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Lipofibromatous hamartoma of median nerve: A review article

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Abstract

Background: Lipofibromatous Hamartoma (LFH) is a rare condition that involves diffuse infiltration of peripheral nerves by normal-appearing fibrous and adipose tissues.

Material and Methods: A literature search in Google Scholar, Medline, Embase, Current Contents, and Science Citation Index yielded original publications from January 1946 to August 2021 in English using the keywords “lipofibromatous hamartoma,” “fibrolipomatous hamartoma,” “lipofibroma,” “fibrolipoma,” “macro dystrophia lipomatosa,” “fibrofatty proliferation,” “intraneural lipoma,” “median nerve,” “median neuropathy,” and “peripheral nerve diseases.”

Conclusion: LFH of the median nerve is a rare condition, with 180 reported cases in the English and French literature between January 1946 and November 2012. One third of cases are associated with macrodactyly, with no gender predominance. Patients are often affected at or shortly after birth and present before age 30 years.

Keywords: Lipofibromatous hamartoma, median nerve

Introduction

Lipofibromatous Hamartoma (LFH) is a rare condition that involves diffuse infiltration of peripheral nerves by normal-appearing fibrous and adipose tissues. To date, there have been fewer than 200 published cases involving the median nerve since the first report in 1953 by Mason^[1]. In 1965, Emmett described this excessive tissue growth as a hamartoma^[2]. It was not until 1969 that Johnson and Bobfigli introduced the term LFH to accurately describe the characteristic histological findings^[3]. Many different names have been used to describe LFH, including fibrolipomatous hamartoma, fibrolipoma, lipofibroma, fibrofatty proliferation, macro dystrophia lipomatosa, and intraneural lipoma^[4-6]. The term macro dystrophia lipomatosa is used to describe LFH associated with macrodactyly, where overgrowth of all mesenchymal elements is present in the affected digits^[7]. Some authors classify this variation as LFH with or without macrodactyly^[8,9] however, the lack of a single descriptive name has complicated the diagnosis of this condition.

The cause of LFH remains obscure; however, commonly proposed etiologies are congenital malformation^[4, 10-14] and trauma^[9, 15]. LFH preferentially affects the median nerve, although there have been reports that include the buccinators, sciatic, plantar, and superficial peroneal, and posterior interosseous nerves^[16-20]. In the upper extremity, LFH may affect the radial nerve and branches, ulnar nerve and branches, and even the entire brachial plexus^[21-23]. Patients often present with a progressively enlarging mass along the median nerve territory in the region between the distal forearm and fingers, with symptoms of nerve compression and carpal tunnel syndrome. In 1 instance, LFH of the median nerve was found in the elbow, an unusual location, where it was associated with thumb paresthesia^[24].

No clear guideline has been established for the diagnosis and treatment of LFH. Historically described approaches include complete excision of the enlarged median nerve and branches, often resulting in debilitating consequences to sensory and motor functions of the hand^[25]. In recent practice, conservative surgery is performed only in selected cases.

This study reviews the presentation, diagnostic modalities, treatment options, and outcomes of median nerve LFH in order to formulate a systematic approach to the evaluation and management of the condition.

Data sources

A literature search in Google Scholar, Medline, Embase, Current Contents, and Science Citation Index yielded original publications from January 1946 to August 2021 in English using the keywords “lipofibromatous hamartoma,” “fibrolipomatous hamartoma,” “lipofibroma,” “fibrolipoma,” “macro dystrophia lipomatosa,” “fibrofatty proliferation,” “intra-neural lipoma,” “median nerve,” “median neuropathy,” and “peripheral nerve diseases.” The bibliographies of the selected articles were manually examined for relevant references.

Etiology

Although there have been suggestions of a congenital origin to LFH, the etiology remains unclear. Cases arising from post-traumatic incidences have been reported, all showing the characteristic fatty infiltrate on biopsy. The pathophysiology of LFH is unknown.

