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Journal Title: Journal of Radiology Case Reports

Volume: Volume 10, Number 11

Publisher: EduRad Publishing | 2016-01-01, Pages 1-7

Type of Work: Article | Final Publisher PDF

Publisher DOI: 10.3941/jrcr.v10i11.2787

Permanent URL: <https://pid.emory.edu/ark:/25593/s33ct>

Final published version: <http://dx.doi.org/10.3941/jrcr.v10i11.2787>

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Accessed July 22, 2024 4:59 AM EDT

Lipofibromatous Hamartoma of the Median Nerve: A Case Report

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Radiology Case. 2016 Nov; 10(11):1-7 :: DOI: 10.3941/jrcr.v10i11.2787

ABSTRACT

Lipofibromatous hamartoma is a rare and slow growing benign fibro-fatty tumor. It is characterized by the proliferation of mature adipocytes within the epineurium and the perineurium of the peripheral nerves. In the upper extremity, it most commonly affects the median nerve. Median nerve involvement commonly leads to pain, numbness, paresthesia and carpal tunnel syndrome. This article presents a case of lipofibromatous hamartoma in an 8-year-old child followed by a discussion of the epidemiology, diagnosis, imaging details and treatment options for this condition.

CASE REPORT

CASE REPORT

An 8-year-old Caucasian male presented to an outpatient orthopaedics center for a follow up evaluation of a mass-like protuberance in the palmar aspect of his right hand (figure 1). According to his parents, the mass was present since infancy and had been increasing in size for two years. Although asymptomatic at the time of presentation, history revealed two instances of numbness and tingling in his right hand over the past two years.

Physical examination revealed a soft, non-mobile mass starting at the patient's distal forearm extending toward the 2nd and 3rd digits and ending distal to patient's thenar crease. The area was non-tender, and sensation to light touch was intact. His median, radial, ulnar, anterior and posterior interosseous nerves were all neurologically intact. He had a normal radial pulse and a brisk capillary refill. There was full range of motion in all fingers, and no neurovascular deficit was noted.

Magnetic resonance imaging (MRI) obtained one year prior to presentation (figure 2) displayed lipofibromatous hamartoma (LFH) of the median nerve. The mass measured 2.0

x 1.0 cm in transverse by anteroposterior dimensions with 12 cm proximal-distal extension. Local mass effect and crowding in the region of the carpal tunnel was noted on imaging.

One month prior to presentation, the patient complained of worsening pain in his right hand once or twice a week in his thumb and thenar eminence. Squeezing or grasping activities like writing and focal compression of the mass aggravated the pain. Rest quickly resolved the discomfort. The patient noted no numbness, tingling or weakness in his right hand. Repeat MRI was obtained (figure 3). In comparison to imaging 11 months prior, the mass had grown to the following parameters: 2.4 cm x 1.2 cm in transverse by anteroposterior dimensions with 12 cm proximal-distal extension.

Surgical intervention was not recommended given the significant morbidity and the patient's minimal symptoms. The patient was encouraged to modify his activities, such as the way he holds a pencil or other objects or to try and alleviate pressure over this area. Otherwise the patient was told to continue observation and return to clinic if any new symptoms should arise. He was scheduled to have a one-year follow-up with a repeat MRI.

DISCUSSION

Etiology and Demographics:

Lipofibromatous hamartoma is a rare and benign fibro-fatty tumor that is characterized by the proliferation of mature adipocytes within the epineurium and the perineurium of the peripheral nerves [1-13]. The etiology LFH is unknown, but it likely has a congenital origin with the vast majority of cases occurring in infants and less commonly in children and young adults [3, 13]. As of a 2013 comprehensive literature review by Tahiri et al., there have been 180 known cases of LFH of the median nerve reported; to the authors' knowledge, there have been approximately 10 more cases published since that time [5]. However, the exact worldwide incidence is not well described in the literature. Despite some discrepancy, many authors have found that males are overall slightly more affected than females, while macrodactyly is more predominant in females [2,6]. The first report was published by Mason in 1953 [13, 14]. Many names have been used to describe LFH in the past including fibrolipomatous hamartoma, fibrolipoma, lipofibroma, fibrofatty proliferation, microdystrophia lipomatosa, intraneural lipoma, perineurilipoma, fatty infiltration of the nerve, fatty or fibrous neoplasm of nerve and lipomatosis of nerve [1, 4-6, 10, 15]. In 1969, the term lipofibromatous hamartoma was introduced by Johnson and Bonfiglio and remains the most accurate description of this condition [16].

