Lipofibromatous hamartoma of a digital branch of the median nerve: a case report and review of the literature

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Abstract

Lipofibromatous hamartoma is an uncommon tumor that causes nerve enlargement due to fatty adipose tissue infiltration in peripheral nerves being the median nerve at the carpal tunnel the most commonly affected site, typically associated with macrodactyly. Here we present a case which affected a digital branch of the median nerve

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Running title: Lipofibromatous hamartoma of a digital branch of the median nerve

Key Clinical Message: Lipofibromatous hamartoma (LFH) is an uncommon and benign tumor that causes nerve enlargement due to fatty adipose tissue infiltration around the bundles of peripheral nerves, being the median nerve at the carpal tunnel the most commonly affected site, typically associated with macrodactyly. Here we present an uncommon case of a LFH which affected a digital branch of the median nerve of the middle finger without macrodactyly.

Key words: Lipofibromatous hamartoma, Median nerve, Median nerve tumor, Macrodactyly, Nerve compression, Peripheral nerve

Introduction

Lipofibromatous hamartoma (LFH) is an uncommon and benign tumor that causes nerve enlargement due to fatty adipose tissue infiltration around the bundles of peripheral nerves. Clinical presentation may vary but the main symptoms are associated with neural compression and the most common affected site is the median nerve. The diagnosis can be made due to the imaging changes observed with ultrasonographic and magnetic resonance image (MRI), but when in doubt, biopsy of the lesion is needed.[1,2] Due to the lack

of clinical studies there is no a standardized treatment, therefore observation, carpal tunnel release, nerve decompression or excision of the lesion can be performed.[1,2] We report a case of an 18- year-old female with a LFH of a digital branch of the median nerve of the middle finger of the left hand and a review of the current literature.

Case report

A 18-year-old female without any medical condition or genetic disorder presented after she had a fall from a horse and had a forced traction of the middle finger of the left hand with the reins of the horse.

The patient presented with insidious burning pain and numbness located in the dorsal part of the proximal interphalangeal joint of the middle finger. No fractures were seen on radiographs, therefore analgesic treatment was provided. After 4 months the pain was constant and worsened with direct pressure of the proximal interphalangeal joint. She denied having a fever or losing weight.

In the physical exam there was a diffuse mass on the ulnar and dorsal side of the proximal phalanx of the middle finger of the left hand of approximate 1 x 1 cm. The mass was not adherent to the skin and it was mobile (figure 1). Range of motion of the metacarpophalangeal joint was 90^{o} of flexion, 0^{o} of extension. The range of motion of the proximal interphalangeal joint was 15^{o} of flexion, 0^{o} of extension and of the distal interphalangeal joint 20^{o} of flexion, 0^{o} of extension. She had pain at palpation and a positive Tinel's sign at the ulnar and dorsal side of the proximal phalanx of the middle finger. There was not hyperthermia, skin changes or any palpable lymph nodes. The sensibility was normal and there were no paresthesias.

Radiographs were performed which showed a circular radiolucent image measuring 6.5 x 5 mm with thinning of the posterior cortical of the proximal phalanx of the middle finger. (figure 2A)

An ultrasound of the finger was performed, which showed vascular trajectories with ecstatic flux and venous morphology that communicate with an arterial branch that condition cortical bone irregularities. (Figure 2B)

An MRI with gadolinium contrast was made, which showed a lesion of 36 x 5 x 11 mm located on the dorso of the proximal phalanx which displaced the extensor tendons. After gadolinium administration, the lesion was enhanced. There was a focal discontinuity of the cortical bone of 1 mm with one ectasical vascular trajectory. (figure 2C, 2D)

An incisional biopsy of bone and soft tissue with a Jamshidi needle was performed. Which showed a benign lesion constituted by fibrofatty tissue.

Definitive surgery was undertaken 2 weeks later in which a complete excisional treatment was decided. We debride and resected an irregular and gray tumor of 4.3 x 3 x 0.4 cm (figure 3A) located on the neurovascular bundle of the ulnar side of the proximal phalanx of the middle finger. (Figure 3B) The bone had a gray-like color on the dorsal part of the proximal phalanx and a focal discontinuity of the cortical bone of 1 mm of diameter. (Figure 3C) A window of 20 x 10 mm was created to observe the medullary canal, which had a yellowish and grey-like tissue which was removed. The medullary canal was obliterated on the proximal part. Finally, the bone window and the surgical wound were closed.

