Evaluation and management of fibrofatty tumors of the extremities: case report

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Patients presenting with enlarging fibrofatty masses in the extremities pose an interesting dilemma to clinicians, as the differential diagnosis in such cases ranges from benign to malignant, and from lesions optimally managed operatively to those managed nonoperatively. The differential diagnosis includes benign lipoma, liposarcoma, lipoblastoma, and fibrolipomatous hamartoma (lipomatosis) of the nerves. The authors present the case of a 14-year-old girl with an enlarging fibrofatty mass of the forearm, initially thought, based on diagnostic imaging, to be a fibrolipomatous hamartoma of the median nerve, but found to be a lipoblastoma without direct nerve involvement based on histopathological examination of the operative specimen. This case serves to illustrate the diagnostic predicament that can exist with such masses.

The authors advocate the need to establish a tissue diagnosis while having a contingency plan for each of the diagnostic possibilities because the management of each lesion is markedly different. In this report, the authors consider the differential diagnosis of fibrofatty masses of the extremities that the peripheral nerve surgeon may encounter, and they highlight the significant differences in management strategies for each possible diagnosis.

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Radiographic studies are often helpful when classifying fibrofatty tumors of the extremities, but they are not always diagnostic. This presents a dilemma with regard to evaluation and management, particularly because each diagnostic possibility has markedly varying management strategies. The differential diagnosis for fibrofatty tumors of the extremities includes benign lipoma, liposarcoma, lipoblastoma, and fibrolipomatous hamartoma (lipomatosis) of the nerves. We present the case of a 14-year-old girl who presented with a fibrofatty mass of the forearm, thought to be a fibrolipomatous hamartoma of the median nerve, but found to be a lipoblastoma without direct nerve involvement based on histopathological examination of the operative specimen. We review the differential diagnosis of fibrofatty masses of the extremities that the peripheral nerve surgeon may encounter and highlight the significant differences in management strategies for each possible diagnosis.

Case Report

History
A 14-year-old girl with no significant medical history presented with a right forearm mass initially noted 6 years prior to presentation. She reported that the mass had doubled in size in the previous 6 months. She denied pain and weakness but indicated she felt an intermittent tingling sensation overlying the mass.

Examination
The patient appeared to be healthy, with full strength in all muscle groups of the right upper extremity. Sensation was intact in response to both light touch and pinprick throughout the forearm and hand. Palpation of the mass elicited pain radiating into Digits 1 and 2. There was a large, soft, rubbery mass visible and palpable deep in the forearm. Radial, ulnar, and brachial pulses were 2+.  

Electrodiagnostic Examination
Nerve conduction studies and electromyography demonstrated no abnormalities.

Imaging
Within the soft tissues of the right forearm was noted a 12.6 × 6.2 × 4.2–cm lobulated fat-containing mass (Fig. 1). The majority of the mass demonstrated signal alterations similar to fat on all MRI sequences. Multiple T1-weighted
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FIG. 1. Left: Sagittal and axial T1-weighted fast spin-echo MR images demonstrating a large hyperintense forearm mass, consistent with a fatty lesion. The lesion contains numerous thin septations. The median nerve (arrow) is seen within the periphery of the mass. Right: Sagittal and axial postcontrast T1-weighted fast spin-echo MR images with fat saturation. The mass appears mostly hypointense due to suppression of fat signal. There are numerous enhancing septations within the mass, as well as a few areas of enhancing nodularity (arrows).

hypointense, T2-weighted hyperintense septations were seen throughout the mass. The surrounding musculature was displaced but was free of abnormal signal. Similarly, no abnormal signal was seen in the adjacent osseous structures. The median nerve was flattened along the course of the mass. Based on the MR images, the lesion was thought to be a fibrolipomatous hamartoma of the median nerve, although the differential diagnosis included lipoblastoma, benign lipoma, and liposarcoma.

