



Fibrolipomatous Hamartoma (FLH) of Median Nerve: A Rare Case Report and Review

Rajni Ranjan¹ · Rakesh Kumar¹ · Madhan Jeyaraman¹ · Sudhir Kumar¹

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Abstract

Introduction Fibrolipomatous hamartoma (FLH) is a benign tumorous condition of adipose tissue. It is a slow growing, rare tumour involving peripheral nerves with uncertain aetiology. The clinical presentation varies as per presenting severity. The gold standard to diagnose this rare entity is MRI and its management is still controversial. In this report, we have described one such rare presentation of FLH involving Median Nerve in an adolescent male who successfully witnessed resolution of symptoms following our surgical intervention for the same.

Case presentation A 17-year-old male presented to our hospital with a history of swelling in his left hand since past 1 year. The detailed clinical evaluation revealed an irregular fusiform swelling extending from the ulnar aspect of wrist proximally to the thenar eminence along with involvement of the base of 2nd, 3rd and 4th fingers of left hand with consistency varying from soft to firm, tinell's sign positive and a terminal restriction of palmar and dorsiflexion movements. Notably, motor weakness and sensory loss were not elicited. The definitive diagnosis was established by MRI of the left hand. With due consent, the patient was undertaken for surgical intervention whereby his carpal tunnel was decompressed and biopsy of the lesion was sent for further histopathological evaluation as per the protocol. The absolute resolution in symptoms was reported by the patient following our intervention.

Conclusion A meticulous clinical and radiological correlation is required to diagnose such rare clinical entity for improvising the functional quality of life. There is no definitive treatment for lipomatosis of nerve. However, a conservative approach is commonly advocated with successful results from decompression as in our patient.

Keywords Fibrolipomatous hamartoma · Median nerve · Carpal tunnel syndrome · Tinel sign

Introduction

The benign adipose tissue tumour of peripheral nerves is extremely rare. Among all the peripheral nerves, median nerve benign adipose tissue tumour constitutes 60–80% of cases [1]. Fibrolipomatous hamartoma (FLH) of median nerve is a slow growing benign tumour with an uncertain aetiology [2]. FLH of median nerve overlaps with the clinical presentation of carpal tunnel syndrome [3]. It is associated with macrodactyly and muscular lipo-macrodystrophy along the course of the nerve [4]. The treatment of FLH is based on the individualised clinical presentation and the

functional needs of these patient. This manuscript outlines a case report of FLH of median nerve and its management in order to understand the natural course of disease with its due management.

Case Report

A 17 year old male presented at our hospital with a history of swelling in his left hand since past 1 year. Initially, it was a peanut sized swelling which progressed with gradual course of time to fill the entire palm extending from the distal forearm of wrist to the thenar eminence including the base of 2nd, 3rd and 4th fingers of left hand. He experienced on and off episodes of tingling sensation and paraesthesias over his 2nd, 3rd and 4th fingers. Notably, there was no history of trauma to his hand. Physical examination revealed an irregular fusiform swelling extending from the ulnar

✉ Madhan Jeyaraman
madhanjeyaraman@gmail.com

¹ Department of Orthopaedics, School of Medical Sciences and Research, Sharda University, Greater Noida, Uttar Pradesh 201 306, India

aspect of wrist proximally to the thenar eminence along with involvement of the base of 2nd, 3rd and 4th fingers of left hand (as shown in Fig. 1). On palpation, the swelling had a variable consistency ranging from soft over the ulnar aspect and firm over the 3rd and 4th metacarpal respectively. Slip sign was positive over ulnar aspect of the swelling. There was no elicitation of motor weakness or loss of sensations. Tinel's sign was positive whereby patient witnessed the tingling sensation in his hand along with pain and burning sensation in his arm. The movements of left wrist revealed a terminal restriction of palmar and dorsiflexion movements. No other cutaneous or soft tissue lesions were additionally noted.

Magnetic resonance imaging of the left hand revealed a gross thickening of median nerve with lipomatous tissue interdigitating between the prominent fascicles with a coaxial cable like appearance from distal forearm to distal end of the metacarpal region. The internal multiple serpiginous low T1 and T2 signal intensity structures represent thickened nerve fascicles surrounded by distributed fat of high signal intensity on T1/T2 W images with suppression on FLAIR sequence. The thickened nerve causing compression and displacement of flexor tendons and convexity of carpal tunnel retinacular sheath without any significant osseous lesions of forearm, carpals and hand (as shown in Fig. 2a–c).

