

Fibrolipoma of multiple nerves in the wrist

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ABSTRACT

We report fibrolipoma involving the median nerve, its palmar cutaneous branch as well as the ulnar nerve in the same hand of a 25-year-old woman. The patient presented with a lump in the wrist with signs of carpal tunnel syndrome. Multiple nerve involvement was detected on magnetic resonance imaging and further confirmed at surgical exploration and decompression. Imaging is recommended in the management of an unusual lump in the wrist.

Keywords: carpal tunnel syndrome, fibrolipoma, fibrolipomatous hamartoma, median nerve, ulnar nerve

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INTRODUCTION

Fibrolipomas of the nerves are very rare. We report a case of fibrolipoma involving multiple nerves in the same limb, viz. the median and ulnar nerves, which would not have been detected if an imaging study was not done. The fibrolipomas were diagnosed after magnetic resonance (MR) imaging was performed for a young patient presenting with a chronic wrist swelling and carpal tunnel syndrome. The wrist swelling was not typical of tenosynovitis and had a rubbery consistency. There was no clinical evidence of ulnar nerve involvement. This was noted incidentally on MR imaging. Therefore, we recommend the use of imaging in the preoperative investigation of a young person presenting with a chronic rubbery wrist swelling and carpal tunnel syndrome.

CASE REPORT

Our female patient was 25 years of age and right-hand dominant. She presented with a long-standing swelling over the volar aspect of the right wrist associated with numbness and weakness in the hand. She had noticed the painless swelling for the past ten years, and it was slowly growing in size. In the last one year, she had noted that her right hand was weaker than the left. There was also numbness in the radial three digits. Clinical examination revealed an obvious swelling over the volar aspect of the wrist measuring 7 cm × 3 cm (Fig. 1), and extending



Fig. 1 Photograph shows a 7 cm × 3 cm swelling over the volar aspect of the right wrist. There is obvious wasting of the thenar muscles in the right hand. There is reduced sensation over the right thumb, index and middle fingers as well as the thenar eminence.

from the mid-forearm to the region of the carpal tunnel. This swelling was non-tender, fusiform in shape and had well-demarcated edges. It had a rubbery consistency. The thickened cords of nerve fasciculi could be felt within the swelling. It was mobile mostly transversely and less longitudinally. There were no naevi, angiomas, café-au-lait patches or macrodactyly. The right abductor pollicis brevis was wasted (Fig. 1), with slight weakness in the abduction of the thumb. Grip strength (Jamar Dynamometer, Asimov Engineering, Lafayette, IN, USA) on the right was 55 lbs (24.9 kg), when compared to 60 lbs (27.2 kg) on the left hand. Pinch strength (pinch gauge, B & L Engineering Santa Ana, CA, USA) was 8 lbs (3.6 kg) on the right and 10 lbs (4.5 kg) on the left hand. Sensation to pin prick was diminished over the right thumb, index and middle fingers, as well as over the thenar eminence. Two-point discrimination (Disk-Criminator™, Baltimore, MD, USA) was 12 mm over the median nerve distribution. Tinel's sign and Phalen's test were positive for median nerve neuropathy. However, there was no weakness in the flexor pollicis longus and flexor digitorum profundus of the index finger. There was no clinical evidence of ulnar nerve involvement or entrapment neuropathy.

MR imaging showed fat displacing the fascicles of the median nerve, its palmar cutaneous branch, as well as the ulnar nerve. This was pathognomonic of

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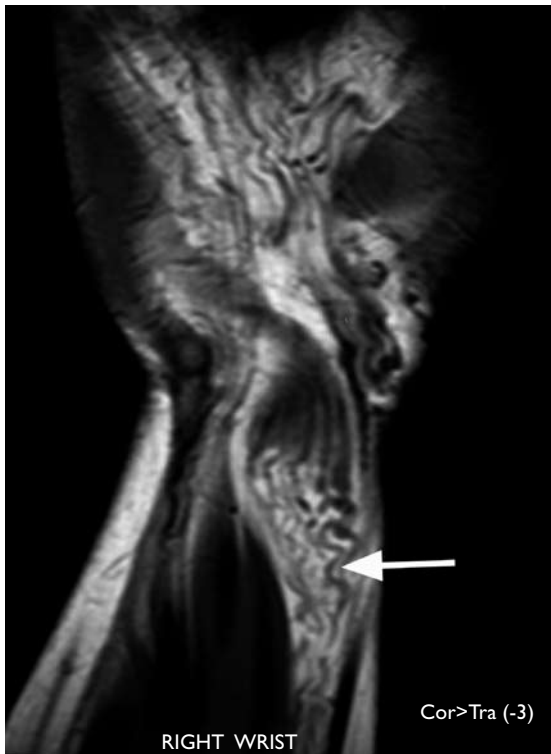


