Clinical Notes

Lipofibromatous hamartoma in the ulnar nerve of the forearm

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Lipofibromatous hamartoma (LFH) is a rare, benign, and slow-growing peripheral nerve tumor. It was first reported by Mason in 1953.[1] The LFH is composed of proliferating mature adipocytes within peripheral nerves, which form a palpable neurogenic mass. The LFH has been termed fibrofatty overgrowth, lipomatous hamartoma, lipofibroma, fibrolipomatous hamartoma, or fatty infiltration of the median nerve. In more than 80% of the reported LFH cases, tumors affecting the median nerve in which sensory and motor deficits were observed.[2] A few cases have been reported in the brachial plexus, ulnar, radial, peroneal, plantar, and digital nerves. The understanding of this peripheral nerve tumor remains limited. In a LFH, it is common to see that over proliferated fibro-fatty tissue penetrates through the epineural and perineural membrane of peripheral nerves. This gives rise to a characteristic appearance on MRI and US images. Here, we report a patient with an extensive LFH in the ulnar nerve of the forearm, and suggest that high-resolution US can provide a reliable evaluation to establish the preoperative diagnosis of LFH.

A 43-year-old male presented with a 6-month history of paresthesia in his left ring and little fingers. On examination, slight atrophy of the hypothenar eminence and a 2 x 2 cm subcutaneous mass on the volar ulnar of the left wrist were observed. The mass was positive for Tinel's sign and appeared to be soft, movable, and nonpulsatile. The US examination revealed enlargement of the ulnar nerve with hypoechoic linear serpentine fibers interspersed between the hyperechoic signal, which is suggestive of fatty infiltration. The mass appeared as a "lotus rhizome" on transverse US scans and presented a spaghetti-like pattern on longitudinal US scans (**Figures 1A & B**). Thus, his clinical presentation, and the US images collectively suggested a diagnosis of LFH.

Surgical exploration was performed under brachial block anesthesia with an upper arm tourniquet. Briefly, a zigzag incision was made extending from the ulnar distal forearm to the wrist. The ulnar nerve was found to be grossly enlarged from the distal forearm to the wrist crease line. Longitudinal fatty streaks were seen within the nerve fibers, which appeared to be yellow and pink. These fatty streaks extended from the distal two-thirds of the forearm to the proximal edge of the Guyon's tunnel. Following division of the ligament of the Guyon's tunnel, the ulnar nerve was also enlarged and infiltrated by fatty tissue. The epineurium was thick. An epineurotomy was performed and revealed severely infiltrated fatty tissue and fibrosis between the ulnar fascicles. The fatty tissue between the nerve fascicles was excised as much as possible for histological examination, and the ulnar nerve was adequately decompressed. Histological analyses showed that the perineurium was infiltrated by mature fibers of nonneoplastic fibro-fatty tissues and that the nerve fascicles were separated by

