

## Lipofibromatous Hamartoma of the Median Nerve with Long-term Follow-up

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Lipofibromatous hamartoma is a rare, benign tumor that most often involves the median nerve. A 16-year-old male with lipofibromatous hamartoma of the median nerve at the wrist level is described. This patient was a child when the mass was first noted. Although there were no symptoms or signs of carpal tunnel compression, the growth of the tumor was progressing. In addition to the release of the carpal tunnel, microsurgical intraneural dissection was done to preserve the thenar motor branch. Then segmental excision of the residual sensory component with sural nerve grafting was performed. Subjectively the patient did not notice the minor motor deficit, however, the patient did experience numbness of fingertips after surgery. There were no scars or trophic ulcers on fingertips at 3 years of follow-up regardless of the inadequate sensory return. Treatment of this benign tumor is still controversial. The relevant reports in the literature are reviewed. (*Chang Gung Med J* 2005;28:111-6)

**Key words:** lipofibromatous hamartoma, median nerve.

Lipofibromatous hamartoma of a nerve is a rare soft tissue tumor. It is a gross enlargement of the nerve caused by large proportions of fat and fibrous tissue, with epineurial and perineurial proliferation. The tumor most commonly affects the median nerve and its branches.<sup>(1)</sup> However, the lesion has also occurred in the ulnar, radial, plantar and peroneal nerve.<sup>(2)</sup> It is usually found at birth or during infancy, and most cases occur before adulthood.<sup>(1)</sup>

The tumor usually grows slowly and presents as asymptomatic swelling. If the lesion at the wrist is large enough, it may cause median nerve compression. A relationship between lipofibromatous hamartoma and macrodactyly has been reported.<sup>(3-5)</sup> Treatment of the benign tumor has been controversial. Some authors have recommended excision of the involved nerve with or without grafting;<sup>(5)</sup> some have recommended microsurgical debulking of the fibrofatty sheath; and others have recommended

minimal treatment with the carpal tunnel release.<sup>(6)</sup> The varying reports in the literature may be confusing to a clinician when deciding which option is the best, especially when the surgical expertise and facilities are not available to treat nerve tumors.

### CASE REPORT

A 16-year-old male presented with a single longitudinal oriented soft tissue mass on the central region of the right proximal palm and distal forearm. The patient had no complaints of any disturbance of sensory or motor functions. The mass had been noticed when the patient was 6 years old. The mass had continued to grow gradually. However the mother did not bring the patient to the clinic until he was 10 years of age owing to lack of symptoms.

On physical examination, the tumor was elastic, soft, and non-tender. No evidence of macrodactyly,

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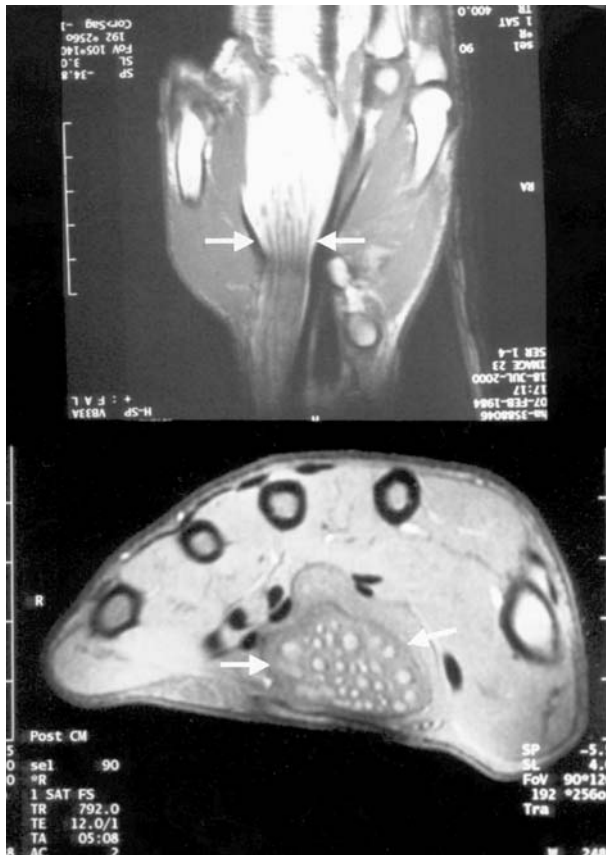
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muscle atrophy or neurofibromatosis was noted. The results of baseline laboratory tests were within normal ranges. Plain X-ray showed no particular findings. The clinical appearance was suggestive of lipoma of the right wrist. However, the magnetic resonance imaging (MRI) revealed diffuse lipomatous infiltration inside the nerve sheath of median nerve resulting in diffuse swelling of the median nerve. No isolated tumor masses were found but diffuse fatty tissue was wrapped around the nerve fascicles (Fig. 1). The impression of the MRI was lipofibromatous hamartoma. Recommendation of surgical excision was given to his parents.

