Case Report

10-Year Clinical Follow-Up After Decompression of Lipofibromatous Hamartoma of the Median Nerve in a 3-Year Old Patient

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10-Year Clinical Follow-Up After Decompression of Lipofibromatous Hamartoma of the Median Nerve in a 3-Year Old Patient

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Abstract: Lipofibromatous hamartoma, first reported in 1953, is a rare, slowly-progressive soft tissue tumor, characteristics of which include enlargement of the affected nerve by epineurial and perineurial proliferation of adipose and fibrous tissues. Out of previously reported 200 cases of lipofibromatous hamartoma of the median nerve, there have been approximately 40 pediatric cases under the age of 18. Herein, we report a case of lipofibromatous hamatoma of the median nerve in a 3-year-old female patient, who was surgically decompressed by carpal tunnel release only and histologically confirmed. The patient was followed-up on outpatient clinic basis annually along with ultrasonography, and the postoperative 10th year follow-up did not show recurrence or any deficits in motor and sensory functions

Keywords: lipofibromatous hamartoma; pediatric carpal tunnel syndrome; median nerve; carpal tunnel release

1. Introduction

Lipofibromatous hamartoma (LFH), first reported in 1953, is a rare, slowly-progressive soft tissue tumor, characteristic of the enlargement of the affected nerve by epineurial and perineurial proliferation of adipose and fibrous tissues.[1,2] Median nerve is the mostly commonly reported nerve of LFH, but there have been previous literatures of LFH occurring on other nerves, such as ulnar, radial, and peroneal nerves. Primary clinical manifestations are an enlarging mass and its nerve compression symptoms when it is large enough. The most common (approximately 1/3) associated anomaly has been reported to be macrodactyly, a condition of which is referred as macrodystrophia lipomatosa.[3] However, the clinical presentations of LFH vary from asymptomatic to characteristic symptoms of compressive neuropathy.

While carpal tunnel syndrome (CTS) is the most common compressive neuropathy in adult population, but it is reported relatively rare in pediatric populations. While CTS is idiopathic in nature in adults, pediatric CTS are mostly secondary to anatomic variations, trauma, or congenital malformations.[4] Second to mucopolsaccharidoses, the primary metabolic disorder causing pediatric CTS, LFH is considered a frequent cause to CTS. The two most discussed etiologies of LFH are repetitive microtrauma from the transverse carpal ligament to the median nerve and congenital anomaly.[5] Differential diagnoses vary from benign to malignant soft tissue tumors, such as ganglion cysts, neurofibroma, schwannoma, and malignant peripheral nerve sheath tumors.[2]

Since the first introduction of LFH in 1954, there have been approximately 200 cases of LFH of the median nerve, and pediatric cases.[6] There have been about 25 cases of pediatric LFH occurred under age of 18.[6,12,13,16–33] However, even though most previous literatures depend on case reports or a small case series with short-term follow-up periods, some reported progressive declines in sensory and motor functions of the affected nerve in a long-term follow-ups of adult patients.[8] Herein, we report a case of a 3-year-old female patient with LFH on the left median nerve, who was

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surgically decompressed via transverse carpal release with epineurolysis and was followed-up annually with ultrasonography assessments until the postoperative 10th year.

2. Case Description

A three-year old female patient without underlying diseases visited the outpatient clinic at our institution with chief complaints of an enlarging mass and discomfort on volar aspect of the left wrist (Figure 1). The mother recalled no history of trauma or congenital anomalies but a progressive enlarging solitary mass on the wrist for the past two weeks with mild pain in the beginning. The patient was otherwise unremarkable in medical conditions without any medications, and his immunizations were up to date.

Figure 1. Clinical image of enlarged volar mass on the left wrist taken at the first visit to the outpatient clinic. No thenar atophy is noted.

Physical examination revealed a solitary, soft, mildly tender, palpable mass sizing approximately 1 × 1 centimeters on the center of the volar aspect of the wrist. No limitation in the wrist and finger ranges of motions, but the patient's agitation escalated with passive movement of the wrist joint. For the pediatric patient with difficulty in expressive language yet, it was difficult to subjectively assess Tinel's sign, Phalen's test, or specific neuromuscular deficits, but no thenar atophies were noted (Figure 1). Gross inspection did not indicate any congenital abnormalities, such as polydactyly, or café-au-lait spots.

Ultrasonographic examination on the first visit to the clinic showed severely edematous and hypertrophied median nerve from the left wrist level down to the palm with low-echogenic intraneural multiple fascicles (Figure 2). In addition, subsequent enhanced magnetic resonance imaging of the left wrist indicated fusiform enlargement the median nerve from the left forearm distal 1/3 area down to the left hand metacarpal area along with characteristic findings of longitudinally-oriented cable-like appearances of the tumor in addition to its invasion into flexor retinaculum Figure 3).

