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Fibrolipomatous Hamartoma of Median Nerve – A Diagnostic Challenge

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Learning Point of the Article:

The characteristic MR imaging features are confirmatory for a definitive diagnosis of fibrolipomatous hamartoma (FLH), which alleviates the need for any unnecessary biopsy.

Abstract

Introduction: Fibrolipomatous hamartoma (FLH) is an uncommon slow-growing tumor of benign etiology, which predominantly affects the median nerve.

Case Report: We report the case study of a 17-year-old male patient, who presented with complaints of a gradually increasing localized swelling over the volar aspect of left hand for 1 year. A contrast-enhanced Magnetic resonance (MR) scan of the left hand was performed which demonstrated characteristic findings. The patient was treated surgically and post-excision histopathological examination confirmed the diagnosis.

Conclusion: The characteristic MR imaging features of coaxial cable-like appearance on axial section or spaghetti shaped enlarged nerve fascicles and fibrous tissue is confirmatory for a definitive diagnosis of FLH, which alleviates the need for any unnecessary biopsy.

Keywords: Fibrolipomatous hamartoma, magnetic resonance imaging, median nerve.

Introduction

Fibrolipomatous hamartoma (FLH) of median nerve is a slow growing tumor with an uncertain etiology [1, 2]. It is a rare benign tumor with characteristic magnetic resonance image (MRI) findings, which may obviate the need for an unnecessary biopsy [3, 4]. The other names for FLH include neurofibrolipoma, perineural lipoma, intraneural lipoma, neural fibrolipoma, and lipomatosis of nerve [5, 6, 7]. Depending on the extent of nerve involvement, the treatment strategy varies, depends on the extent of nerve involvement as severe motor and sensory deficits, have been reported following complete tumor resection [8, 9, 10]. We report a case of FLH in a young male patient, initially diagnosed purely on the basis of imaging features. He was successfully operated and histopathology specimen confirmed the diagnosis.

Case Report

A 17-year-old male patient was referred to the Department of Radiology for imaging of a focal swelling over the volar aspect of left hand. Initially, the swelling was peanut-sized which progressed to involve the entire palm region, extending from the distal forearm of wrist to the nar eminence and base of 2nd, 3rd and 4th fingers of left hand as depicted in (Fig. 1). The patient complained of periodic episodes of tingling and parasthesia over 2nd, 3rd and 4th fingers of right hand. There was no past history of trauma over the hand. On examination, an irregular-shaped, fusiform swelling was noted over the volar aspect of left hand, proximally extending from the ulnar aspect of left wrist to the thenar eminence and base of 2nd, 3rd and 4th digits. The swelling had a variable consistency on palpation, being softer over the ulnar aspect and firm over the 3rd and 4th metacarpals. The overlying skin was pinchable and non-adherent. On

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Author's Photo Gallery


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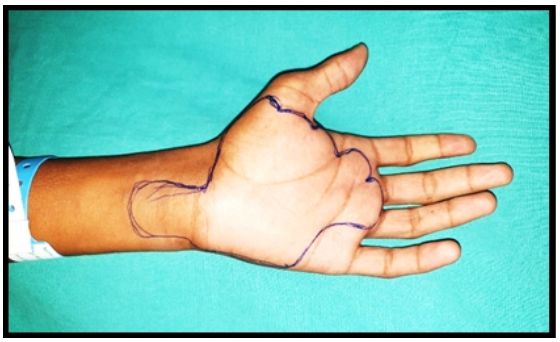

Figure 1: Picture of the left hand of the patient (marked area) showing the extent of the lesion.