Clinical & Imaging Findings:

LFH most commonly affects the median nerve in the upper extremity, accounting for 80% of cases [1, 2, 8, 11, 12]. Ulnar, radial, sciatic, plantar and brachial plexus nerves are other less commonly affected sites of involvement [1, 3, 10]. Involvement of the median nerve typically leads to pain, numbness, paresthesia and carpal tunnel syndrome [1, 8]. Symptoms often arise in the third and fourth decade [1, 8]. The most common characteristic associated with LFH is macrodactyly, which occurs in approximately 30% of cases [1-4, 10].

There have been no clear guidelines established for the diagnosis and treatment of LFH [5]. LFH can be diagnosed by ultrasonography (US), computed tomography (CT) and magnetic resonance imaging (MRI)[5, 8]. US displays an enlarged hypoechoic median nerve containing hypoechoic coaxial cable like bundles encased by an echogenic substratum [1, 3, 4]. MRI is generally the imaging modality of choice to assist with diagnosis and surgical planning [8]. The pathognomonic features seen on MRI are serpiginous hypointense nerve fibers (coaxial cables) interwoven with abundant T1/T2 hyperintense fatty tissue along the median nerve [2, 4, 5, 7]. An MRI alone is adequate to make a diagnosis, thus a biopsy is not necessary [4].

Treatment & Prognosis:

The treatment modality for LFH ranges from observation in asymptomatic patients to prophylactic carpal tunnel release (CTR), biopsy and total nerve resection in symptomatic patients [4, 5]. CTR has proven helpful for many with carpal tunnel symptoms, leading to a complete resolution of symptoms [4] and is the mainstay for treatment with neuropathy [5]. A biopsy should be performed with CTR to

confirm diagnosis. Patients with worsening symptoms may be required to undergo intraneural dissection, nerve resection with grafting and debulking [4]. The literature shows mixed results of early aggressive resection. Tumors that infiltrate the nerve are difficult to extract and hard to access. For these patients, performing an epineurotomy and intraneural dissection have seen good results [4]. There have been reported cases post CTR of recurrence of the mass and progression of symptoms leading to more aggressive repeat surgery [17]. Complete excision of a LFH of the median nerve can result in tremendous loss of neurological function. Louis et al. reported 2 patients with secondary surgery who eventually lost all median nerve function [4, 17].

Treatment options for macrodactyly include debulking overgrown tissue, digital amputation, wedge osteotomy, middle phalangectomy with arthroplasty, and epiphysiodesis [5]. Aggressive surgery should be only be used if conservative management fails or when the benefits of surgery outweigh the risks.

Differential Diagnoses:

Differential diagnoses include traumatic neuroma, neurofibroma, neuroma, schwannoma, ganglion cyst, lipoma, hereditary hypertrophic interstitial neuritis and vascular malformation [2, 3, 5,15,18]. On MRI, LFH is unique in comparison to these differential diagnoses due to its serpiginous, hypointense nerve fibers that are interwoven with abundant T1/T2 hyperintense fatty tissue along the median nerve. neurofibromas and schwannomas may exhibit a target sign. Additionally, schwannomas may also display features such as a split fat sign and fascicular sign. A lipoma exhibits T1 and T2 hyperintensity with complete saturation on fat-suppressed sequences. A ganglion cyst exhibits a fluid mass near a joint or tendon, and hereditary hypertrophic interstitial neuritis shows abnormal foci of high T2 signal intensity on MRI.

