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Clinical Reasoning: A 27-year-old man with hand numbness

Exploring new horizons and reinventing the past

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SECTION 1

A 27-year-old crane operator presented with left hand numbness of 3 months' duration. He reported pain above the left elbow following a trivial trauma prior to symptom onset. There was no involvement of other extremities. On examination, there was no wasting of the hand intrinsic muscles but mild weakness of the left abductor digiti quinti with normal power of long hand flexors. Sensation was impaired over the dorsal and volar medial one and half fingers and palm. Deep tendon reflexes were 2+ with normal neurologic examination of the other extremities. There was tenderness and fullness over the left medial elbow. Systemic examination was unremarkable.

Questions for consideration:

1. What differential diagnoses would you consider?
2. What investigations would you suggest to confirm the diagnosis?

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Table Electrodiagnostic studies of the upper extremities

	Right	Left
Hand temperature	30.2	31.4
Median nerve studies		
Sensory nerve conduction		
Median (digit 3) orthodromic		
Sensory peak latency, ms	3.10	3.15
Sensory nerve action potential, μV	21.8	18.1
Conduction velocity, m/s	57.9	51
Motor nerve conduction		
Median abductor digiti minimi		
Distal latency, ms	3.20	2.95
Motor amplitude (wrist), mV	7.3	10.4
Proximal motor latency, ms	7.30	6.10
Motor amplitude (elbow), mV	7.3	10.7
Conduction velocity, m/s	56.8	69.8
F-wave latency, ms	24.30	23.65
Ulnar nerve studies		
Sensory nerve conduction		
Ulnar (digit 5) orthodromic		
Sensory peak latency, ms	2.50	2.80
Sensory nerve action potential, μV	11.5	7.4
Conduction velocity, m/s	53.7	53.7
Dorsal ulnar cutaneous nerve		
Sensory peak latency, ms	1.90	—
Sensory amplitude, μV	4.1	Absent
Conduction velocity, m/s	40	—
Motor nerve conduction		
Ulnar abductor digiti quinti		
Distal motor latency, ms	2.10	2.8
Motor amplitude (wrist), mV	9.30	6.3
Motor latency (below elbow), ms	5.55	7.10
Motor amplitude (below elbow), mV	8.4	6.6
Conduction velocity (below elbow-wrist), m/s	63.8	46.8
Motor latency (above elbow), ms	7.15	10.3
Motor amplitude (above elbow), mV	8.5	3.0
Conduction velocity (across elbow), m/s	62.5	40.0
F-wave latency, ms	27.70	31.80
Ulnar first dorsal interossei		
Distal motor latency, ms	3.45	2.65
Motor amplitude (wrist), mV	7.2	7.3
Motor latency (below elbow), ms	7.25	7.40
Motor amplitude (below elbow), mV	6.2	7.3
Conduction velocity (below elbow-wrist), m/s	52.6	40.0
Motor latency (above elbow), ms	9.10	10.50
Motor amplitude (above elbow), mV	5.9	4.1

Continued

SECTION 2

The clinical scenario presented is compatible with a left-sided ulnar neuropathy. Other differential diagnoses that need to be considered include involvement of the medial cord or lower trunk of the brachial plexus and a C8-T1 radiculopathy. The clinical sign that confirms the clinical impression of an ulnar neuropathy is sensory loss confined to the dermatomal distribution of the ulnar nerve. The left elbow pain suggests the site of ulnar nerve pathology.

Further evaluation would help narrow the etiologic diagnosis. An elbow joint pathology with compression of the nerve as a result of arthritis, synovitis, osteophytes, or loose articular bodies is common. Other common causes of an ulnar neuropathy at the elbow include cubital tunnel syndrome or compression of the nerve in the retrocondylar groove. Less common causes are nerve compression in the retrocondylar groove as a result of past trauma, ganglia, lipoma, a primary nerve tumor, or presence of a variant anconeous epitrochlearis muscle. Rarely, entrapment of the ulnar nerve in the arm can occur beneath and proximal to the ligament of Struthers. Systemic diseases associated with ulnar neuropathy include acromegaly and leprosy.

The initial investigations should include electrodiagnostic studies and an x-ray of the elbow. X-ray of the left elbow showed no deformity or joint effusion. Electrodiagnostic studies are important for confirming the diagnosis of ulnar neuropathy and help distinguish it from a medial cord or lower trunk brachial plexopathy and a C8-T1 radiculopathy. Furthermore, they assist in localizing the lesion in case of a mononeuropathy and in differentiating axonal from demyelinating pathology. The table shows the nerve conduction study report.

