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Case Report

Radiology Section

Fibrolipomatous Hamartoma of the Median Nerve: A Case Report

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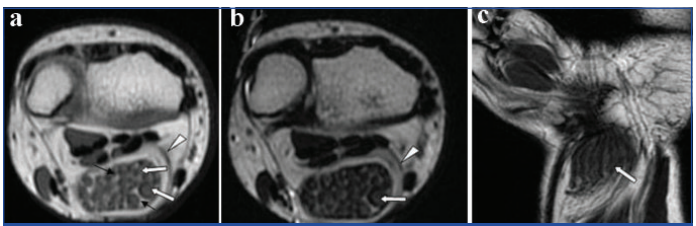
ABSTRACT

Fibrolipomatous hamartoma, also known as fibrofatty overgrowth or perineural lipoma, or neural fibrolipoma, is a rare, benign, congenital lesion that occurs due to an abnormal overgrowth of mature fat and fibroblasts within the perineurium. This leads to the expansion of the nerve and separation of fascicles. It is more commonly observed in the median nerve at the wrist or hand level. Here, the authors presented the case of a 32-year-old female patient, who complained of swelling in the left wrist region for the past two years. The swelling gradually increased in size over one year and caused pain and paresthesia. Magnetic Resonance Imaging (MRI) of the left wrist revealed fibrolipomatous hamartoma of the median nerve. Surgical management was advised, but the patient declined and opted for conservative treatment for two months. Knowledge of this rare condition helps radiologists make an accurate diagnosis and may eliminate the need for invasive biopsy.

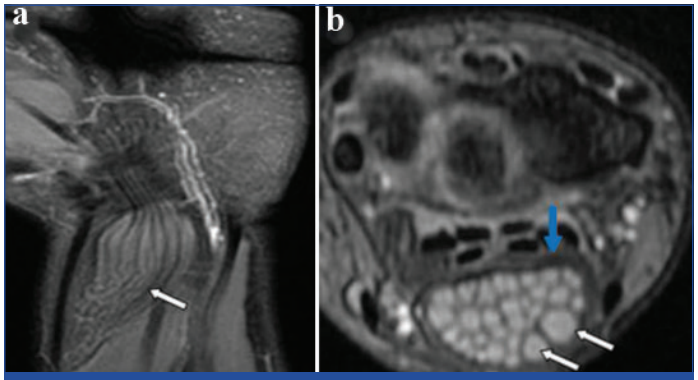
Keywords: Magnetic resonance imaging, Neural fibrolipoma, Paraesthesia, Perineural lipoma

CASE REPORT

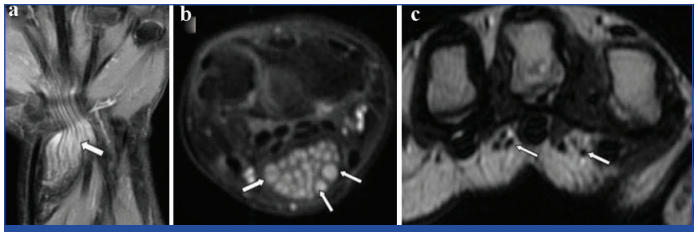
A 32-year-old female presented with complaints of swelling in the left wrist region for the past two years, which had gradually increased in size for one year and had caused pain and paraesthesia. There was no history of trauma to the hand, diabetes mellitus, or hypertension. No other significant past history or family history was noted. On examination, a smooth, fusiform-shaped, tender swelling measuring approximately 2×2 cm with firm consistency was observed over the ventral aspect of the left wrist region, with no associated skin changes. The rest of the physical examination was unremarkable. Blood investigations including complete blood count, C-reactive protein, and serum uric acid were normal. The possible differential diagnoses considered were ganglion cysts, vascular malformations, schwannomas, neurofibromas, and lipomas. A screening ultrasound of the wrist revealed enlarged and hypoechoic nerve fascicles surrounded by echogenic fat tissue, giving a "cable-like appearance" on axial images. MRI of the left wrist, both plain and with contrast, showed diffuse thickening of nerve fascicles of the median nerve, [Table/Fig-1-3] extending from the distal forearm to the wrist and slightly into the distal palmar region. [Table/Fig-3c] There was diffuse and extensive interfascicular and perineural lipomatous infiltration, with the nerve fascicles shows mild diffuse enhancement on post-contrast study. [Table/Fig-3a,b] The diagnosis was fibrolipomatous hamartoma of the median nerve. Surgical management was advised, but the patient declined and opted for conservative treatment with tab etoricoxib 90 mg OD, tablet tolperisone 150 mg BD, tab gabapentin 100 mg+nortriptyline 10 mg HS, and tab pantoprazole 40 mg OD for two months. On follow-up, the patient's symptoms had slightly improved with the above management, and the patient wished to continue with the same treatment due to symptom relief. The imaging findings on MRI and ultrasound of the wrist were characteristic of fibrolipomatous hamartoma of the median nerve. The classical "coaxial cable-like appearance" on MRI eliminates the need for biopsy.



[Table/Fig-1]: Axial T1 weighted (a), Axial T2 weighted (b) and coronal T2 weighted (c) Grossly thickened median nerve in distal forearm and wrist with prominent nerve fascicles (white arrows) shows coaxial cable-like appearance on axial images. Note the perineural lipomatous encapsulation around the median nerve (arrowhead in [Table/Fig-1a,b] with lipomatous tissue interdigitating between the prominent nerve fascicles (black arrows in [Table/Fig-1a]).



