Fibrolipomatous hamartoma of the median nerve: MR imaging findings

Published on 27.06.2013

**DOI:** 10.1594/EURORAD/CASE.11028

**ISSN:** 1563-4086

**Section:** Musculoskeletal system

**Area of Interest:** Musculoskeletal soft tissue Extremities

**Procedure:** Diagnostic procedure

**Imaging Technique:** MR

**Special Focus:** Pathology

**Case Type:** Clinical Cases

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**Patient:** 20 years, female

**Clinical History:**

A 20-year-old woman presented with a large mass in the volar aspect of the left wrist and hand, which had been increasing over the past two years. She had paresthesia in the 2nd, 3rd and 4th fingers. Lymph node tumefaction was not detected, nor was any sign of neurofibromatosis. There was no history of trauma.

**Imaging Findings:**

Magnetic resonance imaging (MRI) demonstrated a fusiform enlargement of the median nerve at the level of the distal forearm, wrist and volar aspect of hand, extending to the digital branches. The tumour measured 6 x 2.5 cm in its transverse diameters and 17 cm in its length. T1 and T2 weighted images (WI) showed tubular low-intensity structures encased in high signal tissue representing fat, which homogeneous drop in signal intensity on fat suppression images. On axial sequences the lesion demonstrated a “coaxial cable-like” appearance while on coronal and sagittal images a “spaghetti string” appearance was noted. This constellation of findings is pathognomonic of fibrolipomatous hamartoma.

**Discussion:**

Fibrolipomatous hamartoma is a benign tumour composed of hypertrophied fibrofatty tissue intermixed with nerve tissue, usually confined to the nerve sheath. This lesion is also known as neural fibrolipoma, lipofibromatous hamartoma, perineural lipoma and intraneural lipoma [1-4].

The aetiology of this disorder has not been definitively clarified. Various hypotheses were suggested: congenital origin, growth by nerve irritation, inflammation or prior trauma [2, 3, 5].

The age of presentation is typically before 30 years of age, often in early childhood and less commonly in early adult age [1-4, 6, 7].

Males and females are equally affected, and there is no familial predisposition [1].

The median nerve is the most commonly affected nerve (80-85% of cases). However, involvement of others nerves has been reported [1-3, 8-10].

Patients typically present a soft, slowly enlarging mass on the volar wrist or distal forearm [4], often present since infancy. It may be asymptomatic, or may produce neurologic symptoms, due to nerve compression, in the affected nerve distribution, such as pain, numbness, diminished sensibility and strength or carpal tunnel syndrome [1, 2, 3, 5].

Frequently fibrolipomatous hamartoma is associated with macrodactyly (20-67% of cases) [1, 2, 4], which results in bony overgrowth, and fat deposition in subcutaneous tissue, tendons, muscles, and nerves [3].

Plain film is non-specific and may be normal in patients without macrodactyly or may show a soft tissue mass related to the fibrofatty tissue [1].

At ultrasound (US) it appears as a soft tissue lesion with smooth round hypo-or anechoic bands (fascicles)
surrounded by hyperechoic tissue (fatty tissue) [1, 2, 4, 10]. Spectral Doppler will demonstrate no flow within the mass [2].

Computed tomography (CT) shows fusiform nerve enlargement which is caused by fatty tissue interposed among serpentine or tubular structures of soft tissue density [2, 3]. Although the sonographic and CT features are characteristic of the lesion [10], MRI is the preferred modality for diagnosis, because MRI features are considered pathognomonic [2]. They consist of longitudinal cylindrical linear or serpiginous bands, about 3 mm in diameter, with low signal intensity on T1 and T2-WI, representing nerve fascicles and surrounding fibrous tissue, surrounded by fat, which appears as high intensity on T1 and T2-WI, with decrease of high signal on fat suppressed images. This leads to a “coaxial cable-like” appearance on axial images and a “spaghetti string” appearance on coronal images [1-3, 6, 7, 10]. Associated intramuscular fat deposition has been described, related to nerve injury and muscle denervation, macroductyly or in the absence of macroductyly [3].

**Differential Diagnosis List:** Fibrolipomatous hamartoma of the median nerve, Intraneural lipoma, Lipomas, Traumatic neuroma, Nerve sheath tumours, Ganglion cysts, Vascular malformations. The previously described pathognomonic imaging findings are not seen in any other soft tissue lesion and should allow confident non-invasive diagnosis, avoiding the need for biopsy.

**Final Diagnosis:** Fibrolipomatous hamartoma of the median nerve

**References:**


Description: Sagittal T2-weighted fast spin echo (FSE) MRI (TR 3500/TE 106) in the location of palpable mass shows a fusiform mass in the expected location of the median nerve (arrow) with a “spaghetti string” appearance. Origin: Department of Radiology, Hospital de São João, Porto, Portugal
Description: Axial T1-weighted SE MRI (TR 781/TE 17) shows a “cable-like” appearance due to low signal intensity circular structures (arrows) of thickened nerve fascicles and surrounding fibrous tissues embedded in the high signal intensity fatty tissue. Origin: Hospital de São João, Department of radiology, Porto, Portugal.
Figure 3

Description: Axial T1-weighted SE MRI (TR 781/TE 17) at level of the carpal tunnel. The mass displaces the flexor tendons leading to the bowing of the flexor retinaculum (arrow). Origin: Department of Radiology, Hospital de São João, Porto, Portugal.
**Description:** Coronal T1-weighted SE MRI (TR 550/TE 17) shows thickened nerve fascicles with high signal fat interspersed, resulting in a “spaghetti string” appearance. **Origin:** Hospital de São João, Department of radiology, Porto, Portugal.
Description: Axial T2-weighted FSE MRI (TR 3340/TE 105) of the wrist. The mass (asterix) displaces the flexor tendons dorsally (arrow). **Origin:** Department of Radiology, Hospital São João, Porto, Portugal
**Description:** T2-weighted fat-suppressed FSE MRI (TR 3800/TE 106), the hyperintense signal of the tissue drops completely and homogeneously (arrow). **Origin:** Department of Radiology, Hospital São João, Porto, Portugal
Figure 7

Description: Axial T1-weighted SE MRI (TR 870/TE 17) at the level of palm shows the mass (asterix).
Origin: Department of Radiology, Hospital São João, Porto, Portugal