Right Upper Extremity Pain and Weakness

Tung Henry Tang, DO and Bryan Tsao, MD

No one involved in the planning of this CME activity have any relevant financial relationships to disclose.
Authors/faculty have nothing to disclose.

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American Association of Neuromuscular and Electrodiagnostic Medicine

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CME Information
Product: CS23 - Right Upper Extremity Pain and Weakness

Course Description
Intended Audience
This course is intended for Neurologists, Physiatrists, and others who practice neuromuscular, musculoskeletal, and electrodiagnostic medicine with the intent to improve the quality of medical care to patients with muscle and nerve disorders.

Learning Objectives
Upon conclusion of this program, participants should be able to:
1. localize (electrodiagnostically) and differentiate an ulnar neuropathy at the forearm from an ulnar neuropathy at the wrist or elbow, a medial cord or lower trunk lesion, and a C8 radiculopathy.
2. identify the mix electrophysiological features of axon loss and demyelinating conduction blocks in a neuropathy of a compressive type.
3. display the principle of wallerian degeneration affecting earlier motor and later sensory fibers in a neuropathy of the compressive type.

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Right Upper Extremity Pain and Weakness

May 2008

CME Available from May 2008 through May 2011

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Presenting Symptom: Right upper extremity pain and weakness

Case prepared by: Tung Henry Tang, DO; Bryan Tsao, MD

Affiliations: Loma Linda University Medical Center

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Appropriate Audience: Residents, clinical neurophysiology or neuromuscular fellows, and practicing physicians.

Learning Objectives: After completing this educational activity, participants will be able to:
1) Localize (electrodiagnostically) and differentiate an ulnar neuropathy at the forearm from an ulnar neuropathy at the wrist or elbow, a medial cord or lower trunk lesion, and a C8 radiculopathy
2) Identify the mix electrophysiological features of axon loss and demyelinating conduction blocks in a neuropathy of a compressive type.
3) Display the principle of wallerian degeneration affecting earlier motor and later sensory fibers in a neuropathy of the compressive type.

Level of Difficulty: Intermediate.
Right Upper Extremity Pain and Weakness

History

This is a 72-year-old right-handed male chronic smoker presenting with right upper extremity weakness progressively worsening for the past 1 month. He has difficulty with grip strength on his right hand. This is made worse with activities such as writing or bowling. He is an avid bowler and notes that he has had to adjust from using his usual 12 lbs. bowling ball to an 8 lbs one. He complains of a deep throbbing and burning pain in the right elbow and forearm region and an intermittent tingling sensation involving in digits 4 and 5 about 1-2 weeks ago. He denies any neck pain. He is asymptomatic in the left upper or bilateral lower extremity.

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

Based on the history, a focal neurogenic process with both sensory and motor involvement in the right upper extremity should be considered as a generalized process is highly unlikely. Pertinent history involves weakness in the intrinsic hand muscles, tingling in digits 4 and 5 and pain in the forearm and elbow. This clinically may be seen in an ulnar neuropathy at various locations, a medial cord lesion, a lower trunk lesion or an intraspinal lesion involving the C8 nerve root segment.

In terms of etiologies:

1. Trauma causing an ulnar neuropathy at various sites should be considered. The ulnar nerve is superficial and quite prone to injury at the elbow and less commonly at the wrist that may cause both motor and sensory symptoms.

2. Cervical degenerative disc disease or a herniated disc are quite common especially in the elderly population and should be in the differential diagnosis. A C8 radiculopathy may result in pain, numbness in the digits 4 & 5 and weakness in intrinsic hand muscles.

3. Mass lesions such as a focal abscess or neoplasm should be considered. IV drug users are more prone to getting abscess. Primary soft tissue or bone cancers in the upper extremities may cause a compressive lesion to neighboring structures such as a focal ulnar neuropathy or brachial plexopathy to the lower trunk or medial cord. Metastatic lesions to the right upper extremity can result in a similar clinical presentation.

4. Peripheral vascular disease are commonly seen in chronic heavy smokers and often quite painful. Focal vasculopathies may infarct neighboring nerves causing motor and sensory deficits. As an example, Buerger’s disease causes a vaso-occlusive ischemia to the affected extremities and highly affiliated with long term tobacco use.

5. Metabolic disorders usually causes a symmetric process involving both sides. However, rare complications of focal neuropathies as a possible complication of diabetes and compressive neuropathies caused by a gouty tophus have been reported.

6. Autoimmune mediated multi-focal acquired demyelinating motor and sensory neuropathy is an extremely rare condition and though less likely, may be an entity to considered with negative imaging studies and nerve conduction studies showing features of conduction block in both motor and sensory responses.
7. Hereditary neuropathy of liability pressure palsy is characterized by recurrent mononeuropathies as a result of trivial or minor trauma at usual compression sites. Though unlikely in this case, may be a consideration if all other differentials have been ruled out and the focal neuropathy becomes recurrent at the usual compression sites.

The etiologies are listed by the order of their likelihood. Additional helpful questions would be to ask about the patient’s past medical history. In addition, a detailed social history including occupation, intravenous drug abuse and the quantity of cigarettes or tobacco use may be fruitful in narrowing down the differential diagnosis.

History, Continued

Further questioning reveals the patient as having a significant past medical history. He has a history of primary lung cancer s/p lobectomy, radiation and chemotherapy and is currently in remission for approximately 1.5 years. He also has prostate cancer and is undergoing medical management. He had an abdominal aortic aneurysm repair 4 years ago. For the social history, he is a retired electrical and plumbing contractor. He was a chronic smoker enjoying mainly cigarettes and cigars since his teenage years with an approximate 50+ pack year history. He denies any intravenous drug abuse.