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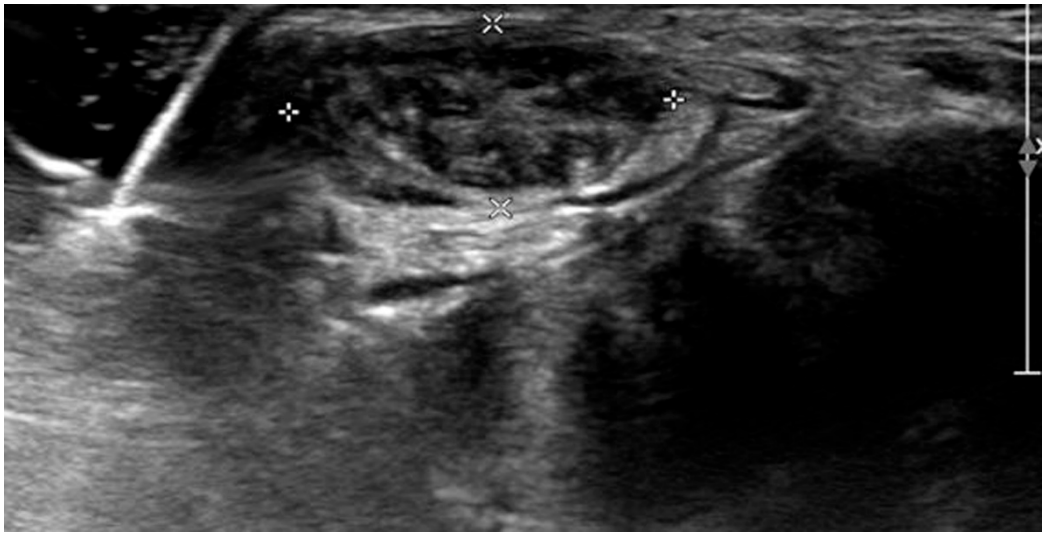

Figure 2. Wrist ultrasonography shows thickened and hypoechoic intraneural fascicles (white crosses).

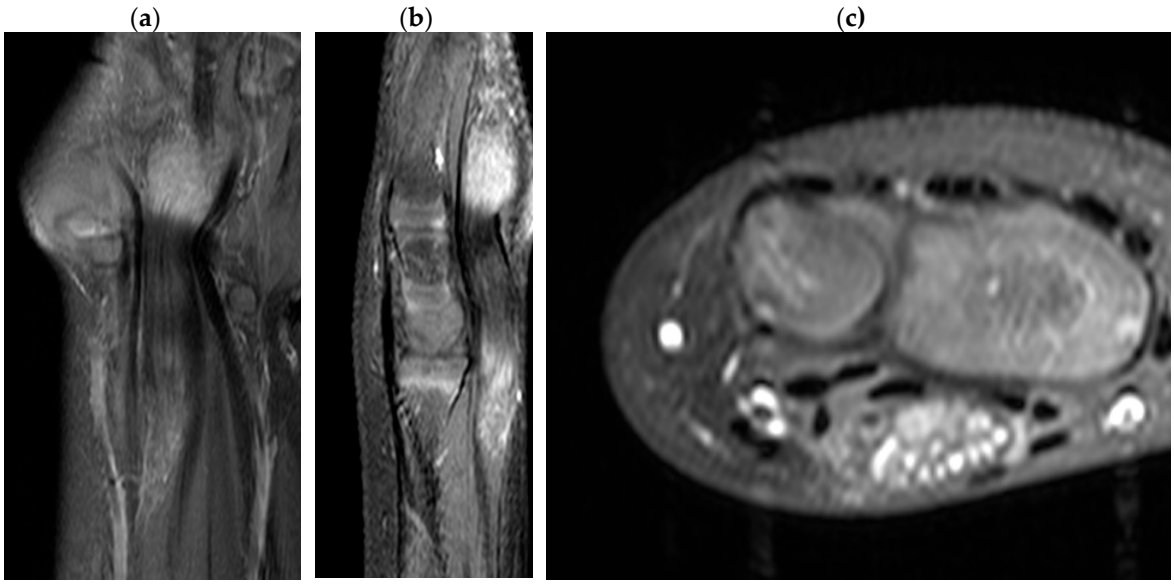

Figure 3. T1-Magnetic resonance imaging of the lipofibromatous harmatoma in the median nerve. (a) coronal view (b) sagittal view (c) axial view.

