Lipofibromatous Hamartoma of the Palmar Cutaneous Branch of the Median Nerve

Jason S. Hoellwarth¹ Robert J. Goitz¹

¹Department of Orthopaedic Surgery, University of Pittsburgh Medical Center, Pittsburgh, Pennsylvania, United States

Address for correspondence Jason S. Hoellwarth, MD, Department of Orthopaedic Surgery, University of Pittsburgh Medical Center, 3471 Fifth Avenue, Suite 911, Pittsburgh, PA 15213, United States (e-mail: hoellwarthjs@upmc.edu).

J Hand Microsurg 2018;10:109–112

Abstract

Keywords

- lipofibromatous hamartoma
- hamartoma
- palmar cutaneous branch
- median nerve
- median nerve tumor

Background Lipofibromatous hamartoma (LFH) is a benign tumor of peripheral nerves, most commonly reported in the median nerve, and often associated with carpal tunnel symptoms. There have been nearly 200 reported cases of LFH in the median nerve, but to the authors' knowledge, this 10-year-old girl represents the first case report involving the palmar cutaneous branch.

Methods An excisional biopsy of the mass was performed via a standard midline palmar incision. The tumor was found encapsulating the palmar cutaneous branch, and thus it was sacrificed.

Results The incision healed without issue, but the patient reported numbness in the palm of her hand as was expected.

Conclusion LFH is a benign tumor, although it can cause symptoms due to mass effect. If the diagnosis is uncertain or a patient is symptomatic, excisional biopsy can be performed even if involving a cutaneous nerve.

Introduction

Lipofibromatous hamartoma (LFH) is a rather uncommon tumor affecting peripheral nerves, most often reported in the median nerve. Although reports of LFH exist in other peripheral nerves, the median nerve is the most commonly reported. It was first reported in 1953 by Levenson and Mason,¹ first identified as a hamartoma by Emmett in 1965,² given its current name of LFH in 1969,³ and fewer than 200 cases have been reported arising from the median nerve.^{4,5} The tumor is most frequently found between the distal forearm and fingers, which can cause symptoms and findings consistent with carpal tunnel syndrome, but case reports exist of LFH of the median nerve in the elbow region.^{6,7} The authors report here a patient who presented with a superficial palm mass that at biopsy was found to arise from the palmar cutaneous branch of the median nerve, and upon pathologic evaluation, the patient was diagnosed with LFH. To the authors' knowledge, this is the first time LFH has been observed in the palmar cutaneous branch in any patient.

Case Report

A 10-year-old right-handed girl was referred to the clinic from her pediatrician to address a mass in her left palm, which was first noticed by the patient just over 1 year prior. She and her mother endorsed that the mass had grown during that time but denied any history of trauma, pain, or sensory changes. On examination, her left hand revealed a moderately well-circumscribed 4- × 2-cm mass in the palm from just proximal of the wrist crease to the thenar crease. The mass felt soft and spongy. Palpating the mass proximally caused the distal aspect of the mass to bulge slightly, and vice versa. She had full range of motion, normal strength, and full normal sensation to all digits. She had no macrodactyly, no tenderness, and Tinel's sign and Phalen's test were both negative. Radiographs of the hand were unremarkable. Magnetic resonance imaging (MRI) identified a mass superficial to the transverse carpal ligament in the palmar subcutaneous tissue that had a nonhomogeneous consistency (►Figs. 1 and 2). Given the uncertain diagnosis and reported increase in size in the past year, the authors recommended excisional biopsy.

received
August 6, 2017
accepted after revision
December 21, 2017
published online
April 3, 2018

©2018 Society of Indian Hand & Microsurgeons

DOI <https://doi.org/10.1055/s-0038-1626682>.
ISSN 0974-3227.

Fig. 1 Axial MRI T1 (A) and T2 (B) weighted representative image through the palm. Arrows indicate the mass.

Fig. 2 Sagittal MRI T1 (A) and T2 (B) weighted representative image through the palm. Arrows indicate the mass.

