



January 1997 EMG Case-of-the-Month

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HISTORY

The patient is a 54-year-old man who experienced sudden onset of a severe headache, nausea, vomiting, and mental confusion six weeks ago. A head CT revealed a diffuse subarachnoid hemorrhage in the area of the circle of Willis. An angiogram showed an intercommunicating artery aneurysm with vasospasm. A ventral craniotomy was performed with anterior communicating artery ligation. The patient did well postoperatively, however, was noted to have absent sensation in the right hand with markedly diminished sensation in the left hand. He had no function of the right hand with only a minor degree of left hand function. MRI of the cervical region revealed no abnormalities. An electrodiagnostic evaluation was requested six weeks following operative intervention to help assess the cause of the patient's bilateral hand dysfunction.

- **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 fully cooperative with the examination. Visual inspection of the hands reveals marked flattening of the palmar arch as well as gross wasting of all hand intrinsic and forearm flexor muscles bilaterally. Manual muscle testing demonstrates shoulder flexion, extension, abduction, and internal/external rotation to be 5/5 on the left and right. Elbow flexion/extension and wrist extension are also 5/5 bilaterally. Finger extensors appear intact but cannot fully extend the interphalangeal joints without metacarpophalangeal joint stabilization in neutral by the examiner. Wrist/finger flexors (superficial & deep) on the right are 0/5 and 2/5 on the left. All hand intrinsic muscles are 0/5 on the right and 2/5 on the left. On the right, all sensory modalities are absent on the entire volar aspect of the hand and dorsal region of the hand medially as well as the medial forearm and arm. Sensation in the remainder of the right upper limb is preserved. On the left, similar abnormal findings are obtained except there is a marked diminution in sensation as opposed to complete absence for homologous areas. Again, sensation in the remainder of the left upper limb is normal. Muscle stretch reflexes of the biceps brachii, brachioradialis, and triceps are normal and symmetric bilaterally. The pronator teres and finger flexor reflexes are absent on the left and right. No abnormal findings were noted in the lower limbs.



- **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

| 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 p wave = positive sharp waves fibrillation = fibrillation potentials recr = recruitment amp = amplitude dur = duration poly = polyphasic potentials | | | | | | | | | |
| R/L | MUSCLE | INSERTION SPONTAN | | | VOLUNTARY | | | | |
| | | activ | p wave | fib | recrt | amp | dur | poly | effort |
| L/R | deltoid | N | 0 | 0 | N | N | N | N | - |
| L/R | biceps brachii | N | 0 | 0 | N | N | N | N | - |
| L/R | triceps | N | 0 | 0 | N | N | N | N | - |
| L/R | extensor carpi radialis | N | 0 | 0 | N | N | N | N | - |
| L/R | extensor digitorum communis | N | 0 | 0 | N | N | N | N | - |
| L | pronator teres | incr | 4+ | 4+ | 0 | N | N | N | - |
| R | pronator teres | incr | 3+ | 3+ | 1 motor units | N | N | N | - |
| L | flexor carpi radialis | incr | 3+ | 3+ | 2 motor units | N | N | N | - |
| R | flexor carpi radialis | incr | 3+ | 3+ | 1 motor units | N | N | N | - |
| L | flexor digitorum superficialis | incr | 3+ | 3+ | 1 motor units | N | N | N | - |
| R | flexor digitorum superficialis | incr | 3+ | 3+ | 0 | N | N | N | - |
| L | flexor carpi ulnaris | incr | 3+ | 3+ | 0 | N | N | N | - |
| R | flexor carpi ulnaris | incr | 3+ | 3+ | 0 | N | N | N | - |
| L | abductor pollicis brevis | incr | 4+ | 4+ | 2 motor units | N | N | N | - |
| R | abductor pollicis brevis | incr | 4+ | 4+ | 1 motor units | N | N | N | - |
| L | first dorsal interosseous | incr | 4+ | 4+ | 2-3 motor units | N | N | N | - |