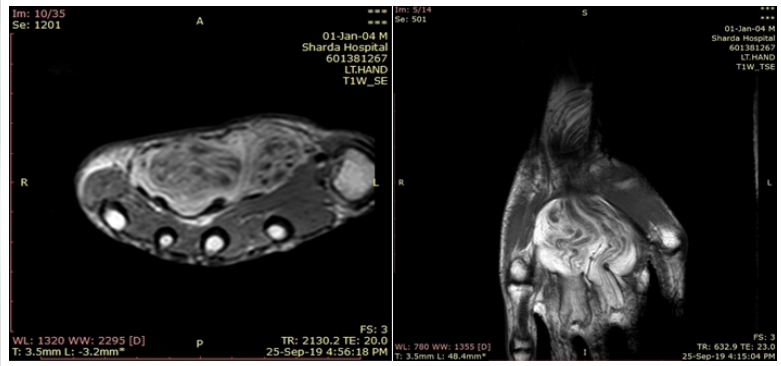

Figure 2: (a and b) T1W axial and coronal sections of the left hand showing a hyperintense lobulated mass lesion, interspersed with hypointense prominent thickened nerve fascicles.

neurological examination, no motor weakness or sensory loss was elicited. The movements of left wrist revealed terminal restriction of palmar and dorsiflexion. No other cutaneous or soft tissue lesions were present.

Non-contrast multiplanar, multi-sequence MRI of the left hand was performed. MR showed gross thickening of the median nerve. Diffuse presence of lipomatous tissue (hyperintense on T1W and T2W sequences, hypointense on SPAIR sequence) was noted inter-digitating between the prominent thickened nerve fascicles (hypointense on T1W and T2W sequences), in a coaxial cable-like pattern, extending from the level of distal forearm up to the distal metacarpal regions of 2nd-4th digits. The thickened nerve was seen causing compression and displacement of the flexor tendons and convexity of the carpal tunnel retinacular sheath. The extension of the lipomatous tissue was seen extending to the 2nd and 3rd inter-metacarpal space. No evidence of any focal osseous lesions was noted. The tendons of the hand appeared normal in signal intensity (Fig. 2-4). The MR features were suggestive of fibro-lipomatous hamartoma of the median nerve with adjacent mass effect.

Under general anesthesia, the patient underwent surgical decompression of carpal tunnel. On extensive dissection, the grossly thickened median nerve was identified as two giant fascicles adherent with fibrolipomatous structures around (Fig. 5). Biopsy of fibrolipomatous structures was taken and care was taken not to injure the giant median nerve bundles. The patient reported absolute pain relief after the surgical decompression of carpal tunnel without any neurological deficit. Histopathological examination of the lesion showed a uniform pattern consisting of lobules of mature adipocytes, separated by fibro-collagenous septae, interspersed with perineural elements as demonstrated in (Fig. 6).

The patient recovered completely from paresthesia after 12 weeks of surgical decompression of the carpal tunnel and he was neurologically stable. The patient performed his activity of daily living without any difficulties. The patient was followed up for 24 months.

Discussion

FLH are slow-growing tumors of benign etiology, which usually affect the infants, and less commonly children and young adults [6, 9, 11, 12]. The median nerve is the most predominantly affected nerve (80%) in FLH, followed by the ulnar and radial nerves, dorsum of the foot and brachial plexus [9, 11]. Some researchers even consider it to be a congenital tumor, [13] while some others believe it to be incited secondary to nerve irritation, inflammation, or prior trauma [14]. FLH presents as a benign tumorous growth of combination of fibrous and lipomatous tissue within the nerve sheath [15, 16]. 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 [1, 8, 9].

The imaging of FLH may include sonography, computed tomography or MRI. The preferred modality for diagnosing

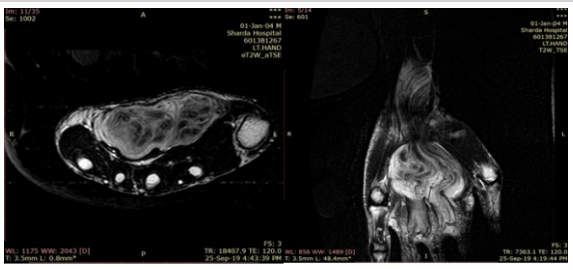

Figure : (a and b) T2W axial and coronal sections of the left hand showing diffuse lipomatous tissue (hyperintense), inter-digitating between the prominent thickened nerve fascicles (hypointense) in a coaxial cable-like pattern.