The histopathological examination showed a benign lesion constituted by fibrofatty tissue which compressed the nerves fascicules with overgrowth of the rest of connective constituents, proliferation of blood vessels, myofibroblastic proliferation and collagen deposition. (Figure 4A and 4B) Immunohistochemistry was performed with S100 which highlighted nerve fascicles compressed by the tumor. No necrosis or atypical mitosis was found. (Figure 4C)

At the latest follow up (7 months) the patient has no pain on the surgery site. Ranges of motion of the finger are preserved. She has no paresthesias but continues with numbness on the dorsal part of the proximal interphalangeal joint of the middle finger.

Discussion

LFH is an uncommon tumor that affects the peripheral nerves, where fibroadipose tissue infiltrates the epineurium and perineurium.[1] The most common age of presentation is in the 30s to 40s. The median nerve is affected in 80% of the cases, but other nerves like the brachial plexus, cranial, ulnar, radial, sciatic, peroneal, plantar and digital nerves can be affected.[2,3] Signs and symptoms depend on the location of the tumor. It can present as a compressive neuropathy including sensory and/or motor deficits on the nerve distribution. Pain and tenderness also may be associated.[2] One third of the cases are associated with macrodactylia. [4]

In 1953, Mason first described the LFH.[5] In 1985 Silverman and Ezinger studied 26 cases of LPH in which the main age distribution was between the 3rd and 4th decade of life. There were 25 cases in the wrist and hand while only one case was at the toe.[6] It most commonly affects male caucassians, but females are most commonly affected if macrodactylia is present.[7] Macrodactyly is a common finding in 20-66% of the cases. [8,9]

The cause of abnormal growth of the nerve remains unknown.[2] It is also unknown why the median nerve is the most commonly affected.[7] Some authors believe that repetitive microtrauma is the reason why mature fat cells and fibroblasts of the epineurium hypertrophy. [2,7.] Other authors believe that there is an unknown congenital abnormality affecting the growth of fibrofatty tissue. [10]

There are no pathognomonic findings for this disease.[2] History of trauma, increasing pain, tenderness, diminished sensation, paraesthesia with enlarging mass causing compression neuropathy may be present. Carpal tunnel syndrome is a late complication seen in some cases. [1,2,4,6,7,10-12]

The differential diagnosis include ganglionic cysts, hereditary hypertrophic neuritis of Dejerine-Sottas syndrome, vascular malformations, traumatic neuromas, schwannomas, neurofibromas and intraneural lipomas, fibromatosis, plexiform neurofibroma.[2,7]

True lipomas can be differentiated because they are sharply demarcated, well-encapsulated masses that normally occur on the surface arising in the perineurium or epineurium and contain no neural elements.[2]

Radiographs may only be helpful to view macrodactyly changes and soft tissue opacity. [12]

Diagnosis of the fibrolipomatous Hamartoma can be made with sonography and MRI without the need of biopsy. If there is familiarity with the features of the ultrasound, the MRI and biopsy may be obviated.[13]

The Sonographic findings show smooth, rounded, thickened hypoechoic or anechoic fascicles surrounded by echogenic fatty tissue. There is no flow on color doppler. [14]

The fusiform nerve enlargement and thickened axonal bundles may be seen on the MRI. Because of the epineural fibrous tissue that encapsulates the bundles, the axial images give a ¨coaxial cable¨ like appearance and ¨spaghetti¨ like appearance on coronal images.[15]

Electromyography and nerve conduction studies can be useful to confirm compressive neuropathy.[14]

In LFH the diffuse involvement of the nerve showing relative bland changes is consistent with a hamartomatous rather than a neoplastic tumour. The lesion is described as hamartoma because of the overgrowth of the connective tissue constituents. The proportion of fibrous tissue to fat varies from case to case[15.] Histologically the lesions are characterized by connective and adipose mature tissue that surrounds and infiltrates a major nerve trunk and infiltrates its perineurium and epineurium. The differential diagnosis include lipomatous neurofibroma, lipofibromatosis, diffuse lipomatosis and intraneural perineurioma.[16]

Due to the context of a benign tumor there are different recommendations for the treatment of this pathology. These include observation, biopsy, intraneural neurolysis and/or carpal tunnel release. [2]

In asymptomatic patients different authors recommend observational treatment as the first option, due to tumoral regression.[12,17]

