

Lipofibromatous Hamartoma of the Median and Ulnar Nerves: A Case Report

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Learning Point of the Article:

In fibrolipomatous hamartoma involving major peripheral nerves, MRI is diagnostic, and conservative surgery with extended neurolysis may preserve function and provide durable symptom relief.

Abstract

Introduction: Lipofibromatous hamartoma (LFH) is a rare, benign tumor of peripheral nerves characterized by fibroadipose infiltration of the epineurium and perineurium. It most frequently involves the median nerve and is rarely seen with concomitant ulnar nerve involvement.

Case Report: We report a 28-year-old male with a 13-year history of a slowly progressive mass over the volar aspect of the left hand and wrist, associated with persistent hypoesthesia in the thumb and intermittent paresthesia in the ring and little fingers. Magnetic resonance imaging (MRI) revealed the pathognomonic “coaxial cable” and “spaghetti” signs involving both the median and ulnar nerves. The patient underwent extended neurolysis of both nerves with carpal tunnel and Guyon’s canal release. Histopathology confirmed LFH without atypia. At 12-month follow-up, he showed sustained improvement in pain and sensory symptoms, with complete return to work.

Conclusion: Concomitant LFH of the median and ulnar nerves is extremely rare. MRI provides pathognomonic diagnosis, and extended neurolysis without tumor resection can achieve favorable functional outcomes.

Keywords: Median nerve, Hamartoma, Ulnar nerve, Peripheral nervous system neoplasms.

Introduction

Lipofibromatous hamartoma (LFH) is a benign, slow-growing, and very uncommon tumor of peripheral nerves, characterized by fibroadipose infiltration of the epineurium and perineurium [1]. It occurs most frequently in the upper extremity, with the median nerve being the most commonly affected site. In pediatric patients, LFH is often associated with macrodactyly [2].

Patients typically present with a long-standing soft-tissue mass associated with pain, swelling, or paresthesia, most often diagnosed in the third or fourth decade of life [1]. On magnetic resonance imaging (MRI), the lesion demonstrates the

characteristic “coaxial cable” appearance on axial view and “spaghetti sign” on coronal view [3].

Treatment depends on the extent of nerve involvement. Radical tumor resection usually requires sacrificing part of the nerve, which, in adults, has been associated with poor functional recovery. Consequently, conservative management with neurolysis and decompression is generally preferred, yielding better outcomes [4]. Importantly, LFH is considered a benign lesion without malignant potential.

Combined involvement of both the median and ulnar nerves is exceedingly rare. We present the case of a young adult with long-standing LFH of the median and ulnar nerves, treated with

Author's Photo Gallery



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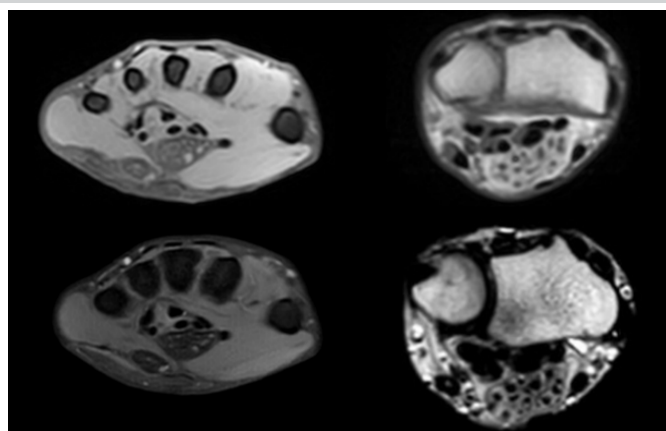


Figure 1: Magnetic resonance imaging images of a transverse cut through the base of the fingers to the wrist showing the median and ulnar nerve with its lipomatous infiltration.

extended neurolysis and achieving good functional recovery.

Case Report

A 28-year-old male presented with a slowly progressive mass over the volar aspect of the left hand and wrist, present for 13 years. He reported a 3-month history of persistent hypoesthesia in the thumb and intermittent paresthesia in the ring and little fingers, without motor compromise.

