

# Fibrolipomatous hamartoma of the median nerve presenting with carpal tunnel syndrome

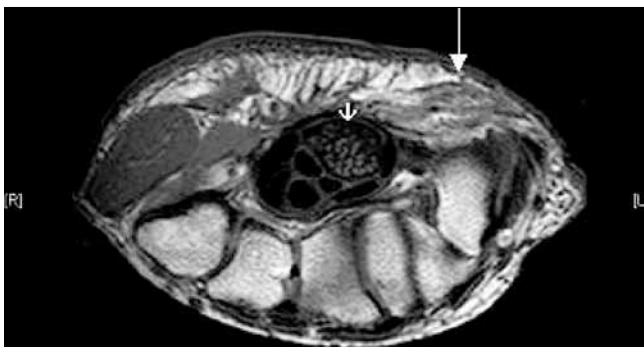
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A 37-year-old gentleman presented with weakness, burning sensation, pain and a progressively increasing swelling in the volar aspect of the right palm and wrist for three years. On examination there was wasting of the thenar eminence with muscle power of MRC Grade II and decreased sensation in the median nerve distribution of the right hand. Nerve conduction study showed no response to stimulation of the right median motor and sensory components. Palmar conduction showed no response to stimulation in right median nerve. Magnetic resonance imaging (MRI) of the wrist and hand showed enlargement of the median nerve with hypointense linear serpentine fibers interspersed between hyperintense signal both in T1W, proton density and T2W [not shown in figure] images suggestive of fatty infiltration, giving the appearance of “coaxial- cable- fibers” in the axial plane [Figure 1] and a “spaghetti pattern” in the coronal plane [Figure 2]. The mass caused displacement of the flexor tendons and bulging of the flexor retinaculum. Nerve thickening was extending proximally within the forearm and distally involving the digital branches [Figure 3]. A diagnosis of fibrolipomatous hamartoma

of the median nerve causing carpal tunnel syndrome was made.

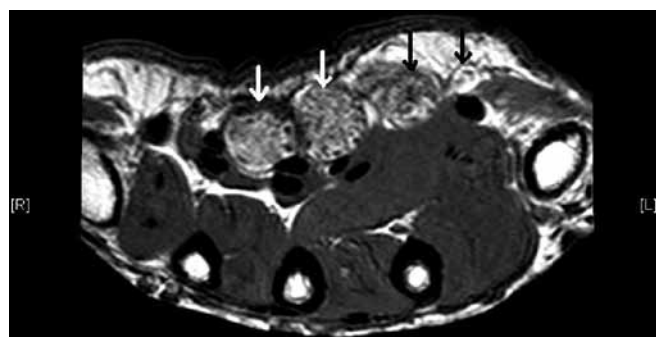
Under regional block anesthesia and tourniquet control, the patient underwent carpal tunnel release by the open extensile approach with the incision placed ulnar to the axis of the flexed ring finger starting proximal to the Kaplan line to cross the wrist crease at 45° angle to as far as 5 cm in the distal forearm. The tensely stretched transverse carpal ligament and the investing fascial layer of the distal forearm were released longitudinally decompressing the nerve. Internal neurolysis and epineurotomy of the thickened median nerve fascicles was carried out under loupe magnification. The wound healed normally in the postoperative period, with absence of pain and burning sensation. Biopsy of limited nerve section was consistent with fibrolipomatous changes of the median nerve showing expansion of the epineurium by mature adipose tissue intercepted focally by bands of fibrous tissue separating the fascicle. There was concentric perineurial fibrosis



**Figure 1:** Axial FSE T1-weighted image through the level of carpal tunnel showing enlarged median nerve with coaxial-cable-like appearance due to hyperintense fat interspersed between the hypointense nerve fascicles [short arrow]. Hyperintensity and atrophy of the thenar muscles also seen [long arrow]



**Figure 2:** PD coronal image of the wrist and hand showing hypointense linear median nerve fibers in hyperintense fat giving spaghetti pattern



**Figure 3:** Axial FSE T1-weighted image through the level of metacarpals showing involvement of the digital branches of median nerve [arrows]

with infiltration of very occasional adipocytes into the perineurium. The endoneurium showed marked loss of myelinated fibers with formation of onion bulbs. On immunohistochemistry the cells forming onion bulbs were positive for S100 protein and negative for epithelial membrane antigen. There was no clinical evidence of improvement in muscle power at the sixth month follow-up although the patient remained symptom-free.

Fibrolipomatous hamartoma of a nerve is a rare soft tissue tumor, which occurs due to fibro-fatty proliferation within the nerve bundles with massive epineurial and perineurial fibrosis leading to fusiform enlargement of the nerve.<sup>[1]</sup> It affects most commonly the median nerve and its branches. Other peripheral nerve involvement like ulnar, radial, plantar, peroneal and sciatic has been described. Most cases occur before the age of 30 years.<sup>[2]</sup> Clinical symptoms consist of gradually increasing swelling, sometimes associated with symptoms of a compression neuropathy. The lesion is also seen to be associated with macrodactyly and macrodystrophia lipomatosa, particularly of a

lower extremity.<sup>[3]</sup> The MR appearance of this condition is unique and characteristic. Thickened nerve fascicles embedded in evenly distributed fat appear as serpentine low-intensity structures surrounded by high-intensity signal in both T1W and T2W images giving the coaxial-cable-like appearance in the axial plane and a spaghetti-like appearance in the coronal planes. This appearance is considered to be pathognomonic of the lesion and routine biopsy is even considered not necessary.<sup>[2]</sup> Management is usually conservative. But where the patient is symptomatic indicating a severe compression element, decompression surgery as of the carpal tunnel in this patient with median nerve involvement has been shown to provide adequate relief of symptoms.<sup>[4]</sup>

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