In symptomatic patients there are different surgical procedures. Carpal tunnel release alone can lower the tumor size, decrease pain severity and improve opposition strength. [16,18]

Patients treated with nerve excision and neurolysis may develop sensitory deficit in 2 points discrimination. [6,7,9-11]

Mohamed et al. Mention that nowadays the standard management for most of the surgeons is restricted to a limited biopsy taking with carpal tunnel release. [7]

Amadio et al. had 14 cases and concluded that microsurgical dissection was not clinically more successful than the radical excision.[8]

Nanno et al. had a similar case in which the ulnar digital nerve of the thumb was affected. They did a limited resection of the tumor to prevent excessive damage to the nerve. At three years of follow-up there was no recurrence of the mass nor neurological deficits.[19]

Steentoft and Solerman had a similar case in which they used a microscope and partially resected the tumor without compromising the digital nerve. At two years follow up the patient had numbness of the index finger.[20]

Conclusions

In conclusion, LFH is a rare condition that has different clinical presentations. Evidence suggests that the diagnosis can be made with an ultrasound and an MRI without the need of a biopsy. In asymptomatic patients the treatment can be observational, on the other hand if the tumor is symptomatic surgical excision can be made. In our case the tumor was symptomatic, it presented in an atypical location and was not associated with macrodactyly, so we decided on a complete excisional treatment. This pathology still needs more research and patient follow-up to understand the pathophysiology and to dictate the best treatment option based on the patient characteristics.

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Figure 1. A: Diffuse mass on the ulnar and dorsal side of the proximal phalanx of the middle finger of the left hand of approximate 1 x 1 cm. B: Purple color of the proximal interphalangeal joint

