

GIANT LIPOFIBROMATOUS HAMARTOMA OF MEDIAN NERVE IN A CHILD

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ABSTRACT

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Lipofibromatous hamartoma (LFH) is a rare fibrofatty tumor of adipocytes within peripheral nerves, affecting mainly children. It typically presents as a palpable mass surrounding the nerves of the upper limbs, causing pain and neurological deficits in the affected nerve distribution. We report the case of a child with a 2-years presentation of a mass in the right wrist associated with pain and paresthesia, who underwent investigation with magnetic resonance imaging (MRI). It showed thickening of the median nerve with spaghetti-like appearance associated with lipomatous tissue in a coaxial cable-like pattern, both features characteristic of LFH. This case illustrates the importance of MRI in the differential diagnosis of limb masses in the pediatric population.

Keywords: *Lipofibromatous hamartoma; Median nerve; Magnetic resonance imaging*

INTRODUCTION

Lipofibromatous hamartoma (LFH) is a rare, benign fibrofatty tumor composed by proliferative mature adipocytes within peripheral nerves, which form a palpable neurogenic mass. It affects mainly nerves of the upper limbs, causing pain, sensory and motor deficits in the affected nerve distribution. Although LFH was first described in the English literature in 1953, fewer than 100 cases have been documented. We describe a rare case of median nerve involvement by LFH in a 6-year-old child.

CASE REPORT

A 6-year-old child presented with tumefaction in the right carpal tunnel region associated with pain and paresthesia for 2 years. The patient underwent radiological investigation with magnetic resonance imaging (MRI) non-contrast multiplanar, multi-sequence of the right hand. It showed gross thickening of the median nerve as well as diffuse presence of lipomatous tissue (hyperintense on T1 and T2 sequences) in a coaxial cable-like pattern, extending from the level of distal forearm up to the distal metacarpal region. In coronal and axial images, the nerve had a spaghetti-like appearance (Figures 1-2).

The thickened nerve was seen causing compression. The tendons of the hand appeared normal in signal intensity. Diagnosis of fibrolipomatous hamartoma arising from the right median nerve was made, based on these characteristic imaging findings: thickening of the median nerve as well as diffuse presence of lipomatous tissue and a coaxial cable-like or spaghetti-like pattern (Figures 1-3).

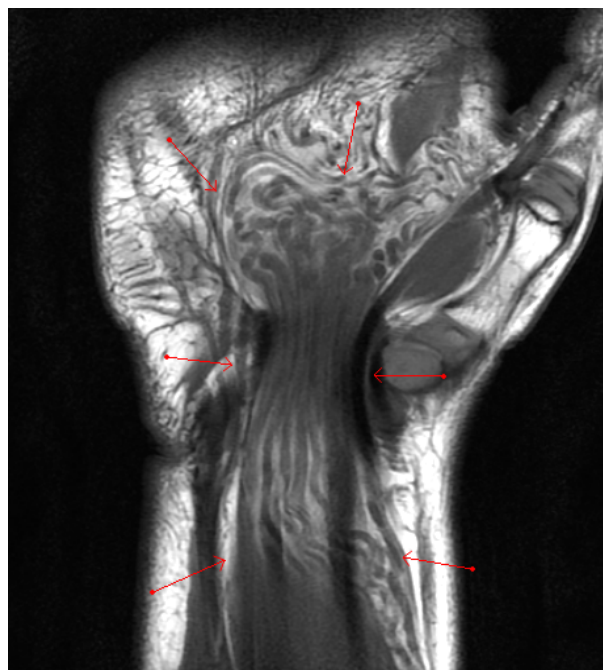


Figure 1: Coronal T1 image showing interposition of the nerve fascicles by fatty tissue, the so-called “spaghetti” sign (arrow).

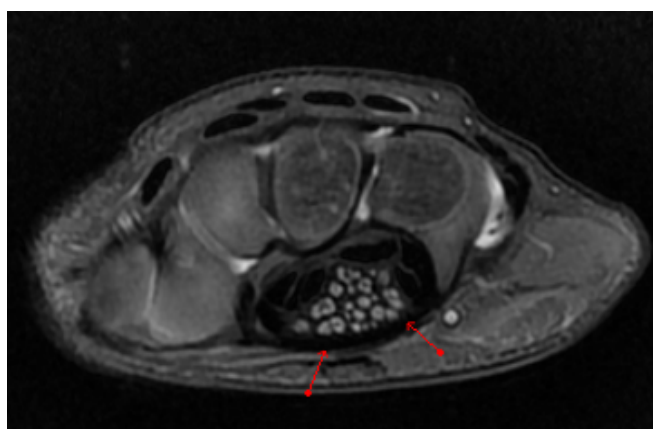


Figure 2: Axial T2-weighted fat saturated image showing diffuse fascicular enlargement of the median nerve with surrounding and interposing hyperintense signal on T1WI with corresponding high signal on the T2FS-weighted images indicated fat, the so-called “co-axial cable” sign (arrow).