Question for consideration:

1. How would you interpret the electrodiagnostic studies?

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Table Continued

	Right	Left
Conduction velocity (across elbow), m/s	63.2	41.9
F-wave latency, ms		
Short segment incremental studies		
6 cm below elbow, ms		6.60
4 cm below elbow		7.15
2 cm below elbow		7.35
At elbow		7.50
2 cm above elbow		7.70
4 cm above elbow		9.90
6 cm above elbow		10.55
Radial nerve studies		
Radial cutaneous sensory nerve		
Sensory peak latency, ms	1.85	2.00
Sensory amplitude, μV	47.4	49.3
Sensory conduction velocity, m/s	55.2	58.1

SECTION 3

Electrodiagnostic studies demonstrated a left-sided ulnar mononeuropathy. Normal medial antebrachial cutaneous potentials make a medial cord or lower trunk brachial plexopathy less likely. Sensory potentials are preserved in vertebral foraminal compression of sensory nerve roots as the lesions are preganglionic. The absent dorsal ulnar cutaneous nerve potential and the presence of normal median compound muscle action potential make the diagnosis of left-sided C8-T1 radiculopathies unlikely.

A comprehensive electrodiagnostic study of the ulnar nerve should include ulnar motor studies with recordings from the abductor digiti quinti and first dorsal interossei and stimulating at the wrist, below and above elbow, axilla, and supraclavicularly.[1] Ulnar sensory studies include recording or stimulating digit 5, digit 4, and the dorsum of the medial hand. Further studies include mixed nerve stimulation at the wrist and recording from below and above the elbow and comparison of conduction velocity between the wrist-to-below-elbow segment and the across-elbow segment.

Further techniques to localize the lesion at the elbow segment include short segment incremental stimulation (inching) studies and needle EMG. These techniques can reveal an abnormality even when routine ulnar nerve studies are normal.[2–4] Inching studies are performed across the elbow by stimulating the ulnar nerve at 2-cm increments starting 6 cm below the elbow to 6 cm above and looking for abrupt change in latency or drop in amplitude between adjacent segments.

Short segment incremental stimulation studies can localize the compression to any of the following sites: proximal to Struthers ligament, the retrocondylar groove, proximal to the humeroulnar arcade, or as the ulnar nerve exits from underneath the flexor carpi ulnaris (FCU). However, the effectiveness of this technique is limited with subluxation of the ulnar nerve, which would make the points of stimulation along the ulnar nerve inaccurate.[5,6] Ulnar subluxation is common, occurring in 10%–20% of the population.

Needle EMG studies mainly help in excluding a lower brachial plexopathy or a C8-T1 radiculopathy. Their main value in localization of ulnar nerve lesions is in differentiating proximal from distal lesions. Proximal lesions are associated with denervation potentials from the FCU and flexor digitorum profundus. The variable exit of the motor branch to the FCU above or below the elbow, selective fascicular sparing (the first dorsal interossei branch is most commonly affected), and mild compression of the ulnar nerve can, however, decrease the sensitivity of EMG studies in localizing the site of compression of the ulnar nerve at the elbow.

Our patient demonstrated unequivocal evidence of a conduction block with more than 50% drop in amplitude when stimulating the ulnar nerve segment from below and above the elbow and recording from the abductor digiti quinti. In addition, there is segmental conduction slowing across the elbow. This is compatible with an ulnar neuropathy at the elbow of demyelinating type. The absence of response from the left dorsal ulnar cutaneous nerve and the slowing in the motor conduction velocity of the wrist to elbow ulnar nerve segment when recording from the first dorsal interossei suggest mixed demyelinating and axonal involvement.

Short segment incremental studies for further localization, however, indicated a more proximal lesion in the above elbow segment. The latency difference and drop in amplitude were greatest between sites 2 cm and 4 cm above the elbow, suggesting localized nerve pathology in that location. Needle EMG was not performed as localization was achieved through nerve conduction studies.

Question for consideration:

1. What investigations would further characterize the ulnar neuropathy at the elbow?

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SECTION 4

Peripheral nerve ultrasound is an important adjunct investigative modality in peripheral nerve disorders, especially in entrapment neuropathies. Its role in detecting and confirming ulnar neuropathies at the elbow has been established.[7,8] The diagnostic yield is highest when electrodiagnostic studies show signs of ulnar neuropathy without being able to localize. Studies have also shown its value in nerve lesions outside the elbow.[9]

The differential diagnoses of neuropathies with nerve enlargement include Charcot-Marie-Tooth disease, hereditary neuropathy with liability to pressure palsies (HNPP), chronic inflammatory demyelinating polyneuropathy, leprous neuropathy, amyloid neuropathy, neurofibromatosis, and primary nerve tumors.

