Clinical Reasoning: A Septuagenarian With Painless Ulceration on the Fingertip

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A 78-year-old non-diabetic non-smoker male presented with painless non-healing ulceration on the middle fingertip of the left hand. Similar episodes have occurred in the past involving the left middle and index fingers resulting in amputation of the tip of index finger. Diagnosis was arrived at by clinical reasoning and confirmed by additional investigations. The readers are presented with a rational step by step diagnostic paradigm in a very rare presentation of a common neurological disorder.

A 78-year-old right hand dominant male was referred for neurological consultation and electrodiagnostic studies, by a hand surgeon concerned about “Neuropathy.” The patient’s main complaint was painless non-healing ulceration on the left middle fingertip of 3 weeks duration with no history of trauma or Raynaud’s phenomenon. On 2 previous occasions similar ulceration healed after prolonged antibiotic therapy by dermatologist. Three years earlier similar ulceration on the left index finger became resistant to therapy resulting in amputation of the fingertip by an orthopedic surgeon. No similar ulceration occurred in the other fingers of left hand, fingers of right hand or the toes. The left index and middle fingers have been numb, and he often accidentally dropped objects that he held in the hand. Tingling of fingers occurred intermittently with nocturnal exacerbations. He is not diabetic and is a non-smoker.

Clinical examination showed swelling, peeling of skin and ulceration on the tip of left middle finger (Figure 1 A). The tip of index finger was missing. Radial artery pulsations were normal, and the fingers
felt warm. There was loss of pain, temperature, and light touch in the left thumb, index and middle finger, radial side of ring finger and radial side of palm, sparing the proximal palm (Figure 1 B). There was wasting and weakness of left thenar muscles. Muscle strength was: 2/5 in Abductor pollicis brevis (APB); 5/5 in Flexor pollicis longus (FPL), Pronator teres (PT), First dorsal interosseous (FDI), Abductor digiti minimi (ADM), Extensor pollicis longus (EPL), Extensor digitorum communis (EDC) and the more proximal muscles. Phalen and Hoffman-Tinel signs were negative.

Questions for consideration:

1. Where is the location of the lesion?
2. What are the differential diagnoses?

Section 2:

Localization: The salient findings are sensory loss, thenar muscle weakness and painless ulcer at the tip of the left middle finger. The sensory loss involves portions of C6 and C7 and C8 dermatomes and may result from lesions at the level of the dorsal root, brachial plexus, or the median nerve. The “split pattern” of sensory loss in the ring finger (Figure 1 B) favors median nerve rather than nerve root/brachial plexus location. The sparing of sensations in the proximal radial palm suggests intact palmar cutaneous branch and points to location at the wrist.

Thenar muscle weakness may occur with lesions at the T1 ventral horn cells, T1 ventral root, lower trunk/medial cord of the brachial plexus or the median nerve. Normal strength in FPL and PT (median nerve) as well as normal strength in FDI (ulnar nerve) and EDC (radial/posterior interosseous nerve) makes it unlikely that the causative lesion is at the C8/T1 roots or the lower trunk / the medial cord of the brachial plexus. Thus, both the sensory and the motor findings localize to the median nerve distal to the innervation of pronator teres and the site of origin of the anterior interosseous nerve (AIN) (in view of the normal FPL).

Digital ulcers occur in scleroderma, Berger’s disease, vasculitis, and herpes simplex infection (herpetic whitlow) as well as in neuropathies. Differential diagnoses of painless ulcers of fingers include syringomyelia, and neuropathies from varied causes involving the median and or ulnar nerves. Focal neuropathies (traumatic, compressive, entrapment, infectious) and diffuse polyneuropathies from multiple etiologies can cause ulceration of fingertips. The selective involvement of index finger (past) and middle finger (current) points to median nerve neuropathy.

Considering both the motor and sensory findings and the painless ulceration of the middle finger, the probable anatomical locations of the median nerve neuropathy are at the carpal tunnel (most likely) or the distal forearm.

A positive Phalen and Hoffman-Tinel sign may have supported clinical localization to the carpal tunnel. A positive timed Phalen test (TPT) with symptoms occurring in 10 seconds or less is reported to predict significant nerve conduction abnormalities in patients with carpal tunnel syndrome. However, occurrence of negative Tinel/Phalen tests as in this patient does not necessarily rule out median nerve entrapment at the carpal tunnel; the overall sensitivity of Phalen sign is reported to be only 85% and the sensitivity becomes even less with longer duration of symptoms.
Questions for consideration:
How to confirm the clinical diagnosis of median nerve neuropathy and its location?

Section 3:
Nerve conduction and needle EMG studies can provide clues to the location as well as the underlying functional pathology (conduction block, demyelination, axon loss). Stimulation of the left median nerve in this patient did not evoke precisely measurable compound muscle action potential (CMAP) over the APB or the 2nd lumbrical. No sensory nerve action potentials (SNAP) could be recorded over the thumb, index, middle or the ring finger on stimulation of the median nerve at the wrist. Hence the nerve conduction study findings failed to provide accurate localization. Needle EMG showed fibrillations in the APB with a few polyphasic motor units on attempted abduction of the thumb. FDI, FPL, PT and EDC were normal. The topography of denervation changes pointed to focal median nerve neuropathy distal to the origin of AIN with significant axon loss, but still the localization was not sufficiently precise for potential surgical intervention.

What additional tests may give more conclusive localization? MR neurography and high-resolution ultrasonography (HRUS) are likely to provide further insight into the location and the underlying cause. Of the two, the HRUS is available at the point of care and has been documented as a sensitive tool in the evaluation of severe distal median nerve neuropathies as in this case. This patient underwent ultrasound imaging of the left median nerve using an 8-18 MHz probe. The findings were (Figure 2 A, B): Significant drop in diameter of the median nerve within the carpal tunnel and marked increase in the cross-sectional area (CSA) at the carpal tunnel inlet (47 mm$^2$ as compared to normal of =/<12 mm$^2$) and normal CSA at the mid forearm. These findings were considered diagnostic of median nerve entrapment at the carpal tunnel.

Discussion:
This case illustrates a very rare presentation of a common disorder, carpal tunnel syndrome (CTS), the most frequent focal neuropathy of the upper extremity. There are only a few case reports of this unusual clinical picture, and the nomenclature has included “Ulcerative and mutilating variant of carpal tunnel syndrome” and “Ulcerative carpal tunnel syndrome.” Among 10,000 patients with CTS whose diagnosis was confirmed by electrodiagnostic studies (EDX) in our facility during the past 15 years, there were only 4 cases with such presentation. The diagnostic features included painless ulcerations confined to index and middle fingers (which receive sensory innervation exclusively from the median nerve), sensory loss in median nerve distribution as well as weakness/atrophy of thenar muscles.

Two mechanisms have been postulated to explain the rare occurrence of finger ulceration in CTS. Loss of sensations can lead to multiple traumata resulting in painless ulceration. Dysfunction of autonomic nerve fibers can also potentially cause hypoperfusion of digits resulting in ulceration. Araujo et al
describe a case in which profound sensory loss persisted although ulcers healed after CT release and argue that sensory loss alone cannot explain the ulceration. Leger and Lavalle performed arteriography and demonstrated “vasospasm caused by the compression of the autonomic fibers of the median nerve”. The occurrence of Raynaud’s phenomenon in patients with carpal tunnel syndrome is also considered supportive of the role of vasospasm. The importance of considering this form of CTS in the differential diagnosis of digital ulcers seen in other conditions like scleroderma has been stressed by Carman and Ernesto.

This case also highlights difficulties in making a conclusive diagnosis of entrapment of median nerve at the carpal tunnel (CT) in advanced cases of CTS using EDX. The atrophy of thenar muscles resulting in loss of CMAP makes it difficult to document focal slowing of motor conduction across the CT. The same problem occurs in sensory conduction study as well when SNAPS are not recordable due to severe involvement of sensory fascicles. Needle EMG is helpful to narrow down the location of the median nerve neuropathy, but still proves to be imprecise as in this case. Under these circumstances HRUS is a handy tool for localizing the median nerve neuropathy more accurately and it may also provide clue to the underlying cause. MR neurography can also be quite helpful but the advantage of HRUS is its instant availability at the point of care and the minimal cost.
References:

8. Chung MS, Gong HS, Baek GH. Raynaud’s phenomenon in idiopathic carpal tunnel syndrome. J Bone Joint Surg (Br) 2000;82-B:818-819
9. Carmen B, Ernesto G. Ulcerating variety of carpal tunnel syndrome mimicking scleroderma. MOJ Orthop Rheumatol 2022;14(5):135-137
**Figure Legends:**

1: Clinical Findings:

1 A: Note the ulceration at the middle fingertip and the short index finger due to prior amputation.

1 B: Note the area of loss of pain and light touch sensations corresponding to the territory of median nerve and sparing that of the palmar cutaneous branch.

2: Ultrasound Findings

2 A: Long axis view showing the left median nerve within the carpal tunnel (arrows); note the marked drop in diameter within the carpal tunnel.

2 B: Short axis view of the left median nerve at the distal wrist crease showing marked increase in cross sectional area (47 mm$^2$ compared to normal of $\leq$12 mm$^2$).
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