Hand & Microsurgery

Case Report
Hand Microsurg 2018;7:45-48
doi:10.5455/handmicrosurg.255406

Lipofibromatous hamartoma of the digital nerve: A case report

Emin Sir[1], Alper Aksoy[2]

ABSTRACT

Lipofibromatous hamartoma (LFH) is a rare, benign lesion of the peripheral nerves, which most frequently involves the median nerve and its digital branches (80 %). It is characterized by gradual proliferation of perineural fibroadipose tissue, which also invades epineurium and perineurium. As an outcome of invasion, the nerve fascicles separate from each other yielding a pathognomonic cable-like appearance on magnetic resonance imaging (MRI). Even though the treatment of LFH is debatable, since it causes symptoms due to nerve compression, decompression surgery, along with the reduction of fibroadipose tissue and limited excision following microsurgical dissection are recommended. We present a case of lipofibromatous hamartoma of the digital nerve.

Key words: *Lipofibromatous hamartoma,digital nerve, magnetic resonance imaging*

Introduction

Lipofibromatous hamartoma (LFH) is a rarely seen benign peripheral nerve tumor. Fatty infiltration, lipofibroma, and intraneural lipofibroma are other terminologies used to define LFH. Since it contains normal connective tissue elements, such as fat and fibrous tissue, it is called hamartoma. It is characterized by gradual proliferation of perineural fibroadipose tissue, which also invades epineurium and perineurium [1].

As an outcome of invasion, the nerve fascicles separate from each other yielding a pathognomonic cable-like appearance on magnetic resonance imaging (MRI) [2]. It frequently becomes apparent on median nerves (80%), and its distal branches [3]. It also involves radial, ulnar, sciatic, cranial, and plantar nerves [4-8]. It is associated with macrodactyly in more than one-third of the cases with median nerve involvement. As a result of its gradual growth, sensory changes, due to nerve damage, may cause pain and loss of motor functions [9]. Although its etiology is not known for sure, some authors indicated its congenital origin [10].

Even though the treatment of LFH is debatable, decompression surgery, along with reduction of the fibroadipose tissue and limited excision, following microsurgical dissection are recommended [11]. Aggressive attempts at excision may result in nerve dysfunction, which may necessitate repair with a nerve graft.

Author affiliations : *Department of Plastic and Reconstructive Surgery, [1]Izmir Training and Research Hospital, Izmir, [2]Acıbadem Konur Hospital, Bursa, Turkey*
Correspondence : *Alper Aksoy, MD, Department of Plastic and Reconstructive Surgery, Acıbadem Konur Hospital, Bursa, Turkey. e-mail: aksoya@gmail.com*
Received / Accepted : *January 10, 2017 / March 10, 2017*

Case Report

A 29-year-old male patient consulted with swelling of the radial aspect of the two digit of his left hand which became manifest nearly three years ago. He complained of incomplete closure of pulp to palm distance on his affected finger and decrease in his sensory function. His personal and family history were unremarkable.

Upon physical examination, a subcutaneous mass with soft consistency measuring nearly 3 x 1 cm was detected. A negative Tinel sign was detected. Results of the static and dynamic two-point discrimination tests were found to be 6.5 and 3.4 mm, respectively. On axial MRI, on the radial aspect of the 2. digit a hypointense lesion measuring 3 x 1 cm along the course of the digital nerve on T1 weighted fat suppression series was detected (Figure 1). On coronal plane, any pathological sign was not detected in bone, tendons, and muscular structure (Figure 2).

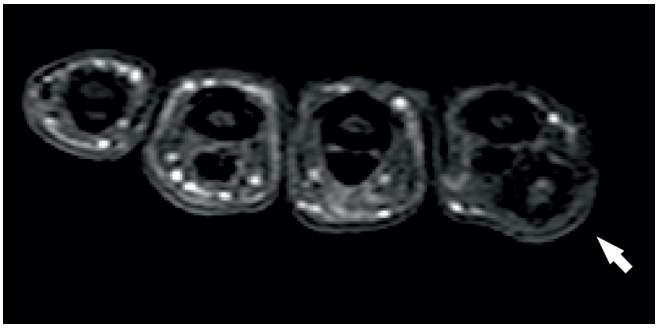

Figure 1. Axial plane MRI, a hypointense lesion digital nerve on T1 weighted fat suppression series (white arrow).