Clinical feature

Patients typically present with gradually enlarging non-tender lesions in the distribution of the affected nerve. Since LFH often involves the median nerve, the presentation of median nerve LFH shares considerable overlap with carpal tunnel syndrome. Affected individuals complain of numbness and tingling along the volar aspect of the wrist and hand. Motor deficits are a late finding. Congenital origin of LFH with or without macrodactyly has been previously suggested, but results have been mixed. Most cases occur within the first three decades of life, with the mean age of 22.3 in isolated cases and 22.0 in cases with macrodactyly [26]. Silverman and Enzinger reported 26 cases of upper and lower extremity LFH, 7 with macrodactyly and 19 without. Combining their work and subsequent studies, it was determined that there is a 2:1 female to male ratio of cases with macrodactyly and a 1:1 ratio in cases without [27]. Complicating the scenario even further is the considerable overlap with Klippel-Trenaunay-Weber syndrome, congenital lymphedema, hypertrophic mononeuritis, and hereditary hypertrophic interstitial neuritis of Dejerine-Sottas [28].

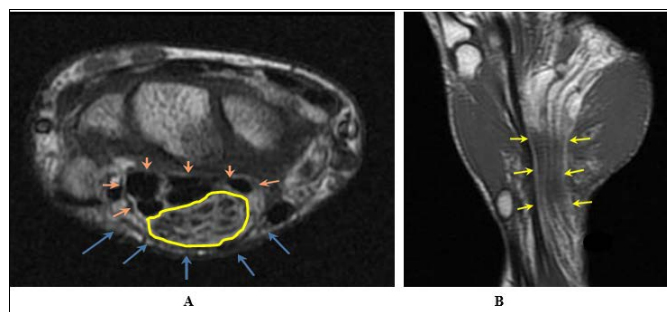
Upper extremity morphology includes mass, hypertrophy, macrodactyly, syndactyly, radial or ulnar deviation of phalanx, neurological symptoms including pain/dysesthesia, paresthesia (numbness, tingling, pins and needles), motor deficits (weakness, impaired function), decreased sensibility, others including café-au-lait spots, port-wine stains, neurofibromatosis.

Personal or family history of patients with LFH includes trauma to affected area, surgery to affected area, Klippel-Trénaunay-Weber syndrome, family with LFH and/or macrodactyly and neurofibromatosis.

On physical examination, the mass is commonly described as soft, firm, nonfluctuant, mobile, and minimally tender. Macrodactyly, when present, follows the course of the median nerve [29]. Syndactyly has also been reported to be associated with LHF [30]. Radial or ulnar deviation of the affected digit is associated with macrodactyly and results from asymmetrical lengthening of the radial and ulnar aspect of the involved digits. There was 1 report of “syndromic” presentation with a “monstrous” enlargement of the entire right upper extremity, clinodactyly, and syndactyly [31]. One female patient reported port-wine stains on the neck, chest, and arm [32].

Some reports describe neurological deficits associated with LFH, such as hypesthesia; decreased grip, pinch and opposition strength; and paresthesia yielding positive Tinel’s

and Phalen’s tests [33]. Functional impairment is caused by either mass effect or muscle weakness. Many patients who present with impaired hand function experienced asymptomatic swelling months to years before consultation.



Magnetic Resonance Imaging (Mri) Findings Of Lipofibromatous Hamartoma (Lfh) Of The Median Nerve

A. Transverse section depicts displacement of flexor tendons (orange arrows) within the carpal tunnel and palmar protrusion of the flexor retinaculum (blue arrows) to an enlarged median nerve (yellow outline).

B. Sagittal section demonstrates fusiform or hour glass enlargement of the median nerve (yellow arrows) with low-intensity nerve bundles embedded in hyperintense adipose tissue.

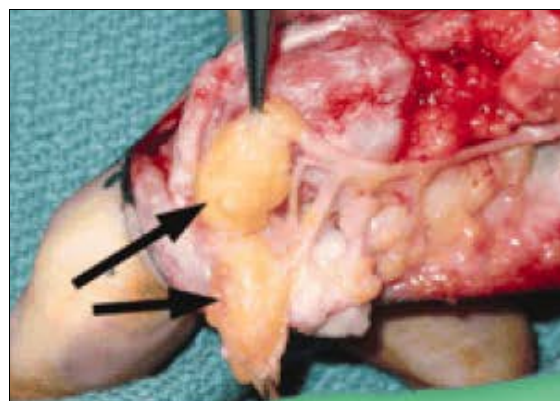


Fig 1: Intraoperative photograph shows a lipofibromatous hamartoma. This shows the typical gross appearance of LFH. Multiple soft, gray-yellow lobulated masses (arrows) are present within epineurial sheath of radial digital nerve of this right index finger. Epineurium of digital nerve displays extensive perineural fibrosis

