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Pearls and Pitfalls in the Imaging of Soft-Tissue Masses in Children

Oscar M. Navarro, MD

There is a broad spectrum of soft-tissue masses in children that can be challenging to diagnose clinically and on imaging. This article reviews the typical clinical and imaging findings of the most common and relevant benign, intermediate and malignant pediatric soft-tissue tumors in the following categories of the 2013 World Health Organization (WHO) classification: adipocytic tumors (lipoma, lipoblastoma, and liposarcoma), fibroblastic/myofibroblastic tumors (nodular fasciitis, myositis ossificans, fibrous hamartoma of infancy, fibromatosis colli, desmoid-type fibromatosis, lipofibromatosis, and infantile fibrosarcoma), pericytic tumors (myofibroma/myofibromatosis), skeletal muscle tumor (rhabdomyosarcoma), nerve sheath tumors (neurofibroma, malignant peripheral nerve sheath tumor), and uncertain differentiation (synovial sarcoma). In general, ultrasound and magnetic resonance imaging are used as first- and second-line imaging modalities, with limited roles for plain radiographs, computed tomography, and fluorodeoxyglucose-positron emission tomography. Many of these tumors have nonspecific imaging findings although there are some key imaging clues that in conjunction with the clinical information allow a specific diagnosis or a narrow differential diagnosis. However, in many instances, histology is required for final diagnosis.
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Introduction

Soft-tissue masses are a common occurrence in children and many are diagnosed clinically based on the clinical history and findings on the physical examination. However, in a good number of cases the diagnostic work-up of these masses requires further investigation with imaging examinations. This can be a challenge for radiologists because soft-tissue masses can be caused by a broad spectrum of conditions, often with nonspecific clinical manifestations, and in many instances also present with nonspecific imaging findings.

This article does not attempt to be a comprehensive review of soft-tissue masses in children but intends to highlight those clinical and imaging findings that are helpful in the diagnosis of selected soft-tissue masses that are more relevant in children and mention some pitfalls that can lead to an erroneous interpretation.

Department of Medical Imaging, University of Toronto, Department of Diagnostic Imaging, The Hospital for Sick Children, Toronto, Ontario, Canada.

Address reprint requests to Oscar M. Navarro, MD, Department of Diagnostic Imaging, The Hospital for Sick Children, 555 University Avenue, Toronto, ON M5G 1 × 8, Canada E-mail: oscar.navarro@sickkids.ca

Classification of Soft-Tissue Masses

The most widely accepted classification system for soft-tissue tumors in children is that proposed by the World Health Organization (WHO), last revised in 2013.[1] This classification includes the following categories of tumors: adipocytic, fibroblastic/myofibroblastic, so-called fibrohistiocytic, smooth-muscle, pericytic (perivascular), skeletal muscle, vascular, chondro-osseous, nerve sheath, uncertain differentiation, and undifferentiated/unclassified.

Imaging Approach

Ultrasound is in general the preferred first-line imaging modality for the investigation of pediatric soft-tissue masses owing to its accessibility and relatively low cost, and to the advantages it offers in children (no need for sedation or use of anesthesia, no use of radiation, and no need for contrast agent injection). Moreover, many soft-tissue masses present as superficial lumps and bumps that can be easily imaged in their entirety with ultrasound.

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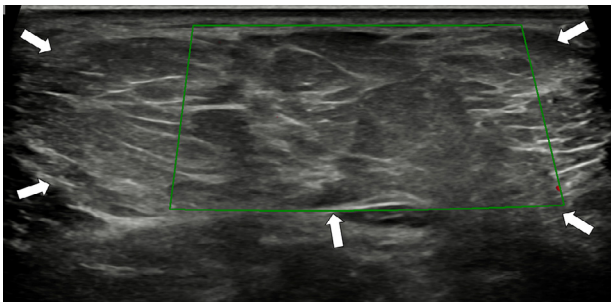

Figure 1 Lipoma in a 6-year-old boy with a slowly growing mass in the left axilla since age 2 years. Transverse color Doppler ultrasound image shows a subcutaneous mass (arrows) that is predominantly hypoechoic and contains multiple hyperechoic lines that course mostly parallel to the skin surface, similar to normal subcutaneous fat. There is no detectable internal vascularity.

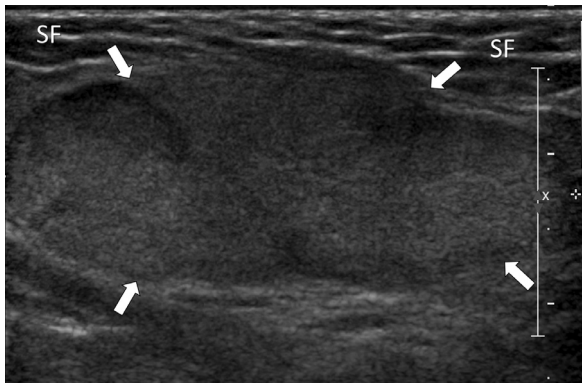

Figure 2 Lipoblastoma in a 16-month-old girl with a growing soft mass in the left inguinal region. Longitudinal ultrasound image shows a well-defined, lobulated deep subcutaneous mass (arrows) that is predominantly hyperechoic as compared to subcutaneous fat (SF).

Ultrasound is usually performed using high-frequency linear-array transducers but it may require additional use of lower frequency curved-array transducers when dealing with larger and deeper lesions. Gray-scale imaging, color or power Doppler, and spectral Doppler analysis are all required in most cases in order to demonstrate the solid or cystic components of the mass, presence of calcifications, type and degree of vascularity, and involvement of one or more soft-tissue planes as well as extension into neighboring structures.

Contrast-enhanced ultrasound has been used experimentally in the evaluation of soft-tissue masses, mainly in adults. It has been found that lack of enhancement or homogeneous enhancement of a mass is typically seen with benign and intermediate lesions. Heterogeneous enhancement can be seen with malignant lesions but does not allow its differentiation from benign or intermediate lesions.[2]

If the ultrasound examination is not conclusive or cannot completely assess the lesion, magnetic resonance imaging (MRI) is often used as a second-line imaging modality. However, in cases of very small superficial lesions that are well

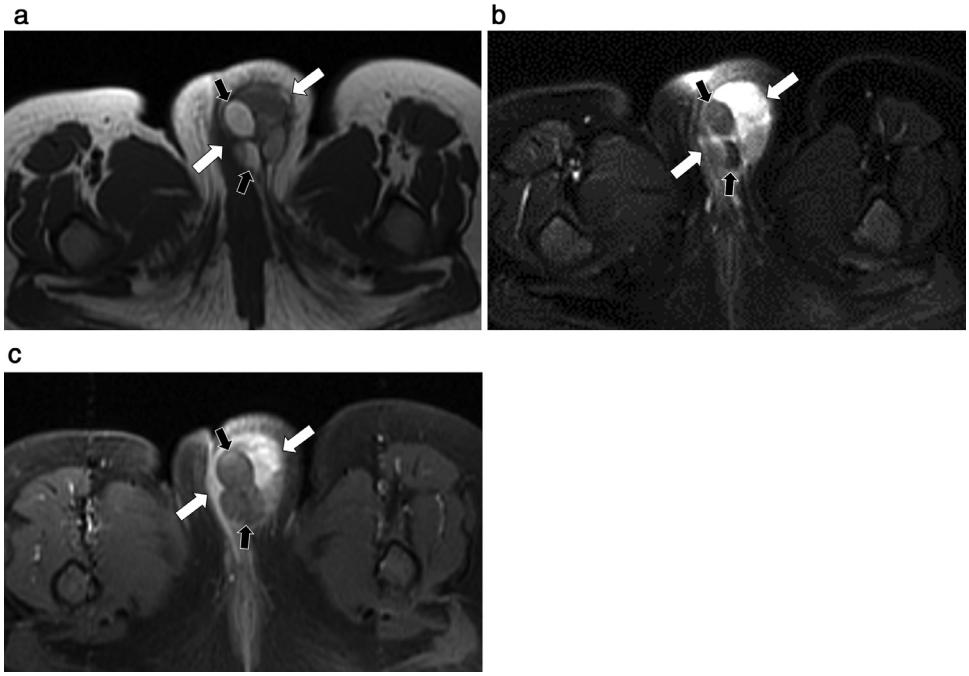

Figure 3 Lipoblastoma in a 13-month-old girl with a left labial mass. (a) Axial T1-weighted, (b) fat-suppressed T2-weighted, and (c) contrast-enhanced fat-suppressed T1-weighted MR images show the usual heterogeneous appearance of lipoblastoma. The left labial subcutaneous mass (white arrows) shows mature adipocytic component (black arrows) with signal intensity similar to subcutaneous fat, that is, hyperintense in (a), hypointense in the fat-suppressed images (b, c), and without significant contrast enhancement (c). The rest of the mass does not show typical features of fatty tissue reflecting the presence of lipoblasts, fibrous septa and myxoid stroma that are also part of lipoblastomas and allow differentiation from lipoma on imaging.

outlined with ultrasound, MRI can be obviated as surgical excision can be the next step for final histologic diagnosis and treatment.

MRI can sometimes be used as first-line imaging modality when there is high suspicion for malignancy, when faced with obviously large and deep lesions or when it is also important to assess the adjacent osseous structures. MRI may also be done as first-line imaging modality when the diagnosis is already established clinically but it is important to assess the lesion extent, particularly prior to interventional radiology procedures or surgery.

