A 60-Year-Old Woman with Symptoms of Combined Carpal Tunnel Syndrome and Cubital Tunnel Syndrome Due to an Elastofibroma Causing Compression of the Median and Ulnar Nerves

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Patient: Female, 60-year-old
Final Diagnosis: Elastofibroma of hand
Symptoms: Hand numbness • hand pain • hand weakness • muscle atrophy
Medication: —
Clinical Procedure: — Plastic Surgery

Objective: Rare disease

Background: Cubital tunnel syndrome results from pressure or stretching of the ulnar nerve, and carpal tunnel syndrome involves the median nerve. Elastofibroma is a rare, benign, slow-growing soft-tissue tumor that commonly occurs as a bilateral infrascapular tumor in elderly women. This report is of a 60-year-old woman who presented with combined carpal tunnel syndrome and cubital tunnel syndrome due to an elastofibroma causing compression of the median and ulnar nerves.

Case Report: We report the case of a 66-year-old woman with left-hand numbness, tingling along the fingers, sleep disturbance, and weakness in pinching or holding objects for an extended period. The clinical examination and nerve conduction studies established the diagnosis of combined carpal tunnel syndrome (CTS) and cubital tunnel syndrome (CuTs) complicated by intrinsic muscle wasting. The patient underwent left carpal and cubital tunnels release surgery and end-to-side anterior interosseous nerve transfer to the motor component of the ulnar nerve. Pathologic evaluation of the entire specimen showed collagen bundles alternating with refractive cylinders stained with Verfoeff-van Gieson elastic stain.

Conclusions: This report is of a rare case of a histologically-confirmed single, peripheral, benign elastofibroma involving compression of the ulnar and median nerves. This case highlights the importance of histopathology in diagnosing rare soft-tissue tumors arising at an uncommon site and presenting with rare symptoms.

Keywords: Carpal Tunnel Syndrome • Cubital Tunnel Syndrome • Muscle Weakness • Nerve Compression Syndromes • Nerve Transfer

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Background

Carpal tunnel syndrome and cubital syndromes are types of peripheral nerve compressions that present with numbness and tingling along their distribution on the upper extremity. Many pathological causes have been described in the literature.

Elastofibroma is an uncommon, slow-growing, tumor-like structure characterized by tough fibroelastic sheets with ill-defined margins [1,2]. It was traditionally described as arising from the connective tissue between the lower portion of the scapula and the chest wall of elderly people [3], hence the initial term of elastofibroma dorsi. However, the classical features of elastofibroma have continuously changed due to publications in the last decade that described elastofibroma at different anatomical locations and a variety of clinical presentations [4-8]. Elastofibroma dorsi (ED) is a misnomer and should no longer be tagged by “Dorsi” as indicated by many other reported sites for elastofibroma like axilla, posterior elbow, ischial tuberosity, greater trochanter, stomach, rectum, omentum, eye, hand, and foot [9,10].

Elastofibroma has immunological, biomechanical, and genetic predisposing factors [1], but apparently all proposed explanations of elastofibroma formation must be related to age progression. It is usually a unilateral lesion, but it is bilateral in about 25% of cases [1,11].

We describe an unusual case of combined carpal tunnel syndrome (CTS) and cubital tunnel syndrome (CuTs) complicated by intrinsic muscle wasting of the hand due to elastofibroma. Decompression of the CTS, CuTs, and ulnar nerve at Guyon’s canal led to an uneventful clinical course and resolution of symptoms.

Case Report

A 66-year-old woman, on her first visit, presented with delayed features of left CTS and CuTs complicated by hand weakness and intrinsic muscle wasting. Clinically, she reported long-standing numbness, tingling along the fingers, sleep disturbance, and hand weakness in pinching or holding objects. Upon examination, she had thenar muscle wasting, first dorsal interosseous muscle wasting, and weak abduction/adduction of the fingers (MRC 3). Tinel’s and Phalen’s signs were positive for CTS. A pre-operative work-up, including an electromyography/nerve conduction study, confirmed the clinical diagnosis of median nerve entrapment at the carpal tunnel and non-localized ulnar nerve neuropathy, as well as MRI were performed to exclude cervical radiculopathy, and any proximal pathology might cause compression neuropathy in the upper limb.

We treated the patient surgically. First, a single incision at the palm that was extended to the midforearm to release the median nerve at carpal tunnel, the ulnar nerve at Guyon’s canal, and for end-to-side anterior interosseous nerve transfer (AIN) to the motor component of the ulnar nerve (Figure 1A, 1B). Second, another surgical incision was made to release the ulnar nerve at the cubital tunnel. Median and ulnar nerves were found tightly adherent to the surrounding structures by tough fibrous tissue, preventing the nerves from gliding. Therefore, external neurolysis was essential to free the nerves.