TEACHING POINT

Lipofibromatous hamartoma is a rare and slow growing benign fibro-fatty tumor that most commonly affects the median nerve. The pathognomonic MR appearance is serpiginous hypointense nerve fibers ("coaxial cables") interwoven with abundant T1 and T2 hyperintense fatty tissue.

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FIGURES

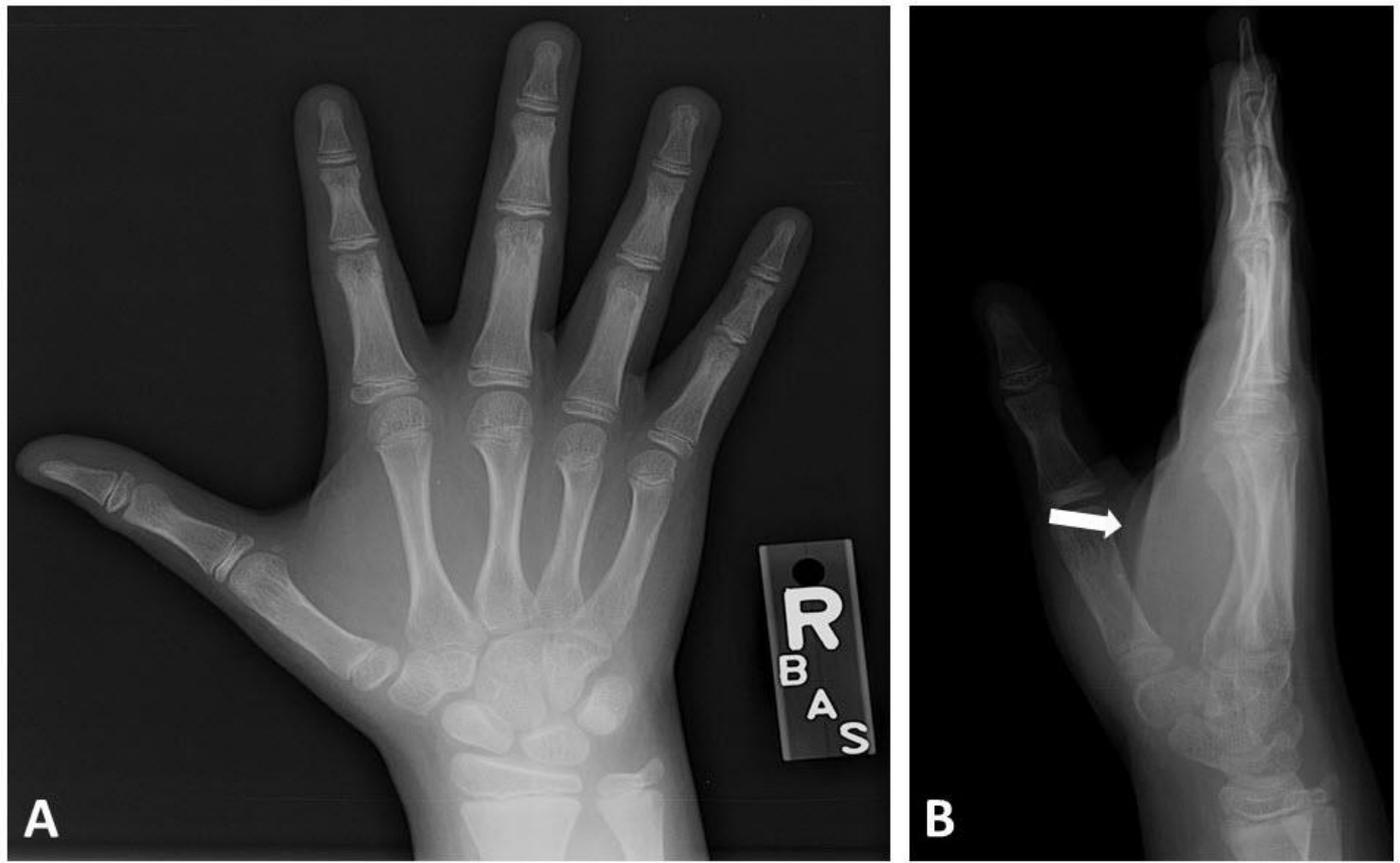


Figure 1: 8-year-old male with LFH of the median nerve.