| | | | | | | | | | |
|-----|---------------------------|------|----|----|-----------------|---|---|---|---|
| R | first dorsal interosseous | incr | 4+ | 4+ | 1 motor units | N | N | N | - |
| L | abductor digiti minimi | incr | 4+ | 4+ | 2-3 motor units | N | N | N | - |
| R | abductor digiti minimi | incr | 4+ | 4+ | 1 motor units | N | N | N | - |
| L/R | C5-T2 paraspinals | N | 0 | 0 | N | N | N | N | - |

| SENSORY NERVE CONDUCTION | | | | | | | | | |
|--|--------------|-----|------|----------------|----|------|------------------|---|------|
| nr = no response | | | | | | | | | |
| NERVE | LATENCY (ms) | | | AMPLITUDE (µV) | | | CONDOC VEL (m/s) | | |
| | R | L | Norm | R | L | Norm | R | L | Norm |
| sural | - | 3.8 | - | - | 29 | - | - | - | - |
| median | nr | nr | - | - | - | - | - | - | - |
| ulnar | nr | nr | - | - | - | - | - | - | - |
| radial | 2.9 | 3.1 | - | 28 | 25 | - | - | - | - |
| lateral antebrachial cutaneous | 2.9 | 2.9 | - | 9 | 10 | - | - | - | - |
| medial antebrachial cutaneous cutaneousc | nr | nr | - | - | - | - | - | - | - |

| MOTOR NERVE CONDUCTION | | | | | | | | | |
|------------------------|--------------|-----|------|----------------|------|------|------------------|----|------|
| nr = no response | | | | | | | | | |
| NERVE | LATENCY (ms) | | | AMPLITUDE (mV) | | | CONDOC VEL (m/s) | | |
| | R | L | Norm | R | L | Norm | R | L | Norm |
| peroneal | - | 4.9 | - | - | 3000 | - | - | 51 | - |
| median | nr | nr | - | - | - | - | - | - | - |
| ulnar | 3.9 | 3.3 | - | 400 | 900 | - | 47 | 48 | - |

- 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

Profound Wallerian degeneration of the median and ulnar nerves bilaterally most likely from the axillary or proximal arm region. There are a few remaining axons intact to most but not all of the muscles innervated by these two nerves. Prognosis for complete functional return of hand intrinsic muscles is poor secondary to the severity of denervation combined with length of neural regrowth required.

DIFFERENTIAL DIAGNOSIS

Distal upper limb weakness and sensory loss with preservation of proximal limb musculature and sensation occurring acutely should generate a limited differential diagnosis. The thought process should involve consideration of central versus peripheral causes. It is improbable that cerebral ischemia would result in such symmetric findings. One may consider a central cord syndrome but the clinical findings of upper limb weakness present in this patient would be very atypical. A syrinx should be considered, but the lack of pain/temperature versus vibration/proprioception dissociation, combined with normal lower limb findings (strength, tone, reflexes) and the absence of pain in the cervical region speaks against a syrinx. All of the above possibilities occur proximal to the dorsal root ganglia, which would result in preservation of the sensory nerve action potentials (SNAP). Since the median and ulnar SNAPs were absent in this patient, one must consider a lesion distal to the dorsal root ganglia to account for the patient's signs and symptoms.

The physical examination strongly suggests that the muscles innervated by the median and ulnar nerves are preferentially, if not exclusively, affected. The pronator teres as well as the abductor pollicis brevis are abnormal, suggesting dysfunction of motor fibers from C6 through T1. Preservation of wrist/finger extension and forearm supination lead one to conclude that the lesion is distal to both the root and brachial plexus locations. If the brachial plexus were involved, then elbow extension and wrist/finger dorsiflexion should be weak, i.e. those functions arise from the same root levels as the pronator teres and flexor carpi radialis muscles. Absent median/ulnar C8/T1 muscle function with complete preservation of shoulder internal rotation (pectoralis major) supports a lesion distal to the brachial plexus. The physical examination strongly suggests that the patient has a bilateral axillary or proximal arm compromise of the median and ulnar nerves where they are in close proximity. This is supported by the wife's observation of significant discoloration in the patient's axillary region, suggesting considerable trauma/hematoma formation with likely axon injury to the median and ulnar nerves.

