



## MSK Case No. 18, November 2008

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**Presenting Symptom(s):** Brachial neuritis

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**Disclosures:** Faculty members have no relevant financial relationships to disclose.

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**Appropriate Audience:** Residents and practicing physicians

**Learning Objectives:** After completing this educational activity, participant will be able to: (1) Evaluate and analyze a case of acute onset muscle weakness and form a differential diagnosis; (2) Develop skills to localize the point of injury to peripheral nerves in the upper extremities.

**Estimated Level of Difficulty:** Moderate

#### *History*

A 52 year old man presents with complaints of acute onset of right arm weakness and pain of 6 weeks duration. He was driving on a trip to Florida and noticed he had difficulty elevating in his arm. This weakness was associated with sharp pain in the shoulder over the deltoid muscle. In Florida the patient went to urgent care and received trigger point injections and had his shoulder placed in a sling. He received physical therapy for rotator cuff tendonitis and states his pain decreased but the weakness remained.

His complaints on presentation were right arm weakness, numbness in the 4<sup>th</sup> and 5<sup>th</sup> digits of his right hand and aching right shoulder region pain after exercise.

PMH: Cervical spondylosis, panic attacks, anxiety, increased cholesterol and asthma.

Medications: Ativan, Prozac, Advair, Darvocet and Zocor which was discontinued 6 weeks ago.

Social history: He is a financial consultant. He has a 30 year pack per day smoking history. His hobbies include horse grooming.

1. Prior to continuing, please develop a list of differential diagnosis in order of likelihood.
2. What additional information would you like to obtain?

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## **Commentary I**

Differential diagnosis at the time of presentation includes cervical radiculitis, rotator cuff tendonitis or tear, traumatic, infectious, neoplastic, psychosomatic disorder or adverse reaction to a drug.

His history of smoking puts him at risk for cancer, pancoast tumor must be considered. Suprascapular neuropathy which can occur spontaneously is in the differential. Less likely but possible are axillary neuropathy or long thoracic neuropathy.

Additional information which would be helpful includes history of any preceding events such as trauma or illness. Information on other areas of pain if they exist, any change since being off the Zocor, weight loss of any significance.

### ***History, Continued***

Two days prior to the onset of his symptoms the patient had a fever of 101.7 degrees Fahrenheit and diarrhea. The next day he states he had a burning sensation in his right thigh with no accompanying weakness. That same day he worked on a horse and had to pull hard on its mane to stabilize it. Four weeks after the onset of his symptoms the patient states he had a brief episode of decreased balance which resolved spontaneously.

### **Studies performed prior to his office visit include:**

MRI of the right shoulder with arthrogram which showed partial thickness tear of the supraspinatus tendon, infraspinatus tendonitis and AC joint hypertrophy causing impingement.

Liver enzymes were normal.

1. If necessary, revise your differential diagnosis.
2. What would the physical examination focus on?

## **Commentary II**

Based on the additional history and results, rotator cuff pathology can be moved up higher in the differential. A post infectious neuropathic process should also be considered. The physical examination will focus on the neuromuscular examination to try and determine which of the possible diagnosis are the most tenable, which can be excluded and what testing will help differentiate the possibilities.

### ***Physical Examination***

The patient is not in acute distress. He smells strongly of nicotine. He has marked atrophy of the right supraspinatus, infraspinatus and deltoid muscles; no scapular winging; and forward displacement of right shoulder. (See Figure 1)

**FIGURE 1**



ROM: Active right shoulder abduction is 25deg, forward flexion is 40deg

Manual Muscle Testing: Right shoulder external rotation = 3/5, shoulder abduction = 3-/5 and internal rotation = 5-/5

All other muscles in both upper extremities = 5/5.

Sensory: no deficit present. Reflexes: Biceps, Triceps and Brachioradialis = 2+ bilaterally

Hawkins sign is positive on the right.

Hoffman and Spurlings signs are negative bilaterally.