Fig. 2 Coronal T1-W MR image shows the classical “cable sign” (arrow) of fibrolipoma in the median nerve of the right wrist. There is a fusiform enlargement of the median nerve especially in the region of the carpal tunnel. Low-intensity nerve bundles are interspersed with the high-intensity fatty hamartomatous components. The latter is serpentine in shape, giving rise to the “cable sign”.



Fig. 3 Operative photograph shows the enlarged serpiginous nerve fasciculi after epineurotomy of the median nerve (*). The ulnar nerve (white arrow) and the palmar cutaneous branch of the median nerve (dark arrow) are also involved with the fibrolipoma distending the epineurium.

fibrolipoma of the nerve. The classical “cable” sign was found on the coronal T1-weighted image (Fig. 2), where the low-intensity nerve fibres were interspersed with the high-intensity hamartomatous components. There was enlargement of the median nerve fascicles and mild increased signal within the nerve fasciculi proximal and distal to the carpal tunnel, suggesting neuritis. The fat within the nerve sheath was most prominent proximal to the carpal tunnel and was also seen distal to the carpal tunnel where the fat extended into the divisions of the nerve sheath to the index, middle and ring fingers.

Intraoperative findings confirmed a well-circumscribed tumour of the median nerve and its branches, and the ulnar nerve. The median nerve tumour was yellowish, firm, fusiform and lobulated. It measured 7 cm × 3 cm extending from the distal forearm to the carpal tunnel. At the carpal tunnel, it was severely compressed and narrowed with minimal lipomatous tissue. Distal to this, it was enlarged in its digital branches including the recurrent motor branch. The palmar cutaneous branch of the median nerve was also affected by the lipomatous tumour as it exits the deep fascia, and this involvement extends to its terminal

branches (Fig. 3). The ulnar nerve involvement was a similar tumour measuring 3 cm × 3 cm just proximal to and within the Guyon’s canal (Fig. 3).

Decompression incisions were done for the median nerve at the carpal tunnel, the palmar cutaneous branch at its exit through the deep fascia, and the recurrent motor branch at its entrance to the muscle. The ulnar nerve was also decompressed at the Guyon’s canal. Epineurotomy of the median nerve was done. Its fasciculi appeared enlarged and oedematous (Fig. 4), lying in a serpiginous manner and encased by the fibrolipomatous infiltrations. A terminal branch of the palmar cutaneous nerve was excised for frozen section histology. It showed mature fat cells surrounding normal nerve fasciculi, with no evidence of malignancy. A further biopsy specimen was taken from a nerve fascicle of the median nerve, together with its adjacent fibrolipomatous tumour. The cut nerve endings were coapted with sutures. The histology on paraffin section later confirmed the diagnosis of fibrolipoma of the nerve with mature fat cells within the epineurium of the nerve. The nerve fasciculi were grossly normal, and there was perineural fibrosis.

At the two-year follow-up, the patient did not



Fig. 4 Operative photograph shows that the nerve fasciculi (arrow) are enlarged, oedematous and serpentine in appearance. They are closely interspersed with fibrofatty hamartomas.

complain of residual swelling, numbness of the hand or pillar pain. On examination, there was a full range of motion at the wrist, and the Tinel's sign was negative. The two-point discrimination improved to 7 mm along the median nerve distribution. The grip and the pinch strength of the right hand also improved to 60 lbs (27.2 kg) and 11 lbs (5 kg), respectively.

