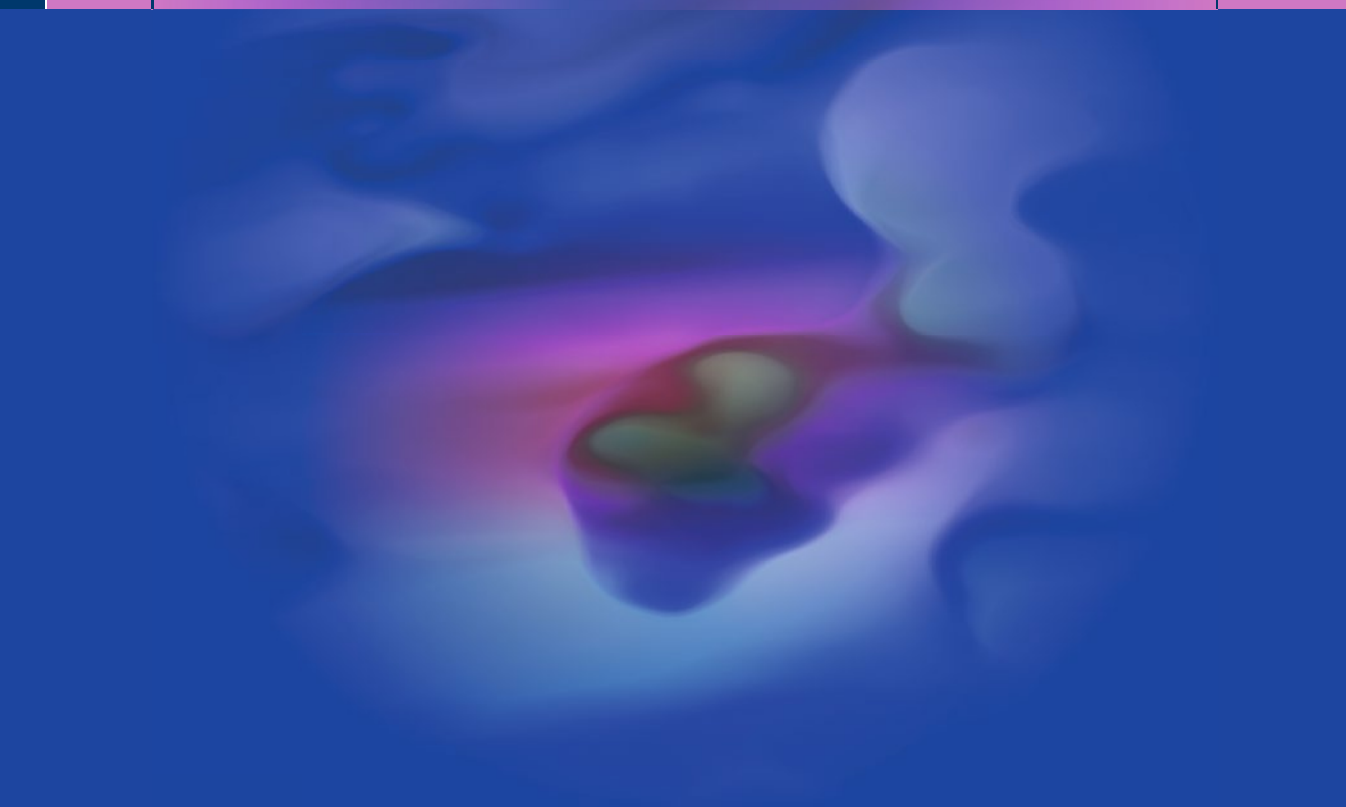
Heung Sik Kang
Sung Hwan Hong
Ja-Young Choi
Hye Jin Yoo

Oncologic Imaging
Soft Tissue Tumors



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To my coauthors, Sung Hwan Hong, Ja-Young Choi, and Hye Jin Yoo, for their passion, excellence, and cooperation.

And to my three lovely grandsons, wishing them a bright and prosperous future.

—Heung Sik Kang, MD

To all the teachers and colleagues who have brought me to this point.

And to my sweet wife, Yun Kyung, and daughter, Jisu, my buttress and love.

—Sung Hwan Hong, MD

To my colleagues and family.

And finally to my mentor, Professor Kang, for his new start.

—Ja-Young Choi, MD

To my colleagues without whom I would not be here.

To my family for their endless support.

And to my cute daughter, whose smile warms my heart with love.

—Hye Jin Yoo, MD

Preface

Diagnostic imaging of soft tissue tumors remains a challenge in musculoskeletal radiology. The difficulty can be attributed to the diversity of soft tissue tumors and the significant overlap in their imaging features. Moreover, confusion persists in the literature regarding the terminology of soft tissue tumors because their taxonomy has evolved over time.