To excise the mass, a longitudinal midline incision was made through the palmar skin approximately at the site of a standard carpal tunnel release. Fibrofatty tissue immediately bulged out from the wound. The mass was well demarcated, not invasive but not encapsulated. However, the palmar cutaneous branch of the median nerve was fully encapsulated by the mass and could not be disentangled and was thus excised with the mass (→ Fig. 3). The wound was closed without issue. The pathology report described a gross appearance of white-tan tissue. Microscopic description was thickened perineurium and lobules of fibroadipose tissue in concentric pattern. Nerve bundles were interposed by adipose tissue and by fibrosis. A diagnosis of LFH was made.

The patient returned for follow-up visit 2 weeks later and had already returned to nearly full activity using her hand for daily tasks without issue. Detailed examination revealed a healing incision with numbness in the palm as expected but no other changes.

Discussion

Two recent articles provide excellent extensive reviews of the literature and discuss details of diagnosis at length.^{4,8} The authors thus focus on essential principles and their specific

Fig. 3 Clinical photograph of the excised mass. Left side of mass is proximal, right side distal.

thought process in their management of this expanding palmar mass in a child.

LFH is a rare tumor of peripheral nerves, which is characterized by an excessive infiltration of the epineurium and perineurium by fibroadipose tissue. There are no unique features obtainable based solely on history taking and physical examination. Patients often report a painless mass, usually slow growing, that may have been with them for years or since childhood. The most common age of presentation is in the 30s to 40s, but case reports of a pediatric median nerve LFH have been published.⁸⁻¹⁰ Young patients are more likely to also exhibit macrodactyly.¹¹ Symptoms are often related to the location of its presentation; when occurring in the median nerve, it may lead to dysfunction of median nerve innervated muscle, sensory impairment of the skin, and most commonly this can mimic carpal tunnel syndrome due to its mass effect. Benign masses such as a ganglion cyst, lipoma, or neurolemmoma may be high on the differential. Malignant tumors such as liposarcoma and malignant peripheral nerve sheath tumors could also be considered. Evidence of rapid growth or fixed masses may suggest a potentially malignant process. An MRI^{12,13} or ultrasound^{14,15} showing fusiform neural cables can lend confidence in the diagnosis of LFH. Indeed, axial MRI images may reveal pathognomonic serpiginous “cable-like” appearance of a peripheral nerve mass.¹⁶

The etiology and pathogenesis of this lesion remain obscure, and the use of numerous pseudonyms in the literature attests to this confusion (e.g., “intra-neural lipoma,”¹⁷ “lipofibroma,”¹⁸ and “fatty infiltration of the median nerve”¹⁹). Indeed, Johnson and Bonfiglio first introduced the term “lipofibromatous hamartoma” (LFH), and it is now considered most appropriate.³ Multiple conditions have been associated with this lesion, and a greater understanding of these associations may clarify the pathogenesis. Patil and Nagle in 2009 reviewed the literature and found that only approximately 88 cases of LFH have been reported and that 33 of these were associated with macrodactyly.²⁰ This association is well addressed in the literature and has prompted speculation that LFH may be associated with neurofibromatosis. Gigantism in patients with neurofibromatosis is associated with the development of osteochondromas, further seeding this speculation of a possible

association. Nevertheless, histologic examination of nerves from patients with neurofibromatosis does not show the fatty infiltration seen in LFH, and patients with neurofibromatosis usually present with numerous hallmarks of the disease. It has been suggested that a dysgenetic disorder similar to neurofibromatosis may ultimately be responsible for LFH.²⁰ Silverman and Enzinger have also postulated that there may be a genetic difference between LFH associated with and without macrodactyly.²¹ Finally, lipomas and vascular tumors have also been associated with LFH. Indeed, Al-Qattan suggested that an unidentified trophic factor that may cause LFH may also be responsible for the associated conditions.²² A greater understanding of all these associations may be key to revealing the pathogenesis of LFH.