FINDINGS: Initial frontal (A) and lateral (B) radiographs demonstrate enlargement of thenar eminence of right hand (white arrow). The actual substance of the LFH is not visible by plain radiography.

TECHNIQUE: Frontal and lateral radiographs of the right hand.

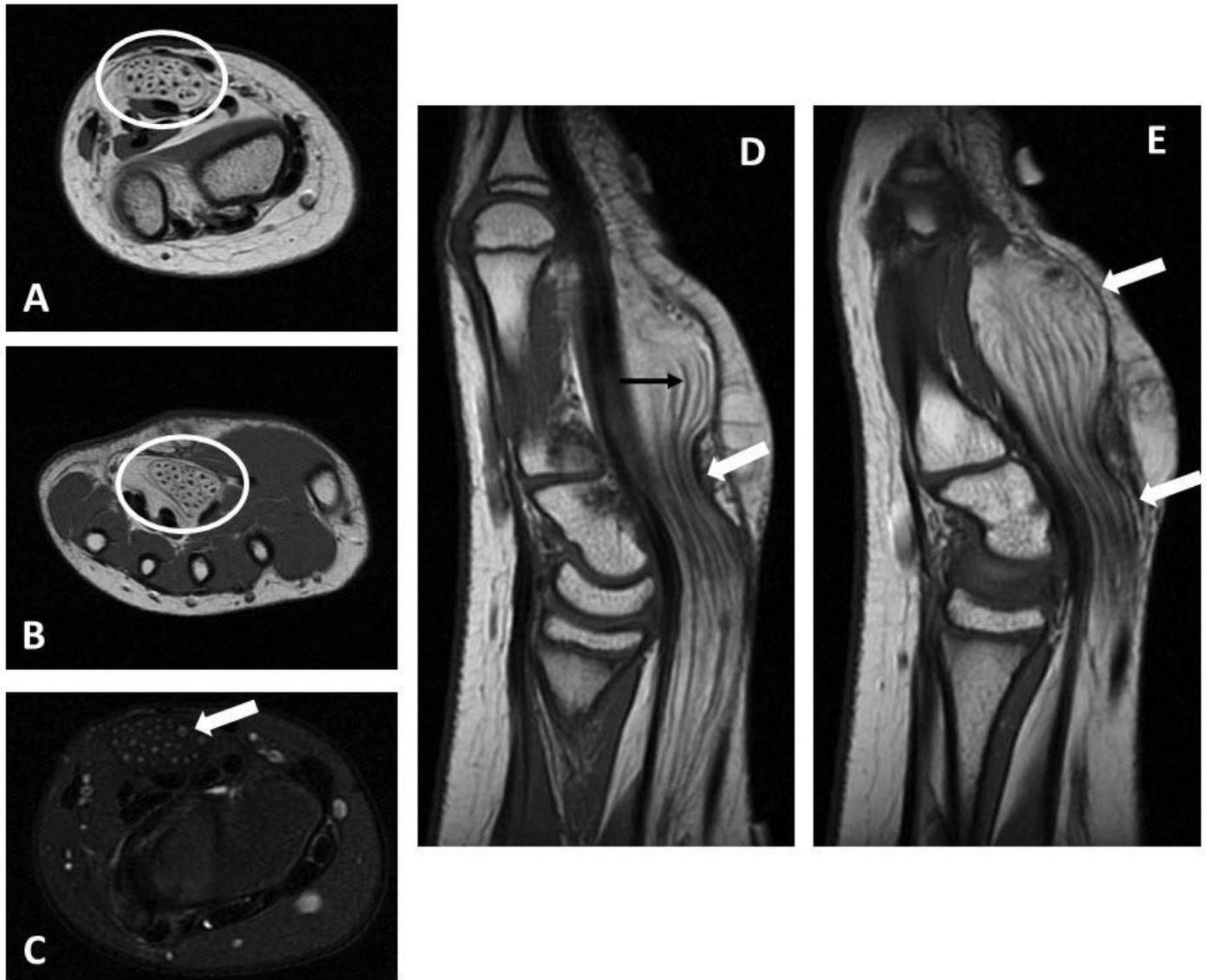


Figure 2: 8-year-old male with LFH of the median nerve.

FINDINGS: (A) and (B) are axial T1-weighted images through the distal forearm and mid-hand respectively. The intrinsically T1-hyperintense fat surrounds and separates the T1-hypointense nerve bundles; the LFH mass, which measures 1 x 2 x 12 cm in total, is circled. (C) Proton density fat-suppressed (PDFS) axial image; the fatty substance