Since the discovery of X-ray, radiologic examination has been widely used to assess bone tumors, whereas soft tissue tumor evaluation was not broadly possible until the introduction of magnetic resonance imaging (MRI) in the early 1980s. Since then, remarkable progress has been made in the imaging of soft tissue tumors. Today, MRI is indispensable in the management of patients with soft tissue tumors, and its utility continues to increase as newly developed technologies become clinically available. As a result, radiology now plays a pivotal role in the detection, diagnosis, and treatment planning of soft tissue tumors.

This book presents a wide range of radiologically illustrated soft tissue tumors classified according to the 2013 WHO classification for soft tissue tumor. Not only radiologists but also other specialists should become well versed in the imaging evaluation of soft tissue tumors, to allow a multidisciplinary approach to treatment. As a radiology textbook, this book aims to familiarize readers with both the imaging findings and pattern analysis of soft tissue tumors. We hope that it will allow readers to improve their understanding of soft tissue tumors and facilitate communication among physicians.

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1.1 Clinical Consideration

The vast majority of soft tissue masses that present to physicians are benign lesions. The incidence of benign soft tissue tumors is estimated to outnumber malignant tumors by a factor of at least 100 (Balach et al. 2011). Unlike their intraosseous counterparts, it is often not possible to establish a meaningful differential diagnosis for soft tissue tumors or reliably determine if they are benign or malignant. In these cases, knowledge of the soft tissue tumor's prevalence, along with the patient's age and the lesion's location, will allow a suitably ordered differential diagnosis (Kransdorf 1995). Embryonal rhabdomyosarcoma occurs most often in children under 10 years of age, while synovial sarcoma is most prevalent in adolescents and young adults. Most soft tissue sarcomas, including undifferentiated pleomorphic sarcoma, liposarcoma, and leiomyosarcoma, dominate in the elderly. The most common tumors in childhood and adolescence are, in order of prevalence, hemangioma, fibrous hamartoma, granuloma annulare, lipoblastoma, fibrosarcoma, and rhabdomyosarcoma. The most common tumors in adults are lipoma, liposarcoma, and myxofibrosarcoma (De Schepper and Bloem 2007).

Lesion locations include subcutaneous, perifascial, intramuscular, intermuscular, and intraarticular/periarticular. The lesion location is important for limiting the differential diagnosis (Walker et al. 2011a). For example, the majority of soft tissue myxomas are intramuscular, whereas myxoid liposarcomas are usually intermuscular lesions (Murphey et al. 2002). Certain tumors are known for their inherent predilections for specific locations, such as elastofibroma, which is typically found in the infrascapular or subscapular area. In a similar fashion, recognizing that a lesion arises from a specific structure (e.g., nerves, vessels, or tendons) can help in lesion characterization (Wu and Hochman 2009). A multifocal or an extensive lesion also limits diagnostic considerations to include angiomatous lesions, neurofibromatosis (NF), fibromatosis, lipomatosis, myxoma (Mazabraud syndrome), metastases, or lymphoma (Walker et al. 2011b).

Clinical history may be relevant in systemic or concomitant diseases, such as melanotic schwannoma in Carney syndrome (cardiac myxoma, spotty pigmentation, and endocrine overactivity), cavernous hemangioma in Maffucci syndrome (enchondromatosis), fibromatosis in Gardner syndrome (intestinal polyposis, osteomata), xanthoma in familial hypercholesterolemia, myxoma in Mazabraud syndrome (polyostotic fibrous dysplasia), amyloidoma in multiple myeloma and neurofibromata, and schwannomata in neurofibromatosis type II and schwannomatosis (De Schepper and Bloem 2007).

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1.2 Imaging Modalities

1.2.1 Radiography

Although the utility of radiographs in evaluating soft tissue lesions is rather limited, some radiographic findings help to detect or characterize soft tissue lesions. At radiography, lipoma or well-differentiated liposarcoma can be appreciated as a low-density mass. If radiography shows a soft tissue mass with multiple phleboliths, it is most likely hemangioma. A soft tissue mass with a central amorphous bone formation is suggestive of extraskeletal osteosarcoma, whereas a soft tissue lesion with peripheral shell-like mineralization is characteristic of myositis ossificans. The presence of multiple punctate or curvilinear calcifications is indicative of an extraskeletal chondroid tumor. Other examples of intratumoral mineralization include pilomatricoma, calcifying aponeurotic fibroma, gouty tophus, and synovial sarcoma. Localized cortical thickening or hyperostosis may occur adjacent to soft tissue hemangiomas or lipomas (Ly et al. 2003; Kim et al. 1999). Any tumor involvement with the adjacent bone may be better evaluated with radiography than with MR imaging. For example, long-term pressure erosion may be easily distinguished from an aggressive permeative destructive pattern on radiography, whereas it may be confusing on MR imaging (Manaster 2013).