The coil selection for the MRI examination is based on the location and size of the lesion. MRI is acquired in at least 2 orthogonal planes using protocols that include T1-weighted images and fluid-sensitive images such as fat-suppressed T2-weighted spin-echo images or short tau inversion recovery (STIR) images. The use of fat suppression in the fluid-sensitive images is important to allow differentiation of the subcutaneous fat from masses or edema, which tend to be hyperintense on these sequences. Fat suppression can also be achieved using Dixon techniques.

MRI is also acquired after the injection of intravenous gadolinium. This can be done with fat-suppressed T1-weighted spin echo images, which may allow further tumor delineation as well as improved demonstration of invasion into adjacent structures and differentiation from adjacent edema. In some conditions, particularly in the evaluation of vascular anomalies, magnetic resonance angiography using a time-resolved technique is extremely useful.[3]

Multiparametric MRI, especially using diffusion-weighted imaging (DWI) and dynamic contrast-enhanced technique have shown potential value in differentiating benign from malignant masses, tumor staging, assessment of tumor extent, and in evaluating tumor response and recurrence after treatment.[4]

MRI findings that have been associated with malignancy include large size (>5-6 cm), absence of low signal intensity on T2-weighted images, heterogeneous signal intensity on T1-weighted images, peripheral and centripetal contrast enhancement, and invasion of adjacent bone and/or neurovascular bundle.[5] However, none of these findings is specific as they can also be seen with benign and intermediate lesions.

Plain radiographs can be used when there is initial clinical uncertainty of a bone or soft-tissue origin. In this setting, radiographs are useful to assess bone involvement and may also demonstrate calcifications within a soft-tissue mass. However, plain radiographs and computed tomography have overall a very limited role in the diagnosis of soft-tissue masses, mainly limited to the evaluation of suspected myositis ossificans.

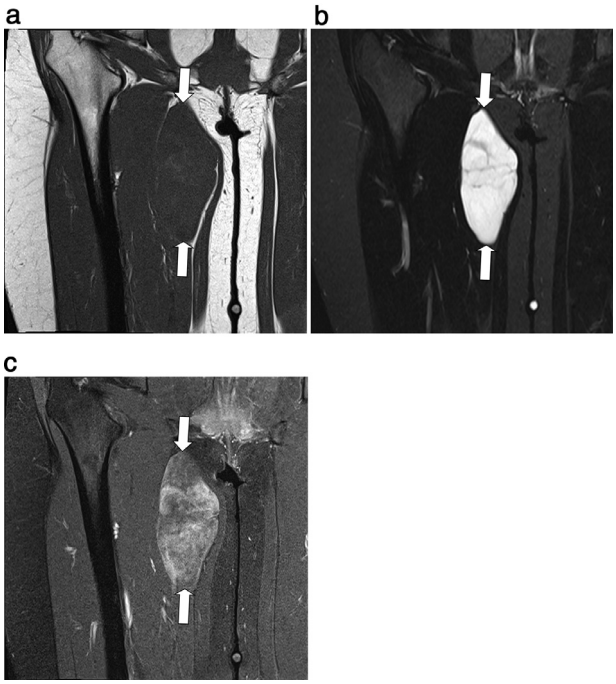

Figure 4 Myxoid liposarcoma in a 16-year-old girl with a growing mass in the right thigh. (a) Coronal T1-weighted, (b) short-tau inversion recovery (STIR), and (c) contrast-enhanced fat-suppressed T1-weighted MR images show a large mass within the adductor magnus muscle that appears predominantly isointense to muscle in (a), predominantly hyperintense to muscle in (b), and with heterogeneous contrast enhancement in (c). There is no recognizable fat on these MR images reflecting the low mature fat content often seen in the myxoid type of liposarcoma. The imaging appearance of this mass is nonspecific requiring biopsy confirmation.

Clinical Correlation

It is important to emphasize that the diagnosis of soft-tissue masses in children can only rarely be done based solely on the imaging findings. Correlation with the clinical history and findings on the physical examination is crucial and mandatory. Information such as patient's age, presence of underlying predisposing conditions, the time the lesion was first noticed (congenital or postnatal), changes in size over long

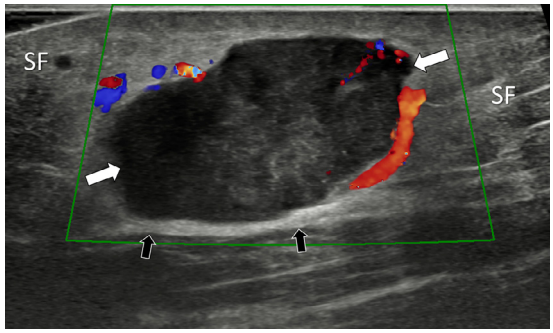

Figure 5 Nodular fasciitis in a 12-year-old boy with a growing mass in the right forearm. Color Doppler longitudinal ultrasound image shows a well-defined, slightly lobulated solid mass (white arrows) that is confined to the subcutaneous plane although abutting the superficial fascia (black arrows). The mass is predominantly hypoechoic compared to the subcutaneous fat (SF), with some posterior acoustic enhancement and low internal vascularity. The ultrasound appearance is not specific and the histologic diagnosis was made after surgical excision.

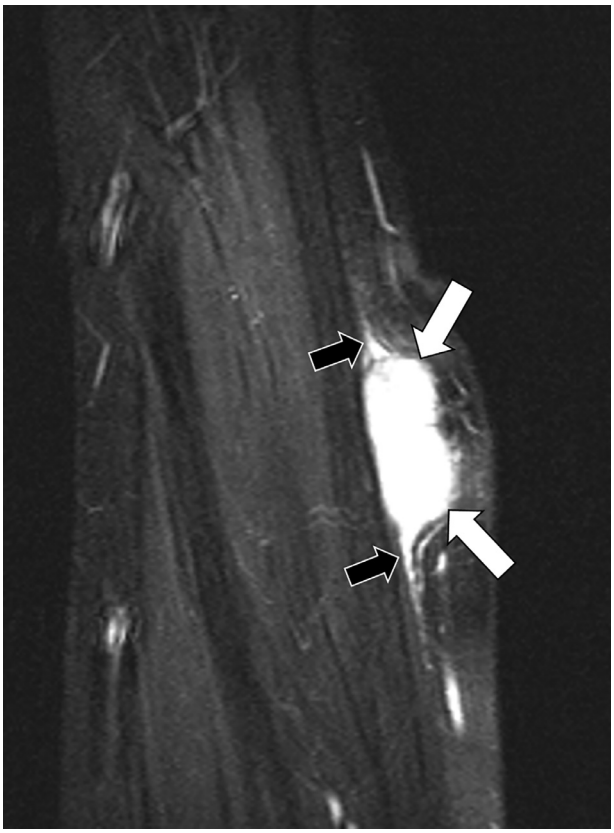

Figure 6 Nodular fasciitis in an 11-year-old girl with a growing mass in the left forearm. Coronal fat-suppressed T2-weighted MR image shows a well-defined, slightly lobulated hyperintense mass (white arrows) in the deep aspect of the subcutaneous plane abutting the superficial fascia. There is increased signal extending along the superficial fascial plane (black arrows) at the superior and inferior margins of the mass reflecting edema. Final histologic diagnosis was confirmed after surgical excision.

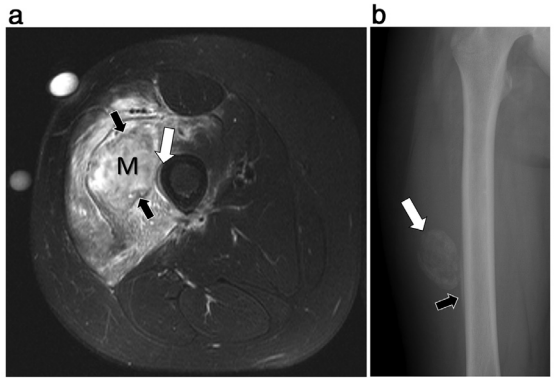

Figure 7 Myositis ossificans in an 11-year-old boy with a palpable mass in the right thigh and no clear history of trauma. (a) Axial fat-suppressed T2-weighted MR image shows extensive increased signal intensity involving the quadriceps femoris muscle, especially the vastus intermedius and vastus lateralis and to a lesser extent the vastus medialis, in keeping with edema. This inflammatory pattern is characteristic of the initial proliferative phase of myositis ossificans. Within the vastus intermedius, there is a poorly defined mass (M). At the periphery of the mass, there are linear areas of low signal intensity in keeping with calcifications (black arrows), which can be easily overlooked and therefore the diagnosis of myositis ossificans may not be suspected. Subperiosteal new bone formation is seen in the adjacent femur (white arrow). (b) Frontal radiograph of the right thigh obtained 12 days after the MRI shows a partly calcified ovoid mass (white arrow) in the soft tissues lateral to the mid-shaft of the right femur in correspondence with the poorly-defined mass seen on MRI (a). The calcifications are more prominent at the periphery of the mass. There is also subperiosteal new bone formation in the femur adjacent to the mass (black arrow). The overall appearance is compatible with myositis ossificans.

period of time (growth, stability, and involution), changes in the size or appearance of the lesion with patient's crying, positioning or other maneuvers, history of pain or bleeding, discoloration of the overlying skin, consistency of the lesion, and presence of ulcerations, hair or vesicles are all of value in establishing a specific diagnosis or in narrowing the differential diagnosis.