FORMULATION OF ELECTROPHYSIOLOGIC STUDIES

The history and physical examination strongly suggest a profound axonal insult to the median and ulnar nerves in the axilla or proximal arm region. The suspected location implies that all muscles innervated by the median and ulnar nerves should demonstrate membrane instability on needle examination while those muscles with similar nerve root levels but innervated by the radial, musculocutaneous, and axillary nerves should be spared. Additionally, nerve conduction studies are anticipated to reveal low amplitude compound muscle action potentials and low amplitude or absent sensory nerve action



potentials for the median and ulnar nerves. Again, the remaining nerves should have normal CMAPs and SNAPs.

It is necessary to perform a detailed needle electromyographic examination of the upper limbs bilaterally to define the extent of axon loss, and if possible confirm the presumptive localization. Muscles must be examined that are clinically weak as well as some that are normal. The possibility exists that those muscles that appear unaffected on manual muscle testing may still show some evidence of axon loss (membrane instability), thus modifying our initial clinical impression of focal lesions. Specifically, a brachial plexus insult may have occurred with a preferential but not exclusive insult to the lower trunks or medial cords. We must also perform a careful examination of the cervical paraspinal region to ensure that injury at or proximal (anterior horn cells: e.g. syrinx) to the root level is not present. Motor and sensory nerve conduction studies are also important. The median SNAP is of particular importance since it is composed of fibers originating from the C6/C7 level in contradistinction to its motor supply of C6/C7/C8/T1. If the median SNAP is absent in association with membrane instability in the median innervated hand intrinsic muscles, a lesion at or distal to the formation of the median nerve is suggested, i.e. not in the brachial plexus. Similarly, sparing of the superficial radial SNAP with absence of the median SNAP again suggests that the lesion is likely located in a peripheral nerve as opposed to the plexus level. Compound muscle action potential assessment provides us some insight into the degree of axon loss. If there are absent or small CMAPs, then profound axon loss has occurred. However, if the CMAPs are relatively preserved despite a significant reduction in motor units, the presence of conduction block should be considered. These findings have important prognostic implications.

ASSESSMENT OF ELECTROPHYSIOLOGIC FINDINGS

There is absence of SNAPs from the median and ulnar nerves as recorded from the digits, as well as absent medial antebrachial cutaneous SNAPs. Normal and symmetric SNAPs, however, are detected for the left and right lateral antebrachial cutaneous and radial nerves. These findings clearly support the supposition of a combined medial/ulnar lesion. Additionally, the absence of the medial antebrachial cutaneous SNAP suggests a proximal lesion location. These findings place the lesion postganglionic and out of the brachial plexus. As noted above, an absent median nerve but present superficial radial nerve SNAP suggests the lesion is distal to the formation of the radial and median nerves, i.e. within peripheral nerves and not brachial plexus.

Needle electromyographic findings of fibrillation potentials and markedly reduced recruitment provide the most conclusive information for localizing the lesion. Denervation isolated to the median and ulnar supplied muscles, (C6-T1) without concomitant abnormalities in the axillary, musculocutaneous, and radial nerve distributions, localized the lesion. The location inferred by the electrical abnormalities is either in the distal axilla or proximal arm, where the median and ulnar nerves are in close proximity. Thus, the motor and sensory nerve conduction studies along with the needle electromyographic findings all point to a combined median/ulnar nerve injury in the proximal arm or axilla. There is an associated insult to the medial antebrachial cutaneous nerve.

The profound reduction in voluntary motor units present in the muscles supplied by the median and ulnar nerves may be a result of significant Wallerian degeneration, severe



conduction block, or some combination of axon loss and conduction block. Profuse numbers of positive sharp waves and fibrillation potentials in association with markedly reduced numbers of voluntary motor units six weeks following symptom onset suggest that there is little conduction block, with primarily axon loss producing both the clinical and electrophysiologic findings. This is certainly the case for the median nerve as there are no obtainable CMAPs or SNAPs. However, the ulnar CMAPs from the hand intrinsic muscles are certainly larger than would be anticipated if only one or two motor units were present. It is likely that some residual conduction block is present, thereby accounting for the moderate discrepancy between only one or two motor units and a CMAP that is about 500 uV and 1000 uV for the right and left sides respectively.