1.2.2 Ultrasonography

Because of its safety, availability, feasibility, and cost-effectiveness, ultrasonography (US) is frequently used as the initial imaging technique in the evaluation of soft tissue masses. One of the primary roles of US is to determine whether a mass is a cyst or a solid tumor. Superficial soft tissue masses are particularly amenable to high-resolution US, which allows a detailed assessment of the distinguishing ultrasound characteristics that enable a specific tumor diagnosis based on recognized ultrasound appearances (Hung et al. 2014). Malignant tumors are usually hypoechoic and often are hypervascular, but the sonographic appearances of solid soft tissue tumors are otherwise usually nonspecific (Hwang and Panicek 2009). US is in most instances the primary imaging modality in the evaluation of soft tissue masses in children and is particularly useful in the diagnosis of vascular lesions (Navarro 2011). US is the most popular imaging modality for biopsies of soft tissue tumors. Sonographic guidance is suitable for targeting viable tissues, avoiding necrotic portions in soft issue tumors, and obviating the risk of neurovascular injury.

1.2.3 CT

Although MR imaging is preferred for evaluating soft tissue tumor imaging, there are distinguishing CT characteristics that can suggest a specific diagnosis, including the lesion's mineralization pattern, density, pattern of adjacent bone involvement, and degree and pattern of vascularity (Subhawong et al. 2010). CT remains superior to radiography and MR imaging in demonstrating soft tissue mineralization, and the mineralization pattern of soft tissue masses can be a clue for the histologic diagnosis. In myositis ossificans, for example, CT is more sensitive than MR imaging in detecting early-stage mineralization with zonal phenomenon. Lesion density can also suggest a histologic diagnosis, particularly in soft tissue masses containing fat density. CT is excellent in delineating the presence and pattern of bone involvement in soft tissue tumors, which are important factors during preoperative planning. CT has also become essential in radiation therapy, and the use of CT has resulted in greater precision in dose distribution, patient positioning, and 3D dose calculation (Pereira et al. 2014).

1.2.4 MR Imaging

MR imaging is the diagnostic modality of choice for soft tissue tumors due to the high tissue contrast and multiplanar capability. MR imaging is well suited for not only the diagnosis but also for the staging, preoperative planning, postsurgical evaluation, and post-therapy surveillance of soft tissue tumors. MR images can be particularly useful for characterizing benign lesions that do

not require imaging follow-up or biopsy, such as lipomas and ganglia. In cases where a soft tissue lesion is indeterminate based on the clinical and imaging features, a biopsy should be considered (Wu and Hochman 2009).

A combination of T1- and T2-weighted images is the mainstay of MR imaging of soft tissue tumors. Fat-suppression techniques are widely adopted to enhance the dynamic ranges and sensitivity of fast spin echo T2-weighted images and gadolinium-enhanced T1-weighted images. A T2*-weighted gradient-echo sequence is helpful in the detection of blood products, such as hemosiderin. The administration of intravenous gadolinium chelates is used to distinguish cystic from solid components to identify viable and necrotic areas, to show the relative vascularity of tumors, and to delineate the true margin of tumors.

Most soft tissue tumors are T1 isointense or hypointense and T2 hyperintense in signal intensity. The presence of T1 hyperintensity or T2 hypointensity in soft tissue tumors is occasionally found and helpful in differential diagnosis when present. An intratumoral T1 hyperintensity is suggestive of fat, subacute hemorrhage, high proteinaceous fluid, or melanin. Fat-suppression techniques are helpful to distinguish fat from the other T1-hyperintense materials. T2-hypointense substances include calcifications/ossifications, hemorrhages, vascular signal voids, or collagenous tissues. The T2-hypointense element can be a clue of some benign soft tissue tumors such as tenosynovial giant cell tumor, fibromatosis, and desmoplastic fibroblastoma. However, various soft tissue sarcomas may also have T2-hypointense components. Fluid-containing lesions exhibit very high signal intensity on T2-weighted images, which allows the specific diagnosis of cystic masses, such as ganglion or bursitis. Myxoid tumors also exhibit very high signal on T2-weighted images because of their high water content. Contrast enhancement is helpful to differentiate these bright T2-weighted signal intensity lesions. Contrast-enhanced MR images also show the vascularity of soft tissue tumors. Although malignant lesions are apt to show rapid and greater enhancement, the presence of contrast enhancement does not distinguish benign from malignant tumors.

1.3 Benign Versus Malignant

MR imaging provides clear advantages in terms of differential diagnosis of soft tissue tumors. There is, however, much controversy regarding the value of MR imaging in the differentiation of benign and malignant soft tissue tumors. MR imaging differentiation between benign and malignant tumors is complicated by the low prevalence of these lesions, the minimal experience of radiologists in non-dedicated hospitals, the ambiguous information on MR signal intensities, the highly variable histological findings, and the natural evolution of the lesions (Garcia-Gomez et al. 2004).

The definite malignant indicators are distant metastasis and adjacent organ invasion. The likelihood of malignancy also increases with the presence of tumor necrosis, neurovascular encasement, and bone invasion. The integrity of the deep fascia may also be a differential factor. The trend toward invasive behavior is greater in malignant than in benign tumors, and the destruction of the deep fascia on MR imaging can be a useful imaging finding in identifying malignant tumors (Liu et al. 2011).

