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CASE REPORT

Lipofibromatous Hamartoma of Median Nerve: A Case Report

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Introduction

Lipofibromatous hamartoma (LFH) is an extremely uncommon, benign slow-growing tumor of the peripheral nerve. Mason first reported this tumor in 1953, since then many cases are reported[1,2]. Median nerve in carpal tunnel is one of the common sites of occurrence[3]. World Health Organization (WHO) designates this tumor as Nervous lipomatosis[4]. It is also known as fibro-fatty proliferation, intraneural lipoma, unusual tumor of the median nerve, and lipomatous hamartoma[5]. We describe a short case report of this rare tumor and the literature is briefly reviewed.

Case Report

A 35- year- old lady presented with two year history of progressive worsening pain and numbness in left hand. She had left hand grip weakness for last 6-months. On local examination there was a firm swelling over the volar aspect of the distal forearm extending across the wrist joint. The swelling was painful on slightest of touch. Neurological examination revealed hypoesthesia in the left hand in the median nerve distribution. There was wasting of the thenar group of muscles and the abductor pollicis brevis was markedly weak. She was operated at another hospital after performing an ultrasonography (USG) of the swelling. A biopsy was performed on exploration and histopathological diagnosis of neurofibroma was made. She then presented to our clinic with severe pain in the left hand which was hampering her day to day activities and night sleep. The magnetic resonance imaging (MRI) revealed a classical serpiginous, cable like appearance of the neural fascicles of the median nerve which had hyperintense signal on T1- weighted sequences and hypointense signals on T2 - weighted sequences. This was suggestive of fatty infiltration of the neural cables (Fig 1a & 1b). The Electrophysiological studies (EMG, NCV) revealed complete left median nerve neuropathy below wrist with denervation in the small muscles of hand supplied by median nerve on the left side. A preoperative diagnosis of Lipofibromatous hamartoma (LFH) involving median nerve was considered prior to surgical intervention. The tumor was excised and defect was bridged with multiple sural nerve grafts (Fig 2a & 2b). The histopathological examination confirmed the tumor to be LFH. It demonstrated diffuse infiltration and expansion of epineurium by the fibro-fatty tissue (Fig 3a & 3b). She was pain free at 3-month follow up. A post operative MRI scan confirmed complete excision of LFH involving the median nerve (Fig 4).

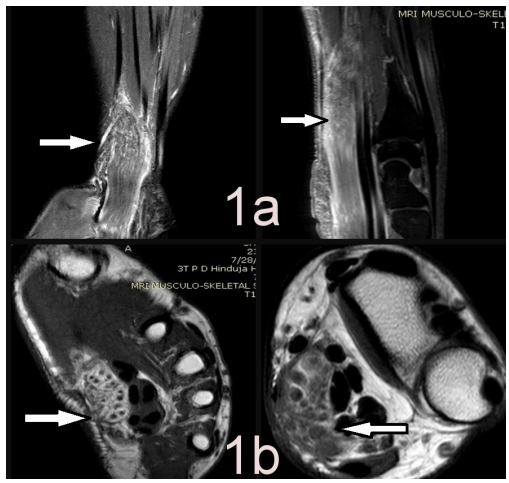

Fig. 1 (a & 1b) Axial & Coronal MR Images of the lesion (White arrows) which show classical serpiginous, cable like appearance of neural fascicles of the median nerve and evidence of fatty infiltration of the neural cables.

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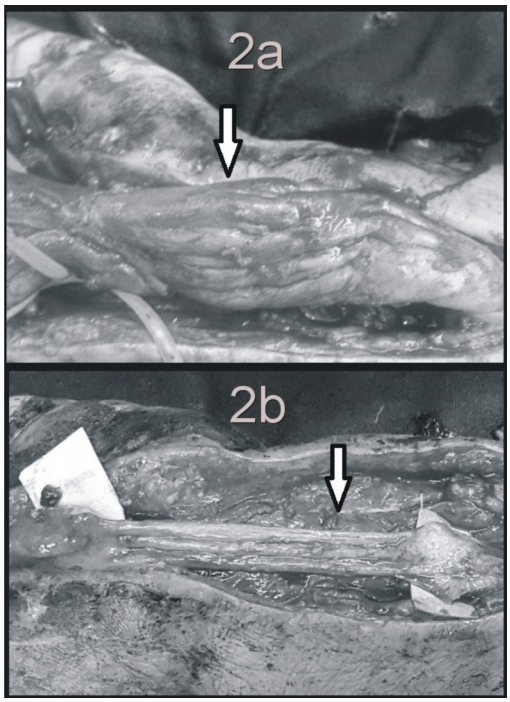

Fig. 2 (a) Intraoperative appearance of an orange yellow, fusiform, sausage like or ropelike enlargement of the median nerve. **(b)** Total excision of the lesion with multiple sural nerve cable grafts.