of the mass here suppresses and the nerve bundles are visible as slightly hyperintense dots (arrow). (D) Sagittal T1-weighted image shows the fatty LFH with cable-like T1-hypointense nerve bundles (black arrow). A white arrow depicts the point of compression as the mass passes through the carpal tunnel. (E) Sagittal T1-weighted image with white arrows demarcating the palmar border of the LFH.

TECHNIQUE: 3 Tesla non-contrast MR imaging of the right wrist with a quad knee coil with FOV=17 cm and slice thickness of 2 mm. T1-weighted imaging with TR=783 and TE=9. PDFS imaging with TR=3050 and TE=50.

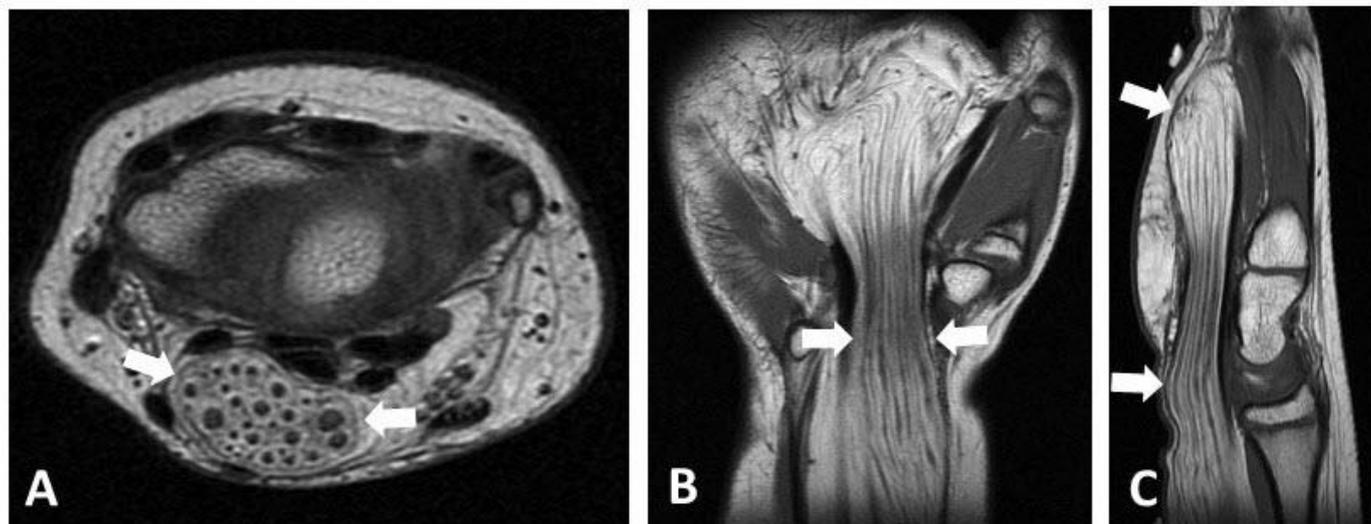


Figure 3: 8-year-old male with LFH of the median nerve.

