

Lipofibromatous hamartoma of the median nerve with macrodactyly of middle finger

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ABSTRACT

Lipofibromatous Hamartoma (LFH) is a rare tumour affecting the peripheral nerves, mostly the median nerve. It is benign and slowly growing. Less than 100 cases have been reported in the literature, about one quarter of them are associated with macrodactyly in the field of the affected nerve.

We reported a case of a 13 years old female with LFH of the median nerve at the level of the wrist associated Macroductyly of the ipsilateral middle finger treated surgically by decompression only.

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Introduction

Lipofibromatous hamartoma (LFH) is a rare, benign and slowly growing tumour affecting the peripheral nerves, characterized by anomalous overgrowth of fibro adipose tissue between and around nerve bundles causing enlargement of the affected nerve,¹ giving the pathognomonic (cable like) appearance on MRI.²

The median nerve and its branches are most commonly affected, but the lesion has also reported in the ulnar, radial, plantar and peroneal nerves.³

Although LFH is considered to be congenital in origin, some consider it as a hamartoma caused by nerve irritation or inflammation.⁴

Affected patients usually presented with gradually enlarging non tender mass in the distribution of the affected nerve. Sometimes, in cases of median nerve lesion, patients presented with symptoms and signs of carpal tunnel syndrome complaining from numbness and tingling along the volar aspect of the wrist and hand and finally with motor deficit which indicate the late stage.⁵

Sometimes, LFH is associated with Macroductyly and lipomatous macro-dystrophy of muscles and subcutaneous fat in the territory of the nerve.⁶

We reported a case of LFH of left median nerve at the level of wrist associated with macroductyly of middle finger in a 13 years old female.

Case report

A 13 years old female presented with a single painless longitudinally oriented mass in the central proximal region of the left palm with macroductyly of the ipsilateral middle finger (Fig. 4a).

The mass was noticed by the family first time at the age of six years, after that the mass and macroductyly started growing slowly without pain or any other associated symptoms. Past medical, surgical, drug and family history was non-significant.

On examination, the 2*4 cm mass was soft, non-tender, longitudinally oriented with slight longitudinal movement by fingers flexion and extension. No any motor or sensory deficit, no muscle atrophy, no any sign of inflammation and no other palpable mass or any pigmented skin lesion elsewhere in the body.

Laboratory investigations were all within normal. Ultrasound shows Isoechoic lesion 2.8 * 2.5 cm at the palm. Hypertrophied muscular or fatty tissue had been suggested. Radiograph was normal apart from enlargement of the middle finger maximally in the distal phalanx (Fig. 1).

MRI reported hypertrophied non-enhancing fatty tissue of mid palm region along the tendon of flexor muscle and extend to the subcutaneous fat with macroductyly of middle fingers, features suggestive of macrodystrophia lipomatosa (Fig. 2). With-out

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Fig. 1. Plain radiograph, Hypertrophied phalanges of the left middle finger.

mentioning any point about median nerve. On these bases, surgical exploration was recommended.

Under general anesthesia, tourniquet around the left arm, a longitudinal incision was made on the wrist and proximal palm, the mass noticed protruding from below the transverse carpal ligament, when the last divided, an enlarged median nerve was observed, then to facilitate exploration, the wound was extended proximally and distally (Fig. 3). The nerve was yellow and pink,

with gross fatty infiltration. Carpal tunnel was decompressed and biopsy taken but nerve debulking and excision of the infiltrating fatty tissue was avoided due to the high risk of neuronal injury.

Histological examination revealed a fibrofatty tissue separating the nerve fascicles confirming the diagnosis of LFH.

Follow up was done in two occasions, the first after one year and the second after two years and nine months from the surgery, the patient was still neurologically asymptomatic and pain free, But with some progression in her middle finger macrodactyly, the matter that affecting her self-esteem increasingly (Fig. 4b,c).

Discussion

LFH is a rare condition caused by hypertrophy of mature fat and fibroblasts in the epineurium, resulting in fibrofatty infiltration leading to peripheral nerve enlargement. The most commonly affected one is the median nerve, in association with macrodystrophia lipomatosa characterized by macrodactyly.⁷ And this was typical in our presented case.