DISCUSSION

Fibrolipoma of the nerve is a rare condition. We report a case of multiple nerves involvement in the same hand, which to our knowledge, has not been reported in the English literature. Most of these fibrolipomas involve single nerves in the hand, commonly the median nerve,⁽¹⁾ and rarely the ulnar nerve.⁽²⁾ Bilateral involvement of the same nerve has been reported by Salon et al.⁽³⁾ Although fibrolipoma can occur as an independent entity, up to a third has an association with macrodactyly.⁽⁴⁾ This has been found to be the most frequent lesion associated with digital hypertrophy in the upper limb.⁽³⁾

The condition is believed to be congenital in origin, although there has not been a known genetic transmission. In our case, it is unclear. Though the patient's parents had noticed a lump over the right wrist since birth, she only noticed the lump at the age of 15 years when it was growing in size. A patient with fibrolipoma of the nerve in the hand usually reports a slow-growing lump for many years. As the lump grows in size, it may be associated with the signs and symptoms of compression neuropathy. Our patient presented late, and there were late clinical signs of advanced median nerve neuropathy. The palmar cutaneous branch of the median nerve was involved, but there was no proximal extension to involve the anterior interosseous nerve. Typically, the fibrolipoma has a "stringy" surface on top of its doughy consistency.

Ultrasonography of the neural fibrolipoma may

reveal alternating hyperechoic and hypoechoic bands.⁽⁵⁾ MR imaging is the most commonly-utilised investigative tool for the diagnosis of fibrolipomas of the nerves.^(4,5) The tumour presents the pathognomonic features known as the "cable sign".⁽⁶⁾ This is best seen on axial T1-weighted images, where multiple low-signal serpentine, tubular nerve bundles are juxtaposed with the high-signal hamartomatous component within an expanded epineurium. Also, onion-ring-like concentric circles of fibrous and fatty tissues surrounding the nerve fibres can be seen on the axial slices. At times, the nerve can appear hypointense on the T1-weighted images, suggestive of fibrous degeneration. The nerve may appear hyperintense due to the neuritis. The exact appearance of the nerve depends on the duration of any compressive neuropathy. MR imaging may reveal the involvement of other nerves in the same region that may not be evident clinically. In our case, it revealed the involvement of the ulnar nerve.

In the literature, the macroscopic appearance of a fibrolipoma is classically a firm, yellowish-orange fusiform mass with an intact epineurium that is well-demarcated from the surrounding tissue.⁽⁴⁾ Within the epineurium, the nerve fasciculi appear enlarged and interspersed with the fibrous and lipomatous tissues.⁽⁷⁾ There are no planes between the nerve fasciculi and the fibrolipomatous infiltrations, and these nerve fibres are often inseparable from the hamartomatous components. Histologically, there is fibrofatty infiltration of the interfascicular planes,⁽⁸⁾ but not into the nerve fasciculi. These fibrous and fatty tissues have been found to be of epineural, perineural and endoneural origins.⁽⁹⁾ The lipomatous component consists of mature fats, and there is often perineural fibrosis.^(10,11) The nerve fasciculi are normal, and the epineurium is intact.

There have been numerous treatment options reported, but the consensus is on conservative surgery. If median nerve compression is present at the wrist, decompression can be done with the division of the transverse carpal ligament, superficial excision of epineural proliferation,⁽³⁾ together with a biopsy of a palmar cutaneous branch.⁽¹²⁾ This simple approach has produced good results.^(3,4,13) Interfascicular microdissection has been attempted, but in most cases, the results have been disappointing, with partial or complete loss of median nerve function.⁽¹⁴⁾ A further option is radical en bloc excision of the tumour-bearing nerve segment with sural nerve graft interposition. However, the results have been poor even in children with their greater potential for nerve regeneration. The resultant motor loss may be treated with an opponensplasty,⁽¹²⁾ but there is no definite solution for sensory loss. Therefore, radical surgery, with

its potential for permanent motor and sensory loss, is not recommended, especially in a young patient who has no functional disability. In our case, we performed a biopsy of a nerve fascicle together with its repair, in addition to carpal tunnel release and decompression of the other sites of entrapment of the palmar cutaneous and recurrent motor branches of the median nerve. The ulnar nerve was also decompressed at the Guyon's canal.

Fibrolipoma of the median nerve is a rare condition, and it may present late with advanced signs of compressive neuropathy. Our patient is a rarer case of multiple nerve involvement in the same hand with only signs of median nerve entrapment neuropathy. The involvement of the other nerves was detected on MR imaging. Therefore, we recommend the use of imaging in the investigation of young patients with carpal tunnel syndrome, and concomitant chronic unusual volar wrist swelling that is not typical of synovitis or a cystic lesion. Imaging would facilitate planning for surgery as well as detecting the subclinical involvement of the other nerves. An elective decompression of these nerves may be done before the presentation of late signs in an advanced disease. The treatment of this benign tumour should be restricted to simple decompression and epineurotomy.

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