Under general anaesthesia, the patient underwent surgical decompression of carpal tunnel. After extensive dissection over palm, the grossly thickened median nerve was identified as two giant fascicles adherent with fibrolipomatous structures around (as shown in Fig. 3a, b). The biopsy of fibrolipomatous structures were taken from the lesion. An utmost care was taken by not injuring the giant median nerve bundles. Patient reported absolute relief in pain after the surgical decompression of carpal tunnel without any neurological deficit.

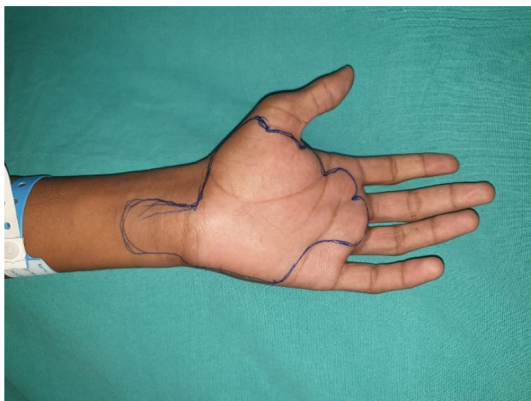


Fig. 1 Clinical image of left hand (marked area) showing the extent of the lesion

Histopathological examination of the lesion revealed a uniform pattern consisting of lobules of mature adipocytes separated by the fibrocollagenous septae. In addition, there has been expansion of epi- and perineurium due to adipose tissue (fat) deposition coupled with perineurial fibrosis. Notably, this aforementioned further warrant IHC markers assessment such as Claudin and EMA for perineurium and S100P for Schwann cells and nerve fibres respectively (as shown in Fig. 4a–c).

Discussion

Fibrolipomatous hamartoma (FLH) of a nerve has been reported as lipofibroma, lipomatosis, fibrofatty overgrowth, fatty infiltration of the nerve and neurolipoma [5]. In 1964, Morley described the clinical entity of an intraneural lipoma [6]. In 1978, classification of benign adipose tumours of peripheral nerve was given by Terzis et al. into (a) well-encapsulated intraneural lipomas, (b) diffusely infiltrating fibrofatty tumours (lipofibromatous hamartomas) and (c) macrodystrophia lipomatosa [7]. In 1994, Guthikonda et al. proposed a new system of classification for neural lipomatous tumours into (a) soft lipoma, (b) intraneural lipoma, (c) lipofibromatous hamartoma and (d) macrodystrophia lipomatosa [8].

In 2013, World Health Organization (WHO) designated fibrolipomatous hamartoma (FLH) as lipomatosis of the nerve [9]. The prevalence of fibrolipomatous hamartoma (FLH) or lipomatosis involving the peripheral nerves were reported in the median [10], radial [11], ulnar [12], brachial plexus [13], suprascapular [14], posterior tibial [15], fibular [16] and sciatic nerves [17].

Being rare tumour, fibrolipomatous hamartoma presents with a benign tumorous growth of combination of fibrous and lipomatous tissue within the nerve sheath. FLH lesion has predilection in males without macrodactyly and towards female sex with macrodactyly [18]. Due to their slow-growing nature, fibrolipomatous lesions of nerve usually present as asymptomatic or minimally symptomatic swelling with or without mild to severe symptoms based on nerve affection [19]. In our case, FLH/lipomatosis affected an adolescent male who presented with the features of carpal tunnel compression whereas the detail evaluation confirmed the involvement of unilateral median nerve without macrodactyly. The review of fibrolipomatous hamartoma of median nerve from 2012 to 2019 were tabulated in Table 1.

The etiopathogenesis of the lipomatosis of nerve lesion remains controversial. Guthikonda et al. proposed that the chronic microtrauma to the median nerve from the carpal ligament or pressure from an abnormally developing flexor retinaculum can initiate a reactive process which culminates in the development of fibrolipomatous hamartoma



Fig. 2 MRI of the left hand. **a** Coronal section and **b** sagittal section showing lipomatous tissue (shown in red arrow) around median nerve and **c** axial section showing T1/T2 W images with suppression

on FLAIR sequence showing thickened nerve fascicles surrounded by distributed fat (shown in red arrow)

[8]. Various studies suggested that an unidentified trophic factor which may cause FLH may also be responsible for the associated conditions like macrodactyly [19–26].

