Lipofibromatous hamartoma of the digital branches of the median nerve presenting as carpal tunnel syndrome: A rare case report with review of the literature

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ABSTRACT
Lipofibromatous hamartoma (LFH) is a rare, benign fibrofatty tumor composed of a proliferation of mature adipocytes within peripheral nerves, which form a palpable neurogenic mass. It affects the median nerve in 66–80% of cases, causing pain and sensory and motor deficits in the distribution of the affected nerve. Patients typically present with gradually enlarging nontender lesions in the distribution of the affected nerve. The lesion is also seen to be associated with macrodactyly. The pathophysiology of LFH is unknown. Treatment of LFH is based on symptoms of the condition. Histopathology is characteristic. We present a case of young male diagnosed as lipofibromatous hamartoma of the median nerve involving the right index finger. The case is presented due to its rarity.

KEY WORDS: Index finger, lipofibromatous hamartoma, macrodactyly, median nerve

INTRODUCTION
Lipofibromatous hamartoma (LFH) of the nerve is a rare and benign tumor characterized by proliferation of mature adipocytes within the peripheral nerves resulting in a palpable mass. It affects the median nerve in majority of the cases, causing carpal tunnel syndrome like features.\(^1,2\) Due to overlapping findings this entity needs to be differentiated from other similar entities. Accurate diagnosis is mandatory for definitive treatment. Only a handful of cases are available in the literature, so the exact treatment pattern is not well documented. The management of the case depends on the clinical presentation. A case of lipofibromatous hamartoma of the digital branches of the median nerve in a young male is presented.

CASE REPORT
A male aged 21 years presented with pain in the right index finger for 4 years. According to the patient, he got hurt while playing with a ball and then he developed a swelling of the right index finger. The swelling was progressively increasing and now he developed pain. He even has limited flexion and extension movements of the finger. There was no H/o numbness. His past medical and family histories were insignificant. There was no evidence of macrodactyly.

Examination showed a bulbous swelling of the proximal phalanx of the right index finger. The mass was adhered both to the skin surface as well as the deeper tissues. The sensations were normal. X-Ray showed a soft tissue mass [Figure 1]. Preoperative fine needle aspiration and cytology (FNAC) suggested lipoma. A decompression surgery was performed and the resected specimen was sent for histopathology.

Grossly, a linear yellowish-white tissue piece measuring 4 cm in length and firm in consistency was received. Microscopy showed multiple mesenchymal components including blood vessels, adipose tissue, fibrous tissue, and nerve bundles. The adipose tissue was arranged in a lobular pattern separated by fibrous septae. The blood vessels were numerous and small in size. At places, thick walled blood vessels were observed with perivascular fibrous tissue proliferation [Figure 2]. The cut – section of the nerve bundles showed increased perineural thickening.

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Adipocytes were seen surrounding the nerve bundles [Figure 3]. The diagnosis of neural fibrolipoma or LFH of nerves was thus made. The postoperative period was uneventful. He recovered his motor deficit significantly 2 weeks after surgery.

**DISCUSSION**

LFH was first described in the English literature in 1953. In 1969, Johnson and Bonfiglio coined the term LFH, accurately describing the entity and its relation to carpal tunnel syndrome. Several terms have been used to describe this condition including fibrolipomatous hamartoma, intraneural hamartoma, neural fibrolipomatosis, and neural fibrolipoma. As per the World Health Organization, this entity is described as lipomatosis of the nerves.

Adipocytes are normally present in the perineurium and epineurium. LFH of the nerve is a rare soft tissue tumor, which occurs due to fibro-fatty proliferation within the nerve bundles with massive epineural and perineural fibrosis leading to fusiform enlargement of the nerve. It affects the median nerve in 66–80% of cases, causing pain along with sensory and motor deficits along the distribution of the affected nerve. While there is an unexplained predilection for the median nerve, involvement of the brachial plexus, ulnar, radial, peroneal, plantar, sciatic, and digital nerves have also been reported. LFH of the nerve is a rare soft tissue tumor, which occurs due to fibro-fatty proliferation within the nerve bundles with massive epineural and perineural fibrosis leading to fusiform enlargement of the nerve. It affects the median nerve in 66–80% of cases, causing pain along with sensory and motor deficits along the distribution of the affected nerve. While there is an unexplained predilection for the median nerve, involvement of the brachial plexus, ulnar, radial, peroneal, plantar, sciatic, and digital nerves have also been reported. Patients typically present with gradually enlarging nontender lesions in the distribution of the affected nerve. There is numbness and tingling along the volar aspect of the wrist and hand. Motor deficits occur late. LFH may be associated with macrodactyly or macrodystrophia lipomatosa, particularly affecting the index and the middle fingers.

Most cases occur within the first three decades of life, with the mean age of 22.3. There is a 2:1 female to male ratio of cases with macrodactyly and a 1:1 ratio in cases without. Although there have been suggestions of a congenital origin to LFH, the etiology remains unclear. Cases arising after trauma have also been documented. It has been suggested that persistent microtrauma to the median nerve from the carpal ligament or external pressure from a flexor retinaculum can initiate a reactive process which induces this tumor. Traumatic neuromas also share similar pathogenesis, but the only difference is that LFH usually extends beyond the carpal ligament. It has also been suggested that like in neurofibromatosis, a dysgenetic disorder may be responsible.

The differential diagnosis includes ganglion cyst, vascular malformation, traumatic neuroma, schwannoma, neurofibroma, and lipoma. LFH shares considerable gross overlap with neurofibromatosis requiring both radiologic and microscopic investigation for accurate differentiation. The importance of this is apparent as LFH is a benign tumor, whereas neurofibromatosis can progress to frank malignancy. Where LFH displays uniform fatty infiltration, intraneural lipomas show focal fatty masses separated from the individual nerve bundles. There are no known laboratory studies which aid in the diagnosis of LFH. FNAC usually suggests a soft tissue tumor as in the present case. Sonographic studies can support the diagnosis of LFH but have to be supplemented with other diagnostic modalities for confirmation. The magnetic resonance appearance of this condition is unique and pathognomonic. Thickened nerve fascicles embedded in evenly distributed fat appear as serpentine low-intensity structures surrounded by high-intensity signal in both T1- and T2-weighted images giving the coaxial cable-like...
appearance in the axial plane and spaghetti-like appearance in the coronal planes.[6]

LFH tumors are irregular, yellow masses. Microscopy demonstrates nerve bundles that are entrapped within a fibrofatty-fusiform mass. LFH involves a disorganized overgrowth of the epineurium, perineurium, and endoneurium with fatty infiltration. There is no inflammation or involvement of the surrounding tissues. Lipomatous masses have been divided into four types according to their location within the parent nerve-soft tissue lipoma, intraneural lipoma, macrodystrophia lipomatosa, and LFH.[9]

Treatment of LFH depends on the presenting symptoms. While some cases cause no neurologic or functional complications, others do. Management is usually conservative.[4] In patients presenting as carpal tunnel syndrome, decompression surgery has been shown to provide adequate relief of symptoms.[3,10] Excision of the nerve is not recommended. Furthermore, extensive intraneural dissection is to be avoided.

CONCLUSION

Because of the infrequency of the diagnosis, no standard treatment protocol has been established. With more reports of LFH, further insight will be gained into appropriate management modalities.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES