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An unusual cause of thenar hypertrophy and carpal tunnel syndrome

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Carpal tunnel syndrome (CTS) is characterized by classic symptoms of paresthesias in the median distribution and weakness that is sometimes accompanied by thenar atrophy. We describe an unusual case presenting with thenar hypertrophy.

A 52 year-old, right-handed man presented with 10 years of intermittent paresthesias in the left median nerve distribution and difficulty snapping the fingers. Driving and typing worsened his symptoms. His examination revealed increased thenar bulk without focal masses (Fig. 1A). A Tinel sign was present. Sensory exam did not elicit significant abnormalities. Mild thenar weakness was noted. Electrodiagnostic studies revealed prolonged distal motor latency (7.9 ms), decreased compound motor action potential amplitude compared to the right (left 5.1 mV, right 9.2 mV) and absent left median sensory nerve action potential. Ulnar studies and F-wave latencies were normal. Needle EMG of abductor pollicis brevis showed increased insertional activity, 1+ fibrillation potentials, 2+ positive sharp waves, 3+ fasciculation potentials, increased motor unit potential (MUP) amplitude, reduced MUP recruitment and increased firing rate. Studies were consistent with left focal median neuropathy at the wrist.

To further elucidate the etiology of thenar hypertrophy, MRI of the hand and wrist was performed to assess structural detail. MRI revealed linear strands of fat infiltrating thenar muscles (Fig. 1B), which was different from diffuse fibrofatty change associated with advanced muscle atrophy. There was globular enlargement of the median nerve with linear cable-like appearance of the nerve fascicles with surrounding fat. The enlargement measured 1.2×1.6 centimeters and was consistent with lipofibromatous hamartoma (LFH) (Fig. 1C).

He underwent decompression of the nerve. Nerve enlargement and swelling was noted at the time surgery but fibrofatty change was not appreciated because of intact epineurium. Three months post-operatively, paresthesias improved. However, imaging revealed no change in the size of the median nerve mass (Fig. 1D).

This case is instructive because thenar hypertrophy led to imaging, which revealed LFH. Thenar hypertrophy could result from either pseudohypertrophy or true hypertrophy related to spontaneous activity, increased work of remaining fibers, and fiber stretching^{2,3}. Chronic denervation and spontaneous activity on needle EMG raises the possibility of true thenar hypertrophy. The focal, band-like pattern of fatty infiltration in thenar muscles suggests extension of LFH. If the fatty replacement were due to severe muscle atrophy, then significant clinical weakness and diffuse fibrofatty change should be present. To our knowledge, we describe the first case of thenar pseudohypertrophy likely due to extension of median nerve LFH.

Besides LFH, localized hypertrophic neuropathy has been reported in hereditary sensory motor neuropathy type 1, Refsum syndrome and diabetic neuropathy⁴. However, a “cable-like” appearance of the median nerve is pathognomonic for LFH and arguably nullifies the need for biopsy⁵. Additionally, LFH have a predilection for the median nerve⁶. The etiology of LFH is unknown. There are many proposed mechanisms including a post-traumatic reactive process, congenital and association with neurofibromatosis⁷.

Surgical treatment of the tumor is debated. Mass excision can lead to sensory deficits⁶. In our case, an initial conservative decompression procedure resulted in improvement in sensory symptoms. LFH of the median nerve should be considered for patients who present with CTS and clinical thenar pseudohypertrophy.

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Abbreviations

CTS	Carpal Tunnel Syndrome
LFH	Lipofibromatous Hamartoma

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Figure 1.

A) Thenar eminence in the symptomatic left hand is appreciably larger compared to the right. B) MR axial T1 weighted non-fat saturated image reveals linear strands of hyperintense fat (arrow) infiltrating the thenar muscles. C) MR axial fat saturated T2 weighted pre-operative image reveals a globular mass within the median nerve with a “cable-like” (arrow) appearance measuring 1.6×1.2 cm. D) MR axial fat saturated T2 weighted post-operative image revealing a globular mass within the median nerve with a “cable-like” appearance without significant change in size (arrow).