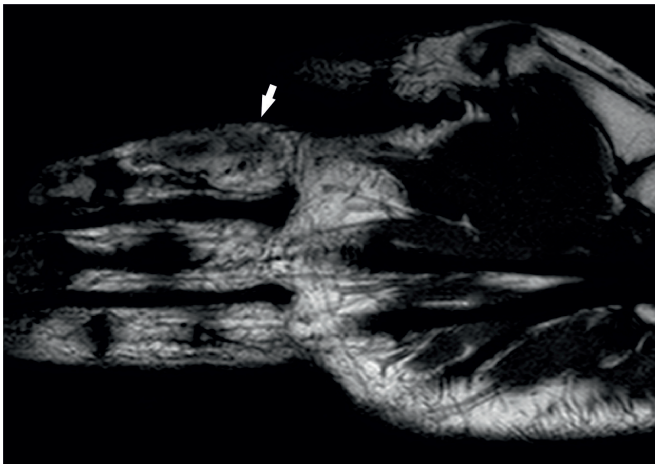

Figure 2. Coronal plane MRI, any pathological signs was not shown bone, tendons, and muscular structure (white arrow).

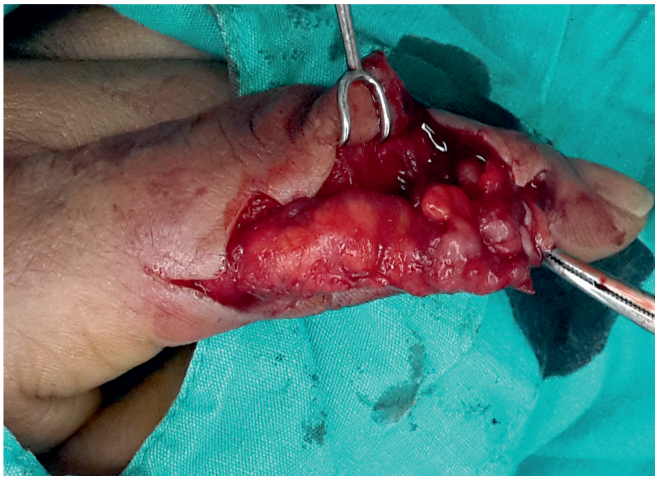

Figure 3. Intra-operative photograph showing encapsulated and hypertrophic digital nerve.

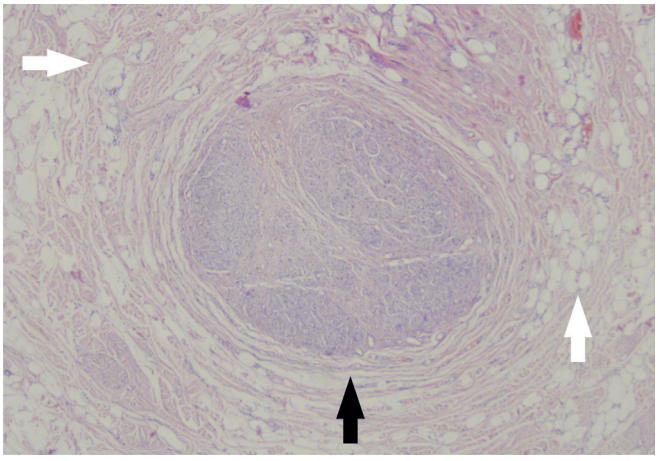

Figure 4. Histopathological examination showing fibrolipomatous tissue with few atrophic peripheral nerve fascicles (black arrow) and fibroadipose tissues (white arrows) (H&E, x200).

The patient was submitted to operation under axillary block anaesthesia. Following surgical exploration, a yellow-coloured, encapsulated, hypertrophic digital nerve extending like a cable was observed (Figure 3). The mass demonstrated extension up to the first interdigital space. Using microscopic dissection, the digital nerve fascicles were accessed. Since the mass encircled nerve branches, İntraneural fascicular dissection and limited excision was performed. Postoperative histopathological analysis of the specimen established the diagnosis of LFH (Figure 4).

The patient was followed up for nearly one year. Any recurrence and neurological damage were not

observed during the follow-up period. Preoperatively observed restriction, and numbness seen on flexion regressed.

Discussion

Lipofibromatous hamartoma of the nerves is a rarely seen benign peripheral nerve tumor progressing with proliferation of the fibroadipose tissue around branches of the affected nerve. It is generally detected as an isolated lesion and in two-thirds of the cases, it may be accompanied by macrodactyly [11]. However, the association between proliferation of the fibroadipose tissue on the periphery of the peripheral nerve, and macrodactyly has not been elucidated so far. If true macrodactyly is present in consistent with the area innervated by the affected nerve this condition is called "macrodystrophia lipomatosa" Macrodactyly can be diagnosed at birth or during early childhood. In macrodactyly, the nerves, phalanges, tendons, vessels, subcutaneous adipose tissue, nails, and skin are enlarged [12].

Morphological characteristics are the most important diagnostic tools. Even though a biopsy is sometimes recommended, care should be exerted during biopsy and biopsy material should be obtained from different points without causing any additional neurological problems. However, a definitive diagnosis cannot be established based on biopsy results [2]. Therefore, we did not prefer to perform incisional biopsy. On histopathological examination invasion of fibroadipose tissue into epineural areas, and among the branches of the affected nerve is observed. In addition, perineural, and endoneural fibrosis can be seen. In chronic lesions, metaplastic bone tissue can be seen dispersed in fibroadipose tissue. MRI is the gold standard diagnostic management of LFH.