FINDINGS: Repeat non-contrast MRI obtained 11 months later in the same patient demonstrating interval growth. (A)-(C) T1-weighted axial, coronal, and sagittal images respectively with arrows delineating the borders (white arrows) of the LFH - which is larger in cross-sectional area than on prior examination. The T1-hypointense coaxial-cable-like nerve bundles are contained within the fatty-substance of the LFH.

TECHNIQUE: 3 Tesla non-contrast MR imaging of the right wrist was performed with a quad knee coil with FOV=17 cm and slice thickness of 2 mm. T1-weighted imaging was performed with TR=783 and TE=9.

Etiology	Lipofibromatous hamartoma is a rare and benign fibro-fatty tumor that is characterized by the proliferation of mature adipocytes within the epineurium and the perineurium of the peripheral nerves. The cause LFH is unknown, but it likely has a congenital origin with the vast majority of cases occurring in the pediatric population. In general, males are slightly more affected than females. Macrodactyly predominated in females.
Incidence	The exact worldwide incidence is not well described in the literature.
Gender Ratio	Many authors have found that males are overall slightly more affected than females, while macrodactyly is more predominant in females.
Age predilection	Congenital. Symptoms usually arise in the third or fourth decade.
Risk factors	Unknown, congenital.
Treatment	The treatment modality for LFH ranges from observation in asymptomatic patients to prophylactic carpal tunnel release (CTR), biopsy and total nerve resection in symptomatic patients.
Prognosis	There is an overall good prognosis.
Findings on imaging	MRI displays serpiginous hypointense nerve fibers (often referred to as coaxial cables) interwoven with abundant T1/T2 hyperintense fatty tissue along the median nerve.

Table 1: Summary table for Lipofibromatous Hamartoma.

Differential Diagnosis	Imaging		
LFH	MRI – Serpiginous, hypointense nerve fibers (often referred to as coaxial cables) interwoven with abundant T1/T2 hyperintense fatty tissue along the median nerve		US Enlarged hypoechoic median nerve containing hypoechoic coaxial cable like bundles encased by an echogenic substratum
Ganglion cyst	MRI - Unilocular/multilocular oval fluid mass nearby a joint or tendon		US – Anechoic/hypoechoic cysts (which may be clustered/multiple) with well-defined margins with posterior acoustic enhancement
Schwannoma	MRI - T1: isointense/hypointense. T2: hyperintense. Split fat sign, target sign, and fascicular sign may be present		CT- Intrinsically isodense or hypodense. Enhancement may be homogeneous or heterogeneous enhancement depending on the size of the tumor
Neurofibroma	MRI – T1 – Hypointense rim, T2- hyperintense rim. Target sign may be present		CT – Hypodense mass
Lipoma	MRI - T1 and T2 hyperintense with complete saturation on fat-suppressed sequences	CT – Hypodense uniform fatty-attenuation mass with approximate density of negative 100 Hounsfield Units and nearly imperceptible capsule	XR – Low density mass effect
Hereditary Hypertrophic Interstitial Neuritis	MRI – Abnormal foci of high T2 signal intensity		

Table 2: Differential diagnosis for Lipofibromatous Hamartoma.

ABBREVIATIONS

CTR = carpal tunnel release
 LFH = lipofibromatous hamartoma
 MRI = magnetic resonance imaging

KEYWORDS

Lipofibromatous hamartoma; Fibrolipoma; Median nerve; Macrodactyly; Hamartoma; Carpal tunnel; Fibrolipomatous hamartoma; Lipofibroma; Magnetic resonance imaging; Microdystrophia lipomatosa; Intra-neural lipoma; Perineurolipoma; Case report

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