The first case of LFH with similar course was reported by Mason in 1953,⁸ the followed by two cases of median nerve also published by Mikhail in 1964.⁹

The term of Lipofibromatous hamartoma was first coined by Johnson and Bonfiglio in 1949 after reviewing the literature and putting up their own case with detailed histological description of the lesion.¹⁰

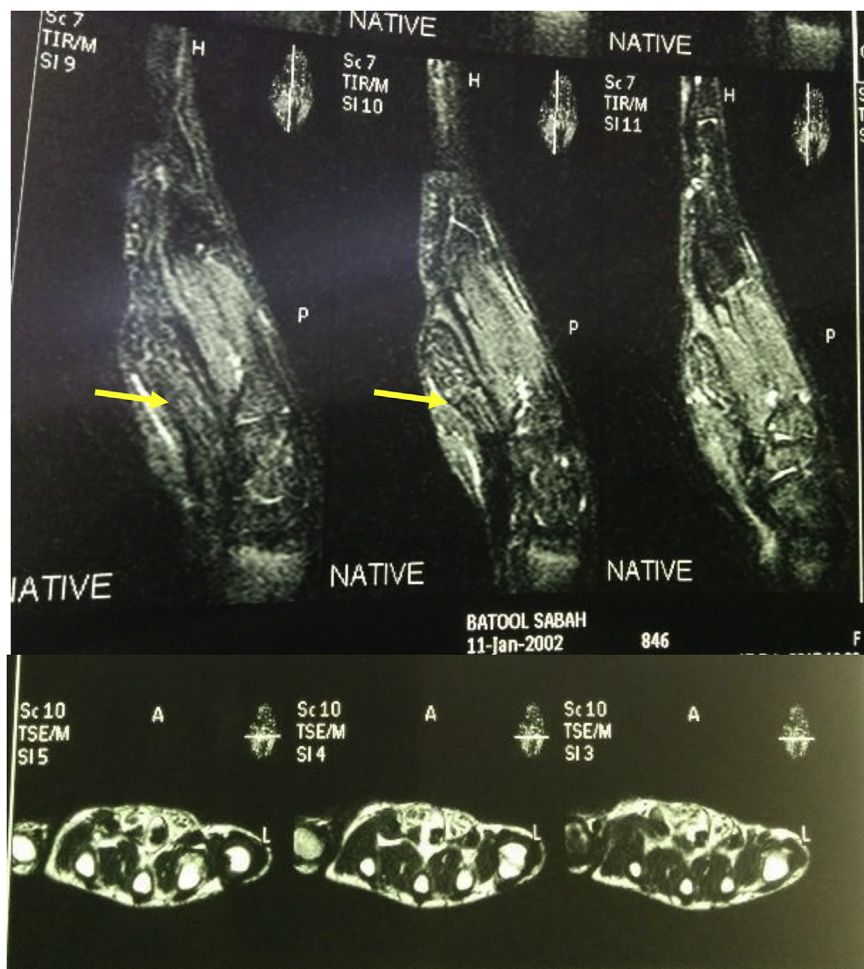


Fig. 2. Sagittal and axial section MRI, Enlarged cable like median nerve.

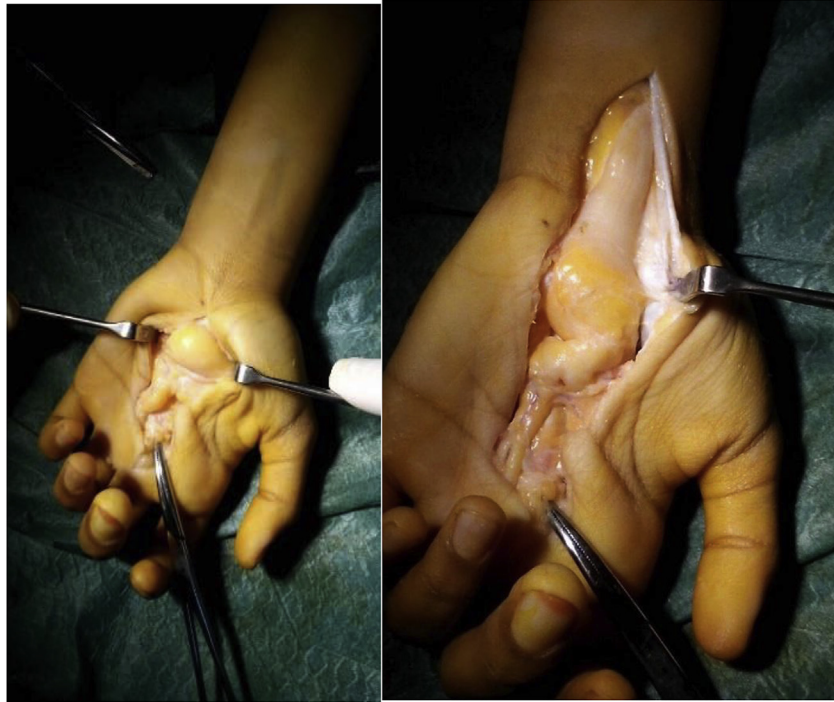


Fig. 3. Intraoperative picture, enlarged median nerve.