Such massive Wallerian degeneration in the median nerve, with absence of the SNAP and CMAP, portends a poor prognosis for functional recovery. If the lesion is in the axillary region as suggested by the clinical and electrophysiologic data, then the median nerve must regrow approximately 36 inches, connoting yet worse prognosis for the median supplied hand intrinsic muscles. If nerve growth is about one inch a month, then it may take three years for the nerve to reach the thenar muscles. Because the lesion is likely a result of compression, thus not disrupting the nerve's connective tissue elements, a "lesion in continuity" is present; the endoneurial tubes are intact and aligned. Hence, the axons are assured an accurate path back to their original muscles. On the other hand, the long distance and time prior to reconnection between nerve and muscle means that the distal endoneurial tube may become stenotic secondary to natural shrinkage of an empty axon tube. This could impede axon regrowth several years down the road when the advancing nerve front finally reaches the nerve's termination at the muscle. Also, the muscles will atrophy considerably and may undergo significant fibrosis by the time the nerve arrives. There may be little muscle left for the nerve to supply. An alternative method of reinnervation is through the process of collateral sprouting. Fortunately, a single remaining axon may have the capability of forming about five times the original number of terminal axons. In this case, though, there are very few motor units detected six weeks following the neural insult. Because the negative factors are likely to outweigh the positive, the patient's prognosis is most likely poor for those muscles innervated by the median nerve, with a progressively worse prognosis the more distal the muscle.

Similar prognostic statements apply to the ulnar nerve with the exception that small but present CMAPs were recorded from the abductor digiti minimi bilaterally. This suggests that, despite the detection of only one or two motor units, some degree of conduction block is present in the ulnar nerve. If this is the case, and assuming the block will resolve, the patient has a slightly better prognosis for recovery than for the median nerve. The factors of muscle atrophy, long distance, and endoneurial tube shrinkage continue to apply and may moderate the effects of conduction block. It could be concluded that the patient may eventually have slightly better function of ulnar compared to median innervated hand intrinsic muscles. The overall prognosis for a return to premorbid function is very poor and the patient will most likely not recover significant functional skills in the area of fine motor dexterity.

It is not possible to state with assurance that the vest restraint was the sole factor producing the axillary/proximal arm lesions observed. There is considerable suspicion for this, though, given: highly symmetric lesions, hematoma formation in the axilla and proximal arm as observed by the wife, insult localized to the median and ulnar nerves where they are in close proximity in the axilla/proximal arm, combative and confused patient



requiring vest restraint. The author tried on the vest restraint and had a colleague pull the vest with resistance. A very uncomfortable sensation in the axilla/proximal arm was experienced as the vest acted as a sharp edge, clearly compressing the neurovascular structures in the axilla.

ADDITIONAL HISTORY

Contact was made with the surgeon who performed the operation. The position required for the operative intervention did not place the patient's cervical spine, brachial plexus, or ulnar nerves at risk for compression. The patient's wife was present in the examination room. On questioning, she confirmed that the patient tolerated the surgery well, but was very combative for several days following surgery requiring, vest restraints in bed. When the patient's combative stage resolved (about 4-5 days), the wife noted that her husband no longer had functional use of his hands despite his ability to follow commands quite well. About a week later the wife began to assist the nursing staff in caring for her husband in preparation to take him home, and noted purplish discolorations in his axillary region bilaterally, which extended down his arms and anterolateral thoracic regions. These findings had resolved at the time of the present examination.

FORMULATION OF AN IMPRESSION

The lesion location and degree of severity are supported by the clinical and electrophysiologic findings. There is little doubt that the median and ulnar as well as the medial antebrachial cutaneous nerves were injured bilaterally in the axilla or proximal arm. The degree of injury was severe as evidenced by the absence of SNAPs and absent or significantly reduced CMAPs. Florid membrane instability in association with only a few voluntary motor units are commensurate with the nerve conduction studies. The degree of axon loss combined with the proximal lesion location all suggest a very poor functional outcome for the median and ulnar innervated hand intrinsic muscles. The final outcome will depend upon the patient's nervous system to regrow across the lesion site and reach the various muscles prior to their irreparable atrophy or fibrosis.

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