Adipocytic Tumors

Adipocytic tumors are characterized by the presence of intralesional fat. This category includes benign tumors such as lipoma and lipoblastoma and malignant tumors such as liposarcoma. It is important to emphasize that intratumoral fat is not exclusive of adipocytic tumors but can also be seen in other conditions including fibroblastic/myofibroblastic tumors such as fibrous hamartoma of infancy and lipofibromatosis as well as vascular anomalies such as involuting hemangiomas, fibroadipose vascular anomaly, intramuscular fast-flow vascular anomaly and PTEN hamartoma of soft tissue.[6] A summary of clinical and imaging features of fat-containing soft-tissue masses of nonvascular origin in children is listed in the Table.

Detection of fat on imaging is crucial to suggest the diagnosis of these fat-containing lesions. On ultrasound fat can have variable echogenicity. Pure fat is hypoechoic but it becomes progressively more echogenic as the number of interfaces with other tissues increases depending on the location and content of the mass. This explains the relatively hypoechoic appearance of normal subcutaneous fat and many superficial lipomas as opposed to the more heterogeneous and increased echogenicity of intramuscular lipomas and other fat-containing tumors.[6]

On MRI, the detection of intralesional fat is generally easier than with ultrasound as intratumoral fat shows similar signal intensity to subcutaneous fat, appearing hyperintense to muscle on T1-weighted and T2-weighted images and hypointense on fat-suppressed images.

Lipoma

Lipoma represents approximately two-thirds of adipocytic tumors in children. At histology, they are comprised of mature adipocytes. Lipomas can present at any age as slowly-growing soft masses, more commonly in the trunk, neck and proximal limbs. Superficial lipomas are usually small (<5

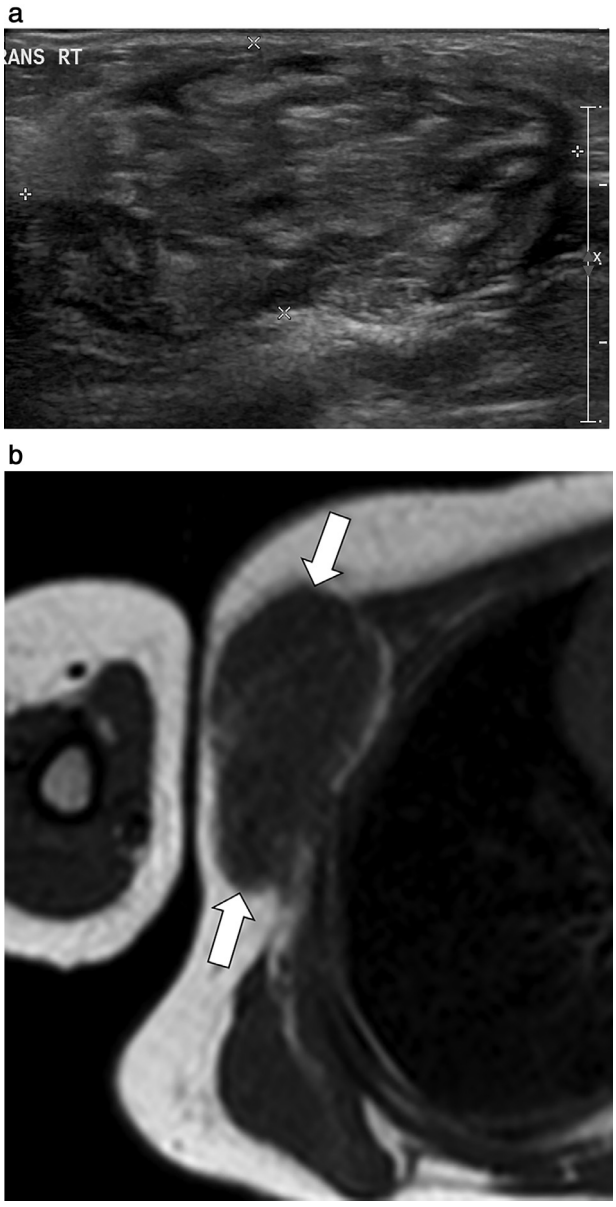

Figure 8 Fibrous hamartoma of infancy in an 11-month-old girl with recently noticed right axillary mass. (a) Transverse ultrasound image of the right axilla shows a subcutaneous mass (between calipers) of heterogeneous echogenicity containing hyperechoic areas representing mature adipose tissue separated by hypoechoic bands and a partially seen hypoechoic rim representing fibrous tissue. (b) Axial T1-weighted MR image shows a relatively well-defined subcutaneous mass (arrows) in the right axilla that is predominantly isointense to muscle although containing numerous small pockets of hyperintense fatty tissue.

cm) whereas deeper lipomas tend to be larger at time of diagnosis. Lipomatosis refers to a more infiltrative presentation usually with invasion of multiple soft-tissue planes, more commonly seen under the age of 2 years.[6,7]

On ultrasound, lipomas can have variable echogenicity. However, more than 50% of subcutaneous lipomas tend to be isoechoic to adjacent subcutaneous fat often with a thin echogenic capsule as well as hyperechoic lines[6] (Fig. 1). Intramuscular lipomas are usually heterogeneous with hyperechoic components. Regardless of their location, lipomas generally show low or absent internal vascularity on Doppler interrogation.[6]

MRI is quite specific for diagnosis of lipomas since these tumors show similar signal intensity to subcutaneous fat on all sequences. They may have a thin fibrous capsule and septa that can show mild contrast enhancement but most of the lesion does not show significant enhancement.[6,7]

Lipoblastoma

Lipoblastoma is a benign tumor representing approximately 30% of adipocytic tumors in children.[6] They typically present under the age of 3 years and are only rarely seen in children older than 8 years. The age of presentation is a useful discriminating feature in the differentiation of lipoblastoma from liposarcoma. At histology, lipoblastomas show variable proportion of maturing adipocytes, lipoblasts, fibrous septa, and myxoid stroma. They are usually of slow growth but faster than lipomas and are mostly confined to the subcutaneous plane, more frequently in the extremities, but have

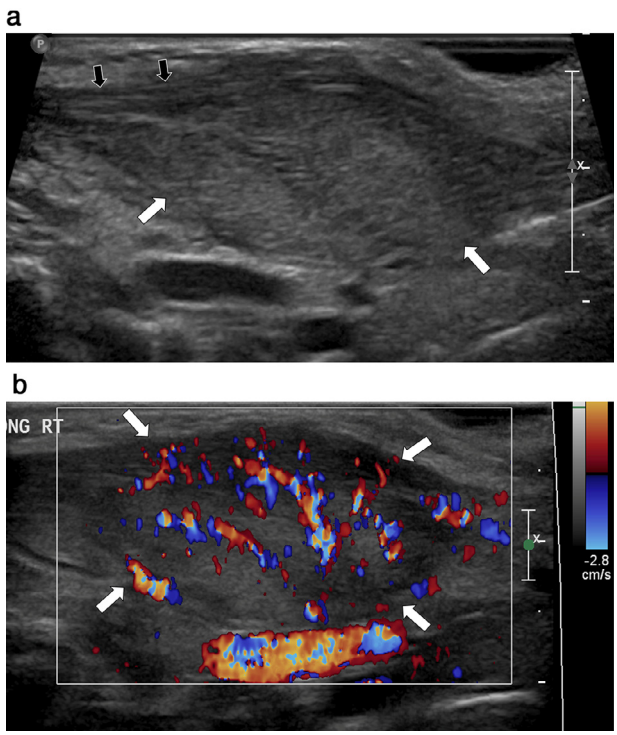

Figure 9 Fibromatosis colli in a 22-day-old boy with right-sided neck swelling for 2 days. (a) Longitudinal ultrasound image shows fusiform enlargement of the right sternocleidomastoid muscle (white arrows). The expanded muscle has heterogeneous echogenicity with loss of the normal fibrillar pattern except anteriorly and superiorly (black arrows). (b) Longitudinal color Doppler ultrasound image shows increased vascularity within the right sternocleidomastoid muscle. However, hyperemia is not a constant finding in this entity.

been reported in many other locations.[6] Lipoblastomatosis refers to a more infiltrative presentation that can involve the deeper soft tissues and adjacent musculature.

On ultrasound, lipoblastoma can have varying appearances although it is more frequently a relatively homogeneous hyperechoic mass (Fig. 2) with low internal vascularity.[7]

The MRI appearance of lipoblastoma varies depending on the histology of the mass. With increasing number of mature adipocytes, there is a corresponding larger amount of fat signal within the mass. However, when lipoblasts and myxoid stroma predominate, the lesion shows larger nonfatty components that appear hypointense on T1-weighted images, hyperintense on fat-suppressed T2-weighted images, and with varying degrees of enhancement after gadolinium injection[6-8] (Fig. 3).

In practice, in a young child, the demonstration of a fat-containing tumor that does not have typical findings of a lipoma is more likely to represent a lipoblastoma. However, presentations in specific locations increase the likelihood of alternate diagnoses, such as fibrous hamartoma of infancy in the axilla, and lipofibromatosis in the hands and feet.