Lower extremity: Manual Muscle testing = 5/5 strength bilaterally,

Intact proprioception, mild decreased vibration sense on the right, Babinski is not present. Bilateral patellar and achilles reflexes are normal. Tandem gait is normal.

1. At this point, review your differential diagnosis and revise as appropriate.
2. What tests would you order?

### **Commentary III**

The patient's atrophy is confined to three muscles; the deltoid, supraspinatus and infraspinatus. These muscles are in the distribution of the Axillary and Suprascapular nerves. The nerve fibers originate in the C5, C6 nerve roots and are carried via the upper trunk of the brachial plexus. Other muscles innervated by the same nerve root as the above mentioned nerves are not involved.



At this point a cervical nerve root lesion is less likely and a Brachial plexus origin of the weakness more likely. The patient's smoking puts him at risk for a lung lesion such as a pancoast tumor which remains in the differential, although it will more frequently affect the lower trunk of the brachial plexus.

The most appropriate tests to order are: EMG, MRI of the C-spine and brachial plexus and chest x-ray.

**Lab Results 1**

- Chest x-ray revealed left upper lobe nodule.
  - MRI of the Cervical spine revealed mild multilevel cervical spondylosis most remarkable at C4-5 where there is moderate central canal stenosis.
  - MRI of the Brachial plexus revealed diffuse atrophy and abnormal T2 hyperintensity within the right supraspinatus, infraspinatus and deltoid muscles. This was suggestive of edema in those muscles which may be seen in the setting of denervation.
1. What further steps should be taken?

**Commentary IV**

At this point the overall likelihood of Cervical radiculopathy is decreased because there is apparently no involvement of the other muscles innervated by C5-6 nerve roots. The MRI of the cervical spine is not remarkable. The MRI of the brachial plexus is suggestive of denervation. An EMG and NCS is warranted for further investigation. The chest x-ray revealed a left lung lesion which needs to be further investigated. Note: the patient's weakness is on the right.

1. What tests would you perform at this point?

**Lab Results 2**

- CT-scan chest: No pulmonary nodules, changes compatible with early emphysema, minimal scars at the lung base.
- EMG and NCS

SENSORY NERVE CONDUCTION STUDIES							
All Studies are performed antidromically							
NERVE	SIDE	STIM SITE	RECORD	cm	AMPL	LAT	CV
Lat Ante Brach Cutan	Left	Lateral Biceps	Lateral forearm	12	9.5	1.4	
Lat Ante Brach Cutan	Right	Lateral Biceps	Lateral forearm	12	9.8	1.6	
Median	Left	Wrist Mid Palm	3 <sup>rd</sup> digit	14	3.5	5.2	26.9
			3 <sup>rd</sup> digit	7	4.3	2.5	
Median	Right	Wrist Mid Palm	3 <sup>rd</sup> digit	14	12.2	4.4	31.8
			3 <sup>rd</sup> digit	7	15.4	2.2	
Radial	Left	Wrist	Base 1 <sup>st</sup> digit	10	6.0	2.6	
Radial	Right	Wrist	Base 1 <sup>st</sup> digit	10	6.8	2.7	
Ulnar	Left	Wrist	5 <sup>th</sup> digit	14	11.7	8.3	16.9
Ulnar	Right	Wrist	5 <sup>th</sup> digit	14	NR	NR	



MOTOR NERVE CONDUCTION STUDIES							
NERVE	SIDE	STIM SITE	RECORD	cm	AMPL	LAT	CV
Axillary	Left	Erb's point	Deltoid		7.2	4.2	
Axillary	Right	Erb's point	Deltoid		5.4	16.6	
Median	Left	Wrist Elbow	Abd Poll Brevis	8 28.5	4.6 5.2	4.4 9.7	53.8
Median	Right	Wrist Elbow	Abd Poll Brevis	8 29	7.1 6.9	3.4 8.4	58.0
Musculocutaneous	Left	Erb's point	Biceps		8.2	5.1	
Musculocutaneous	Right	Erb's point	Biceps		8.4	5.1	
Suprascapular	Left	Erb's point	Supraspinatus		5.5	2.3	
Suprascapular	Right	Erb's point	Supraspinatus		1.0	2.6	
Ulnar	Left	Wrist Elbow	Abd Dig Minimi	8 26	6.3 5.9	2.8 7.6	54.2
Ulnar	Right	Wrist B Elbow A Elbow	Abd Dig Minimi	8 22.5 10	7.9 7.3 5.9	3.0 7.3 9.1	52.3 55.6