Figure 3: Sagittal T2-weighted fat saturated image showing diffuse fascicular enlargement of the median nerve with surrounding and interposing hyperintense signal on T1WI with corresponding high signal on the T2FS-weighted images indicated fat, the so-called “co-axial cable” sign (arrow).

DISCUSSION

LFH was described as most commonly in infants than in children and young adults. It usually presents as a slowly growing, asymptomatic mass, but pain and paresthesia may develop if it causes nerve compression or entrapment, which are our case's presenting symptoms. It involves mainly the median (80%), ulnar and or radial nerves and/or dorsum

of the hand; this predilection remains unexplained. Also, macrodactyly of the involved limb or digit can be present in 66% of the cases¹. The WHO classification of soft tissue tumors categorizes LFH as an adipocytic tumor, due to hypertrophy of mature fat and fibroblasts in the epineurium². Table 1 presents a list of 15 cases obtained in a PubMed search of articles between 2012 and 2022 using the terms (Fibrolipomatous hamartoma) AND (median nerve).

Tabela 1: PubMed literature review using the terms (Fibrolipomatous hamartoma) AND (median nerve), from 2012 to 2022.

Author (reference), year	Age, years	Sex	Clinical presentation	Imaging methods used	Treatment	Follow-up
Pallewate and Samarasinghe ³ , 2021	17	Female	Swelling	USG, CT and MRI	Incisional biopsy	N/A
Aladel et al. ⁴ , 2021	36	Female	Pain, paresthesia, swelling, macrodactyly. Positives Tinel's and Phalen's sing	X-ray, MRI	Conservative management	N/A
Rai et al. ⁵ , 2021	17	Male	Swelling and paresthesia	MRI	Carpal tunnel release and excisional biopsy	Complete remission
Michel et al. ⁶ , 2021	26	Male	Swelling	X-ray, USG, MRI	Carpal tunnel release	Complete remission
Azeemuddin et al. ⁷ , 2018	31	Male	Swelling, pain, paresthesia, reduced mobility of the middle finger	X-ray, MRI	Excision of the middle finger and nerve alograft	Complete remission
Ranjan et al. ⁸ , 2021	17	Male	Swelling, paresthesia. Positive Tinel's and Slip sign	MRI	Carpal tunnel decompression	Complete remission
Parry, Wani, and Shaheen ⁹ , 2021	32	Male	Pain, paresthesia and swelling	MRI	N/A	N/A
Maldjian ¹⁰ , 2013	54	N/A	Swelling. Previous renal transplant	CT	Conservative management	N/A
Shekhani, Hanna, and Johnson ¹ , 2016	8	Male	Swelling, paresthesia.	X-ray, MRI	Conservative management	N/A
Kitridis et al. ¹¹ , 2018	15	Male	Pain, swelling and reduced mobility. Positive Tinel's and Phalen's sign	X-ray, MRI	Carpal tunnel decompression and excisional biopsy	Complete remission
Ha et al. ¹² , 2012	55	Female	Swelling, paresthesia.	USG, MRI	Excisional biopsy	Complete remission
Palmer, Sritharan, and Sampson ¹³ , 2017	3	Male	Swelling, reduced mobility, paresthesia	X-ray, USG	Ressection of the median nerve and nerve alograft	Complete remission
Okubo et al. ¹⁴ , 2012	Case 1: 56	Female	Swelling	MRI	Excisional biopsy	Complete remission
	Case 2: 17	Male	Paresthesia, swelling	MRI	Carpal tunnel release	Complete remission
	Case 3: 15	Male	Paresthesia and swelling	MRI	Carpal tunnel release	Complete remission

MRI is an important tool in the diagnosis of LFH, as it not only confirms the diagnosis but also provides a detailed assessment of nerve involvement preoperatively. A characteristic feature on MRI is the appearance of serpentiform nerve fascicles surrounded by fibrofatty tissue within the expanded nerve sheath. Distribution of fat between fascicles is asymmetric^{3,15}. On coronal images, the nerve has a spaghetti-like appearance that is pathognomonic for LFH. It is necessary to mention and consider the use of ultrasound of the patient's median nerve, as it is also a method that can demonstrate the coaxial aspect of lipofibromatous hamartoma, widely available, low cost, fast acquisition and suitable for delimitation of extensive injuries. In our case, due to the suspicion of malignancy due to the size and progression of the lesion, the investigation was carried out directly with MRI. In this context, our case draws attention due to the size of the mass and

the uncommon age group of the patient. Its report reinforces the need to include LFH in the differential diagnosis of masses in the region of the upper limbs with clinical signs of nervous involvement, with MRI being the exam of choice^{4,13}.

CONCLUSION

LFH is a rare condition that occurs in the younger age group and has a predilection for upper extremity nerves. Pathognomonic imaging features are present on cross-sectional imaging, especially MRI, and may represent histopathological structure.

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None.

Conflicts of interest

None.

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