Liposarcoma

Liposarcoma is a rare malignant tumor in the pediatric age, usually seen in children older than 10 years although it has been occasionally documented in younger patients.[6-8] Most pediatric liposarcomas are of the myxoid type which at pathology characteristically have a high water content (myxoid matrix) and relatively small amount of fat (10%-25% of tumor content).[6] They are more commonly seen in the lower extremities, although they have been described in other locations, frequently arising from the intermuscular fat. Clinically, they generally present with a nontender enlarging mass.[6,9]

On ultrasound, myxoid liposarcoma has a variable appearance more often well-defined, hypoechoic but solid-looking, with posterior acoustic enhancement.[9]

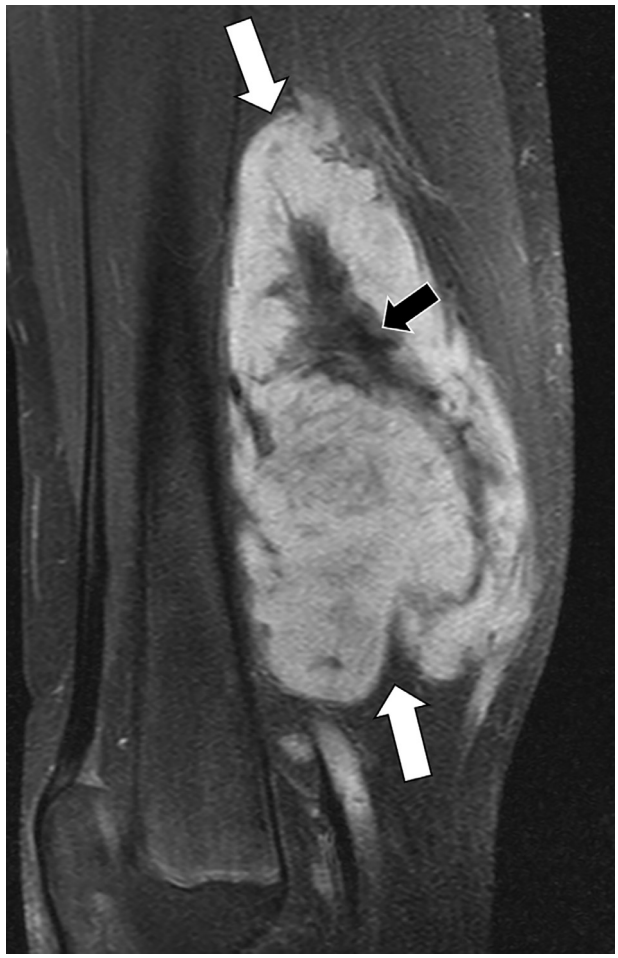

Figure 10 Desmoid-type fibromatosis in an 11-year-old girl with an enlarging mass in the left thigh. Sagittal contrast-enhanced fat-suppressed T1-weighted MR image shows a large mass (white arrows) involving the adductor longus and biceps femoris muscles. The mass shows avid enhancement except for an irregular central area (black arrow) that was hypointense on all sequences and corresponds to collagen-rich fibrous tissue, a useful clue in the diagnosis of desmoid fibromatosis.

Table Common Clinical and Imaging Features of Nonvascular Fat-Containing Soft-Tissue Masses in Children

Tumor Type	Age	Diagnostic Clues
Lipoma	Any age	Variable echogenicity on ultrasound Same signal as subcutaneous fat on all MRI sequences Typically in trunk, neck, proximal limbs
Lipoblastoma	Typically <3 years	Often hyperechoic on ultrasound Variable amount of fat signal on MRI Typically in limbs
Liposarcoma	Typically >10 years	Nonspecific imaging appearance with small amount or no visible fat Typically in lower limbs
Fibrous hamartoma of infancy	Typically <2 years	Alternating fibrous and fatty tissue on ultrasound and MRI Typically in trunk, especially axilla
Lipofibromatosis	Typically <3 years	Ill-defined mass with >50% fat Typically in limbs, especially hands and feet

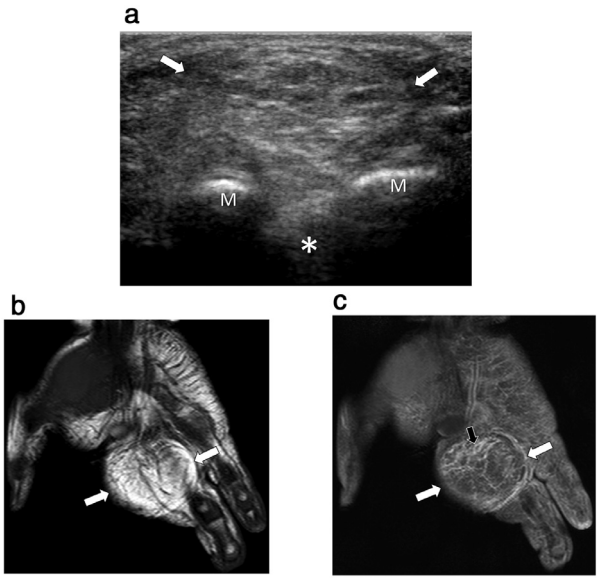

Figure 11 Lipofibromatosis in a 17-month-old girl with a slowly growing mass in the hand since birth. (a) Transverse ultrasound image shows an ill-defined heterogeneous, predominantly hyperechoic mass (arrows) in the dorsal subcutaneous tissues of the hand in between the third and fourth metacarpals (M). The mass is not completely imaged as it extends into the palmar aspect of the hand (*). The increased echogenicity of the mass suggests presence of fat. (b) Coronal T1-weighted and (c) contrast-enhanced fat-suppressed T1-weighted MR images show the mass (white arrows) in the right hand with a predominantly mature adipocytic component mostly similar to the signal intensity of the subcutaneous fat in (b) although with some increased peripheral and focal septal enhancement (black arrow) in (c). The congenital presentation and the location in the hands suggests the diagnosis of lipofibromatosis although lipoblastoma should be considered in the differential diagnosis.

On MRI, myxoid liposarcoma is usually predominantly hypointense on T1-weighted images and hyperintense on T2-weighted images containing fatty septa or small fatty nodules. The mass shows variable patterns of enhancement after gadolinium injection[9] (Fig. 4). It is important to emphasize that in a child a mass with a predominantly fatty component is therefore unlikely to represent a liposarcoma.

Fibroblastic/Myofibroblastic Tumors

Nodular Fasciitis

Nodular fasciitis is a benign self-limited neoplastic lesion that can occur at any age but is more commonly seen in adolescents.[10] It is a solitary lesion, usually presenting in the face or neck, trunk, and upper extremities. It may or not be tender and typically shows rapid growth. It is more frequently subcutaneous but can arise in other soft-tissue planes and may involve more than one soft-tissue plane.[10,11]

On ultrasound, nodular fasciitis has a variable appearance, more often well-defined, heterogeneous but relatively hypoechoic, broadly based at the superficial fascia and sometimes with a "fascial tail," which refers to extension of tumor along the fascial plane. Larger lesions may have posterior acoustic enhancement (Fig. 5). On color Doppler interrogation, there is low to moderate vascularity.[11]

On MRI, nodular fasciitis is a well-defined mass, isointense to muscle on T1-weighted images, heterogeneously hyperintense on T2-weighted images (Fig. 6), and with heterogeneous enhancement after gadolinium injection. In less than half of the cases, the lesion may exhibit an "inverted target" sign, with a hypointense peripheral rim and a hyperintense center on T2-weighted images, the latter showing lack of enhancement on postgadolinium images. The "fascial tail" may also be evident on MRI with associated perilesional edema along the fascial plane.[11]

Myositis Ossificans

Myositis ossificans is a benign self-limited fibrous proliferation with metaplastic mature lamellar bone formation in the soft tissues.[12] Clinically, it is divided in 3 types: posttraumatic myositis ossificans, nontraumatic myositis ossificans, and myositis ossificans progressiva.

Posttraumatic myositis ossificans is the most common type although patients may not necessarily recall the trauma. It is more frequent in children older than 10 years of age, commonly affecting the extremities, especially the quadriceps femoris and brachialis muscles.[11] Clinically, it presents with a firm, painful mass, and with erythema of the overlying skin mimicking malignancy or infection.

The imaging appearance of myositis ossificans reflects its histologic evolution with an initial proliferative phase in the first 3 weeks after trauma, an intermediate calcification and ossification phase 3-7 weeks after the trauma, and a mature ossification phase more than 7 weeks after the trauma.

In the initial phase, the imaging appearance can be difficult to differentiate from a soft-tissue sarcoma. MRI may be useful by showing marked edema of the muscles around the mass, a more inflammatory pattern than would be expected for a sarcoma (Fig. 7). The lesion is isointense to muscle on T1-weighted images, hyperintense on T2-weighted images, with diffuse or peripheral enhancement after gadolinium injection. Adjacent subperiosteal new bone formation may be evident but the bone cortex appears intact.[11]

With progression into the intermediate phase, peripheral calcifications become evident, which are easier to recognize on plain radiographs (Fig. 7) and CT, and can be detected even earlier with ultrasound.[13] On MRI, calcifications may appear as hypointense foci on T2-weighted images but initially may be more difficult to recognize compared to the other imaging modalities. With progression into the mature ossification phase, the peripheral calcification is more evident on MRI and is associated with decrease of the perilesional edema.[11]

Fibrous Hamartoma of Infancy

Fibrous hamartoma of infancy is a benign tumor comprised of mature fibrous and adipose tissue as well as an immature mesenchymal component.[14] It is usually seen in children younger than 2 years of age and up to 20% are congenital.