**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
Right	Biceps	N	0	0	N	N	N	N	N
Right	Deltoid	N	3+	Psw 4+	N	dec 3+	inc	inc	inc
Right	Supraspinatus	N	0	0	N	dec 3+	N	N	N
Right	Infraspinatus	N	0	Psw 2+	N	dec 3+	N	N	N
Left	Biceps	N	0	0	N	N	N	N	N



Left	<b>Deltoid</b>	N	0	0	N	N	N	N	N
Left	<b>Supraspinatus</b>	N	0	0	N	N	N	N	N
Left	<b>Infraspinatus</b>	N	0	0	N	N	N	N	N
Left	<b>Rhomboid</b>	N	0	0	N	N	N	N	N
Right	<b>Upper Cerv Paraspinals</b>	N	0	0	N	N	N	N	N
Right	<b>Mid Cerv Paraspinals</b>	N	0	0	N	N	N	N	N
Right	<b>Lower Cerv Paraspinals</b>	N	0	0	N	N	N	N	N
Left	<b>Upper Cerv Paraspinals</b>	N	0	0	N	N	N	N	N
Left	<b>Mid Cerv Paraspinals</b>	N	0	0	N	N	N	N	N
Left	<b>Lower Cerv Paraspinals</b>	N	0	0	N	N	N	N	N

**Commentary V**

The EMG/NCS findings revealed bilateral ulnar neuropathy based on absent sensory response (the right ulnar sensory study was most likely a volume conducted motor response) and bilateral Carpal tunnel syndrome are likely present. Corresponding motor abnormalities were not present.

This is not related to the proximal problem which would only affect the right median sensory amplitude whereas the left was more affected.

The abnormal motor units seen in the right deltoid, supra and infraspinatus muscles point to brachial plexus pathology.

1. What is the final diagnostic impression?

***Final Diagnostic Impression***

The patient has a right idiopathic brachial plexopathy (IBP). There were incidental abnormalities in median and ulnar sensory conduction studies which may indicate bilateral carpal tunnel syndrome and ulnar neuropathy.

1. What treatment would you initiate for the patient?

**Commentary VI**

The patient was placed on an intensive physical therapy program whose focus included:

- Passive and active ROM of the shoulder stabilizers.
- Strengthening the rotator cuff muscles as well as the deltoid. Preventing harmful compensatory biomechanics and functional electrical stimulation of atrophied muscles.
- Ability to attain independence in ADL'S.
- Ability to use the computer and drive a vehicle to prevent disability with regards to employment.

The patient was given a figure of 8 clavicle harness to prevent shoulder impingement due to the forward displacement of the humeral head secondary to his muscle atrophy.

The patient was educated on his diagnosis and realistic goals were set.

10 months from his initial presentation the patient had made the following recovery:

**1. Strength:**

- Right shoulder abduction improved from 3-/5 to 4/5.
- Right shoulder extension improved from 3-/5 to 5/5.
- Right shoulder external rotation improved from 3-/5 to 4-/5.
- Right shoulder flexion improved from 3-/5 to 4/5.
- Right shoulder internal rotation improved from -5/5 to 5/5

**3) Active ROM:**

- Right shoulder flexion improved from 50 deg to 165 deg.
- Right shoulder abduction improved from 30 deg to 175 deg
- Right shoulder extension improved from 35 deg to 60deg
- Right shoulder external rotation improved from 20 deg to 75 deg
- Right shoulder internal rotation improved from 60 deg to 80 deg

The patient continues to make improvement and never went on disability. He continues to be advised to stop cigarette smoking.