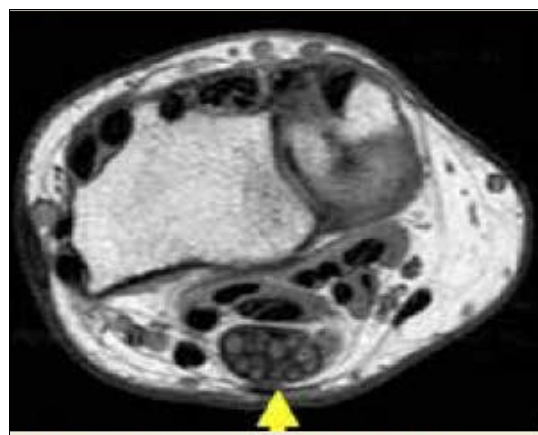


Fig 2: Axial T1-weighted imaging of the wrist through Guyon’s canal which is grossly distended with fat (arrow head) and thickened fascicles of the ulnar nerve. The patient presented with paresthesia in the ulnar nerve distribution. EMG confirmed conduction delay.



Fig 3: Axial T1-weighted section of arm showing LFH of the radial and median nerves (arrows)

Differential diagnosis

There is no consensus on the diagnosis and treatment of LFH. However, various features of LFH on physical examination, diagnostic imaging, and histology can facilitate its recognition. Benign tumors mimicking LFH of the median nerve include ganglion cyst^[34], Déjerine Sottas disease (Charot-Marie-Tooth type III)^[35], Klippel-Trénaunay-Weber syndrome^[36], lipoma^[37], and traumatic neuroma (spinal and terminal neuromas)^[38]. Tumors with malignant potential, such as neurofibroma^[39] and schwannoma^[40, 41] may also present in a similar fashion. Finally, it is important to exclude malignant conditions like liposarcoma^[42] and malignant peripheral nerve sheath tumors^[43]. Macroscopically, neurofibromatosis is difficult to distinguish from LFH with macrodactyly because neither is encapsulated and both may present with mesenchymal overgrowth following a nerve distribution^[44]. However, the lack of neurocutaneous manifestations and family history differentiate LFH from neurofibromatosis type I (von Recklinghausen disease).⁴⁵ Magnetic resonance imaging (MRI) is used to distinguish plexiform neurofibroma from LFH wherein signals depict neural overgrowth in the place of fat^[46]. The two can be further distinguished on histology: LFH is predominantly associated with proliferation of fibrofatty tissue, which infiltrates between nerve fibers, whereas neurofibromatosis is associated with tumefaction. Klippel-Trénaunay-Weber syndrome is not usually linked to LFH. The 2 conditions have been reported together on only 2 occasions.

Management

Investigations

There are no known laboratory abnormalities associated with LFH. Common imaging modalities used include radiography, ultrasound, and MRI. X-ray may show soft tissue swelling in the affected area with or without macrodactyly. In LFH with macrodactyly, there may also be bony hypertrophy with osteoarthritic changes. Ultrasound depicts a fusiform mass with longitudinal nerve bundles and alternating hypoechoic and hyperechoic bands. MRI of LFH shows fusiform or hourglass-shaped enlargement of the median nerve on coronal section, with displacement of the flexor retinaculum and adjacent tendons on transverse views^[47-49]. On transverse section, this soft tissue mass has a round or oval contour or may appear multilobulated. T1- and T2-weighted images show characteristic findings of low-intensity serpentine nerve

bundles embedded within abundant hyperintense adipose material, with fine fibrous tissue septa coursing along the median nerve. LFH of the median nerve with or without macrodactyly has pathognomonic MRI appearances that are “coaxial cable-like” on axial plane and “spaghetti-like” on coronal or sagittal cuts^[50, 51]. Computed tomography (CT) depicts a disc-shaped bulging mass in the carpal tunnel, with displacement of the flexor retinaculum and tendons. The mass’ density shows fibroblastic components.