[Table/Fig-2]: Short Tau Inversion Recovery (STIR) coronal (a), and axial images (b) Shows encapsulated thickened nerve fascicles with interdigitation fat suppression between the nerve fascicles within the carpal tunnel (white arrows). Note bowing and convexity of superficial carpal tunnel reticular sheath (blue arrow).



[Table/Fig-3]: Fat saturated post-contrast coronal (a) and, axial (b) Enhancement of thickened median nerve and fascicles in the carpal tunnel. [Table/Fig-3c]: Axial T2 weighted at the palm level showing involvement of interdigital branches of the median nerve in the palm region (c) (arrows).

DISCUSSION

Fibrolipomatous hamartomas are benign tumors that commonly affect infants and are less common in children and young adults [1-3]. The median nerve is the most commonly affected nerve (80% of cases), followed by the ulnar and radial nerves, as well as the nerves over the dorsum of the foot and brachial plexus [4]. The median nerve is formed by the lateral cord (ventral roots of C5-C7) and medial cords (ventral roots of C8 and T1) of the brachial plexus [2].

Motor supply is provided to the flexor muscles of the forearm (anterior compartment), excluding the flexor carpi ulnaris and ulnar head of the flexor digitorum profundus. In the hand, it supplies the muscles of the thenar eminence and the radial two lumbricals. Sensory supply is responsible for the skin of the palmar and distal dorsal aspects of the lateral three and a half digits and adjacent palm, as well as the palmar and distal dorsal aspects of the thumb and radial half of the 2nd digit. It also supplies the palmar and distal dorsal aspects of the adjacent sides of the 2nd to 4th digits and the central palm.

Fibrolipomatous hamartomas typically present as a slowly growing mass on the volar aspect of the wrist or in the distal forearm region [1], sometimes accompanied by pain, numbness, decreased sensations, paresthesia, and carpal tunnel syndrome, particularly when associated with nerve entrapment or compression [3]. Macrodactyly of the digit may occur when the digital branches are affected (macrodystrophia lipomatosis) [1,3]. The postulated pathogenesis involves an abnormal overgrowth of mature fat and fibroblasts within the perineurium, leading to the expansion of the nerve with separation of fascicles and fusiform nerve enlargement [3]. The findings of the present case are compared with similar studies/case reports in [Table/Fig-4] [5-8].

Imaging modalities used for fibrolipomatous hamartomas include ultrasound, Computed Tomography (CT), and MRI. The MRI is the preferred modality for diagnosis. Ultrasound shows a characteristic hyperechoic tissue (echogenic fatty tissue) encasing smooth, round hypoechoic or anechoic fascicles, with absent vascularity within the mass on color and spectral doppler examination [9]. On CT and MRI, the lesions appear as fusiform enlargements of the nerve, caused by the thickening of nerve fibers, resembling serpentine or tubular structures. The MRI features of fibrolipomatous hamartomas are pathognomonic, showing a "coaxial cable-like" appearance on axial images and a "spaghetti-like" appearance on coronal images [1]. On T1-weighted imaging, neural bundles are hyperintense compared to surrounding muscle tissue, while on T2-weighted imaging, the fatty components are hyperintense and the fibrous components are hypointense to adjacent muscle [10].

Authors	Year of publication	Study	Follow-up
Pallewatte AS and Samarasinghe EC [5]	2021	Illustrated characteristic clinical, radiological, and histopathological features of fibrolipomatous hamartoma in a teenage female	Diagnosis of fibrolipomatous hamartoma of the left median nerve was made, based on the imaging findings. A biopsy was done which confirmed the same. This study demonstrates that diagnosis can be made confidently on imaging alone especially with MRI, without the need of biopsy
Mam P et al., [6]	2014	Showed a rare case of fibrolipomatous hamartoma found in the great toe of a 25-year-old patient	Surgical excision was performed and a pathologic diagnosis of fibro lipomatous hamartoma was made
Philip L et al., [7]	2015	Reported an unusual case of a fibro lipomatous hamartoma that arose in a nuchal nerve	Identified the lesion based on the histological findings
Nanno M et al., [8]	2011	Showed an unusual case of fibromatous hamartoma in a 43-year-old woman involving ulnar digital nerve	Preoperative MRI evaluation findings and postoperative histopathological findings were consistent with fibrolipomatous hamartoma. After three years of surgery, there was no recurrence of mass or neurological deficits
Present study	2023	Fibrolipomatous hamartoma of the median nerve in a 32-year-old female.	MRI plain and contrast study was done which showed findings consistent with fibrolipomatous hamartoma of median nerve. Patient was advised surgical management, but the patient was not willing for surgical management, hence was managed conservatively for two months

[Table/Fig-4]: Comparison of the case findings with published literature [5-8].

CONCLUSION(S)

Fibrolipomatous hamartoma of the median nerve is a rare congenital abnormality that commonly affects the median nerve, exhibiting classical sonographic and MRI imaging characteristics. Understanding this rare condition enables radiologists to make an accurate diagnosis, potentially eliminating the need for invasive biopsy. MRI, with its inherent soft tissue contrast and excellent spatial resolution, offers the advantage of assessing the extent of nerve involvement and detecting any involvement of other peripheral nerves.

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