***Final Discussion***

The etiology of idiopathic brachial plexopathy (IBP, also known as neuralgic amyotrophy and Parsonage-Turner syndrome) is unknown. The classic presentation of acute onset of pain lasting 1 to 2 weeks with no clear trauma should make one suspect this diagnosis. Weakness is delayed in its onset, and the pain usually will abate spontaneously. The pain is severe, often awakening the patient from sleep. Early on, muscle weakness may be difficult to detect on physical exam because of the prominent pain. However as the pain subsides, significant underlying weakness becomes apparent and muscle atrophy follows. Although paresthesias and sensory loss may also be present, it is not unusual to find only mild or minimal sensory abnormalities on examination.

It has been postulated that the idiopathic version is due to an immune mediated inflammatory reaction.

The frequency in the USA is 1-2:100,000. 10 to 20% of affected patients have residual disability after 2 years. Male to female ratio is 2:1 to 4:1. The right upper extremity is more frequently involved than the left.

The idiopathic form has been linked to certain events and illnesses such as:

- (1) Viral, bacterial and parasitic infections.
- (2) Vaccinations e.g. tetanus, influenza and diphtheria.
- (3) Childbirth.
- (4) Trauma unrelated to the shoulder.
- (5) Medical investigative procedures e.g. administration of radiologic dye.
- (6) Systemic illness e.g. systemic lupus erythematosus (SLE), Ehlers Danlos.

There is an inherited form [Hereditary neuralgic amyotrophy (HNA)] which is autosomal dominant and due to mutations in SEPT9 gene which is on the chromosome 17q25. The inherited form affects males and females equally. It is characterized by recurrent attacks and is often bilateral. HNA is the first monogenetic disease caused by mutations in a gene of the septin family. The journal of Nature Genetics published an article in 2005 from the Department of Neurology, Munster, Germany. The article reported three mutations in the gene septin 9 in six families with HNA linked to chromosome 17q25. Septins are implicated in formation of the cytoskeleton, cell division and tumorigenesis.

One of the most striking features of brachial neuritis is the speed at which significant atrophy occurs and the muscles which are affected.



In a case report published in the American journal of radiology in 2007 it was stated that MRI of the brachial plexus is sensitive for detecting signal abnormalities in the muscles of the shoulder girdle and this is suggestive of denervation. The supraspinatus and infraspinatus muscles are the most commonly affected. The deltoid, serratus anterior, biceps and triceps are also commonly affected [listed in descending order of frequency]

Uncommonly cranial nerves can be affected; the phrenic nerve can become involved and lead to respiratory failure requiring ventilation.

Only 10-20% of people with IBP have some residual disability after two years.

Physical therapy to regain back as much muscle strength and range of motion is the main course of treatment. Bracing with a figure of eight clavicle harness prevents secondary shoulder impingement due to weakness of the shoulder stabilizers. Patient education is necessary to set realistic and hopeful expectations to treatment outcome.

### ***Bibliography***

1. Kuhlenbaumer G, Hannibal MC, Nelis E, et al “Mutations in SEPT9 causes hereditary neuralgic amyotrophy”. Nat Genet. 2005 Oct;37(10):1044-6.
2. Scalf RE, Wenger DE, Frick MA, et al “MRI findings of 26 patients with Parsonage-Turner syndrome”. AJR Am J Roentgenol. 2007 Jul;189(1):W39-44
3. Greiciane Gaburro Paneto, Iuri Drumond Louro, The Gale group;Inc 2005. Gale encyclopedia of Neurological disorders.
4. Nardone R, Bernhart H, Pozzera A, et al “Respiratory weakness in Neuralgic amyotrophy: report of two cases with phrenic nerve involvement”. Neurological sciences 2000 Vol 21, Number 3.
5. Joel A. DeLisa, Bruce M. Gans et al “Physical medicine and rehabilitation, principles and practice, 4th edition”.
6. David C. Preston and Barbara Shapiro “Electromyography and Neuromuscular disorders”.