Upon the clinical and radiologic diagnosis of lipofibromatous harmartoma, the patient was scheduled for the decompression of median nerve with a biopsy. A zig-zag incision on the volar aspect of wrist was made from 10 centimeters proximal from the transverse carpal ligament down to metacarpal joints area. Surgical exploration found a fusiform enlargement starting from 2 centimeters proximal to transverse carpal ligament to the level of thenar crease, surrounding the transverse carpal ligament and the median nerve (Figure 4). After the release of transverse carpal ligament, longitudinal epineurolysis along the median nerve revealed abundant fibro-adipose tissue embedded in the nerve fascicles of the median nerve with severe adhesion. Hypertrophied epineurium was partially resected, and fibro-fatty tissue surrounding the epineurium were biopsied (Figure 4).

(a) (b) (c)

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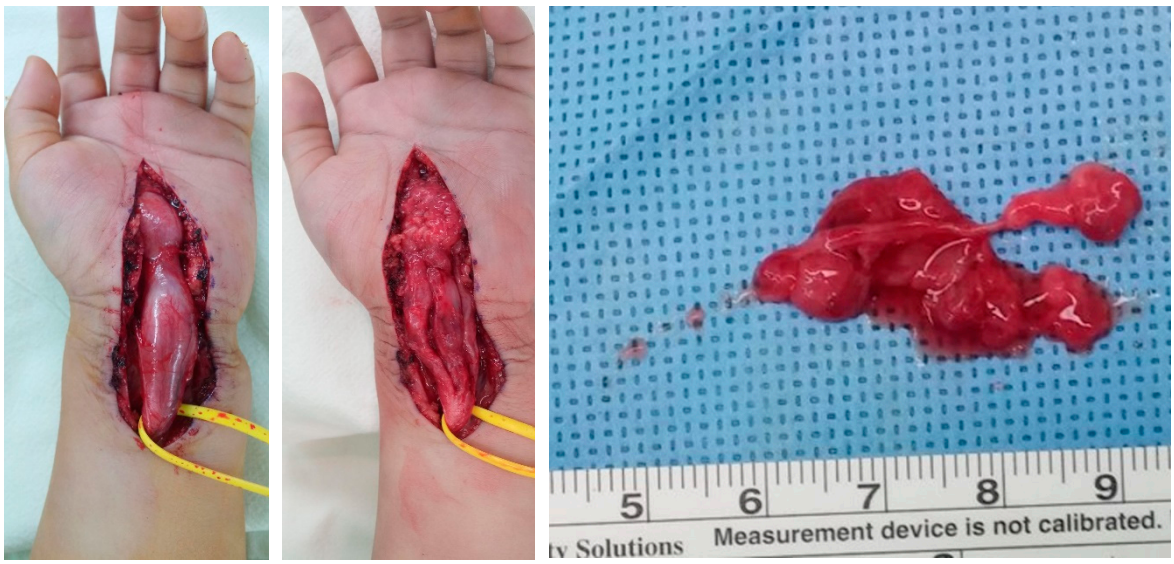

Figure 4. Intraoperative clinical images. (a) Enlarged median nerve with fibro-adipose tissue proliferation (b) Decompressed median nerve with carpal tunnel release and epineurolysis and the partial excision of the mass for biopsy (c) Biopsied specimen from the fibro-fatty soft tissue mass.

Histologic findings showed abundant thickened nerve bundles and accumulated fibrous and adipose tissues around epineurium, and it confirmed the diagnosis of lipofibromatous harmartoma in the median nerve (Figure 5).