Although benign tumors tend to be well delineated and some malignant tumors have ill-defined margins, several studies have concluded that the margin of a soft tissue mass on MR imaging is of no statistical relevance in predicting malignancy (De Schepper et al. 2000). In fact, most soft tissue tumors have well-defined margins regardless of whether they are benign or malignant. The administration of a contrast agent provides further information on the MR imaging characteristics of soft tissue tumors; however, it does not permit the discrimination between benign and malignant lesions when evaluated qualitatively. Dynamic contrast enhancement MR imaging can be used to differentiate malignant from benign soft tissue tumors (Tuncbilek et al. 2005).

In superficial soft tissue tumors, which are defined as masses located within the subcutaneous layer, the following various imaging features are known to be related to malignancy: lobulation, hemorrhage, necrosis, fascial edema, skin thickening, and skin contact. However, size was not found to be an important determining factor for malignancy, with a significant proportion of malignant

superficial sarcomas measuring less than 5 cm in maximal diameter (Calleja et al. 2012).

A study to assess the accuracy of MR imaging in predicting malignancy revealed that the absence of low signal intensity on T2-weighted images, a mean diameter greater than 33 mm, and a heterogeneous signal on T1-weighted images were the most sensitive indicators of malignant soft tissue tumors. MR imaging features with the highest specificity were evidence of necrosis, bone or neurovascular involvement, or metastases and a mean diameter greater than 66 mm (De Schepper et al. 1992).

A simplified systemic MR imaging approach has been proposed to help predict the benign or malignant nature of soft tissue tumors (Chung et al. 2012). The combination of the following three parameters arranged in order resulted in a higher diagnostic value for malignancy: signal intensity (heterogeneity on T2-weighted images), size (≥50 mm), and depth (deep relative to the superficial investing fascia).

1.4 Illustrations: Diagnostic Approach

1.4.1 Location of Soft Tissue Tumors

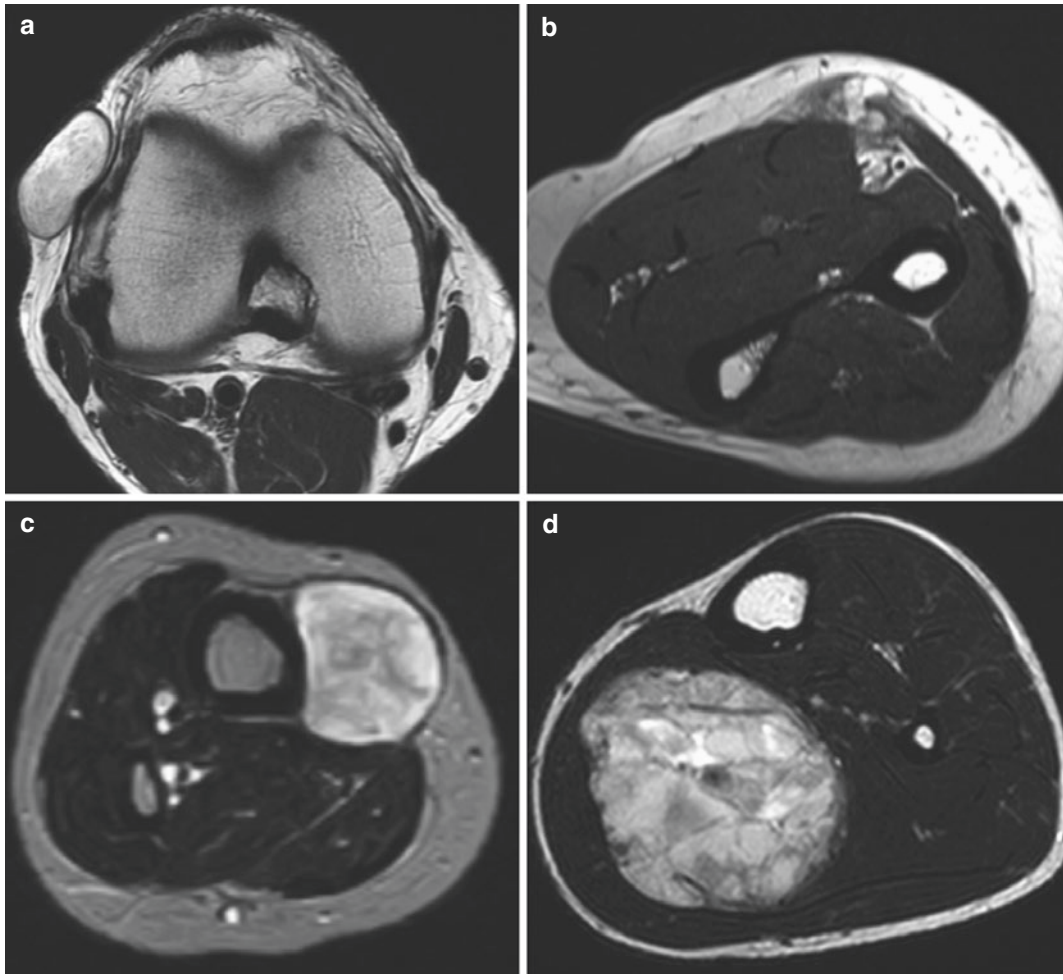

Fig. 1.1 Location of soft tissue tumors. Axial T2WIs show subcutaneous (**a**), perifascial (**b**), subfascial (**c**), and intramuscular (**d**) masses