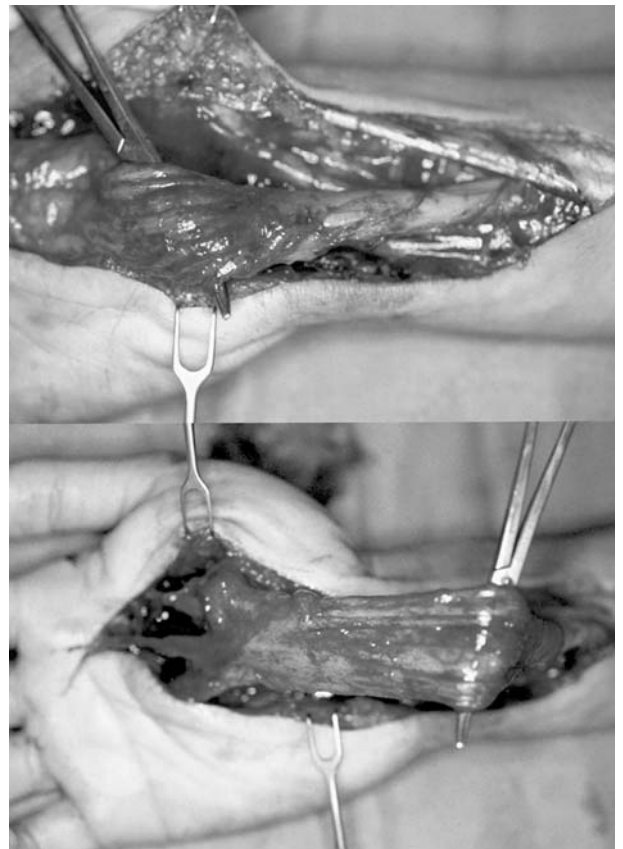
Upon surgical exploration, a 13x2x2 cm fusiform enlarged median nerve with fibrofatty tissue

proliferation involving the neural sheath was observed extending from the distal forearm to the mid palm (Fig. 2). No discrete masses were found in the extraneural or intraneural areas. Release of the carpal tunnel and epineurectomy were done. We identified the thenar motor branch near the neuromuscular junction distally, followed by retrograde microsurgical intraneural dissection to keep the motor element intact. Thirteen cm of the residual sensory part was excised. Using left sural nerve, the resulting 13 cm gap was bridged using interfascicular nerve grafts from the end of the median nerve to its distal branches. One strand of graft was sutured to the branches to the thumb, long finger and ring finger. The other one was connected with the branch to index finger.