LFH is manifested on MRI as dispersed nerve fascicles surrounded by fibromatous adipose tissue beneath the dilated nerve sheath. The nerve is thickened in a fusiform shape along its entire course. Asymmetric distribution of fat between fascicles is seen. On coronal T1-weighted images, serpiginous structures with low signal densities represent thickened nerve fascicles. These pathognomonic findings have been defined as "cable-like" on axial or spaghetti-like images on coronal planes. MRI is important in the confirmation of the diagnosis of LHF [13]. In addition, it is required for the preoperative evaluation of the extent of nerve involvement. Since digital nerves are relatively thinner, any pathognomonic image cannot be observed. Therefore, LFH of the digital nerve may pose difficulties during establishment of diagnosis based on MRI findings [14].

Treatment of LFH is controversial. It has been reported that surgery affects motor and sensory functions adversely with resultant emergence of severe neurogenic pains after resection. Asymptomatic cases, which can be detected only on MRI, should be only observed, while loss of motor and sensory functions should necessitate surgical intervention [3]. Since majority of cases involve median nerve, total excision of the lesion, and then repair with nerve graft yield worse outcomes [15]. LFM may recur in 33 - 60% if incomplete resection. [11]. Nowadays, for symptomatic LFH, primary surgical alternatives are mainly decompression of the affected nerve, preservation of the nerve fascicles following exploration using microsurgical methods, excision of fibroadipose tissue and maintenance of nerve continuity without requiring repair with a nerve graft. Therefore we preferred this method in our case.

At the end of the one year of follow-up, we did not observe any loss in motor and sensory functions.

LFH is a benign tumor of the nerves and it may remain asymptomatic for years. Therefore, its treatment is debatable, symptomatic nerves should be decompressed, and since its total excision will lead to severe neurological problems, it should be considered that preservation of neural structure with partial resection after microsurgical dissection will result in more improved outcome.

Conflict of interest statement

The authors have no conflicts of interest to declare.

References

1. Silverman TA, Enzinger FM. Fibrolipmatous hamartoma of nerve. A clinocopathologic analysis of 26 cases. Am J Surg Pathol 1985;9:7-14.
2. Nilsson J, Sandberg K, Søe Nielsen N, Dahlin LB. Magnetic resonance imaging of peripheral nerve tumours in the upper extremity. Scand J Plast Reconstr Surg Hand Surg 2009;43:153-9.
3. Warhold LG, Urban MA, Bora FW Jr, Brooks JS, Peters SB. Lipofibromatous hamartomas of the median nerve. J Hand Surg Am 1993;18:1032-7.
4. Hauck RM, Banducci DR. The natural history of a lipofibromatous hamartoma of the palm: a case report. J Hand Surg Am 1993;18:1029-31.
5. Herrick RT, Godsil RD Jr, Widener JH. Lipofibromatous hamartoma of the radial nerve: a case report. J Hand Surg Am 1980;5:211-3.
6. Gouldesbrough DR, Kinny SJ. Lipofibromatous hamartoma of the ulnar nerve of the elbow: brief report. J Bone Joint Surg Br 1989;71:331-2.
7. Chiao HC, Marks KE, Bauer TW, Pflanze W. Intraneural lipoma of the sciatic nerve. Clin Orthop 1987;221:267-71.
8. Berti E, Roncaroli F. Fibrolipomatous hamartoma of a cranial nerve. Histopathology 1994;24:391-2.
9. Al-Qattan MM. Lipofibromatous hamartoma of the median nerve and its associated conditions. J Hand Surg Br 2001;26:368-72.
10. Amadio PC, Reiman HM, Dobyns JH. Lipofibromatous hamartoma of nerve. J Hand Surg Am 1988;13:67-75.
11. 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 2010,125:120e-2e.
12. Tsuge K, Ikuta Y. Macrodactyly and fibro-fatty proliferation of the median nerve. Hiroshima J Med Science 1973;22:83-101.
13. Zeng R, Frederick-Dyer K, Ferguson NL, Lewis J, Fu Y. Fibrolipomatous hamartoma of the inferior calcaneal nerve (Baxter nerve). Skeletal Radiol 2012;41:1323-6.
14. Nanno M, Sawaizumi T, Takai S. Case of fibrolipomatous hamartoma of the digital nerve without macrodactyly. J Nippo Med Sch 2011;78: 388-92.
15. Paletta FX, Senay LC Jr. Lipofibromatous hamartoma of median nerve and ulnar nerve: Surgical treatment. Plast Reconstr Surg 1981;68:915-21.