CASE REPORT

Lipofibromatous hamartoma of the superficial peroneal nerve: Two case reports

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A B S T R A C T

Lipofibromatous hamartoma, a rarely occurring nerve hamartoma, can present as an acrochordon, cutaneous cyst or other soft tissue tumor and is usually seen within the first three decades of life. The lesion presents as a slowly growing mass that is largely composed of fat and fibrous tissue with epineural and perineural proliferation. Although such tumors are rare, it is important for physicians to be aware of this disorder and recognize its signs because patients may present with what appears to be a benign skin tumor. If left untreated, the lesion may result in nerve compression and eventually lead to the development of peripheral neuropathy. Here, we present two cases of lipofibromatous hamartomas that presented over the ankle and dorsal foot, respectively, that appeared as simple and benign tumors upon initial inspection. Patients were without symptoms or neurological deficits, and diagnosis was not made until histopathological examination of the biopsied specimens. We also discuss the clinical manifestations, histopathological findings, and the management of lipofibromatous hamartomas.

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Introduction

Lipofibromatous hamartoma (LFH), also known as neural fibrolipoma or lipomatosis of the nerve, is a relatively rare, benign, slow-growing, and subcutaneous soft tissue mass that usually affects the nerves of the upper extremities; it is rarely found in the foot. LFHs are composed of adipocytes and fibrous tissue, both around and within the peripheral nerve. It has long been considered a hamartoma because the fatty, fibrous, and neural components are all essentially mature tissue. LFHs most commonly occur in the median nerve and its branches, followed by the ulnar nerve. Cases involving the radial, digital, and plantar nerves have also been reported. LFHs arising from the sciatic nerve or superficial peroneal nerve are extremely rare. Neural fibrolipomas are associated with the overgrowth of bone and macrodactyly in about one-third of cases and are known as macrodystrophia lipomatosa. Here, we report two cases of LFHs that involved the superficial peroneal nerve.

Case reports

Case 1

A 30-year-old male presented with numbness and radiating pain in the first and second toes of the right foot when walking for 1 year prior to examination. The patient denied having a history of trauma or macrodactyly of the toes. Physical examination revealed a 2 cm × 1 cm subcutaneous soft mass with faint erythema and mild tenderness over the right ankle area; another, smaller mass (1 cm × 0.5 cm) was found beneath the larger mass (Figure 1A). In order to establish a diagnosis, the smaller mass was biopsied. Microscopically, scattered hypertrophic nerve bundles, prominent perineural fibrosis, and focal proliferation of the adipocytes in the deep dermis were noted (Figure 1B, 1C, 1D). In addition, a mild increase in the number of small vessels at the level of the mid-dermis was seen. Based on the histopathological findings, a diagnosis of LFH was made. The smaller lesion was excised, and the pain experienced during walking improved. No recurrence was noted in the 8-month follow-up period. The patient declined intervention for the larger lesion because there were no specific symptoms.

Case 2

A 26-year-old male presented with a mass on his right dorsal foot that he reported had been rapidly growing for the previous...
6 months. He denied any history of trauma. Examination revealed a 2.5 cm × 1.5 cm, soft, non-tender, subcutaneous mass over the anterolateral aspect of the right dorsal foot (Figure 2A). No neurological deficits were noted. The initial clinical impression was angiofibromatous hamartoma or another type of soft tissue tumor, so an incisional biopsy was performed. Microscopically, there was a large caliber nerve fiber that contained thickened perineurial fibrous tissue that was surrounded by fatty tissue proliferation (Figure 2B). A small number of nerve fibers were scattered within the adjacent area, demonstrating concentric perineural fibrosis, that were embedded in a small amount of adipose tissue (Figure 2C). These findings were compatible with a diagnosis of LFH. The patient had no postoperative complications, and at 7-month follow-up had no signs of tumor recurrence.