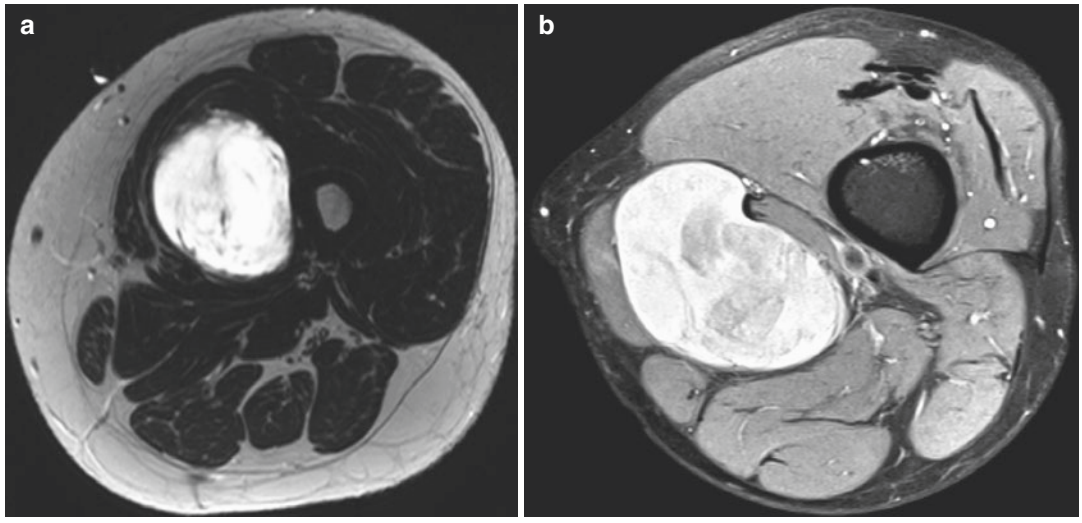

Fig. 1.2 Intra-versus intermuscular location. Axial T2WI (**a**) shows an intramuscular myxoma in the left vastus medialis. Axial FS PDWI (**b**) shows an intermuscular mass proven to be a myxoid liposarcoma in the left thigh

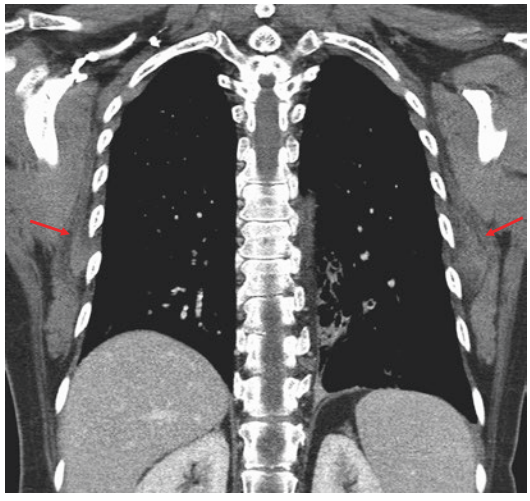

Fig. 1.3 Elastofibroma dorsi. There are bilateral symmetrical soft tissue masses (*arrows*) in the chest wall between the rib cage and serratus anterior. This tumor almost always arises in this very specific location

1.4.2 Soft Tissue Tumor in Clinical Syndrome

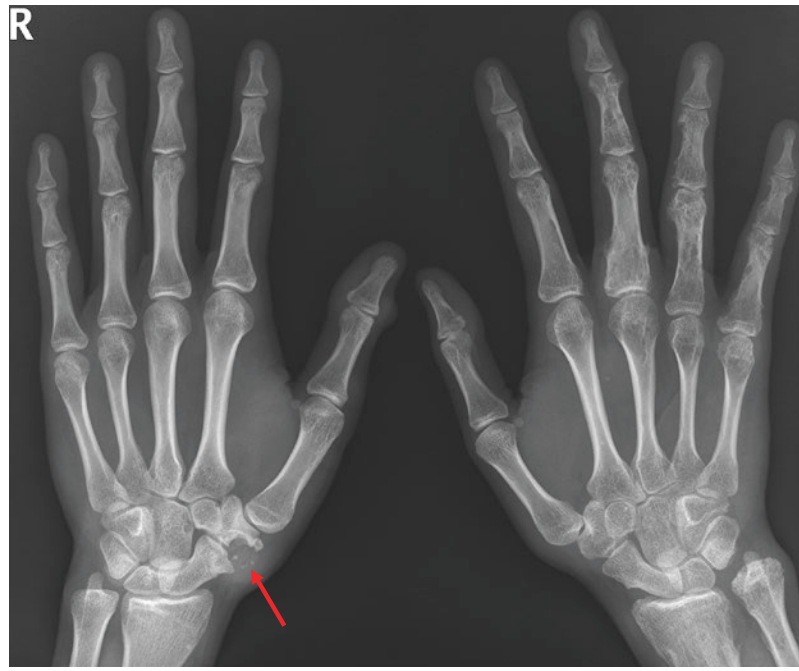

Fig. 1.4 Soft tissue hemangioma in Maffucci syndrome. AP radiograph of both hands shows multiple enchondromas in the left hand and wrist. There is a soft tissue hemangioma (*arrow*) with multiple calcific foci and adjacent bone erosions on the radial side of the right wrist