The resected specimen consisted of 2 tan-white, irregular, rubbery, fatty membranous tissue fragment measuring 3×1.5×0.5 cm in aggregate. The specimen was embedded in its entirety.

Microscopic examination (Figure 2A-2C) revealed collagen bundles alternating with numerous acidophilic, refractive cylinders containing a central dense core, with interdigitation of benign

Figure 1. Median nerve (A) looks like a flat sheet after releasing fibrous tissue surrounding the nerve. The median nerve and ulnar nerve (B) were tightly adherent to the surrounding structures by tough fibrous tissue, preventing nerves from gliding.
adipose tissue. The refractive cylinders stained with Verhoeff elastin stain, confirming elastofibroma. A report was issued, and further investigation was recommended.

At her most recent follow-up visit, 6 months after surgery, she reported complete relieve from numbness and tingling, and showed great improvement in hand function and power, as indicated by hand grip, and pinching on the dynamometer scale and pinch scale, respectively.

Discussion

Elastofibroma is considered a degenerative, tumor-like, fibro-elastic lesion, and was first described in 1961 by Jarvi and Saxon. Most of the elastofibroma patients are in their 5th to 8th decades of life, with a 2:1 female-to-male ratio [3,12-14]. Elastofibroma in the upper extremity is a rare presentation. In the literature, only 1 case of elastofibroma was reported in the finger pulp [10].

From our review to the literature [1-16], elastofibroma is found to be a space-occupying tumor-like structure with clinical features elicited by applying pressure on the surrounding structures. However, our intra-operative findings were different, a tough fibrous tissue were adhesively surrounding and compressing both median and ulnar nerves at carpal, cubital tunnel as well as in Guyon’s canal, a pathology that can easily be missed leading to serious complications of irreversible hand wasting.

Based on these findings, elastofibroma in the upper extremity requires special surgical, radiological, and pathological approaches when complicated with compressive neuropathy. Surgically, the goal is to release any potential anatomical areas of nerve compression and to provide adequate nerve gliding. To achieve these goals, an extended single incision was made to release the median nerve at carpal tunnel and ulnar nerve at Guyon’s canal along with decompression of the ulnar nerve at the cubital tunnel. We recommend performing meticulous external neurolysis to release surrounding adherent fibrous tissue and to avoid future traction neuropathy. In addition to distal end-to-side AIN nerve transfer to the motor component of the ulnar nerve that boost the motor component of ulnar nerve with extra fascicles helping in nerve function recovery. Intrinsic muscles wasting of the hand indicate a long-term compression of the ulnar nerve at the cubital tunnel. Muscle wasting is reversible within a window of 12-18 months. Therefore, AIN nerve transfer is useful to protect and preserve distal motor end plates until the native axons fully regenerate. The goal of nerve transfer is to augment the regenerating nerve with additional axons and to quickly reinnervate target muscles.

Figure 2. Elastofibroma of the hand. (A) collagen bundles alternating with refractive cylinders containing a central dense core (×100). (B) Higher magnification (×400). (C) Refractive cylinders are positive with elastic stain (×200).
Radiological work-up for cervical spine is not routine in idiopathic CTS unless it is indicated by patient symptoms, or as in our case when the patient had combined long-standing CTS and CuTs complicated by muscle wasting. We recommend performing cervical MRI when a post-operative elastofibroma diagnosis is reached to rule out possible multilocal compression neuropathy proximally.

Pathological confirmation is essential; the biochemical composition of elastofibroma is very characteristic, consisting of elastin fibers, but with an amino acid composition slightly different from that of normal elastic tissue. The collagen deposited in the lesion has a mixture of types I, II, and III; the presence of type II collagen is perplexing because this is normally present only in articular cartilage and some ocular structures. Type II collagen is synthesized by spindle cells laid down around the pre-existing elastic fibers [17]. Histopathologically, elastofibroma is characterized by the presence of numerous elastic fibers, which are large, coarse, and deeply eosinophilic in nature. Additionally, they appear fragmented, giving the appearance of beads on a string [1,2,8].

Conclusions

This report is of a rare case of a histologically-confirmed, single, peripheral, benign elastofibroma involving compression of the ulnar and median nerves. This case highlights the importance of histopathology in diagnosing rare soft-tissue tumors arising at an uncommon site and presenting with unusual symptoms.

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