Histological examination exhibited nerve bundles in an expansive fibrofatty stroma surrounded by



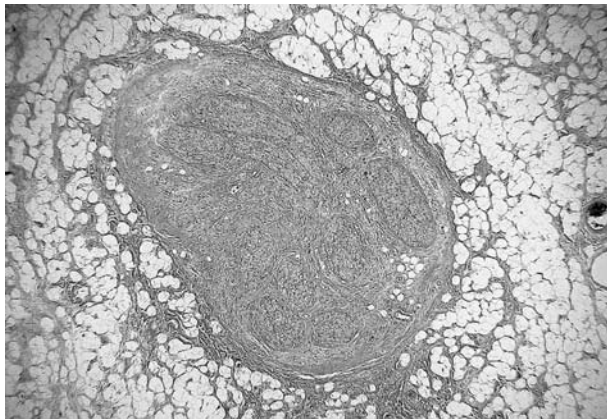
**Fig. 1** (Above) Coronal T1-weighted MRI (TR 400/TE 12) of the wrist showed serpiginous low-intensity structure representing thickened nerve fascicles (arrows) surrounded by evenly distributed fat. (Below) Axial T1-weighted MRI of the wrist revealed diffuse lipomatous infiltration inside the nerve sheath of the median nerve (arrows) resulting in coaxial-cable-like appearance.



**Fig. 2** The enlarged fusiform median nerve with fibrofatty tissue proliferation extended from the distal forearm to mid palm.

epineurium. Fibroadipose tissue overgrowth was also seen outside the perineurium (Fig. 3). The diagnosis of lipofibromatous hamartoma was confirmed.

The patient is presently 19 years of age. Three years after the surgery, the hand reveals complete

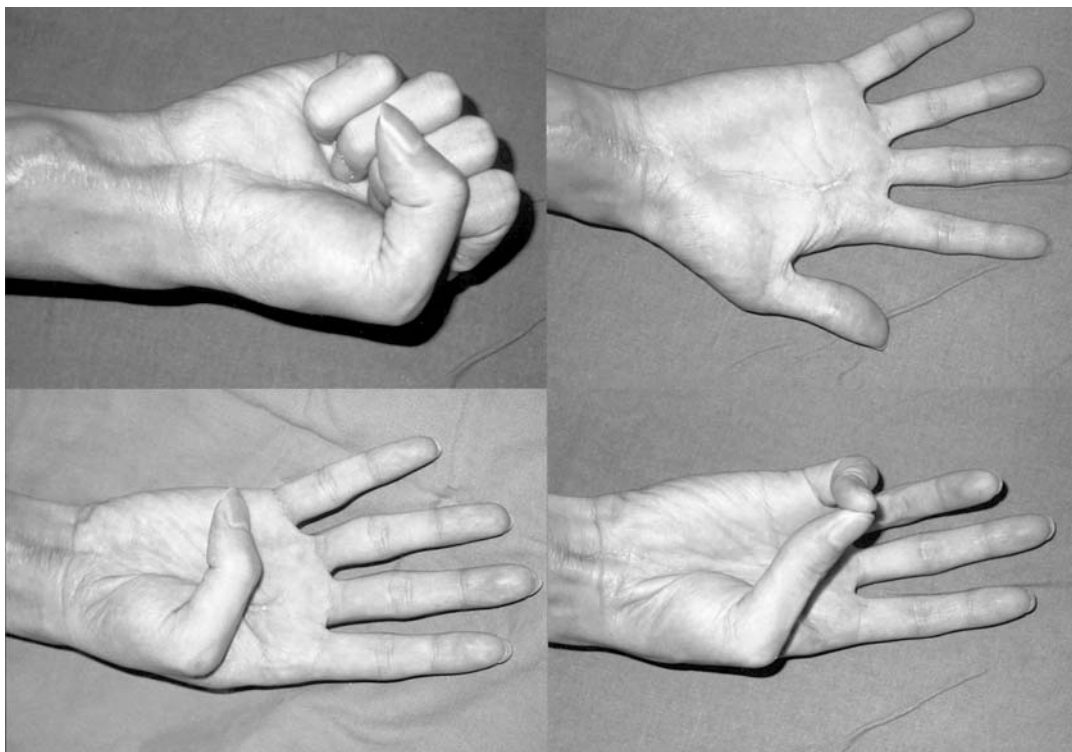


**Fig. 3** Microscopic section showing fibroadipose tissue overgrowth outside a nerve fascicle. Onion bulb-like hypertrophic change due to an increase of perineurial cells. (Original magnification x 200)

range of motion of all the fingers. The patient has acceptable opposition function and grip strength. The tip of the thumb can reach the distal palmar crease at the base of the little finger (Fig. 4). It meets the final stage (stage 10) of Kapandji's method for clinical evaluation of thumb opposition.<sup>(7)</sup> Grip strength was 100% and power pinch strength was 80% of that of the uninvolved side. Sensory recovery of the hand was incomplete on the fingertips of the thumb, index and long fingers of right hand; static two-point discrimination (2PD) was greater than 15 mm. Detailed sensibility of the hand is recorded in Table I. The static 2PD was 4 mm on the fingertips of the uninvolved side. There were no scars or trophic ulcers on fingertips despite the inadequate sensory return 3 years postoperatively. The patient uses the hand well in his job as a radiology technologist.

## DISCUSSION

Lipofibromatous hamartoma of the nerve represents a very uncommon benign tumor that typically



**Fig. 4** (Above) Flexion and extension of the hand 3 years after surgery. (Below) Thumb can reach the distal palmar crease at the base of the little finger; thumb and little finger opposition 3 years after surgery.

involves the median nerve. Involvement of the nerves of the lower extremities is very rare. Silverman and Enzinger reported 26 cases of lipofibromatous hamartoma of the nerve, 25 cases involved the hand, wrist, palm, and finger, only one case involved the foot. In 22 of the 26 cases, the lesions involved were in the distribution of the median nerve.<sup>(3)</sup> Amadio et al. also reviewed 17 cases of lipofibromatous hamartoma of the nerve and four cases were located in the lower extremities. Eleven cases were in the median nerve or its branches.<sup>(5)</sup> Involvement of the digital nerve, ulnar nerve, peroneal nerve and medial plantar nerve have also been reported in the literature.<sup>(2,6,8)</sup> Lipofibromatous hamartoma may be associated with the rare congenital disorders (Proteus syndrome) or Klippel-Trenaunay-Weber syndrome.<sup>(5,9)</sup> Very rarely bilateral involvement or the presence of metaplastic bone has been reported.<sup>(10,11)</sup>

A lipofibromatous hamartoma affecting the median nerve typically presents in children or young adults as a palmar enlargement. It may be asymptomatic or lead to compression neuropathy with the primary complaint of severe pain, sensory changes and thenar motor weakness.<sup>(3)</sup> The tumor is generally considered to be congenital in origin. However, some believe it to be a hamartoma incited to grow by nerve irritation or inflammation.<sup>(12)</sup>

Lipofibromatous hamartoma of a nerve typically appears as a well-encapsulated, pale, yellow-white fusiform enlargement of the nerve, which is not adherent to surrounding tissue. Microscopic examination reveals the epineurium expanded by adipose tissue and delicate confines of the epineurium. Onion bulb-like hypertrophic changes due to an increase of perineurial cells have also been seen. Associated endoneurial fibrosis may be present.<sup>(1)</sup> An association between lipofibromatous hamartoma and macrodactyly has been reported but our patient did not present with macrodactyly. Fibrofatty overgrowth of the nerve, in the absence of macrodactyly, has not been reported to involve nerves other than the median nerve.<sup>(3,5)</sup>