Discussion

The development of LFH includes the proliferation of mature fatty and fibrous tissues within the epineurium and perineurium of a major nerve. The components of the lesion may vary in proportion, thus leading to a variety of histological expression patterns that can make diagnosis difficult. Lesions affecting the lower limbs are exceptional rare. Previous reports have documented extremely rare cases arising from the superficial peroneal nerve (Table 1).4,6,7,20,21 Both of the cases presented here were located over the anterolateral aspect of the dorsal foot, corresponding to the location of the superficial peroneal nerve (Figure 3). In case 1, the patient complained of numbness and pain radiating into the first and second toes when walking, which
corresponds to the distribution of the medial cutaneous branch of the superficial peroneal nerve.

Differential diagnosis for LFH on physical examination includes tenosynovitis, ganglion cyst, lipoma, and the presence of neurogenic tumors such as traumatic neuroma, schwannoma, or neurofibroma. However, the combination of a mobile, soft, slowly growing mass that follows the distribution of a nerve and causes pain only after compression seems to be unique for LFHs. A true lipoma should be differentiated both clinically and pathologically. Lipomas rarely induce compression neuropathy, even when surrounding a peripheral nerve. In addition, lipomas are encapsulated and easily separated from the surrounding tissue. By contrast, the infiltration of fibroadipose tissue within and between the nerve fascicles often occurs with LFHs, excluding the possibility of their complete removal. Microscopically, lipomas may contain remarkable fibrous tissue, the so-called fibrous lipomas, but the neural elements and characteristic fibroadipose tissue that separate nerve bundles are always absent.

The pathogenesis of LFH is unknown. Some tumors are believed to be congenital without an underlying family history. Antecedent trauma and chronic nerve irritation are regarded as precipitating factors. These may explain its greater prevalence in patients who are young and its predilection for the extremities, which are easily damaged. Neither of the two cases presented were associated with Proteus syndrome or Klippel-Trénaunay-Weber syndrome, both of which have been linked to LFH.

Several reports have indicated that magnetic resonance imaging (MRI) may demonstrate the pathognomonic features of LFHs, including the serpentine-like appearance of fibroadipose tissue intermingled with nerve fascicles, a coaxial cable-like appearance on axial sections, and a spaghetti-like appearance on coronal images. However, MRI is not always reliable. Talal et al. reported a LFH that was initially misdiagnosed as a ganglion cyst on MRI. We believe that MRI should be initially performed for typical cases, such as lesions involving the median nerve that typically result in carpal tunnel syndrome, and this may eliminate the need to perform a biopsy. As for cases that do not have typical presentations, diagnostic biopsy may be essential for diagnosis even with the use of imaging studies. In case of a suspicious recurrence, MRI is also recommended.

The treatment options for LFHs are still controversial. Conservative treatments and invasive surgery, including debulking, external or internal neurolysis, or radical excision with or without fascicular grafts, have been described. Treatment should be individualized based on the clinical presentation. For LFHs causing prominent neuropathy, open biopsy with external nerve decompression can be performed and this may prevent further nerve injury. Nevertheless, for small lesions without neuropathy, limited excision is the mainstay of treatment (this can also be performed for diagnostic purposes). The two patients described here both had good clinical outcomes without complications following simple excision. Based on our experience, conservative excision with preservation of the nerve function is recommended when internal neurolysis with meticulous microdissection under high magnification is unavailable or impossible.

Most patients will seek treatment from orthopedic doctors or surgeons; only a small number of patients will visit a dermatologist. The cases seen by dermatologists are usually small lesions that do not present prominent compression neuropathies. Dermatologists should become familiar with this condition because, although rare, if untreated the mass will continue to grow and eventually lead to neuropathy.
In conclusion, this study describes two rare cases of LFHs of the foot. Awareness of LFHs and knowledge of their clinical and histopathological characteristics are necessary for correct diagnosis and timely and proper intervention. MRI is recommended for lesions with a typical presentation, or if there is a suspicious recurrence and may obviate the need for diagnostic biopsy and the risk of iatrogenic nerve damage. Long-term follow-up is mandatory due to the possibility of disease progression or recurrence.

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References