MRI remains the gold standard for diagnosing this lipomatosis of nerve [1, 4, 6, 29, 30]. In our case, MRI of left hand suggested gross thickening of median nerve with lipomatous tissue interdigitating between the prominent fascicles with a coaxial cable like or spaghetti-like appearance from the distal forearm to the distal end of the metacarpal region. MRI guides in deciding for planning up a surgical intervention and biopsy. Capelastegui et al. concluded that

MRI has a positive predictive value of 94% in diagnosing lipomatous lesion of median nerve [27]. Histologically, the lesion revealed a uniform pattern consisting of lobules of mature adipocytes separated by the fibrocollagenous septae with perineural elements in between which were suggestive of fibrolipomatous hamartoma.

The management of FLH lesion/lipomatosis of nerve remains challenging and controversial. Various literature suggests surgical intervention ranging from carpal tunnel decompression to radical excision of the mass lesion. Talal et al. stated that the carpal tunnel decompression

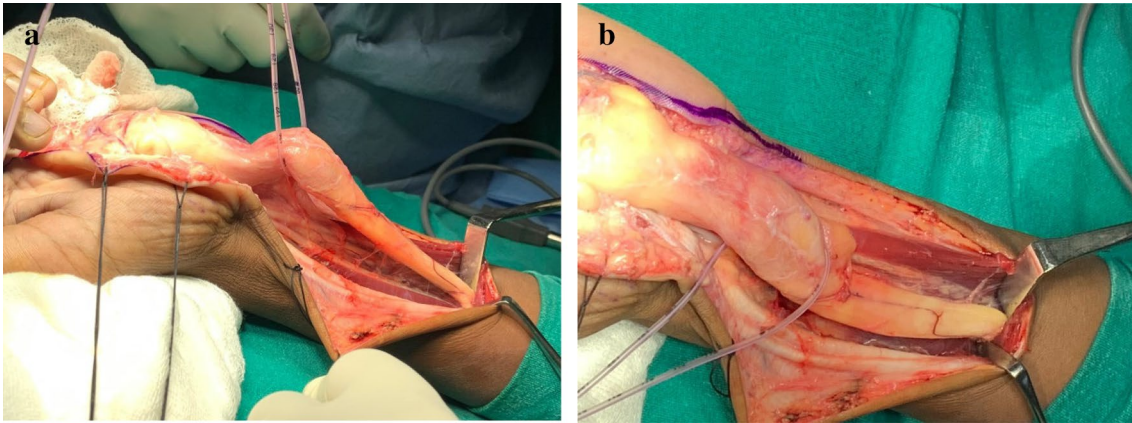


Fig. 3 a, b Intraoperative images showing the dissected giant median nerve at carpal tunnel and double fascicles of median nerve respectively