1.4.3 Radiographic Evaluation

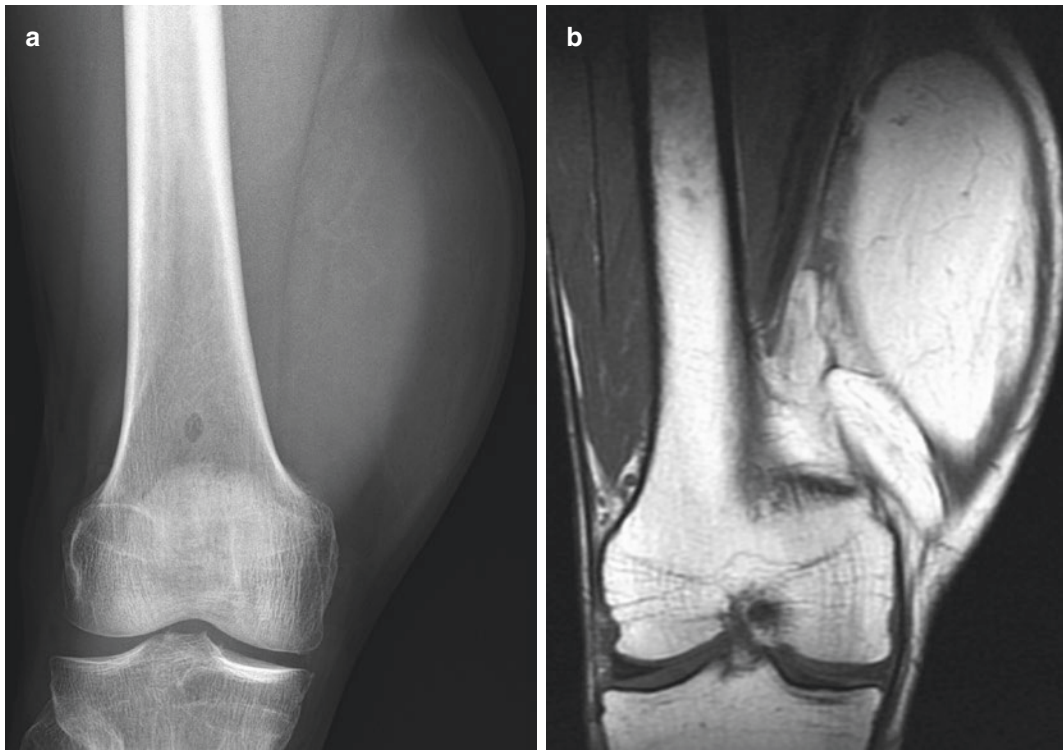

Fig. 1.5 Well-differentiated liposarcoma. AP radiograph of the right knee (**a**) shows a large soft tissue mass with low density indicating fat component. Coronal T1WI (**b**) shows a large fatty mass on the medial side of the right distal thigh