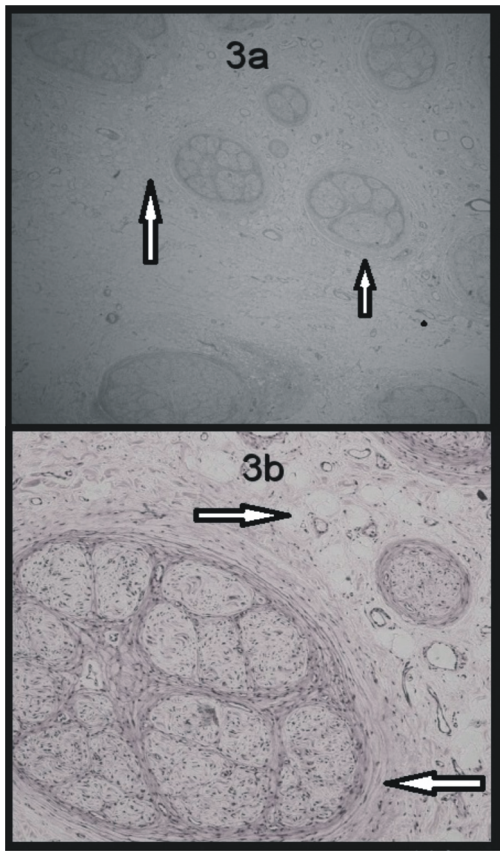

Fig. 3 (a&b) Microscopic view of diffuse infiltration and expansion of the epineurium by adipose tissue.

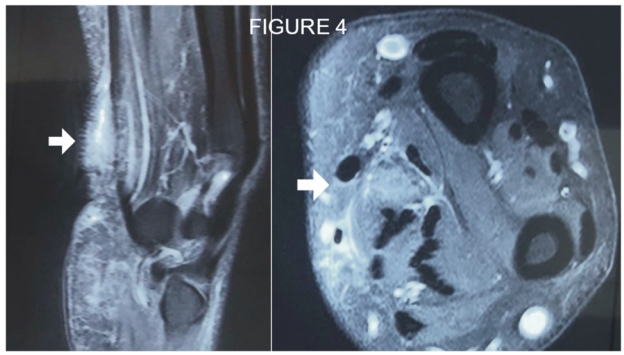

Fig. 4 Post contrast T1 weighted saggital and axial sequences show postop changes at the operative site without any evidence of tumour.

Discussion

LFH is a rare benign overgrowth of fibroadipose tissue within a nerve sheath. It comprises of mature fibrous, fatty and neural elements and hence termed hamartoma[1,2,3]. The etiology is unknown and its predilection for the median nerve is unexplained. LFH typically presents in 3rd to 4th decade of life with history of painful swelling and a progressive neuropathy[6]. LFH has also been reported in the radial nerve, ulnar nerve, nerves on the dorsal aspect of foot, brachial plexus, and cranial nerves. Macrodactyly is present in approximately two thirds of the cases[7].

Macroscopically it is orange yellow, fusiform, sausage like or rope like enlargement of involved nerve. Cross sections show classical epineural and perineural fibrofatty infiltration compressing individual nerve bundles[7]. MRI shows the characteristic cable sign on axial images and spaghetti-like appearance on coronal sections[8].

The treatment of LFH is highly controversial especially in presence of preserved or minimally deficient neurological function. In median nerve LFH with relatively preserved neurological function, carpal tunnel decompression or internal decompression with excision of fibrofatty tissue and neural decompression is an ideal surgical option. However, often internal decompression is not possible without significant neurological insult postoperatively due to vascular compromise. In patients with severe preoperative neurological deficits involving median nerve, complete excision with nerve grafting is advocated. Post-operative neurological worsening in such cases is not severe and there is significant relief in pain and parasthesia as was also noted in our patient. Presence of Martin-Gruber anastomosis in the distal forearm helps in reducing the neurological deficits

postoperatively. In our patient the median nerve function was markedly affected prior to surgery and it did not worsen further following the surgery.

Conclusion

LFH is a benign condition and awareness of this condition by clinician, radiologist and a pathologist is essential to avoid misdiagnosis as it happened in our case. Management is controversial since the lesion cannot be completely excised without sacrificing the involved nerve. It is therefore essential to take decision based on the clinical signs and symptoms and natural history of the disease progression.

Conflicts of Interest Disclosure

The authors have no conflict of interest or any financial disclosures to make.

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