Marom and Helms revealed that MR imaging characteristics of lipofibromatous hamartoma are pathognomonic and may obviate the need of biopsy for diagnosis.<sup>(12)</sup> The imaging features are coaxial-cable-like in appearance on axial planes and spaghetti-like in appearance on coronal planes. Serpiginous

low-intensity structures representing thickened nerve fascicles surrounded by evenly distributed fat, high signal intensity on T1-weighted sequences (Fig. 1) and low signal intensity on T2-weighted sequences. The specific findings were not seen in any other type of median nerve abnormality images so far.<sup>(12,13)</sup> However, in the present case, the final diagnosis was confirmed using results of biopsy as suggested by a number of other authors.<sup>(1)</sup>

Clinically, the differential diagnostic consideration includes intraneural lipoma, diffuse lipomatosis, traumatic neuroma, fibromatosis, hemangioma, plexiform neurofibroma and ganglion cyst. It is not difficult for the differentiation because of the particular gross and histological findings.<sup>(2)</sup> Since the lipofibromatous hamartoma also has the unique magnetic resonance appearance, which is helpful to exclude other diagnoses.

Various surgical procedures have been used in the treatment of lipofibromatous hamartoma of the median nerve at the wrist. The recommendations include carpal tunnel release with or without superficial removal of epineurial proliferation, microsurgical interfascicular dissection, excision of the involved nerve segment with or without nerve grafting.<sup>(5,6)</sup> Warhold et al. reported that the patients treated with carpal tunnel decompression had decreases in the tumor sizes, alleviation of pain and improvement in opposition strength. The report also showed severe and sensory deficits with attempted surgical excision of the masses, although other authors reported minimal disability in young children, presumably due to cerebral compliance.<sup>(1)</sup> In the series of Amadio et al., all cases of nerve excision and neurolysis had abnormal two-point discrimination post-operatively even though the discrimination was normal preoperatively.<sup>(5)</sup> Due to the extensive involvement of the nerve with a large swelling, we chose microsurgical intraneural dissection in the present case to preserve thenar motor branch and wide excision of the tumor with the entrapped nerves. The

**Table 1.** Two-Point Discrimination Test of the Involved Hand

	Thumb	Index	Long	Ring (radial)	Ring (ulnar)	Little
Static(mm)	>15	>15	>15	13	4	4
Moving(mm)	>15	>15	>15	8	3	3



defect was replaced with left sural nerve grafting. At the follow-up examination 3 years after surgery, our patient still had numbness of the thumb, index and long fingers. Currently, treatment is often restricted to a limited biopsy with release of the carpal ligament. Surgeons should avoid unnecessary extensive resections sacrificing important nerve branches.<sup>(9)</sup>

From this report we concluded that when the diagnosis of lipofibromatous hamartoma is uncertain, MR imaging must be done. There is no definite therapy for lipofibromatous hamartoma. Decompression and exploratory operation with limited excision biopsy for final diagnosis is the ideal treatment. Microsurgical intraneural dissection to preserve thenar motor branch and immediate nerve grafting for the defect resulted in loss of acute sensibility in the fingertips. Surgeons should consider this complication before a more radical resection of the nerve is contemplated.

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## 正中神經脂纖維缺陷瘤及長期追蹤

許聰政 陳大正<sup>1</sup> 陳宏基

脂纖維缺陷瘤 (Lipofibromatous hamartoma) 是一種罕見的良性腫瘤，常好發於正中神經。本文報告一位16歲男性確認診斷為右腕部正中神經脂纖維缺陷瘤，其主訴於孩童時發現右腕腫塊。雖然沒有任何腕隧道壓迫的症狀，但腫瘤生長仍不斷進行。手術方式為打開腕隧道、保留支配魚際肌的運動神經枝後，切除病灶處的正中神經，並以腓神經從事神經移植。追蹤病人術後結果發現，右手運動功能良好但正中神經支配之指尖有麻木感，不過未發現有任何反覆受傷引起之疤痕或潰瘍。此良性腫瘤之治療方式仍頗多爭論，文中將整理相關論述。(長庚醫誌2005;28:111-6)

**關鍵字：**脂纖維缺陷瘤，正中神經。

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