnostic findings there is strong consideration of the diagnosis of Hereditary Neuropathy with liability to Pressure Palsies (HNPP).

The patient went back to see his referring physician, and no further follow up was obtained.

Commentary IV

Hereditary Neuropathy with liability to Pressure Palsies (HNPP) is characterized by repeated focal pressure neuropathies such as carpal tunnel syndrome and peroneal palsy with a foot drop. Recovery from the acute neuropathy is often complete. When recovery is not complete, the resulting disability is usually mild. Some affected persons may have mild to moderate polyneuropathy. The clinical diagnosis of HNPP is established in an adult with recurrent focal neuropathies and a positive family history consistent with autosomal dominant inheritance. The presence of mild underlying polyneuropathy, with or without symptoms, is further support for the diagnosis. HNPP is inherited in autosomal dominant manner.

Each child of an affected individual is at 50% risk of inheriting the mutation. Molecular genetic testing for a contiguous gene deletion of chromosome 17p11.2 that includes the PMP22 gene detects about 80% of patients. The remaining 20% of patients have a variety of point mutations in the PMP22 that may lead to frame shifts or other functional changes in the protein. PMP22 gene is the only gene known to be associated with HNPP. Genetic testing is available for both deletions and frame shift mutations. Prenatal testing is clinically available, although requests for prenatal testing for typically adult conditions such as HNPP are not common. 80% of individuals with HNPP have inherited the gene mutation from an affected parent, and 20% of patients have a de novo mutation.⁽⁴⁾ Occasionally neither parent shows signs of the disorder and the family history is negative because of mild and undiagnosed symptoms in a parent who has the mutated gene. Sural nerve biopsy often shows evidence of demyelination and "tomaculous" (focal sausage-like enlargement of the nerve) change.⁽⁷⁾ Tomaculous change is not specific and has been noted in other neuropathies.

HNPP prevalence is estimated to be 2-5 cases per 100,000 populations.⁽¹⁾ Actual prevalence may be higher due to under diagnosis, with a prevalence in Finland of 16/100, 000.⁽²⁾ Males and females are equally affected.

Physical exam often shows evidence of previous nerve palsy such as focal weakness, atrophy and sensory loss. Many patients (50-80%) have absent ankle reflexes and 15-30% have diffusely reduced tendon reflexes. 20% have a mild to moderate pes cavus foot.

Some individuals with HNPP over a period of decades develop symptoms and signs of a diffuse polyneuropathy.⁽³⁾ The proportion of persons who follow this progressive clinical course is unknown. HNPP is characterized by recurrent sensory or motor neuropathy in a single nerve beginning in adolescence or young adulthood. The most common presenting symptom of HNPP is the acute onset of a non painful focal sensory and motor neuropathy in a single nerve (mononeuropathy). Some persons experience transient sensory phenomena without weakness. History of actual physical compression of the nerve may or may not be present. The first attack is usually in the second or third decade with a broad range of first to seventh decades (mean: 37 years; range: 2-70 years). Radial nerve palsy has been reported in a two year old with HNPP. Occasional episodes have been reported during



pregnancy, perhaps related to physiologic changes such as soft tissue swelling and edema. The nerve palsies are often recurrent over a period of many years, but some persons have a single episode and some individuals who have a disease causing mutation are asymptomatic.

The most common sites of focal neuropathy include the peroneal nerve at the fibular head, the ulnar nerve at the elbow, the median nerve at the wrist causing carpal tunnel syndrome; the brachial plexus and radial nerves are also sometimes involved. Transient sensory symptoms and hand pain have been reported.

Full recovery over a period of days to months occurs in about one half of episodes. Incomplete recovery is fairly uncommon, but the remaining symptoms are rarely severe. Poor recovery correlates with a history of prolonged focal compression of the nerve

Electrophysiologic studies show evidence of delayed nerve conduction velocity at the site of compression. Prolongation of distal nerve conduction latencies (e.g. the median nerve at the wrist) occurs in essentially in all patients whether symptomatic or asymptomatic. General motor nerve conduction velocities are usually normal (greater than 40m/s), but a few patients have electrical evidence of a mild diffuse polyneuropathy.

According to Mouton et al (1999), the key electrophysiologic diagnostic features are bilateral slowing of sensory and motor nerve conduction at the carpal tunnel with conduction block in at least one peroneal nerve.⁽⁵⁾ Li et al (2002) found evidence of prolonged distal motor latencies in the median and peroneal, but not ulnar or tibial nerves.⁽⁶⁾

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