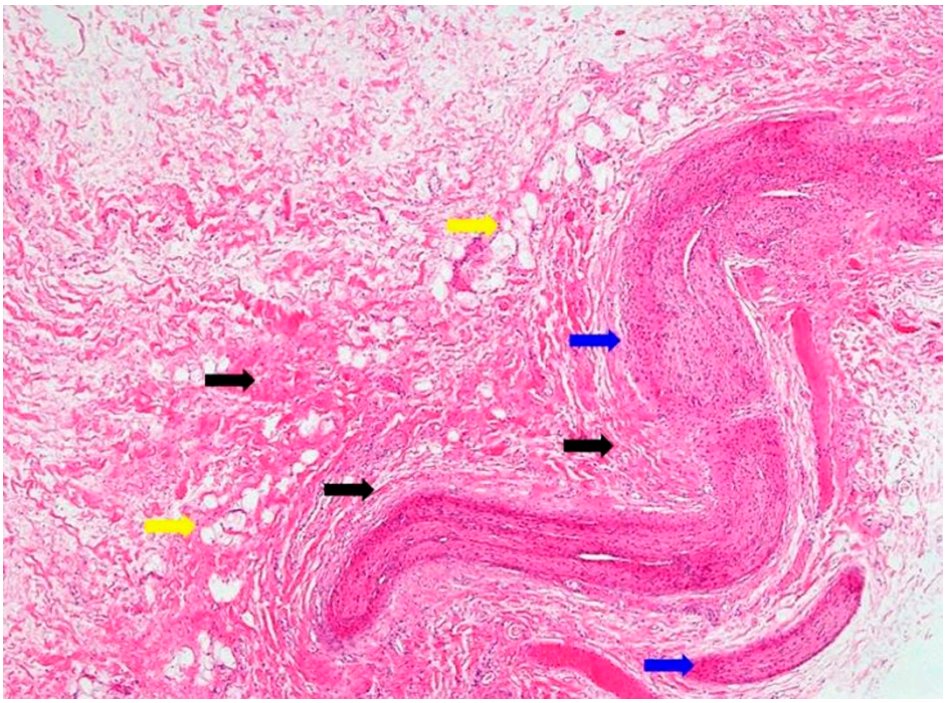

Figure 5. Characteristic histologic findings in lipofibromatous hamartoma (H&E stain, magnification x40). The mass is composed of thickened nerve bundle (blue arrow), surrounded by accumulated fibrous tissue (collagen bundle, black arrow) and adipose tissue (yellow arrow).

Postoperatively, the patient's recovery was uneventful without any complications of pain, discomfort, or decreased joint movement. The patient was followed-up annually with the wrist sonographic examination (Figure 6). At the 10th-year follow-up, the patient remained asymptomatic with full opposition function and grip strengths without sensory deficits or further enlargement of

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the hamartoma in the median nerve, and electromyography and nerve conduction study confirmed no neurologic deficits.

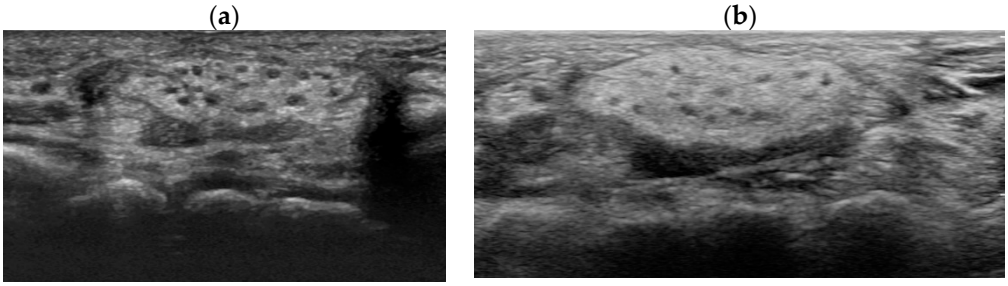

Figure 6. Ultrasonographic follow-up of left wrist at the postoperative (a) 5th and (b) 10th years.

3. Discussion

Since the first introduction in 1953, LFH is a slowly growing soft tissue tumor, which has been reported in 25 pediatric cases of median nerve involvement (Table 1). Its characteristic clinical manifestations involving median nerve show pain, paresthesia (i.e. numbness, tinging, pin/needles), and motor deficits. Neurologic deficits, if present, are mostly irresponsive to conservative management and indicated for surgical management.[2]

Table 1. Pubmed/MedLine Literature Review of Lipofibromatous Hamartoma of the Median Nerve in Pediatric Population (age under 18).

	Publication	Age (Year)	Gender	Laterality	Macrodactyly	Treatment	Follow-up
1	1987[16]	4	M	L		Biopsy	-
2	1987[17]	14	F	L		CTR & Partial debulking	-
3	1991[18]	3	M	L		CTR	-
4	1998[19]	3	M	L		CTR & Excision	1Y
5	2000[20]	3	M	L		CTR & Partial excision	-
6	2006[12]	16	M	R	3rd digit	CTR, Debulking, Nerve graft	4Y (Recur)
7	2006[12]	9	F	R		CTR & Partial excision	10M
8	2006[21]	14	M	L		CTR, Segmental Excision, Nerve Graft	3Y
9	2008[22]	5	F	R	3rd, 4th digits	Stripping of nerve	
10	2008[23]	4	F	L		-	-
11	2009[24]	8	F	R	2nd digit	CTR & Partial excision	8Y
12	2009[25]	8	M	R		CTR & Partial excision	-
13	2009[26]	15	F	R		Observation	-
14	2009[26]	18	F	R		Observation	-
15	2012[27]	17	M	R		CTR & Biopsy	6M
16	2012[27]	15	M	R		CTR & Biopsy	4Y
17	2014[28]	3	M	Both		CTR & Biopsy	
18	2016[6]	8	M	R		Observation	1Y