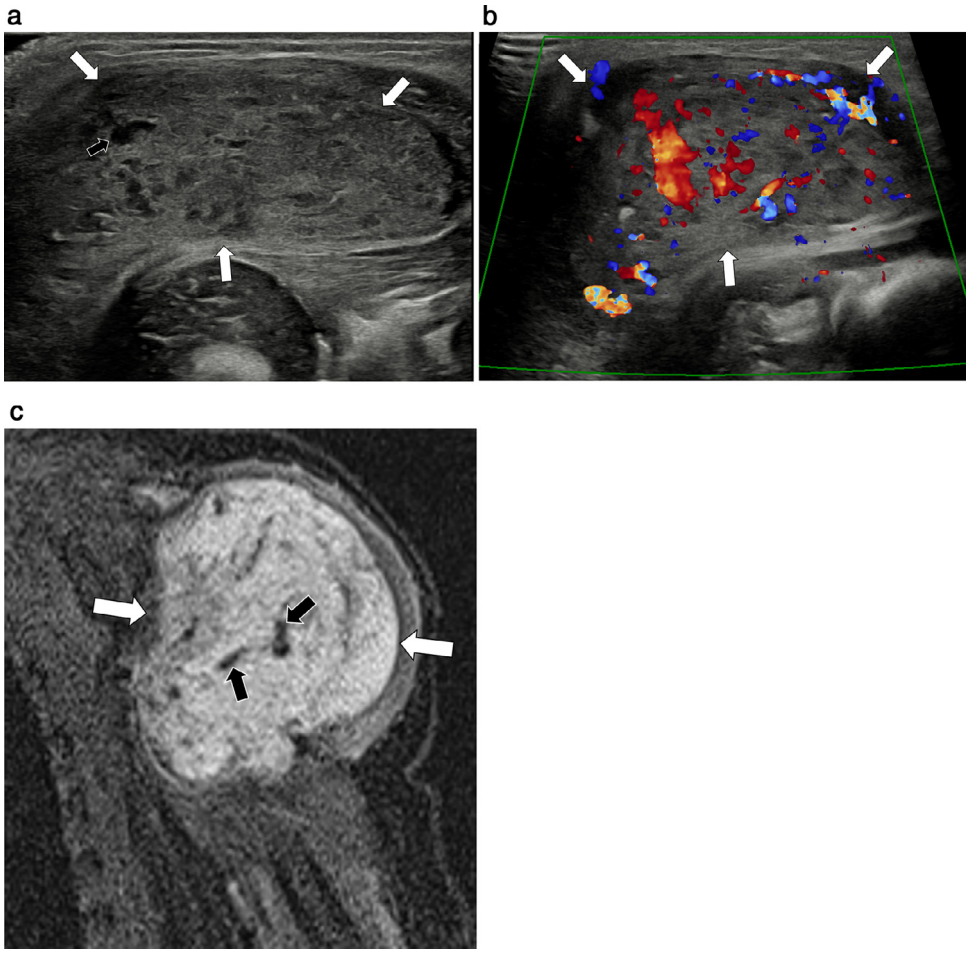

Figure 12 Infantile fibrosarcoma in a 9-day-old girl with a congenital shoulder mass. (a) Transverse gray-scale and (b) color Doppler ultrasound images show a solid intramuscular mass (white arrows) with heterogeneous echogenicity including small anechoic spaces suggestive of necrosis (black arrow). There is high vascularity within the mass (b). (c) Coronal short-tau inversion recovery (STIR) MR image shows a large mass (white arrows) within the deltoid muscle that is hyperintense compared to normal muscle and shows linear hypointense foci that represent signal void from high-flow vessels (black arrows). Some of the findings, particularly the high internal vascularity on color Doppler ultrasound and the high-flow vessels on MRI may lead to the diagnosis of congenital hemangioma, however, areas of necrosis and intramuscular location should alert to the diagnosis of infantile fibrosarcoma at this patient's age. Histologic diagnosis was confirmed with biopsy.

There is a male predominance of 2:1 compared with females. It is usually a poorly defined subcutaneous tumor more frequently seen in the trunk, particularly in the axilla, but it has also been described in the extremities.

On ultrasound, fibrous hamartoma of infancy is often ill-defined and hyperechoic compared to muscle. It contains trabeculated hypoechoic bands and a peripheral hypoechoic rim and shows low vascularity on color Doppler interrogation[6-8] (Fig. 8).

On MRI, fibrous hamartoma of infancy is also often ill-defined and the appearance reflects the variable amounts of fat in relation to fibrous and mesenchymal tissue. The presence of intralesional striated pattern of hyperintense bands of fatty tissue alternating with isointense bands when compared with muscle is suggestive of this entity.[6-8] This pattern is better recognized on T1-weighted images (Fig. 8).

Fibromatosis Colli

Fibromatosis colli is a benign self-limited fibromatous proliferation that affects the sternocleidomastoid muscle, the right more often than the left, usually presenting between 2 and 8 weeks of age. The etiology is not clear but there is often a history of birth trauma, difficult delivery or breech delivery suggesting an association with muscle injury.[8,9] Clinically, there is firm swelling in the affected side in some cases causing torticollis.

Ultrasound is the imaging modality of choice and usually the only imaging modality needed for diagnosis. Fibromatosis colli appears as fusiform enlargement of the sternocleidomastoid muscle, more often affecting the lower two-thirds of the muscle. The affected area may appear hyperechoic, hypoechoic or heterogeneous with loss of the normal fibrillar pattern of the muscle and with variable vascularity on color Doppler interrogation[8] (Fig. 9).

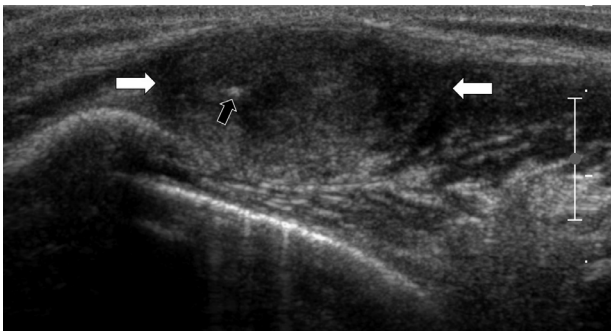

Figure 13 Solitary myofibroma in a 4-month-old boy with a firm nodule on the back since birth. Longitudinal ultrasound image shows a solid, ovoid mass (white arrows) within the muscle plane of the posterior chest wall. The mass is heterogeneous, predominantly isoechoic to muscle with irregular hypoechoic areas that may represent necrosis. There is a small intralesional hyperechoic focus in keeping with calcification (black arrow).

Fibromatosis colli can be accurately and safely diagnosed using ultrasound, largely based on the combination of imaging appearance and clinical context, particularly the age of the patient. The presence of a mass involving the sternocleidomastoid muscle outside of the typical age range of fibromatosis colli should alert to the diagnosis of other conditions with special consideration for malignant neoplasms.

Desmoid-Type Fibromatosis

Desmoid-type fibromatosis is a tumor of the musculoaponeurotic tissues characterized by proliferation of myofibroblastic-type cells with associated collagenous stroma and vascular network. Owing to its high rate of recurrence, this tumor is considered an intermediate, locally aggressive tumor, although it does not have metastatic potential. It presents more frequently in the extremities, abdominal wall and mesentery although other locations

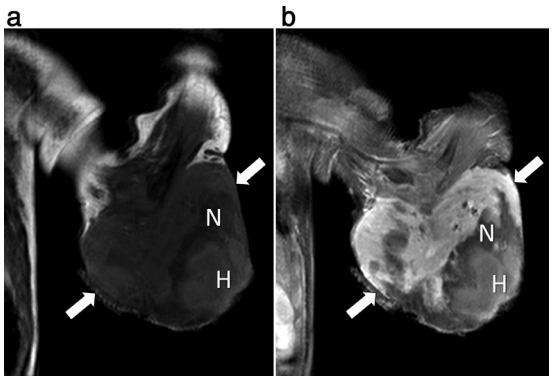

Figure 14 Solitary myofibroma in a newborn with a large ulcerated mass in the left elbow. Coronal T1-weighted (a) and contrast-enhanced fat-suppressed T1-weighted (b) MR images show a large subcutaneous mass with heterogeneous signal intensity. In (a), the mass is predominantly isointense to muscle although with hyperintense areas due to hemorrhage (H) and hypointense areas due to necrosis (N). In (b), a large part of the mass shows avid enhancement except for the areas of necrosis (N).

including head and neck have been described in children. Most desmoid-type fibromatosis are sporadic but a minority is hereditary, particularly in patients with familial adenomatous polyposis. In this latter group the abdominal wall and intraabdominal locations are more common and tumors tend to be larger and can be multifocal.[15] There are 2 age peaks of presentation, the first occurring at 4-5 years and the second in the third decade of life.[11] Clinically, desmoid-type fibromatosis usually presents as a growing painless mass but depending on its location and proximity to a neurovascular bundle it may result in functional impairment and pain.[15]

On ultrasound, desmoid-type fibromatosis appears as a solid mass, more often with well-defined margins and posterior acoustic enhancement. The echogenicity is variable, appearing hypoechoic or isoechoic to muscle and can be homogeneous or heterogeneous without internal necrosis or calcifications. On color Doppler interrogation, there is variable internal vascularity from absent, especially in smaller lesions, to moderate.[16]

On MRI, the appearance of desmoid-type fibromatosis varies depending on the amount of intralesional collagen or cellularity. The collagen-rich areas (fibrous tissue) are hypointense on T1- and T2-weighted images with absent to mild enhancement after gadolinium injection whereas those that have more cellularity are isointense to muscle on T1-weighted images and hyperintense on T2-weighted images and show moderate to avid enhancement after gadolinium injection[11,15] (Fig. 10). Extension along the fascial plain may result in the "fascial tail sign." Linear hypointense bands on T1- and T2-weighted images that show lack of enhancement on postgadolinium images within the tumor, result in the "band sign," also helpful to suggest the diagnosis of desmoid-type fibromatosis.