Electromyogram (EMG) and nerve conduction studies (NCS) on the median nerve yield abnormal results, showing decreased sensory and motor conduction, fibrillations in distal muscles, signs of chronic denervation, and findings consistent with compressive neuropathy^[52, 53].

Biopsy and histopathological examination are the only definitive means of LFH diagnosis. Histological analysis shows interlacing collagen, fibroblasts, mature adipocytes, and occasional capillaries, which separate nerve fascicles and infiltrate the space between the epineurium and the perineurium^[54]. The nerve sheath may be thickened and fibrotic. Nerve fibers appear normal and there are no mitotic figures, inflammation, or myelin degeneration^[55].

We suggest an approach to the diagnosis of LFH of the median nerve that focuses on the presenting symptom of an isolated palmar mass because this is the initial complaint in 88% of patients. Routine nerve biopsy, although diagnostic, is not recommended because it may cause functional sequelae. MRI, conversely, offers pathognomonic features that confidently identify the lesion, bypassing the need for additional invasive techniques. However, care must be taken to rule out alternative diagnoses of potentially malignant tumors. Characteristics suggestive of malignancy may include a rapidly growing firm mass and invasion into adjacent structures. Signs of central hemorrhage or necrosis, which appear as inhomogeneous enhancements on MRI, raise suspicion for malignant peripheral nerve sheath tumors, but these may also be present in benign neurofibromas and schwannomas. Family history is important in the case of neurofibromatosis-1 because the disease may have associations with malignant peripheral nerve sheath tumors, Klippel-Trénaunay Weber syndrome, and schwannomas. Histological findings are limited to perineural and endoneural fibrosis with axons normal in size or atrophic and fatty infiltration around the nerve branches.

Treatment plan

Treatment goals for LFH with or without macrodactyly are 4-fold: symptom prevention, symptomatic relief, function, and aesthetics.

There is no definitive cure for LFH of the median nerve, and patient management varies case by case. Traditionally, complete tumor excision was performed to eliminate the risk of malignancy. Patients would often be left with devastating sensory and functional impairments and the formation of painful neuroma^[56]. Excision of the hamartomatous nerve along with the overlying skin it innervates has been proposed to minimize sensory loss. This procedure has demonstrated superior results in children, possibly owing to increased neuroplasticity in the younger population. Interestingly, 2 authors reported a Martin Gruber anastomosis discovered intraoperatively, and this preserved the patients’ hand function following aggressive resection of the median nerve main trunk^[57].

Indications for surgery

Indications for surgical intervention vary case-by-case. Due to the intimate nerve involvement, LFH is often accompanied by a degree of neurologic morbidity. If the risk of nerve damage

is low and nerve involvement is minimal, surgical debulking for cosmetic reasons can be undertaken. However, in the face of advanced nerve involvement, indications for intervention are progressive and unrelenting neurological deficits. LFH most commonly affects the median nerve in 66 to 80% of cases, causing pain, sensory and motor deficits in the affected nerve distribution^[58]. There have also been cases of LFH affecting the brachial plexus, ulnar, radial, peroneal and plantar nerves^[59, 60]. There is no explanation of why the median nerve is most commonly affected. A fundamental knowledge of the anatomical distribution of nerves helps distinguish LFH from other hand tumors as LFH only involves the nerve. In addition on physical exam, there are soft, palpable nodules along the nerve.

Medical management

Treatment of LFH is based on symptoms of the condition. While some cases cause no neurologic or functional complications, others do. There is no role for medical management and surgery is reserved for those with neurologic deficits. Current treatment has shifted toward a more conservative approach. Expectant management can be offered to patients who present with an asymptomatic swelling without severe neurological impairment. Spontaneous regression is, however, uncommon and the lesion may continue to enlarge, eventually progressing to compressive neuropathy.

Surgical management

Since first appearing in the literature in the 1950's, there has been no widespread consensus on the surgical treatment of LFH; rather, surgeons have adopted a case-by-case approach. The problems which complicate the approach are threefold, each with multiple treatment options. The first problem to be addressed is the treatment of carpal tunnel syndrome. While it is widely accepted that carpal tunnel decompression should be undertaken for relief of paresthesias, some strictly consider waiting for the tumor to become symptomatic prior to surgery, mostly in younger children. In cases of associated macrodactyly, three options exist. The first includes staged debulking, the second is an epiphysiodesis/epiphysectomy and the third is no treatment. In terms of debulking, in a review of eight cases, three reported a decrease in mass size from one to three-year follow-up, four cases noted no change from two to seven years out and one case reported an increase in tumor mass following surgery^[61].