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19	2017[13]	3	M	R		CTR, Debulking, Nerve graft	1Y
20	2018[29]	18	F	R		CTR	-
21	2018[29]	16	F	R	Thumb	CTR & Intraneural dissection	-
22	2018[30]	10	F	R		CTR	2M
23	2018[31]	15	M	R		CTR & Partial excision	3Y
24	2019[32]	13	F	L		CTR	2Y9M
25	2023[33]	6	F	L		CTR & Microsurgical interfascicular dissection	1Y
26	**Case**	3	F	L		CTR & Biopsy	10Y3M

M: Male, F: Female, R: Right, L: Left, CTR: Carpal tunnel release, Y: Year(s), M: Month(s).

In the diagnosis of LFH, MRI plays a critical role by offering pathognomic radiologic features, low-intensity serpentine nerve fibers embedded in abundant high-intensity adipose and fibrous tissues, also known as "cable-like appearances".[9] In addition, ultrasonography also serves an essential tool for initial diagnosis and postoperative non-invasive radiologic follow-ups. Ultrasound imaging of LFH is characteristic of the enlarged cross-section of the affected nerve with hypoechoic fascicles embedded in the hyperechoic fibrous and adipose tissues, and recent ultrasonography has shown to provide as equivalent diagnostic evidence as MRIs.[10] While MRIs are beneficial in assessing the extent of the lesion in the initial diagnosis, ultrasonography serves a critical role in serial postoperative follow-ups, as seen in the current case description. However, biopsy and histologic examinations are the only definitive measures for the diagnosis of LFH, which are characteristic of intertwining collagen, fibroblasts, and adipose cells separating nerve fascicles and infiltrating the space between the epineurium and perineurium without inflammation or myelin degeneration.[2]

In treatment of LFH, there is no standard treatment consensus, and the patient management varies depending on the extent of the soft tissue lesion. Historically, the lipofibromatous lesions were complete excised surgically with devastating sequelae of motor and sensory deficits. However, the current treatment approach has been shifted toward more conservative. Patients with asymptomatic swelling without severe neurologic deficits are frequently closely observed, but surgical treatments for the patients with motor and sensory impairment aim to symptomatic relief from compressive neuropathy without invasive debulking. There have been several previous literatures on techniques for nerve dissections, but their postoperative functions are frequently disappointing.[8,11] In addition, nerve grafting after debulking and interaneural dissection have shown some positive results, but their results only showed short clinical follow-ups.[12,13] Consequently, tumor excision is generally reserved for the cases with progressive and recurring pain and neurologic deficits even after carpal tunnel release and nerve decompression.[14] An interesting previous literature on Martin-Gruber anastomosis, a neural anastomosis between median and ulnar nerves, showed preserved hand and digital functions following radical excision of the main trunk of the median nerve.[15]

It is critical to consider patient's history, physical examination, and radiologic assessment all together before the decision on surgical treatment. As seen in the current case, when the patient is in the toddler age and incapable of expressive language, clinical assessment of certain signs and symptoms are often limited and challenging even in the postoperative follow-ups. Furthermore, due to its nature of rare entity, it is in reality difficult to compare among various surgical techniques for their effectiveness with a long-term follow-up under well-structured study design. Currently available literature on pediatric LFH lacks long-term follow-up outcomes, and they are mostly followed-up for less than a few years. Therefore, the current case report provides valuable clinical and radiologic outcomes without tumor or symptom recurrences through annual follow-ups

consecutively for 10 years. For the 10 years of postoperative period, the patient was able to maintain intact motor and sensory functions without any pain or discomfort after the nerve decompression with the carpal tunnel release and epineurolysis of the median nerve.

4. Conclusions

Due to its rarity of LFH, the precise understandings of epidemiology, treatment, and prognosis are limited in the previous literature. The current case of LFH of the median nerve is the first to be reported in South Korea, and the high index of clinical suspicion for LFH is imperative especially in pediatric carpal tunnel syndrome. Even though very little is known about the predictive prognostic values of surgical techniques, 10-year consecutive follow-up of 3-year-old patient with LFH showed satisfactory outcomes after decompression with carpal tunnel release and epineurolysis. Furthermore, increased multidisciplinary clinical awareness of the tumorous condition is critical for best practice management

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