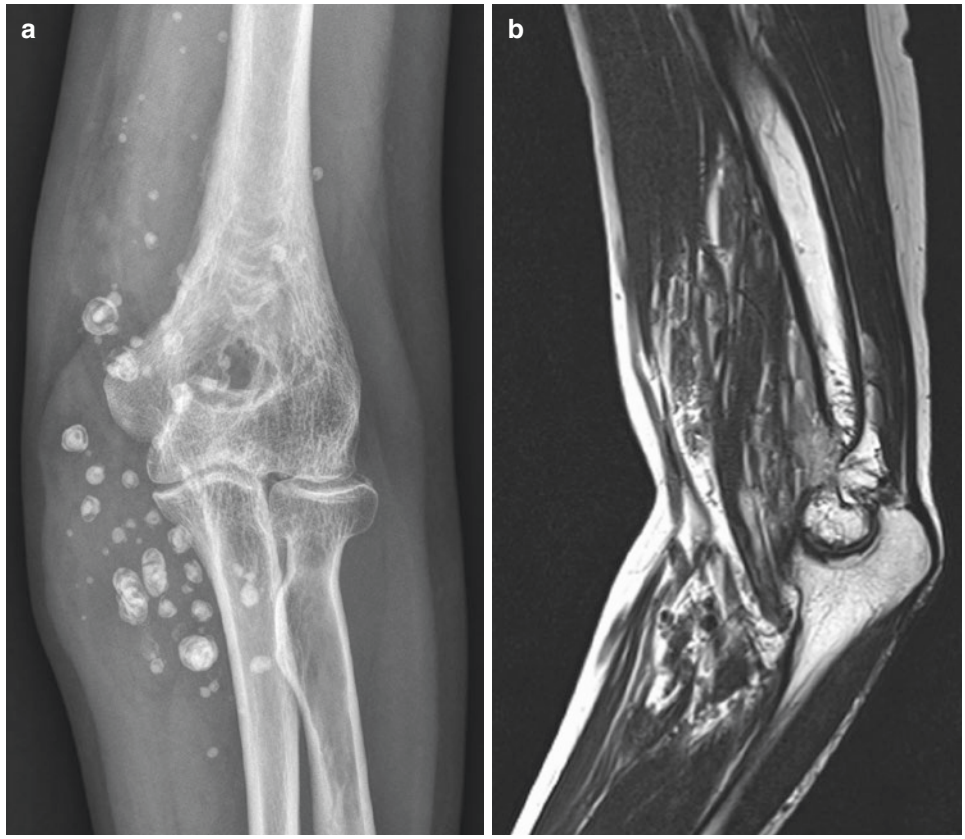

Fig. 1.6 Hemangioma. AP radiograph of the left elbow (**a**) shows a large soft tissue mass with numerous phleboliths. Sagittal T2WI (**b**) shows a complex and infiltrative soft tissue mass with multiple fluid-fluid levels on the left anterior elbow

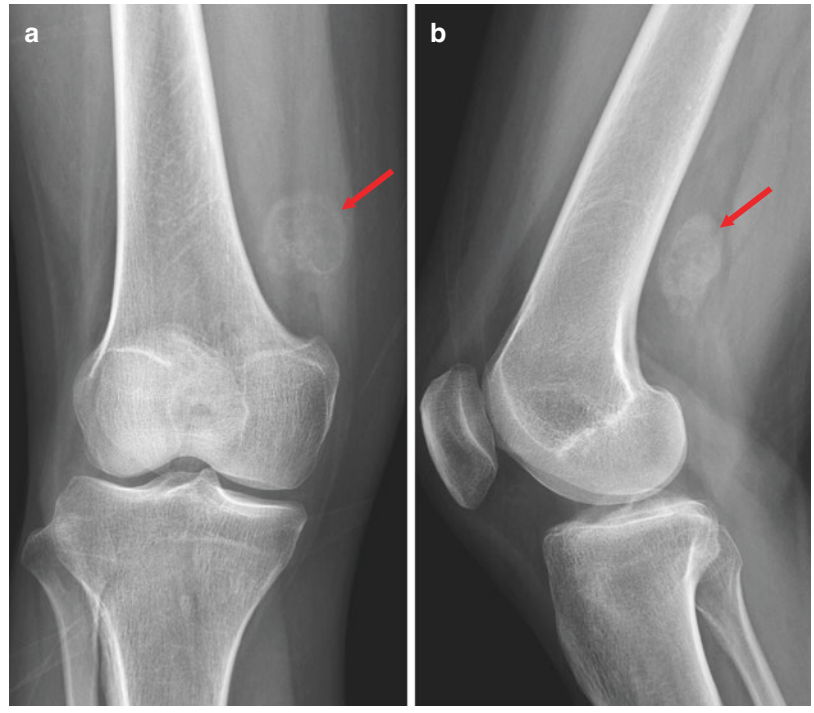

Fig. 1.7 Myositis ossificans. AP (**a**) and lateral (**b**) radiographs of the right knee show a mineralized soft tissue mass (*arrow*) on the posteromedial side of the right distal femur. The mineralization pattern is consistent with myositis ossificans, which exhibits a characteristic zonal phenomenon