Operation

Although significant thought was given to the literature recommending conservative management for fibrolipomatous hamartoma, the rapidly enlarging nature of the mass ultimately prompted exploration of the median nerve, decompression at the pronator teres, and biopsy of the mass for tissue diagnosis. The median nerve was identified at the antecubital fossa and then dissection was carried distally following the nerve. Dissection revealed that the median nerve was adjacent to and compressed by the mass; the mass did not appear to originate from the nerve. The lesion appeared well circumscribed and noninvasive. A biopsy sample was sent for frozen section analysis, which showed no malignant features or nerve elements, but the analysis was otherwise unable to establish a more definitive diagnosis. Based on these findings, fibrolipomatous hamartoma of a nerve and liposarcoma were thought to be unlikely. Thus, we proceeded with en bloc resection of the mass, which was easily separated from surrounding structures.

Pathological Examination

Final pathological examination revealed a benign fatty tumor with an infiltrative pattern and mature, single vacuolated adipocytes (Fig. 2). Occasional foci had myxoid mesenchymal-appearing cells, and one nodule in particular exhibited chondroid morphology with calcification. Only rare multivacuolated adipocytes were seen. No nerve fibers admixed with fatty lobules were identified. The overall findings were diagnostic of mature lipoblastoma.

Follow-Up

Immediately following surgery, the patient had full strength in the median innervated muscles. Given the final diagnosis of mature lipoblastoma, no adjuvant treatment was pursued. Rather, the patient is being followed up clinically and radiographically for signs of recurrence. At 3 months following resection, she continued to have full strength in the median nerve–innervated muscles and showed no clinical signs of recurrence.

Discussion

Patients presenting with enlarging fibrofatty masses in the extremities pose a diagnostic and therapeutic quandary, as the differential diagnosis ranges from benign to malignant and from lesions optimally managed operatively to those managed nonoperatively. The differential diagnosis includes benign lipoma, liposarcoma, lipoblastoma, and fibrolipomatous hamartoma of nerves. Other conditions/lesions that can be confused with fibrolipomatous hamartomas include ganglion cyst, traumatic neuroma, neurofibroma, and Dejerine-Sottas syndrome. However, these lesions are not typically fibrofatty. In our patient, the imaging characteristics and clinical scenario were thought preoperatively to be most consistent with a fibrolipomatous hamartoma of the median nerve.

Fibrolipomatous hamartomas comprise hypertrophied nerves secondary to fibrofatty ingrowth interspersed between thickened nerve bundles. These should be distinguished from intraneural lipomas, in which a well-encapsulated fatty mass displaces nerve fibers that then course along the periphery of the mass. These lesions occur most commonly in association with the median nerve. Most fibrolipomatous hamartomas grow over time, with some doubling in size within a 2-year period.
Younger age has been shown to be associated with higher growth rates. Controversy exists over optimal management of fibrolipomatous hamartoma, though more recently, consensus has suggested that attempts at resection may actually worsen pain and neurological function and do not change the growth pattern.10 The potential for worsening pain and neurological function following resection is thought to occur for two reasons. First, because fibrolipomatous hamartomas are hypertrophied nerves secondarily to fibrofatty infiltration, resection of the mass necessarily means resection of nerve tissue. Second, resection is thought to induce a vigorous fibrotic reaction that can secondarily damage nerve elements. Consequently, a conservative approach is often favored but may include biopsy if the diagnosis is unclear. There are several radiological features that are thought to be pathognomonic for fibrolipomatous hamartoma of a nerve including a coaxial-cable-like appearance on axial imaging and a spaghetti-like appearance on coronal imaging.11 Nerve bundles throughout the lesion appear hypointense on both T1- and T2-weighted images.4 Fat distribution and content varies from lesion to lesion, but the fat is typically distributed either between nerve fibers or surrounding nerve fibers peripherally. In our patient, the pathognomonic features were not present, but the lesion was still thought to most likely be fibrolipomatous hamartoma of the median nerve. Thus, because the diagnosis was in doubt but fibrolipomatous hamartoma was in the differential diagnosis, our initial plan was to proceed with exploration, decompression, and biopsy of the lesion to confirm diagnosis and then manage the patient conservatively if fibrolipomatous hamartoma was confirmed.