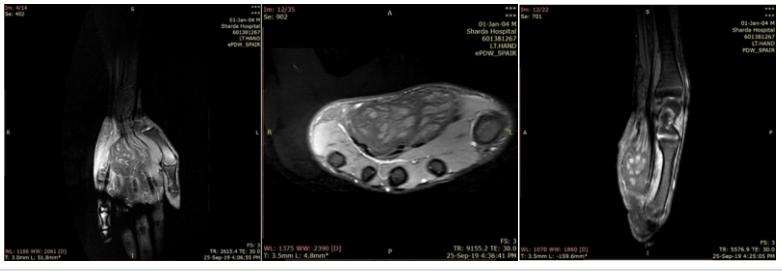

Figure 4: (a-c) PDW SPAIR coronal, axial and sagittal sections of the left hand showing hypointense lipomatous tissue with thickened cable-like nerve fascicles.

Table 1: Review on diagnostic findings of FLH of the median nerve.

Author (year)	Diagnosis	USG findings	MRI findings
Toms et al. [18]	Lipofibromatous Hamartoma of the upper extremity	Sonograms of enlarged median nerve acquired in sagittal and axial planes showed hypoechoic cable-like neural bundles separated by hyperechoic fat.	Axial T1W MR image through wrist at level of distal carpal row shows Guyon's canal distended with and thickened fascicles of ulnar nerve. Coronal T1W MR image of hand show that tumor is predominantly lipomatous and extended from Guyon's canal. Axial T1W MR image of wrist at level of proximal carpal row showed enlarged median nerve. Coronal T1W MR image show lipofibromatous hamartoma of brachial plexus at level of formation of terminal branches. Axial T1W image through arm showed lipofibromatous hamartoma of radial and median nerves. Axial T1W MR image through wrist showed enlarged median nerve and divided flexor retinaculum. Signal intensities of nerve were reversed from typical pattern seen in lipofibromatous hamartoma, with neural fascicles of high fat intensity and intervening substratum of intermediate soft-tissue intensity
So et al. [29]	Fibrolipomatous hamartoma of median nerve		Axial T1W image at the level of the metacarpal base showed coaxial cable-like mass in the position of the median nerve. There were hypointense thickened neural fascicles interspersed by hyperintense fat infiltrate. Mature fat was also found eccentrically in the periphery of the tumour Axial T1W image at the level of the metacarpal shaft and axial T2W image, with fat saturation, at the level just distal showed signal intensity of the thickened neural fascicles which were of low to intermediate signal intensity in both T1-weighted and T2-weighted images with fat saturation
Diwakar et al. [7]	Neurofibrolipoma of median nerve	"Cable like" pattern of median nerve	Axial T2W images showed "Cable like" pattern of median nerve Coronal T1W image showed "Spaghetti like" pattern Fat suppression images showed fat suppression
Choi et al. [30]	Fibrolipomatous hamartoma of digital branch of the median nerve without macrodystrophy		Axial T1W and coronal T2W images show enlargement of the digital branch of the median nerve and hypointense nerve bundles embedded within fat Axial fat-suppressed T1 weighted image with gadolinium enhancement show non-enhancing hypointense fat tissue around the digital branch of the median nerve
Gilet et al. [11]	Fibrolipomatous hamartoma of the median nerve		Axial T1W pre and post gadolinium and T1W fat-suppressed fast spin echo (FSE) MRI showed a fusiform mass in the expected location of the median nerve that is continuous with the nerve proximally and distally On the non-fat-suppressed images the high signal represents the fatty tissue infiltrating and enlarging the median nerve which drops completely with fat suppression. The fat is interspersed within thickened nerve fascicles which are low intensity. The mass displaces the flexor tendons Axial T2W fat suppressed FSE MRI shows intermediate signal longitudinally oriented cylindrical structures representing the nerve fascicles. There is fusiform enlargement of nerve bundles caused by fatty infiltration
Rajadurai et al. [31]	Fibrolipomatous hamartoma of median nerve		T1W axial images showing high signal elongated mass with a coaxial cable like appearance seen within the enlarged median nerve extending to the thenar branches displacing the T1W axial images showing high signal elongated mass with a coaxial cable like appearance seen within the enlarged median nerve extending to the thenar branches displacing the flexor tendons posteriorly T2W fat saturated images showing the enlarged mass with thickened low signal fascicles within interspread by fat which is suppressed in this sequence T2W coronal images showing longitudinally oriented serpentine structures with "spaghetti string" appearance T1W post Gadolinium axial images showing longitudinally orientated thickened neural fascicles which enhance post gadolinium administration.