Lipofibromatosis

Lipofibromatosis is now considered within the category of intermediate, locally aggressive tumors without metastatic potential. It is comprised of fibroblastic cells and adipocytes, most commonly arising from the subcutaneous or deeper soft tissues and shows an infiltrative growth pattern.[6] It is usually diagnosed in children under the age of 3 years and in 18% of cases is congenital.[11] It has a predilection for hands and feet but has also been described in the remainder of the extremities and less commonly in the trunk and neck.[6]

On imaging, the appearance of lipofibromatosis varies depending on the amount of fat and fibrous tissue present within the mass, which in general is at least 50% fat. On ultrasound, it is usually an ill-defined, predominantly hyperechoic mass (Fig. 11) as compared to muscle denoting the fatty component and with low vascularity on color Doppler interrogation.[6,8]

On MRI, lipofibromatosis is usually a poorly defined mass with variable signal intensity, predominantly hyperintense on T1-weighted images with thin septa or nodules that are isointense to muscle (Fig. 11). On fat-suppressed T2-weighted images, it is heterogeneous although of relatively low signal reflecting the fatty component. After gadolinium

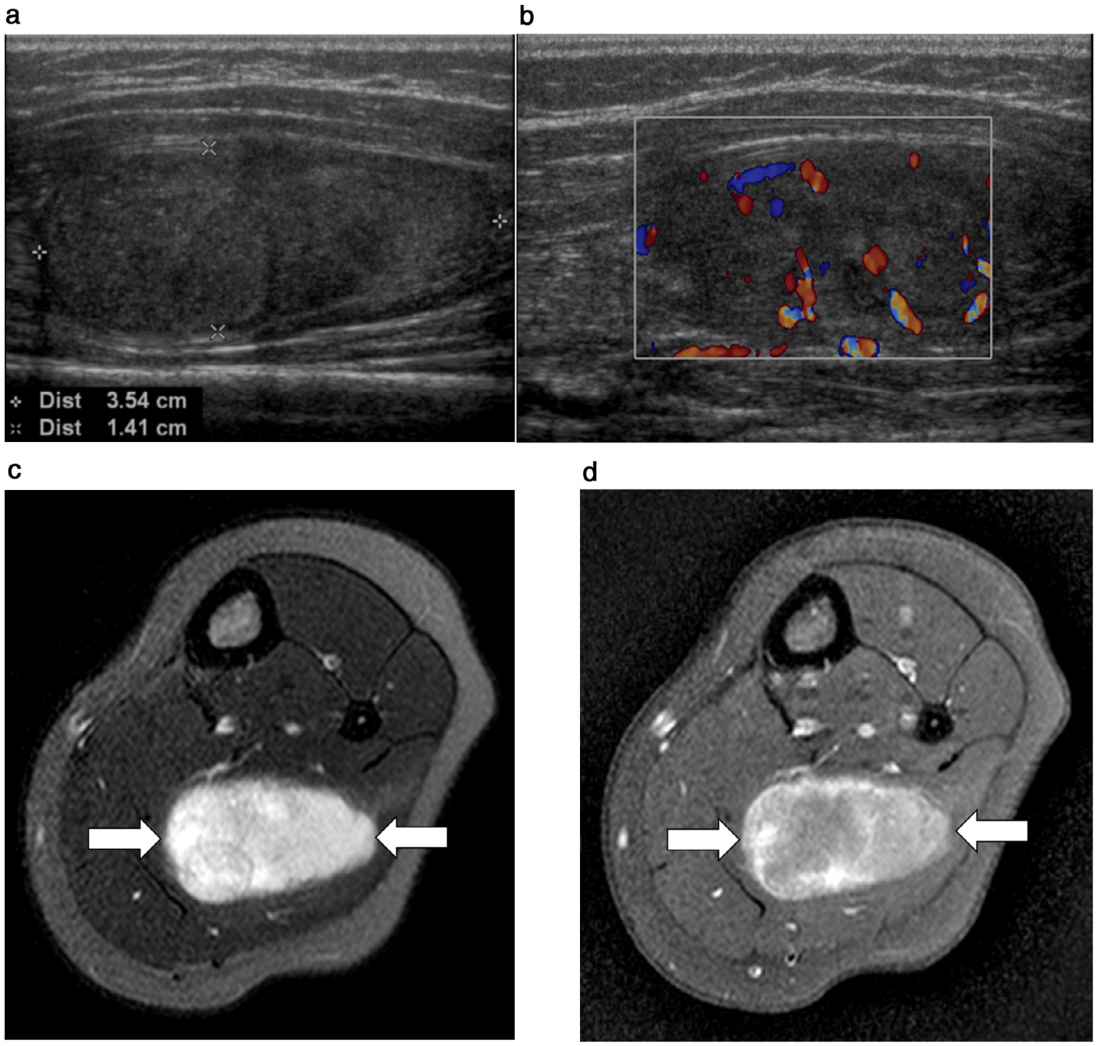

Figure 15 Alveolar rhabdomyosarcoma in a 3-year-old boy with recently noticed painless mass in the right calf. (a) Longitudinal gray-scale and (b) color Doppler ultrasound images show a well-defined solid intramuscular mass (between cursors). The mass is of heterogeneous echogenicity and shows moderate internal vascularity. (c) Axial fat-suppressed T2-weighted and (d) contrast-enhanced fat-suppressed T1-weighted MR images show a well-defined mass (arrows) within the lateral head of the gastrocnemius muscle. The mass is heterogeneously hyperintense in (c) and shows heterogeneous enhancement in (d). The imaging appearance of the mass is nonspecific but worrisome for a malignant neoplasm.

injection, there is mild enhancement of the nonfatty component[6] (Fig. 11).

Infantile Fibrosarcoma

Despite its name, infantile fibrosarcoma, also known as congenital fibrosarcoma, is not a "true" sarcoma but is now considered a rarely metastasizing intermediate tumor.[1] At histology, there is proliferation of spindle cells in a fascicular growth pattern with dense cellularity resembling the adult-type fibrosarcoma although these are 2 distinct entities with a much indolent course and better survival rate for the infantile form.

Most cases are diagnosed in children younger than 2 years and nearly half are congenital.[11] It is the most common nonbenign soft-tissue tumor in children less than 1 year of age.[17] Clinically, it often presents as a rapidly growing painless firm mass, more commonly in one extremity but occasionally may arise in the head and neck and trunk.[11,18] There are several case reports of infantile fibrosarcoma mimicking the clinical presentation of hemangiomas and other vascular anomalies because in infantile fibrosarcoma the overlying skin is usually red or blue-red in color, may appear tense with superficial telangiectasia and may have ulceration with bleeding.[19,20]

On ultrasound, infantile fibrosarcoma is often a heterogeneous mass with moderate to high internal vascularity (Fig. 12). This imaging appearance may also lead to the incorrect diagnosis of congenital or infantile hemangioma. In addition to having a complete and reliable clinical information available to better interpret the sonographic findings, one has to look for imaging clues that help in the differentiation of sarcomas from hemangiomas.[21] The involvement of

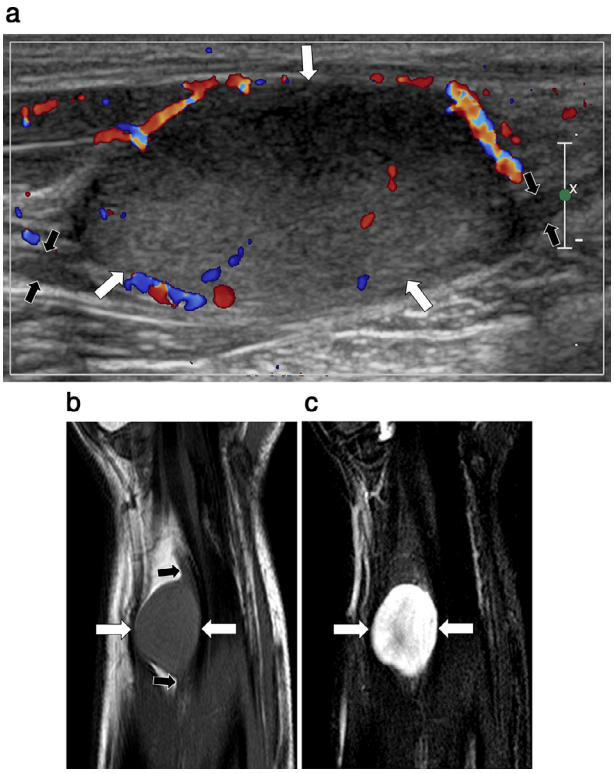

Figure 16 Localized neurofibroma in an 11-year-old girl with neurofibromatosis 1 and new swelling in the forearm. (a) Longitudinal color Doppler ultrasound image shows a well-defined, slightly heterogeneous mass (white arrows) in between muscles in the anterior compartment of the forearm following the course of the median nerve (black arrows). There are some prominent vessels at the periphery of the mass but otherwise the mass has low internal vascularity. (b) Coronal T1-weighted and (c) fat-suppressed T2-weighted MR images show a well-defined, somewhat ovoid mass (white arrows) along the course of the median nerve (black arrows). The mass is iso- to slightly hyperintense to muscle in (b) and shows a target sign in (c) with a hyperintense periphery and an irregular hypointense center.

muscles by the mass and the presence of intralesional necrosis are useful indicators that the lesion is less likely to be a hemangioma (Fig. 12).