Intraneural lipomas may be confused with fibrolipomatous hamartoma of a nerve. While in fibrolipomatous hamartoma the fat is usually between nerve bundles or surrounding them, the nerve fibers typically course on the periphery of a well-encapsulated fatty mass in intraneural lipoma. Similar to fibrolipomatous hamartoma, the most common location for intraneural lipoma is within the median nerve.13 The distinction is important, however, because the management is wholly different. Total excision of intraneural lipomas can be undertaken when symptomatic without posing undue risk to the involved nerve.

Liposarcoma should also be considered in the differential diagnosis of fibrofatty masses of the extremities. Liposarcoma, while uncommon, has a predilection for the extremities and the retroperitoneum. Some imaging features can be suggestive of liposarcoma rather than a benign lipoma, including calcification, thickened irregular septations, associated nonadipose tissue, and swirling or nodularity.6 Sarcoma guidelines suggest that 4 features are important in assessing the likelihood of malignancy: 1) increasing size, 2) size greater than 5 cm, 3) location deep to the deep fascia, and 4) associated pain.5 When these features are present, involvement of a sarcoma specialist should be considered, as treatment typically involves resection with clear margins followed by postoperative radiotherapy.

Finally, as was the diagnosis in our case, lipoblastoma should be considered in the differential diagnosis of fibrofatty masses of the extremities. Lipoblastomas are rare, benign mesenchymal tumors of embryonic fat origin. Most lipoblastomas are diagnosed in patients under the age of 3 years and have a predilection for the extremities.3 Whereas lipoblastomas usually occur in individuals prior to age 3 years, liposarcomas rarely occur prior to age 10 years and usually occur in adulthood with a peak incidence at 50–65 years of age.1,8,9 MRI features that can help distinguish lipoblastomas from other fatty masses include nonenhancing cystic changes and enhancing soft-tissue nodules. Lipoblastomas are benign-behaving lesions but can grow rapidly. In general, the treatment of choice is...
complete excision. Unlike liposarcoma, negative margins are less important, though subtotal resection may increase the likelihood of local recurrence. While no adjuvant therapy is needed, follow-up is important, given the possibility of local recurrence.

Diagnosis of fibrofatty masses of the extremities can sometimes be difficult without tissue diagnosis. Our case demonstrates some of the diagnostic difficulty, highlights recent literature pertaining to the differential diagnosis, and suggests a potential management strategy. At 14 years of age, our patient was older than the typical age for lipoblastoma and younger than the typical age for liposarcoma. The rapid growth rate suggested that a typical, benign lipoma was unlikely. The mass was intimately involved with the median nerve, although the typical MRI features of coaxial-cable-like appearance on axial imaging and the spaghetti-like appearance on coronal imaging usually seen in fibrolipomatous hamartoma of the median nerve were absent. Taken together, fibrolipomatous hamartoma of the median nerve was thought to be most likely. However, this case illustrates the diagnostic quandary that can exist with such masses. We advocate obtaining tissue diagnosis while having a contingency plan for each of the diagnostic possibilities, as the management of each lesion is markedly different. In the case of fibrolipomatous hamartoma of a nerve, resection should not be attempted; rather, the nerve should simply be decompressed. In the case of a lipoblastoma, a conservative attempt at complete resection should be made, and postoperative follow-up for local recurrence for 5 years should be observed. Similarly, resection should be attempted for symptomatic intraneural lipomas, but postoperative follow-up is unnecessary. Finally, in the case of liposarcoma, a sarcoma specialist should be involved from the outset. Treatment involves complete resection with negative margins and usually includes postoperative adjuvant radiotherapy.

References
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