FLH: Fibrolipomatous hamartoma, MRI: Magnetic resonance imaging

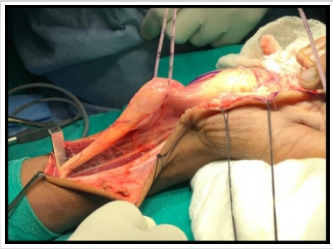

Figure 5: Intra-operative image showing the grossly thickened median nerve at the carpal tunnel.

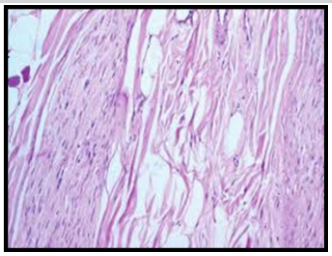

Figure 6: Histopathological evaluation of the lesion showing lobules of mature adipocytes interspersed by fibro-collagenous stromal elements with perineural tissue in between.

FLH is MRI, which remains the gold standard for diagnosis [7, 14]. Plain radiography examination of FLH is non-specific and it may be normal or may sometimes show mild prominence of the soft tissue which is related to fibrofatty tissue [17, 18]. The sonographic examination demonstrates the presence of hyperechoic fatty tissue which is surrounded by smooth round hypo-to-anechoic fascicles [19]. The spectral Doppler evaluation does not demon¬strate any significant vascularity within the mass [11, 18].

On cross-sectional imaging, FLH exhibit fusiform enlargement of the involved nerve, which is caused by thickening of nerve bundles and adjacent diffuse fibro-fatty proliferation. Thickened nerve bundles appear as tubular or serpentine structures on all imaging modalities. Thickening of the neural fascicles is caused by perineural and endoneural fibrosis [18, 20, 21]. The mature fat cells infiltrate interfascicular connective tissue [18]. The perineural and endoneural fibrosis is responsible for the low signal intensity of thickened nerve fascicles, which are seen as tubular or serpentine structures oriented horizontally along the length of the nerve [9, 11]. On axial images, the enlarged nerve has a coaxial cable-like appearance, due to its thickened nerve fascicles and is interspersed with infiltrating fat, which is the pathognomonic feature of FLH [11, 19, 22]. Another characteristic pathological feature of FLH is growth of the tumor along the branching pattern of the nerve [13, 23, 24]. The differential diagnosis of FLH includes intraneural lipoma, schwannoma, tenosynovitis, ganglion cyst, traumatic neuroma, and vascular malformation [6, 25, 26, 27].

The treatment strategy of FLH is still controversial and is largely dependent on the extent of the nerve involvement [1, 6, 12, 28]. Severe motor and sensory deficits have been reported following complete tumor resection. Conventional treatment involves carpal tunnel decompression by excision of the transverse carpal ligament, followed by biopsy of the enlarged nerve. The review on diagnostic findings of FLH of the median nerve is tabulated (Table 1).

Conclusion

FLH is an uncommon, slow-growing tumor of benign etiology, with a predilection for the median nerve. The characteristic MRI features of coaxial cable-like appearance on axial section or spaghetti shaped enlarged nerve fascicles and fibrous tissue is confirmatory for a definitive diagnosis, which alleviates the need for any unnecessary biopsy.

Clinical Message

Although a rare clinical entity of FLH of median nerve, the pathophysiology and management remains controversial, MRI remains the gold standard in the diagnosis of FLH of median nerve.

Declaration of patient consent : The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's parents have given their consent for patient images and other clinical information to be reported in the journal. The patient's parents understand that his names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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