Then in the following 40 years, only 100 cases of LFH of median nerve have been documented, 25% of it was associated with macrodactyly, mostly in female.¹¹

It is most commonly occur in male Caucasians, but predominance in females is noted in presence of macrodactyly.¹²

It is unknown why median nerve is the most commonly involved, there are suggestions that long standing micro trauma to the median nerve from the carpal ligament is the initiative cause. This theory is similar to that of traumatic neuroma pathogenesis, however LFH is usually noted beyond the carpal ligament¹³ and this what been observed intraoperatively in our case, abnormally enlarged median nerve was extended about 10 cm proximal and few centimeters distal to the transverse carpal ligament.

On the other side, if the repetitive microtrauma is the only responsible mechanism, so the lesion should be expected in other sites within the body such as the lateral femoral cutaneous nerve under the inguinal ligament, but this is not the case.¹³

While other authors explained the condition by congenital abnormal growth of adipose tissue of the nerve sheath.¹⁴

LFH is very rarely reported in the lower limbs. Silverman and Enzinger reported 26 cases, from which 25 cases were in the hand and wrist while only one case in the foot. In 22 of the 25 cases, the median nerve was affected.¹⁵

Amadio et al. also reviewed 17 cases of LFH of the nerve, only 4 of them were located in the lower limbs, while 11 cases were in the median nerve or its branch-es.¹⁶

In LFH, perineurium and epineurium surrounding nerve branches become infiltrated by disproportionate amount of fibroadipose tissue, causing fusiform enlargement of the specific nerve¹⁴ and giving the typical MRI picture of coaxial cable appearance on axial view and spaghetti appearance on coronal and sagittal views.¹⁴

On clinical aspect, LFH of median nerve usually presents in children or young patients as a palmar mass, either asymptomatic or causing compression neuropathy with severe pain, paresthesia and thenar motor weakness.¹⁵

The differential diagnoses are: intraneural lipoma, diffuse lipomatosis, traumatic neuroma, fibromatosis, haemangioma, plexiform neurofibroma and ganglion cyst. The differentiation is usually easy due to the gross and histological characteristics.³

Ultrasonographic study shows smooth, rounded and thickened hypoechoic or anechoic fascicles surrounded by echogenic fatty tissue compatible with the histological findings. There is no intralésional flow can be seen in color Doppler.¹⁷

The diagnosis can be made by the aid of sonography and MRI without taking biopsy.¹⁷ But this need a very high index of suspicion and clinical orientation.

Microscopic findings are fibrofatty expansion of the epineural space with splaying of the nerve bundles, perineural and endoneurial fibrosis. Sometimes, in long standing cases, metaplastic bone is seen in the fibrofatty tissue.¹⁵

Many operative procedures have been used in the treatment of LFH of the median nerve, the recommended procedures are carpal tunnel release with or without superficial removal of epineural proliferation, microsurgical interfascicular dissection or excision of the affected nerve segment with or without grafting.¹⁶

Warhold et al. wrote that carpal tunnel release alone can lower the tumour size, decrease pain severity and improve opposition strength. They reported sensory deficit after attempted surgical excision of the tumour although other authors reported minimal disability in young children, may be due to cerebral compliance.¹⁸

In their survey, Amadio et al. shows that every case treated by nerve excision and neurolysis developed an abnormality in 2 points discrimination even though it was normal preoperatively.¹⁶

Nowadays, the standard management for most of the surgeons is restricted to a limited biopsy taking with carpal ligament release.¹⁹

In our case, we preferred to do only decompression and take biopsy, avoiding debulking due to the high expected risk of neuronal injury.

Finally, no definite treatment is available for LFH.



Fig. 4. Clinical hand picture a: Before surgery, b: 12 months after surgery, c: 21 months after surgery.

Decompression and biopsy taking for final diagnosis is the ideal current management.¹⁹

Disclosures

The authors have no financial disclosures or conflicts of interest to declare.

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