Histologically, LFH shows the presence of mature fibroadipose tissue intermingled with nerve fascicles, thus expanding the epineurium. An onion bulb-like appearance is seen due to increased perineurial cells and perineurial fibrosis.²³ The architecture of the tumor makes complete excision very challenging, and the surgical management remains controversial. Indeed, nonoperative expectant management has been shown to be reasonable for patients with minimal symptoms or deficits.¹⁵

The most common treatment for symptomatic median nerve LFH is a carpal tunnel release, because most patients present with carpal tunnel syndrome secondary to mass effect.⁵ This is often combined with biopsy of the adipose mass to confirm diagnosis; excision of the tumor is not always done given its intimacy with the proper median nerve and concern for potential significant neurologic deficits. Although carpal tunnel release alone can help many patients, sometimes symptoms can recur²⁴; debulking the tumor has unpredictable outcomes with some authors reporting minimal functional sequelae and others documenting permanent neurologic deficit.²⁵⁻²⁸ In this 10-year-old girl, given the small area of pure sensory function of the palmar branch of the cutaneous nerve, the authors elected to resect the nerve to minimize the likelihood of a recurrence and accept a potential local sensory deficit that she endorsed on follow-up visit.

Conclusion

The patient presented in this study represents the first case of LFH occurring in the palmar cutaneous branch of the median nerve, based on the authors' review of literature. In this patient, although the cutaneous nerve had to be sacrificed to excise the hamartoma and some palmar numbness resulted, no functional deficit occurred. The authors would encourage future clinicians to consider LFH in the differential diagnoses for slow-growing, painless, nonmalignant presenting masses that appear to be in the subcutaneous tissue of the palm. An MRI or ultrasound is a useful imaging adjunct to assist with diagnosis. However, if doubt remains in the diagnosis or the patient experiences symptoms, excisional biopsy can be performed and most patients can anticipate an uneventful recovery.

Ethical Approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. For this type of study, formal consent is not required. The institutional review board was consulted, and no approval is necessary for single patient case reports.

Funding

None.

Conflict of Interest

None.

References

- 1 Levenson RM, Mason DG. Mixed adenoma (hamartoma) on the liver; report of a case. *Ann Intern Med* 1953;38(1):136-141
- 2 Emmett AJ. Lipomatous hamartoma of the median nerve in the palm. *Br J Plast Surg* 1965;18:208-213
- 3 Johnson RJ, Bonfiglio M. Lipofibromatous hamartoma of the median nerve. *J Bone Joint Surg Am* 1969;51(5):984-990
- 4 Tahiri Y, Xu L, Kanevsky J, Luc M. Lipofibromatous hamartoma of the median nerve: a comprehensive review and systematic approach to evaluation, diagnosis, and treatment. *J Hand Surg Am* 2013;38(10):2055-2067
- 5 Agarwal S, Haase SC. Lipofibromatous hamartoma of the median nerve. *J Hand Surg Am* 2013;38(2):392-397
- 6 Nilsson J, Sandberg K, Dahlin LB, et al. Fibrolipomatous hamartoma in the median nerve in the arm - an unusual location but with MR imaging characteristics: a case report. *J Brachial Plex Peripher Nerve Inj* 2010;5:1
- 7 Ha JF, Teh BM, Abeysuriya DTD, Luo DYW. Fibrolipomatous hamartoma of the median nerve in the elbow: a case report. *Ochsner J* 2012;12(2):152-154
- 8 Senger J-L, Classen D, Bruce G, Kanthan R. Fibrolipomatous hamartoma of the median nerve: a cause of acute bilateral carpal tunnel syndrome in a three-year-old child: a case report and comprehensive literature review. *Plast Surg (Oakv)* 2014;22(3):201-206
- 9 Sirinoglu H, Sönmez A, Sav A, Numanoglu A. Lipofibromatous hamartoma of the median nerve. *Ann Plast Surg* 2010;65(2):174-176
- 10 Frykman GK, Wood VE. Peripheral nerve hamartoma with macrodactyly in the hand: report of three cases and review of the literature. *J Hand Surg Am* 1978;3(4):307-312
- 11 Kini JR, Kini H, Rau A, Kamath J, Kini A. Lipofibromatous hamartoma of the median nerve in association with or without macrodactyly. *Turk Patoloji Derg* 2018;34(1):87-91
- 12 Marom EM, Helms CA. Fibrolipomatous hamartoma: pathognomonic on MR imaging. *Skeletal Radiol* 1999;28(5):260-264
- 13 De Maeseneer M, Jaovisidha S, Lenchik L, et al. Fibrolipomatous hamartoma: MR imaging findings. *Skeletal Radiol* 1997;26(3):155-160
- 14 Cavallaro MC, Taylor JA, Gorman JD, Haghighi P, Resnick D. Imaging findings in a patient with fibrolipomatous hamartoma of the median nerve. *Am J Roentgenol* 1993;161(4):837-838
- 15 Nardella D, Sohawon S, Carlier A. Lipofibromatous hamartoma of the median nerve. Three case reports. *J Plast Reconstr Aesthet Surg* 2009;62(9):e314-e317
- 16 Nilsson J, Sandberg K, Sørensen Nielsen N, Dahlin LB. Magnetic resonance imaging of peripheral nerve tumours in the upper extremity. *Scand J Plast Reconstr Surg Hand Surg* 2009;43(3):153-159