On MRI, the findings are nonspecific. Infantile fibrosarcoma is usually a large, heterogeneous mass that appears isointense to muscle on T1-weighted images and hyperintense on T2-weighted images (Fig. 12) with heterogeneous enhancement on the postgadolinium images. Areas of necrosis and hemorrhage can be evident particularly in the larger masses, and less commonly hypointense foci can be seen reflecting calcifications and fibrous tissue.[4,18] DWI shows diffusion restriction.[4]

Pericytic (Perivascular) Tumors

Myofibroma/Myofibromatosis

Myofibroma refers to the solitary presentation and myofibromatosis to the multicentric presentation of this tumor that is now included in the category of pericytic (perivascular) tumors.[1] At histology, myofibroma shows nodules formed by myofibroblasts and undifferentiated cells arranged around hemangiopericytoma-like vessels. Most are diagnosed in children under the age of 2 years and more than 50% of solitary cases and more than 90% of myofibromatosis are congenital.[19] Solitary myofibromas present almost equally in the skin/subcutaneous plane and in the musculoaponeurotic plane, with a minority presenting in bone. Multicentric myofibromatosis involves both soft tissue and bone with a subgroup of approximately 15%-20% of cases involving also deep soft tissues and viscera. The natural history of myofibroma is of growth during the first year of life followed by a slow spontaneous regression that may take years. Despite this self-limited course, multicentric myofibromatosis with visceral involvement has a poor prognosis with relatively high mortality rates.[8] Clinically, myofibromas present as firm, rapidly growing flesh-colored or purple nodules in the head and neck and trunk.[11]

On ultrasound, myofibromas can have variable echogenicity although they are more often hypoechoic. They may contain fluid pockets that represent areas of necrosis and hemorrhage, which are central in small lesions but eccentric in larger lesions. They may show calcifications (Fig. 13) and exhibit varying degrees of vascularity on color Doppler interrogation.[8,11]

On MRI, they are often isointense to muscle on T1-weighted images and predominantly hyperintense on T2-weighted images with areas of necrosis or hemorrhage (Fig. 14). They may show diffuse or heterogeneous enhancement after gadolinium injection or show a target sign in cases with central necrosis.[8]

Skeletal Muscle Tumors

Rhabdomyosarcoma

Rhabdomyosarcoma is the most common pediatric soft-tissue sarcoma representing approximately half of all cases in children under 10 years of age.[22] Despite its presumed origin in mesenchymal cells committed to skeletal muscle differentiation, rhabdomyosarcoma can arise in many different body sites even in those that normally do not have striated muscle. Most cases are sporadic although there are predisposing conditions including neurofibromatosis 1 (NF1), Beckwith-Wiedemann syndrome, Costello syndrome and Li-Fraumeni syndrome.[23] Common locations are the head and neck, genitourinary tract, and the extremities. Approximately 10%-20% have metastatic disease at time of presentation, more often in the lungs, bone, bone marrow or lymph nodes.[22] In children, the 2 most common histological types are embryonal and alveolar. The embryonal type represents approximately 75% of cases, usually in children younger than 5 years, more frequent in the head and neck and in the genitourinary tract and infrequent in the extremities.[4] The alveolar type represents approximately 20% of cases and is the most common type found in the extremities.[4,8] The embryonal type has in general a better prognosis than the alveolar

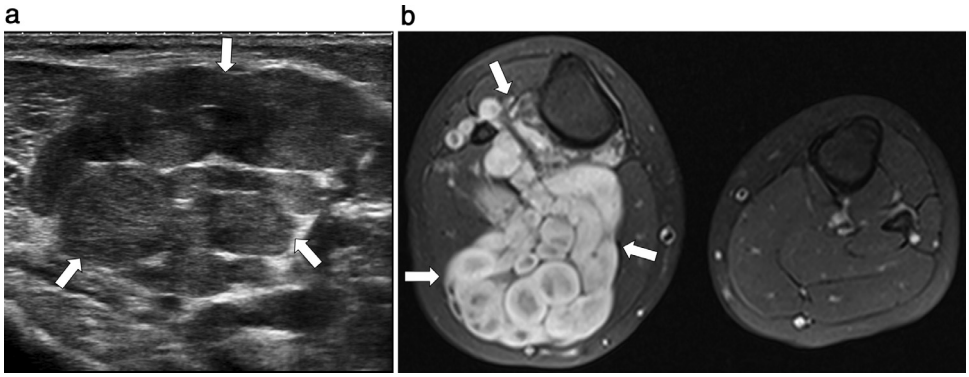

Figure 17 Plexiform neurofibroma in a girl with neurofibromatosis 1 and a slowly growing lump in the upper calf. (a) Longitudinal ultrasound image at 3 years of age shows a solid mass (arrows) comprised of multiple hypoechoic nodules of varying size separated by hyperechoic tissue. (b) Axial fat-suppressed T2-weighted MR image at 8 years of age shows increase in size of the plexiform neurofibroma, which appears as a well-defined lobulated mass (arrows) involving predominantly the posterior muscle compartments and to a lesser extent the anterior and lateral muscle compartments of the right lower leg. The mass is comprised of multiple nodules of varying size, some of which are more rounded and others more elongated in shape resulting in a "bag of worms" appearance. The more rounded nodules exhibit the characteristic "target sign" (hyperintense periphery and hypointense center). There is associated overgrowth of the right lower leg clearly evident when compared with the left lower leg.

type and the extremities are considered an unfavorable site. The clinical presentation of rhabdomyosarcoma in the extremities is usually with a rapidly growing painless mass or less commonly with a small or with a nonpalpable mass but with enlarged lymph nodes.[22]

On ultrasound, rhabdomyosarcoma is usually a well-defined, heterogeneous, predominantly hypoechoic intramuscular mass with varying degrees of internal vascularity on color Doppler interrogation[8] (Fig. 15).

On MRI, rhabdomyosarcoma has a nonspecific appearance, usually iso- to slightly hyperintense to muscle on T1-weighted images and hyperintense on T2-weighted images with heterogeneous enhancement on postgadolinium images (Fig. 15). Intralesional cystic components may be present.[8] DWI shows restricted diffusion.[4]

Nerve Sheath Tumors

Neurofibroma

Neurofibroma is a nonencapsulated benign tumor that in addition to peripheral nerve cells contains other cell types as well as an extracellular matrix and collagen.[23] They are inseparable from the nerve they arise from. Three forms are recognized: localized, diffuse and plexiform. The localized form represents about 90% of cases, usually presenting as solitary nodules affecting skin and subcutaneous tissues.[24,25] The diffuse form is uncommon, usually found in the subcutaneous tissues. These 2 forms are not necessarily related to NF1. On the other hand, plexiform neurofibroma is pathognomonic of NF1. Plexiform neurofibroma shows growth along the nerve sheath of a major nerve trunk extending into its branches and therefore can result in very large masses.

On ultrasound, localized neurofibroma usually presents as a well-defined hypoechoic nodule with posterior acoustic enhancement and may therefore mimic a cyst.[24,26] Small hypoechoic extensions on either side of the nodule can be seen representing the originating nerve[25] (Fig. 16). A "target sign" with a hypoechoic periphery and a hyperechoic center may be present reflecting the myxomatous and

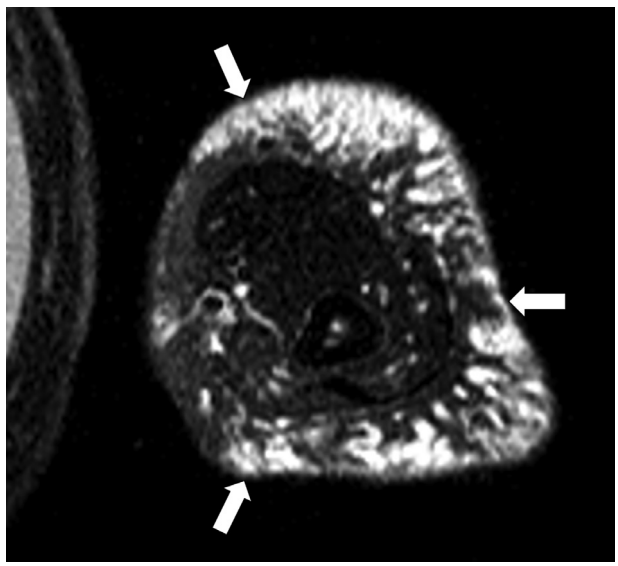

Figure 18 Superficial plexiform neurofibroma in a 6-year-old girl with neurofibromatosis 1 and cutaneous neurofibromas in the left arm. Axial fat-suppressed T2-weighted MR image shows an extensive and ill-defined lesion involving nearly the whole circumference of the arm (arrows). The lesion involves the skin and subcutaneous tissues showing predominantly high signal intensity although with multiple hypointense bands resulting in a reticular appearance. There is absence of the target sign typically seen in deep plexiform neurofibromas (Fig. 17). Without a history of neurofibromatosis 1, superficial plexiform neurofibroma can be confused with a low-flow vascular malformation.