Further characterizations include the following:

1. Localizing the site, length, and pattern of enlargement
2. Differentiating a focal neural enlargement involving one nerve vs a generalized disease process involving multiple nerves
3. Demonstrating preservation or loss of fascicular architecture

Nerve enlargement with preservation of fascicular architecture is seen in Charcot-Marie-Tooth disease and acromegaly. There is diffuse enlargement of the nerves not restricted to entrapment sites. This is in contrast to HNPP, where the nerve enlargement tends to be pronounced at usual sites of entrapment. The pattern and length of enlargement can be helpful, with focal nodular enlargement being commonly associated with neurofibromatosis as opposed to diffuse fusiform swelling seen in leprosy. Entrapment neuropathies result in focal nerve enlargement with loss of fascicular architecture at the site of entrapment.

Our patient demonstrated fusiform swelling of the ulnar nerve at the elbow, which extended proximally up to the midarm with alteration of fascicular architecture and nerve echogenicity (figure; video on the *Neurology*® Web site at Neurology.org). The latter 2 features suggest nerve edema as a result of an inflammatory process. In addition, there was enlargement of asymptomatic nerves of both the upper extremities, including the right ulnar nerve at the elbow, the right dorsal ulnar cutaneous nerve, and both superficial radial sensory nerves. The presence of nerve tenderness, enlargement of asymptomatic nerves, and preferential involvement of the superficial cutaneous nerves makes the diagnosis of pure neuritic leprosy highly probable.

Question for consideration:

1. What would your next line of management be?

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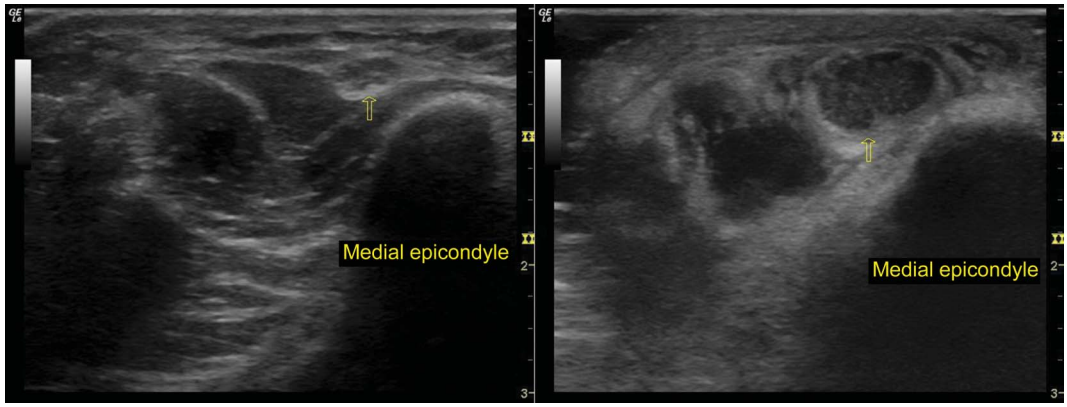
Figure Sonographic images of ulnar nerve at the sulcus ulnaris



Normal-sized ulnar nerve, with a cross-sectional area of 0.09 cm^2, at the sulcus ulnaris on the left; and a grossly enlarged nerve, cross-sectional area of 0.43 cm^2, on the right.

SECTION 5

Left dorsal ulnar cutaneous nerve biopsy revealed solid nests and sheets of foamy, vacuolated cells and histiocytes with accompanying chronic inflammatory infiltrate. There were numerous acid-fast bacilli on the FITE stain confirming the diagnosis of leprosy. Leprosy can be diagnosed based on the triad of enlarged nerves, localized patches of skin anesthesia, and positive acid-fast bacilli on tissue samples. In the absence of typical skin patches, as in our patient, leprosy is diagnosed based on enlarged nerves and demonstration of acid-fast bacilli in nerves or skin. Our patient was started on rifampicin, dapsone, and clofazamine with oral prednisolone.

This case demonstrates the role of peripheral nerve ultrasound in aiding the diagnosis of an Old World disease like leprosy. Its value in detecting the involvement of asymptomatic nerves with normal electrodiagnostic studies can be of significant value in narrowing the differential diagnoses.[10]

AUTHOR CONTRIBUTIONS

Drs. Vijayan, Punzalan, and Wilder-Smith performed the initial diagnostic assessment and investigations. Dr. Vijayan, C.Y. Chuen, and Dr. Wilder-Smith helped in compilation of the text, literature search, and editing of the manuscript.

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