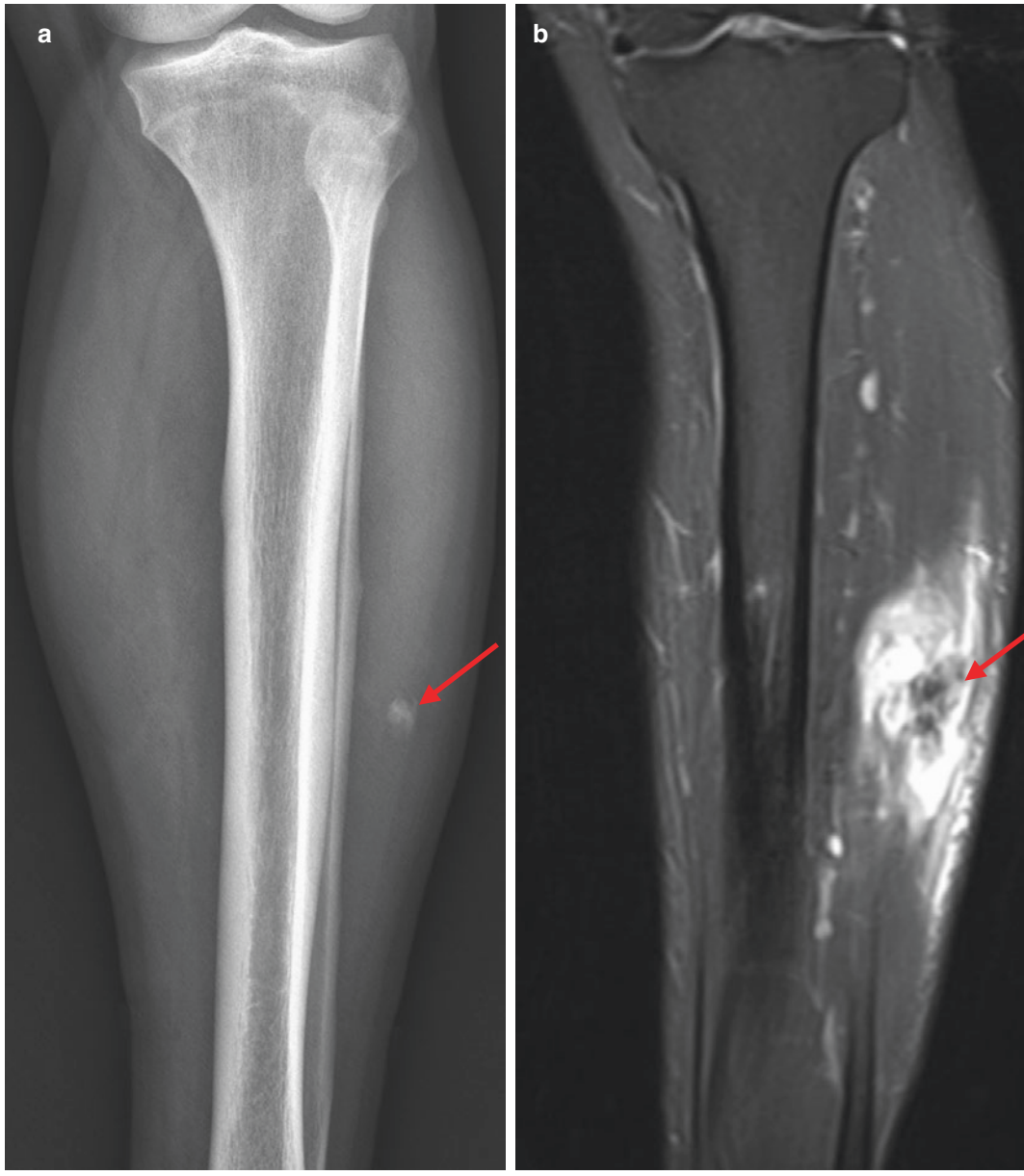

Fig. 1.8 Synovial sarcoma. AP oblique radiograph (**a**) shows a nonspecific calcification (*arrow*) on the lateral side of the left mid lower leg. Coronal postcontrast FS T1WI (**b**) shows a poorly demarcated enhancing soft tissue mass with a central low signal calcification (*arrow*), one of the well-known features of synovial sarcoma

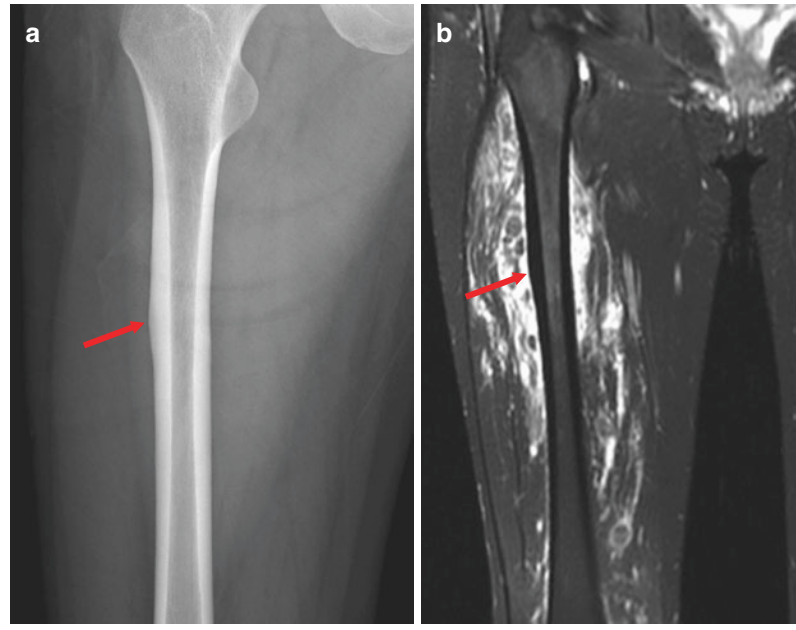

Fig. 1.9 Cortical hyperostosis adjacent to soft tissue hemangioma. AP radiograph of the right thigh (**a**) shows a segmental cortical thickening (*arrow*) on the lateral side of the right femur proximal diaphysis. Coronal FS T2WI (**b**) shows an extensive soft tissue hemangioma with cortical hyperostosis (*arrow*) in the right proximal femur