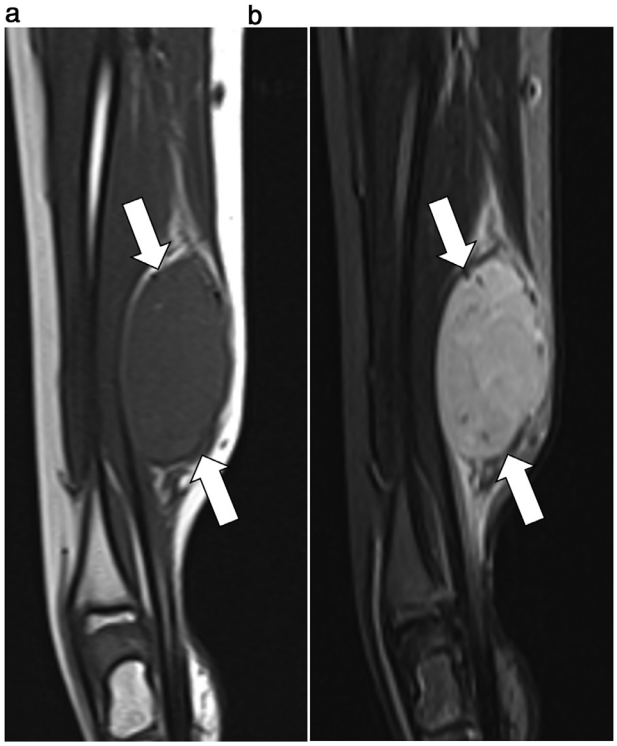

Figure 19 Malignant peripheral nerve sheath tumor in a 10-year-old boy with a rapidly-growing mass in the forearm. (a) Sagittal T1-weighted and (b) short-tau inversion recovery (STIR) MR images show a well-defined ovoid lesion in the anterior muscle compartment of the forearm along the course of the median nerve. The mass is isointense to muscle in (a) and is predominantly hyperintense to muscle with thin hypointense septa in (b). Apart from the close relation to the median nerve, the appearance of the mass is nonspecific and final diagnosis required biopsy.

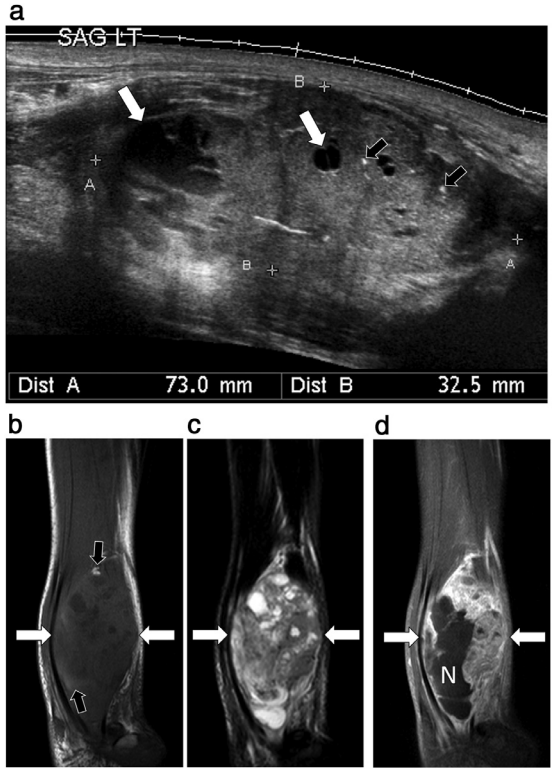

Figure 20 Synovial sarcoma in a 14-year-old boy with progressive swelling of the forearm for 12 months and now with paresthesias. (a) Longitudinal extended field-of-view ultrasound image shows a well-defined lobulated intramuscular mass (between cursors) within the anterior compartment of the left forearm. The mass is heterogeneous, predominantly isoechoic to muscle and contains cystic spaces suggestive of areas of necrosis (white arrows). There are a few small hyperechoic foci in keeping with calcifications (black arrows). (b) Coronal T1-weighted, (c) fat-suppressed T2-weighted and (d) contrast-enhanced fat-suppressed T1-weighted MR images show a large heterogeneous mass (white arrows) originating in the flexor pollicis longus muscle. It shows numerous cystic areas that are hypointense in (b), hyperintense in (c) and do not enhance in (d). Foci of hemorrhage appear hyperintense in (b) (black arrows). The enhancement of the mass is heterogeneous with a large area of necrosis (N) posteriorly characterized by lack of enhancement (d). The imaging appearance of the mass is nonspecific but worrisome for a malignant neoplasm.

fibrocollagenous histological components respectively.[24,26] The diffuse form appears as an ill-defined subcutaneous hyperechoic mass containing interconnecting hypoechoic tubular or nodular structures.[24] The plexiform type appears as multiple neurofibromas adjacent to each other following the course of a nerve[26] (Fig. 17).

On MRI, localized neurofibroma appears as a round or fusiform mass with tapered ends reflecting its contiguity with the originating nerve (Fig. 16). If large, it can have a fascicular appearance. If intramuscular, it may appear surrounded by fat ("split fat sign").[23,24,27] It is generally isointense to muscle on T1-weighted images. It is hyperintense on T2-weighted images, sometimes with a "target sign" seen as a hyperintense rim and a hypointense center (Fig. 16) correlating with the ultrasound appearance. After gadolinium injection, it may show avid enhancement although the enhancement is more heterogeneous in larger lesions.[23,24] In the diffuse form, the ill-defined subcutaneous network of neurofibromas is hypointense on T1-weighted images, hyperintense on T2-weighted images and with avid enhancement on postgadolinium images.[24] The appearance of plexiform neurofibroma depends on its location. Deep plexiform neurofibroma are seen deep to the superficial fascia and appear as lobulated masses that are isointense to muscle on T1-weighted images and hyperintense on T2-weighted images sometimes described as a "bag of worms" with presence of the "target sign" (Fig. 17). Superficial plexiform neurofibroma has a more infiltrative appearance with extension to the skin in a reticular or branching pattern, with smaller nodules and absence of the target sign (Fig. 18).[27] Superficial plexiform neurofibroma can therefore be confused with venous or microcystic lymphatic malformations.

In general, the diagnosis of benign peripheral nerve sheath tumors is not a challenge because most patients already have a known diagnosis of NF1. However, NF1 patients are at risk for other non-neurogenic neoplasms including soft-tissue rhabdomyosarcoma, which may be suspected in the presence of a mass without typical imaging findings of nerve sheath tumors.

Malignant Peripheral Nerve Sheath Tumor

Malignant peripheral nerve sheath tumor (MPNST) is a malignant spindle cell tumor that arises from either a peripheral nerve or its nerve sheath or more frequently from a preexisting benign peripheral nerve sheath tumor, especially a plexiform neurofibroma in the setting of NF1.[28] MPNST often originates from large nerves such as the sciatic nerve and brachial plexus but can be seen in other locations as well. Clinically, MPNST presents with an enlarging mass, with change in the consistency of a preexisting mass from soft to hard, or with neurologic symptoms, including pain, sensory deficits or weakness.[28]

On ultrasound, MPNST appears as a heterogeneous mass that may contain areas of necrosis, hemorrhage and calcifications without visualization of the "target sign." [28] On color Doppler interrogation, there is moderate vascularity with abnormal vessels.

On MRI, MPNST shows some of the features described in benign neurofibromas and therefore it is difficult to differentiate from its benign counterparts (Fig. 19). Findings concerning for malignancy include size >5 cm, heterogeneous appearance, poorly defined margins, cystic changes, infiltration and invasion of fat plane, absence of target sign, peritumoral edema, and peripheral enhancement.[28]

FDG-PET is increasingly used in the work-up of MPNST as may allow differentiation of benign from malignant nerve sheath tumors and may detect recurrent or metastatic disease.[28]

Tumors of Uncertain Differentiation

Synovial Sarcoma

Synovial sarcoma is a tumor that arises from cells of unknown lineage that undergo variable epithelial differentiation, resembling but not related to synovium. In children, it is often seen in the second decade. Although it can occur in many different sites in the body, the most common location is in the lower extremities, more typically juxtaarticular, arising from tendon sheaths, bursae and joint capsules.[28,29] Clinically, it often presents with a painless mass that sometimes has been present for months or years.

On ultrasound, synovial sarcoma is usually a heterogeneous mass that may have areas of necrosis and eccentric calcifications (Fig. 20), with variable degree of vascularity on color Doppler interrogation.

On MRI, synovial sarcoma is usually a well-demarcated mass, isointense to muscle on T1-weighted images, heterogeneously hyperintense on T2-weighted images, and with heterogeneous enhancement after gadolinium injection (Fig. 20). When small, synovial sarcomas may mimic benign para-articular cysts as they may appear diffusely and markedly hyperintense on T2-weighted images.[4,30] In nearly one third of cases, nonfat-suppressed T2-weighted images can depict the "triple sign", characterized by intratumoral areas with signal intensity similar to fluid, areas with signal intensity similar to fat, and areas of lower signal intensity resembling fibrous tissue.[29] Fluid-fluid levels can also be seen in 18% of cases.[29] In one series, 71% of cases were in intimate relation with bone, either abutting bone (50%) or causing cortical thinning or invasion (21%).[28] DWI shows restricted diffusion of the more cellular components of the tumor.[4]

Conclusion

The imaging diagnosis of soft-tissue masses in children can be challenging as imaging findings are often nonspecific. Ultrasound and MRI are often used as first and second line imaging modalities. Use of appropriate imaging protocols may allow identification of diagnostic clues, for example, recognizing presence of fat or fibrous tissue within a mass. It is imperative to correlate the imaging findings with the appropriate clinical information in order to establish a specific diagnosis or propose a reasonable differential diagnosis. However, in many instances the final diagnosis requires histologic confirmation.

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