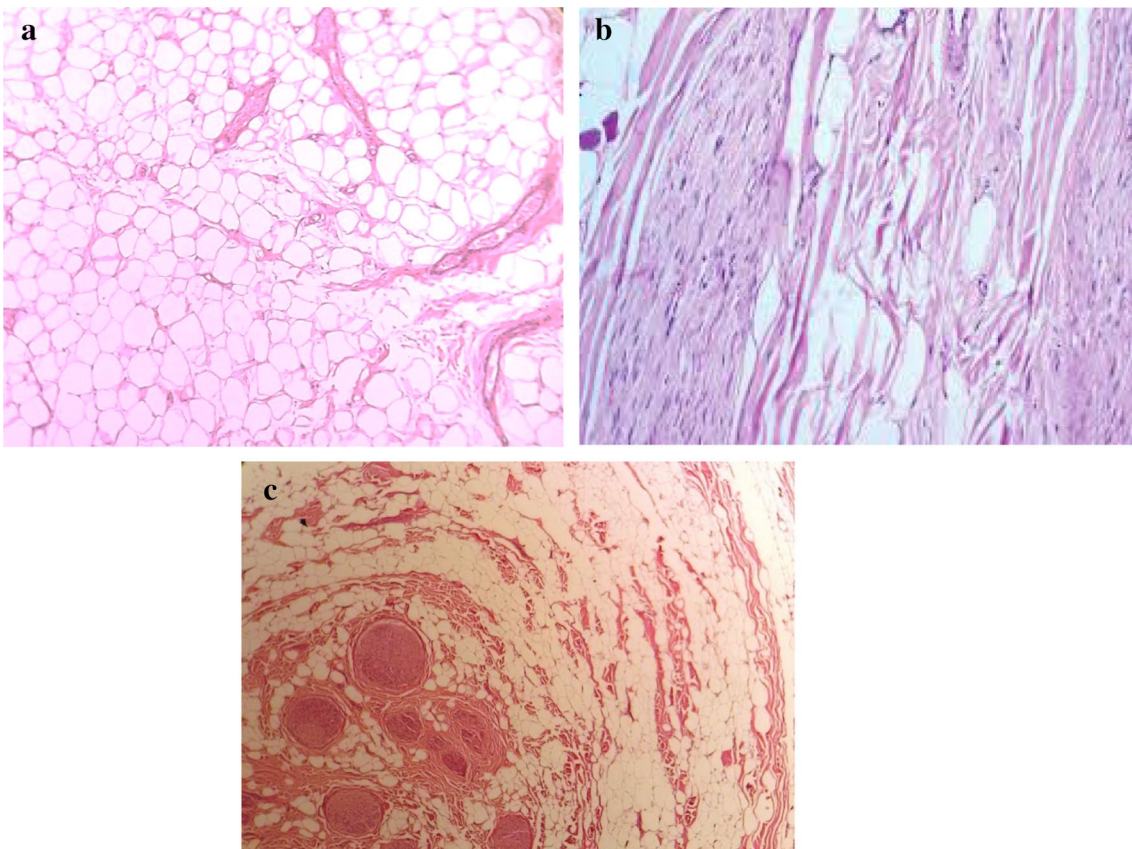


Fig. 4 a–c HPE ($\times 40$) of the lesion showing lobules of mature adipocytes interspersed by fibrocollagenous stromal elements with epi- and perineural tissue in between

provided complete resolution of symptoms and relief [28]. Paletta et al. reported poor results with radical mass excision with fascicular cable graft repair [29]. Various studies stated that the minimal surgical technique of decompression and debulking of the fibrofatty sheath yields more positive results [2, 18, 20, 30]. Our patient had complete resolution

of symptoms following biopsy of the lesion and decompression of carpal tunnel respectively.

Being a rare benign tumour of nerve sheath, MRI is the gold standard in diagnosing fibrolipomatous hamartoma of median nerve. A meticulous clinical and radiological correlation is required to diagnose such rare clinical entity

Table 1 Review of fibrolipomatous hamartoma of median nerve from 2012 to 2019

Author and year of the study	No of FLH cases	Clinical presentation	Imaging modality	Management
Jennifer FH et al. (2012) [20]	1	Diffuse swelling around right elbow with tingling of right thumb since 4 years; No macrodactyly	USG and MRI of elbow	Microsurgical dissection of neural elements
Okubo et al. (2012) [21]	3	Diffuse swelling of wrist and hand without any macrodactyly; No motor involvement	MRI of wrist and hand	Carpal tunnel release and biopsy
Woosun Choi et al. (2012) [22]	2	Soft lump over palm without neurological involvement and macrodactyly	MRI of wrist and hand	Debulking surgery
Bansal A et al. (2015) [3]	1	Paraesthesia of right hand with macrodactyly	MRI of hand	Fasciotomy and biopsy
Ranjan Agarwal et al. (2016) [23]	1	Swelling and pain of right index finger with symptoms of carpal tunnel syndrome; No macrodactyly and no motor involvement	FNAC of the swelling	Decompression surgery with biopsy
Kini JR et al. (2018) [4]	5	Swelling and pain over forearm, wrist and hand with macrodactyly in three cases and without macrodactyly in two cases	MRI of wrists and hands	Surgical carpal tunnel decompression with debulking and biopsy
Muhammad A et al. (2018) [24]	1	Swelling over the wrist and reduced mobility of the middle finger with macrodactyly	MRI of wrist	Wide margin excision of the right middle finger with nerve grafting
MAM Saeed et al. (2019) [25]	1	Painless swelling over left palm with macrodactyly of ipsilateral middle finger	MRI of wrist and hand	Surgical decompression of carpal tunnel
Current study	1	Swelling of left palm with paraesthesia and without macrodactyly	MRI of wrist and hand	Carpal tunnel release and biopsy

to improvise the functional quality of life. There is no definitive treatment for lipomatosis of nerve. However, a conservative approach is commonly advocated with successful results from decompression as in our patient.

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Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical standard statement This article does not contain any studies with human or animal subjects performed by the any of the authors.

Informed consent For this type of study informed consent is not required.

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