On examination, a palpable lobulated mass was found on the volar side at the base of the middle and ring fingers, extending proximally to the wrist at the level of the flexor retinaculum.

Ultrasound revealed thickening of the median and ulnar nerves from the base of the fingers to the wrist. MRI (Fig. 1) demonstrated diffuse thickening with adipose infiltration in the median nerve from the wrist to the distal metacarpus, with narrowing at the carpal tunnel and pathognomonic coaxial cable and spaghetti signs. Similar but less intense, findings were noted in the ulnar nerve.

A multidisciplinary evaluation was performed by the hand surgery and musculoskeletal tumor teams. The patient underwent extended neurolysis of both the median and ulnar nerves with release of the carpal tunnel and Guyon's canal (Fig. 2). Intraoperative biopsy revealed fibroadipose tissue without atypia, confirming LFH. Postoperatively, the patient experienced significant reduction in pain and sensory symptoms, with return to work at 2 months and sustained improvement at 12-month follow-up (Fig. 3).

Discussion

LFH is a rare peripheral nerve tumor characterized by fibroadipose infiltration of the epineurium and



Figure 2: Clinical photograph, characteristic fusiform enlargement of the median nerve at the distal third of the forearm and distally, extending into the carpal tunnel.

perineurium [1]. It most commonly involves the median nerve, though rare cases have been described in other nerves including the ulnar nerve [2,5]. Simultaneous involvement of both nerves in the same patient is extremely rare [2].

The etiology remains unclear, with theories ranging from congenital origin [6] to mechanical or inflammatory triggers [4]. Clinically, patients often present with paresthesia, dysesthesia, or soft-tissue swelling, and symptoms of carpal tunnel syndrome are frequent [7].

MRI is the gold standard for diagnosis, showing the classic coaxial cable and spaghetti appearances [3,8]. Histopathology confirms fibroadipose tissue infiltration between nerve fascicles, but biopsy is not always necessary given the characteristic imaging findings [1]. Differential diagnosis includes intraneural lipoma, traumatic neuroma, and vascular malformations [3].

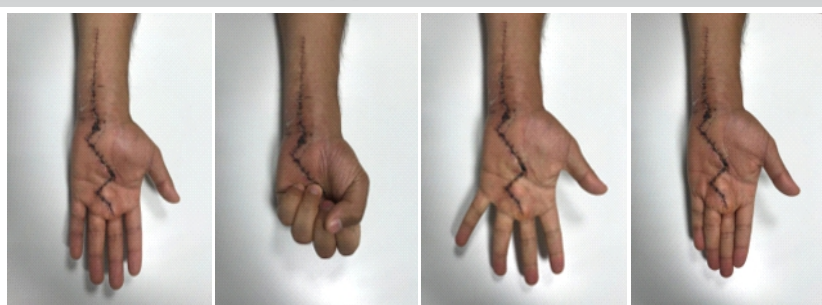


Figure 3: Clinical photograph, 8 weeks after surgery, showing total recovery of medial and cubital nerve function.

Surgical strategies vary. Radical resection risks significant neurological deficits [9], while decompression with or without limited biopsy has shown good outcomes [6,9]. Recent series, such as that by Prabhu et al. [10], suggest that even excision with grafting may preserve function, but conservative management remains preferred in adults.

In our case, extended neurolysis with decompression achieved excellent pain relief and functional recovery, supporting conservative surgery as the optimal approach.

Conclusion

LFH of the median and ulnar nerves is extremely rare. MRI provides a pathognomonic diagnosis. Conservative surgery with extended neurolysis and decompression, without tumor resection, represents a safe and effective approach that preserves function.

Clinical Message

Fibrolipomatous hamartoma of the median and ulnar nerves is extremely rare. MRI provides a pathognomonic diagnosis, and conservative surgery with extended neurolysis – without tumor resection – offers a safe approach with good functional outcomes.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her 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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