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

With a history of both primary lung and prostate cancer, a metastatic neoplasm causing a compressive neuropathy or brachial plexopathy should be given high consideration. This should be followed by a vaso-occlusive ischemia in the upper extremity given his long term smoking history and previous abdominal aortic aneurysm. Traumatic and degenerative diseases should remain in the differential since they are common entities. Metabolic, autoimmune and hereditary etiologies are highly unlikely in his patient and extensive workups should be reserved.

Physical Examination

MSE: Awake and oriented x3. Speech fluent. Comprehension, naming and repetition are intact.
Motor: Weakness in digits 4 & 5 finger flexors, wrist flexion and finger spreading on the right side (4-/5). Extension of wrist and all digits are normal. Flexion of thumb is normal. Atrophy noted in first dorsal interosseus muscle and hypothenar eminence on the right. Normal tone. No Pronator drift.
Sensation: Decreased to light touch, pinprick and temperature sense in the right digits 4 & 5. No sensory deficits are noted in the arm or forearm region, though a deep boring pain is elicited to firm pressure in the proximal right forearm.
Reflexes: Symmetric in bilateral upper extremities and lower extremities.
Coordination: FNF and HS intact.
Gait: Narrow based, non-ataxic gait with normal stride and arm swing.

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 physical exam supplements the patient’s clinical symptoms involving both motor and sensory deficits. The exam is consistent with a right ulnar neuropathy. C8 radiculopathy or a lower trunk lesion is less likely as other non-ulnar innervated C8 and T1 muscles such as FPL, APB, EIP (only C8) tested are not clinically affected. Medial cord lesion may mimic an ulnar neuropathy, but would expect to have some deficits in the distal C8 and T1 non-ulnar innervated muscles (APB and FPL) but sparing the radial innervated C8 muscles. In addition, sensory deficits would be expected if the medial antebrachial cutaneous nerve off of the medial cord is affected.

If necessary, revise your differential diagnosis based on the additional physical findings. Design your approach to the electrophysiologic examination based on the existing data.

Electrophysiologic Data

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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.)
6 weeks after first EDX: Sensory

| Ulnar | Right | Wrist | Fifth | 13 | NR |

Make the final revisions of your diagnostic impressions.

Diagnostic Impression

Right ulnar neuropathy in the forearm with features of initial demyelination as evidenced by conduction block then later axon loss involving both motor and sensory fibers, severe in degree electrically.

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

Commentary V

This is an unusual presentation of an ulnar neuropathy at the forearm from a large compressive mass lesion. MRI with contrast reveals a 4.1 x 4.4 x 7.0 cm subcutaneous heterogenous mass located in the right forearm between flexor carpi ulnaris and flexor digitorum superficialis, superficial to flexor digitorum profundus intimately related to the ulnar artery and nerve without bony involvement. Core biopsy confirmed the mass to be a metastatic undifferentiated large cell carcinoma from the lungs.

The patient presents with right upper extremity weakness for 2 months and sensory symptoms including burning pain and tingling in digits 4 & 5 at 1 month prior to the initial electrodiagnostic study. NCS of the ulnar motor responses recording from both the hypothenar eminence and first dorsal interosseus shows evidence of acquired demyelination with conduction block between the wrist and elbow stimulation sites. There is also evidence of axon loss as the ulnar CMAPs of both hypothenar and first dorsal interosseous muscles are low when stimulating at the wrist. However, ulnar sensory response at digit 5 and the dorsal ulnar cutaneous response on the right are within the normal range. Needle exam shows severe recruitment with minimal fibrillations solely in selected ulnar innervated muscles. We suspect the pathophysiology may be consistent with a mixed demyelination (conduction block) and early axon loss (preserve sensory with motor responses effected) at this point. Limited repeat NCS of the ulnar sensory response 6 weeks later shows no response due to tolerance. If full repeat of NCS/EMG, we would expect to find more decrement or no response of CMAP recording from the hypothenar and first dorsal interosseus muscles. More dense fibrillation potentials and abnormal neurogenic motor unit potentials in ulnar innervated muscles would be expected.

Metastasis of large cell carcinoma of the lung is a rare entity and accounts for approximately 10% of all lung non-small cell carcinomas. Common metastatic sites include supraclavicular lymph nodes, brain, bone, adrenal gland and liver. Based on an extensive literature search, this is the only reported case of a large cell carcinoma of the lung metastasizing to soft tissue presenting as an ulnar neuropathy at the forearm. The median survival is reported to range from 28-41 weeks from the time of diagnosis despite aggressive therapies with external beam radiation and chemotherapy. Only 10-15% survive past 1 year.

Unfortunately, our patient was referred by an outside community facility and lost to follow-up in our neurology clinic.

In summary, the patient has a right ulnar neuropathy at the elbow from an extrinsic metastatic compressive biopsy proven undifferentiated large cell carcinoma of the lungs. Electrodiagnostically, there
are features of initial acquired demyelination followed by axon loss. We likely have performed the NCS/EMG during the sub-acute phase of the wallerian degeneration (within the first weeks) as the ulnar motor responses are affected while the sensory responses are preserved. The follow-up NCS 6 weeks later revealing the absence of the ulnar sensory response at digit 5 confirmed the completion of the wallerian degeneration.

Bibliography