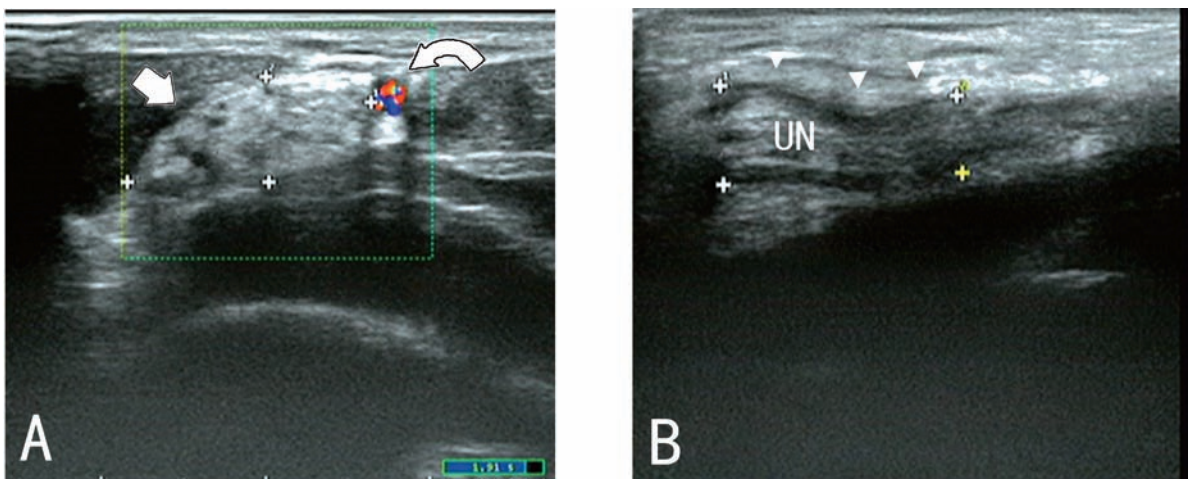

Figure 1 - Patient ultrasound showing **A)** A transverse 5-12 MHz US scan obtained at the forearm shows the typical 'lotus rhizome-like' appearance of the swollen ulnar nerve (UN) (straight arrow and + signs), and nearby the ulnar artery (curved arrow). **B)** A longitudinal 5-12 MHz US scan within the Guyon's tunnel shows a 'spaghetti pattern' appearance and abrupt narrowing of the UN between the thickened floor of the tunnel and the retinaculum (arrowheads and + signs). Proximal to the Guyon's tunnel, the nerve appears swollen with an absent fascicular pattern.

adipose tissue. A diagnosis of LFH was made based on the histological results. After operation, his symptoms were obviously improved. No sign of recurrence was observed at the 12-month follow-up examination.

Lipofibromatous hamartoma of the nerve is a rare soft tissue tumor, which occurs due to fibro-fatty proliferation within nerve bundles with massive epineural and perineural fibrosis, leading to fusiform enlargement of the nerve. The LFH is slow-growing, benign, and often found in the median nerve, causing pain, tenderness, diminished sensation, paresthesia, or thenar motor weakness. Sometimes, patients with LFH may develop secondary carpal tunnel syndrome. Cases of extraneural fibromas causing compression neuropathy of peripheral nerves have also been reported in the literature. The LFH patients usually suffer from gradually increasing swelling, which sometimes is associated with symptoms of compression neuropathy. Most LFHs occur in the first 3 decades of life. Rare cases have been reported later in life. Histologically, LFH should be differentiated from neurofibroma, traumatic neuroma, perineurioma, sclerosing perineurioma, and intraneural lipoma.

Before 2010, only 88 cases of LFH had been reported in the literature.[3] Since November 2011, a total of 180 cases have been reported in the English or French literature.[2] Most cases involved the median nerve in the distal forearm, wrist, or palm, and presented with carpal tunnel syndrome. Involvement of the ulnar nerve is rare.[4,5] Here, we report a patient with an extensive LFH in the ulnar nerve of the forearm and the Guyon's tunnel. Our patient presented progressive weakness and paresthesia in the ulnar nerve distribution of his left hand. The patient had a positive Tinel's sign on the ulnar nerve. These symptoms were consistent with Guyon's tunnel syndrome. Based on clinical symptoms and physical examination, it is extremely difficult to differentiate LFH from some entities such as neurilemmomas, neurofibromas, and intraneural lipomas. The US examination showed a characteristic coaxial cable-like appearance, which is suggestive of LFH. Therefore, a diagnosis of LFH was suggested by the US findings and confirmed by the histological examination on the surgical tissues. Awareness of the existence of such a rare tumor secondary to Guyon's tunnel syndrome may lead to early evaluation and treatment.

The diagnosis of LFH can be made using MRI or US. Current high-resolution US allows sonologists to visualize most major nerves, appreciate fascicular structure of the nerves, distinguish extraneural tumors from endoneural tumors, and easily monitor the progression of nerve lesions through the whole clinical course.[6] The characteristic appearance of LFH on MRI and US images is excessively proliferated fibroadipose tissue infiltrating the epineural and perineural components of the peripheral nerves. In our study, the typical lotus rhizome-like appearance of the swollen ulnar nerve and a spaghetti-like nerve organization were found. The preoperative diagnosis of LFH was made based on the US findings. Therefore, according to the US images, surgery was guided to the target site for a proper operation and provided a biopsy for diagnosis confirmation by histological examination. Although MRI is generally considered a more reliable approach to reach the diagnosis of LFH, current high-resolution US could also provide characteristic manifestations for this condition. Moreover, compared with MRI, high-resolution US is widely accessible due to its relatively low cost. It is also convenient to perform, and has fewer limitations due to the patients' physical condition.

In conclusion, our experience on this case suggested that US can be applied for preoperative evaluation of LFH because it is capable of revealing the characteristic features of LFH and providing sufficient diagnostic information. Therefore, high-resolution US could be an alternative diagnostic imaging technique to MRI for patients with suspected LFH, especially for those who have limited access to an MRI system.

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