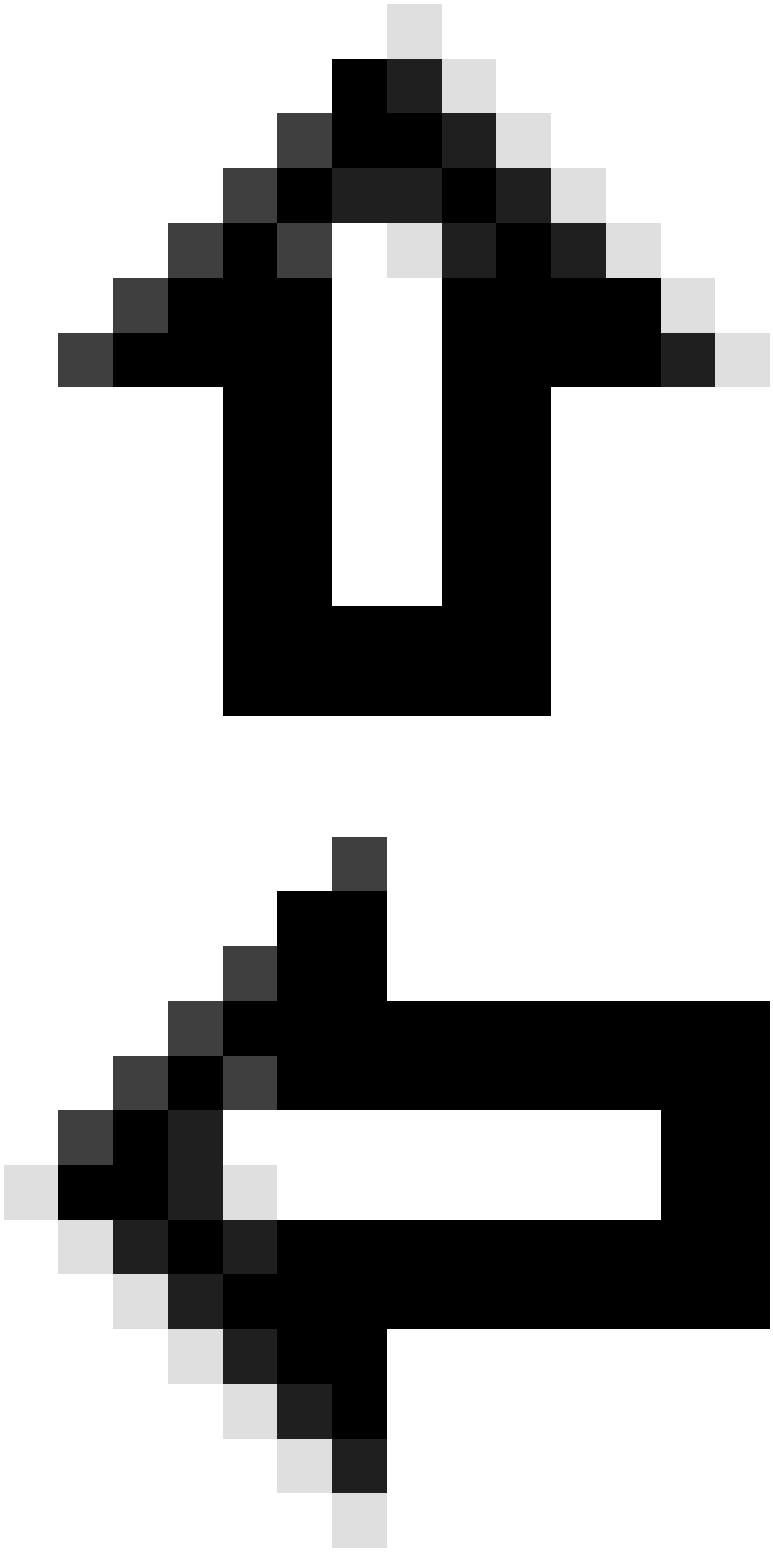

Figure 2. A: Anteroposterior, lateral and oblique radiographs of the middle finger showing a circular radiolucent image with thinning of the posterior cortical (). B: Ultrasound of the middle finger. Dorsal side of the middle finger with ectasial venous flux communicating with an arterial branch can be seen (*). C: MRI of the middle finger without gadolinium. A tumor of 36 x 5 x 11 mm located on the dorsal aspect of the proximal interphalangeal joint which displaced the extensor tendons can be seen (). D: MRI of the middle finger after gadolinium administration. The lesion enhances, there is a focal discontinuity of the cortical bone of 1mm (), with one ectasical vascular trajectory ().

Figure 3. A: Irregular and gray tumor of 4.3 x 3 x 0.4 cm located on the ulnar side of the proximal phalanx of the middle finger. B: Tumor within the neurovascular bundle of the ulnar side of the proximal phalanx of the middle finger. C: gray-like color on the dorsal part of the proximal phalanx and a focal discontinuity of the cortical bone of 1 mm of diameter.

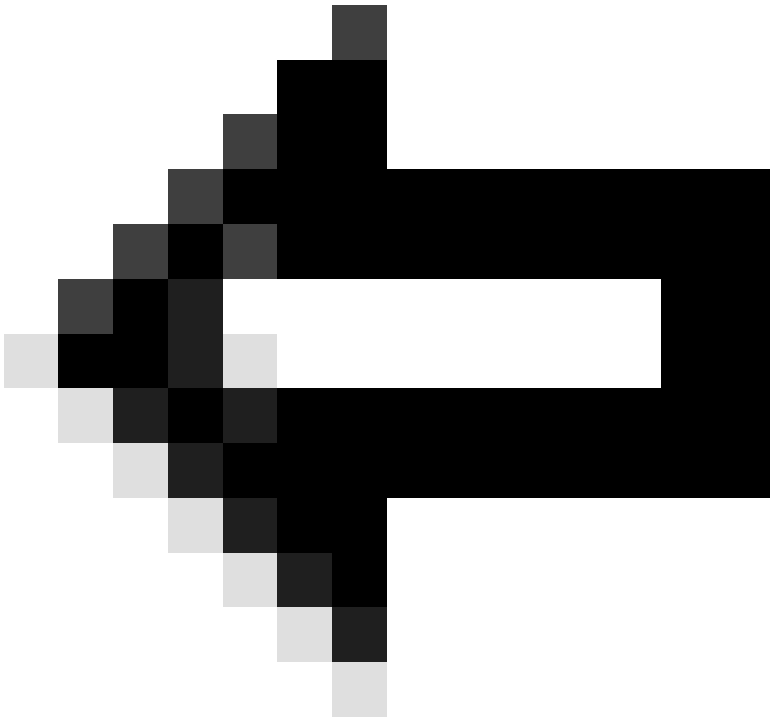

Figure 4 A: HE 4x. Histopathological findings. There is a fibrofatty tissue and connective proliferation () compressing nerves bundles (). B: HE 40x. Nerves fascicules () compressed by overgrowth of connective tissue (). C: S100 immunohistochemistry, highlights the Schwann cells in fascicules nerves ().