- 17 Haverbush TJ, Kendrick JI, Nelson CL. Intraneural lipoma of the median nerve; report of two cases. *Cleve Clin Q* 1970;37(3):145–149
- 18 Rowland SA. Lipofibroma of the median nerve in the palm. *J Bone Joint Surg Am* 1967;49(7):1309–1313
- 19 Yeoman PM. Fatty infiltration of the median nerve. *J Bone Joint Surg Br* 1964;46:737–739
- 20 Patil VS, Nagle S. Lipofibromatous hamartoma of the median nerve: a case report and review of the literature. *Indian J Plast Surg* 2009;42(1):122–125
- 21 Silverman TA, Enzinger FM. Fibrolipomatous hamartoma of nerve. A clinicopathologic analysis of 26 cases. *Am J Surg Pathol* 1985;9(1):7–14
- 22 Al-Qattan MM. Lipofibromatous hamartoma of the median nerve and its associated conditions. *J Hand Surg [Br]* 2001;26(4):368–372
- 23 Warhold LG, Urban MA, Bora FW Jr, Brooks JS, Peters SB. Lipofibromatous hamartomas of the median nerve. *J Hand Surg Am* 1993;18(6):1032–1037
- 24 Louis DS, Hankin FM, Greene TL, Dick HM. Lipofibromas of the median nerve: long-term follow-up of four cases. *J Hand Surg Am* 1985;10(3):403–408
- 25 Bains R, Kotwal A, Saeed W. Recurrent carpal tunnel syndrome in a child due to fibrolipomatous hamartoma of the median nerve successfully treated by limited excision and decompression. *J Plast Reconstr Aesthet Surg* 2006;59(12):1394–1397
- 26 Ulrich D, Ulrich F, Schroeder M, Pallua N. Lipofibromatous hamartoma of the median nerve in patients with macrodactyly: diagnosis and treatment of a rare disease causing carpal tunnel syndrome. *Arch Orthop Trauma Surg* 2009;129(9):1219–1224
- 27 Clavijo-Alvarez JA, Price M, Stofman GM. Preserved neurologic function following intraneural fascicular dissection and nerve graft for digital and median nerve lipofibromatous hamartoma. *Plast Reconstr Surg* 2010;125(3):120e–122e
- 28 Hsu C-C, Chen D-J, Chen H-C. Lipofibromatous hamartoma of the median nerve with long-term follow-up. *Chang Gung Med J* 2005;28(2):111–116