In many cases, both neurolysis and excision of the main trunk of the median nerve can lead to abnormal two-point tactile discrimination and a loss of sensory distribution postoperatively in adult patients. Furthermore, micro-dissection of the median nerve has led to ischemic complications in one reported case and has been unsuccessful in others^[62-53]. One case reported success in median nerve excision in the presence of a Martin-Gruber anastomosis^[64-65]. Amadio reported cases of significant debulking of tissue followed by distal digital nerve excision as nerves affected by LFH have shown decreased EMG function as in our case presentation. Since there is considerable cutaneous overlap in the hand, Amadio suggests debulking followed by rotation of the excess skin from the dorsal aspect to the volar aspect of the affected digits to restore sensation^[66]. While this does not fully restore the dermatomal distribution of sensation, it does offer a degree of sensation which would be lost with either no treatment or nerve excision minus the dorsal to volar rotation. Open carpal tunnel release was performed with the aim to

decrease motor and sensory impairments and to provide symptomatic relief from compressive median neuropathy, and even prophylactically in asymptomatic patients demonstrating "marked enlargement" of the median nerve. Recently, a successful endoscopic carpal tunnel release has been reported. Division of the flexor retinaculum with concurrent forearm fascial release also yielded positive results. Several authors describe decompression and debulking of the neural sheath.⁶⁷ Microsurgical dissection generated disappointing results, although in some cases, hand function was improved. Intra fascicular dissection with nerve graft improved hand function in 1 patient. Surgery performed on 14 patients with macrodactyly for functional and cosmetic concerns included digital amputation (8 cases), epiphysiodesis (4 cases), and wedge osteotomy (3 cases). Digital amputation is usually reserved for severe cases refractory to debulking procedures. In the growing child, epiphysiodesis can be performed to limit longitudinal growth, but this did not prevent circumferential expansion. Tan *et al.* have suggested that middle phalangectomy with arthroplasty may result in a functional joint^[68].

Follow up

Follow-up was reported in 116 cases (64%), ranging from 3 weeks to 26 years^[69]. Symptomatic relief was most successful in patients undergoing carpal tunnel release, and neurological symptoms improved after surgical decompression. However, long-term follow-up often showed a progressive decline in sensory and motor functions, regardless of treatment modality.

Summary

LFH of the median nerve is a rare condition, with 180 reported cases in the English and French literature between January 1946 and November 2012. One third of cases are associated with macrodactyly, with no gender predominance. Patients are often affected at or shortly after birth and present before age 30 years.

Presenting symptoms

- Tumor along the distal median nerve distribution, with or without macrodactyly
- Compressive median neuropathy, carpal tunnel syndrome.

Differential diagnoses

- Ganglion cysts
- Lipoma
- Hemangiomas
- Traumatic neuroma

Diseases with malignant potential:

- Neurofibroma
- Schwannoma
- Liposarcoma
- Malignant peripheral nerve sheath tumor

Diagnosis

- MRI is the single recommended diagnostic modality, with characteristic findings
- X Fusiform median nerve swelling with bulging flexor retinaculum, displacing flexor tendons within the carpal tunnel
- X Tortuous low-intensity signal nerve bundles embedded in hyperintense fatty tissue

- Coaxial cable sign on axial plane and spaghetti-like features on coronal and sagittal sections

Biopsy, though diagnostic, is not routinely recommended owing to potential functional deficits. It shows normal-appearing nerve fibers embedded in abundant mature adipocytes with interlacing fibrous septa. Invasive diagnostic procedures are not warranted in the presence of pathognomonic MRI findings.

Treatment

- Asymptomatic patients: observation or prophylactic carpal tunnel release for large tumors
- Neurological symptoms: carpal tunnel decompression with or without neurolysis
- Macroductyly: ray or digital amputation, wedge osteotomy, middle phalangectomy with arthroplasty or epiphysiodesis
- Options to be tailored to individual needs
- Timing of treatment for macroductyly requires careful balancing of cultural, psychosocial, functional, and aesthetic factors

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