World Health Organization Classification of Bone and Soft Tissue Tumors: Modifications and Implications for Radiologists

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ABSTRACT

The working group of the World Health Organization (WHO) for classification of tumors of soft tissue and bone met in 2002. The consensus of this conference led to modifications in the nomenclature primarily for soft tissue neoplasm, leaving osseous tumors largely unaltered. The most significant changes in nomenclature involved the group of fibrous and lipomatous malignancies. This article reviews the modifications of this nomenclature and the justification for these changes. The WHO suggested replacement of the term *malignant fibrous histiocytoma* (MFH) with undifferentiated high-grade pleomorphic sarcoma and combining myxoid and round cell liposarcoma under the umbrella of myxoid liposarcoma. The imaging appearances of the fibrous and lipomatous malignancies is reviewed and emphasized in this article. It is important for radiologists involved in evaluation of these lesions to have an understanding of the current nomenclature. This allows improved uniformity in our discussions with pathologists and orthopedic oncologists in our team approach in the diagnosis and treatment of these patients.

KEYWORDS: World Health Organization, soft tissue sarcoma, liposarcoma, fibrosarcoma, malignant fibrous histiocytoma

The working group of the World Health Organization (WHO) for classification of tumors of soft tissue and bone convened in April 2002. The consensus of this conference for nomenclature of these lesions was published in 2002.[1] This article will review and familiarize radiologists with modifications in the nomenclature of soft tissue and bone tumors as a result of the WHO working group.[1] The ultimate goal is to improve uniformity and understanding among radiologists, pathologists, and orthopedic oncologists in the discussion of these lesions and treatment of patients. Bone tumor classification was largely unaltered by the WHO.[1] However, there was significant modification in classification of soft tissue tumors. The most prominent changes in nomenclature occurred in the group of fibrous and lipomatous malignancies. Thus this article focuses on the modification in terminology of the fibrous malignancies and liposarcoma and the imaging appearances of these lesions.

FIBROUS MALIGNANCIES

The term *malignant fibrous histiocytoma* (MFH) has fallen out of favor for a variety of reasons.[2–5] This lesion

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was originally defined as a pleomorphic spindle cell malignancy demonstrating both fibroblastic and facultative histiocytic differentiation.[6–9] However, more recent evaluation has not revealed true histiocytic differentiation. The morphological pattern seen with pleomorphic MFH is shared by a large variety of poorly differentiated primary malignant neoplasms. Some believe with newer refinement in immunohistochemistry staining and molecular analysis that the designation of lesions as MFH will continue to decline and be replaced by more specific fibroblastic or myofibroblastic lineage of neoplasms.[10]

These discussions have led to the WHO to suggest use of the terminology *undifferentiated high-grade pleomorphic sarcoma* to replace pleomorphic MFH, *undifferentiated pleomorphic sarcoma with giant cells* for MFH (giant cell type), and *undifferentiated pleomorphic sarcoma with prominent inflammation* for inflammatory MFH.[11] In addition, the terminology *myxofibrosarcoma* is preferred, as opposed to myxoid MFH. In my experience, these designations are cumbersome at best, and the majority of pathologists have retained the previous nomenclature of MFH in their daily practice. These terms are also more familiar to our surgical oncology, orthopedic oncology, and medical oncology colleagues. The additional designations by the WHO of other fibrous malignancies include fibrosarcoma (adult and infantile), low-grade fibromyxoid sarcoma, myxoinflammatory fibroblastic sarcoma, low-grade myofibroblastic sarcoma, sclerosing epithelioid fibrosarcoma, and inflammatory myofibroblastic tumor (Table 1).[1,12–29] These add further complication for radiologists and orthopedic oncologists in the use of this nomenclature. The nonuniformity in these designations is further illustrated by the fact that this nomenclature is reserved for soft tissue neoplasms, whereas intraosseous lesions have maintained use of the terminology of MFH and fibrosarcoma.

In my opinion, radiologists that frequently are involved in the evaluation of these fibrous malignancies should have at least a superficial understanding of these nuances of nomenclature. This may become important because there are prognostic variations within this diverse group of lesions with treatment implications. In the future, various imaging characteristics may also be detected that may allow distinction of these lesions. However, to the best of my knowledge, no significant differences in the intrinsic characteristics of these lesions have been described.[30]

Table 1 WHO Fibrous Malignancies

Inflammatory myofibroblastic tumor
Low-grade myofibroblastic sarcoma
Myxoinflammatory fibroblastic sarcoma
Fibrosarcoma (adult and infantile)
Myxofibrosarcoma
Low-grade fibromyxoid sarcoma
Sclerosing epithelioid fibrosarcoma
Undifferentiated high-grade pleomorphic sarcoma
Undifferentiated pleomorphic sarcoma with giant cells
Undifferentiated pleomorphic sarcoma with prominent inflammation

The fibrous malignancies are the most common soft tissue sarcomas, accounting for 20 to 50% of all cases.[31,32] As a group, the fibrous malignancies most commonly affect middle aged to older patients (40 to 70 years of age).[33] Lesions are most common in the deep soft tissues of the lower extremity, particularly the thigh. The clinical presentation is nonspecific, with a painless and enlarging soft tissue mass.

Cross-sectional imaging of these fibrous malignancies typically reveals a large heterogeneous intramuscular or, less commonly, subcutaneous soft tissue mass in a middle aged to older patient.[33–36] The intrinsic imaging characteristics are nonspecific but lack the presence of adipose tissue, and lesion margins are frequently relatively well defined (Fig. 1). These well-defined margins are seen at gross pathological examination as well and correspond to the pseudocapsule associated with both benign and malignant soft tissue neoplasms. This emphasizes that the definition of the margin should not be used radiologically in determining the likelihood of benignity or malignancy of a soft tissue neoplasm.

On magnetic resonance (MR) imaging, signal intensity of the soft tissue fibrous malignancies is typically intermediate to low intensity on T1 weighting and intermediate to high signal intensity on T2 weighting (Fig. 1).[33] The signal intensity variation is likely related to the various components of fibrous tissue with high collagen content (low signal intensity on all pulse sequences), myxoid tissue (low signal intensity on T1 weighting and high signal intensity or T2 weighting) and hemorrhage (variable signal intensity).[32,33] This intermixture of tissue frequently causes significant heterogeneity on all MR pulse sequences in these fibrous malignancies. Calcification is present in 5 to 20% of MFHs and may also be a cause for low signal intensity on all MRI sequences.[32,33] However, calcifications are superiorly detected and characterized by computed tomography (CT) or radiographs.[37] Involvement of the underlying bone by cortical and marrow invasion is not rare (~5 to 10%) in my experience.[32,33] Low signal intensity may predominate on long TR images, presumably related to prominent collagen content in some lesions. This feature has been reported in sclerosing epithelioid fibrosarcoma as well.[38]

The imaging of the myxoid varieties of the fibrous malignancies, including myxoinflammatory fibroblastic sarcoma, myxofibrosarcoma (myxoid MFH), and low-grade fibromyxoid sarcoma, often reflects the high water

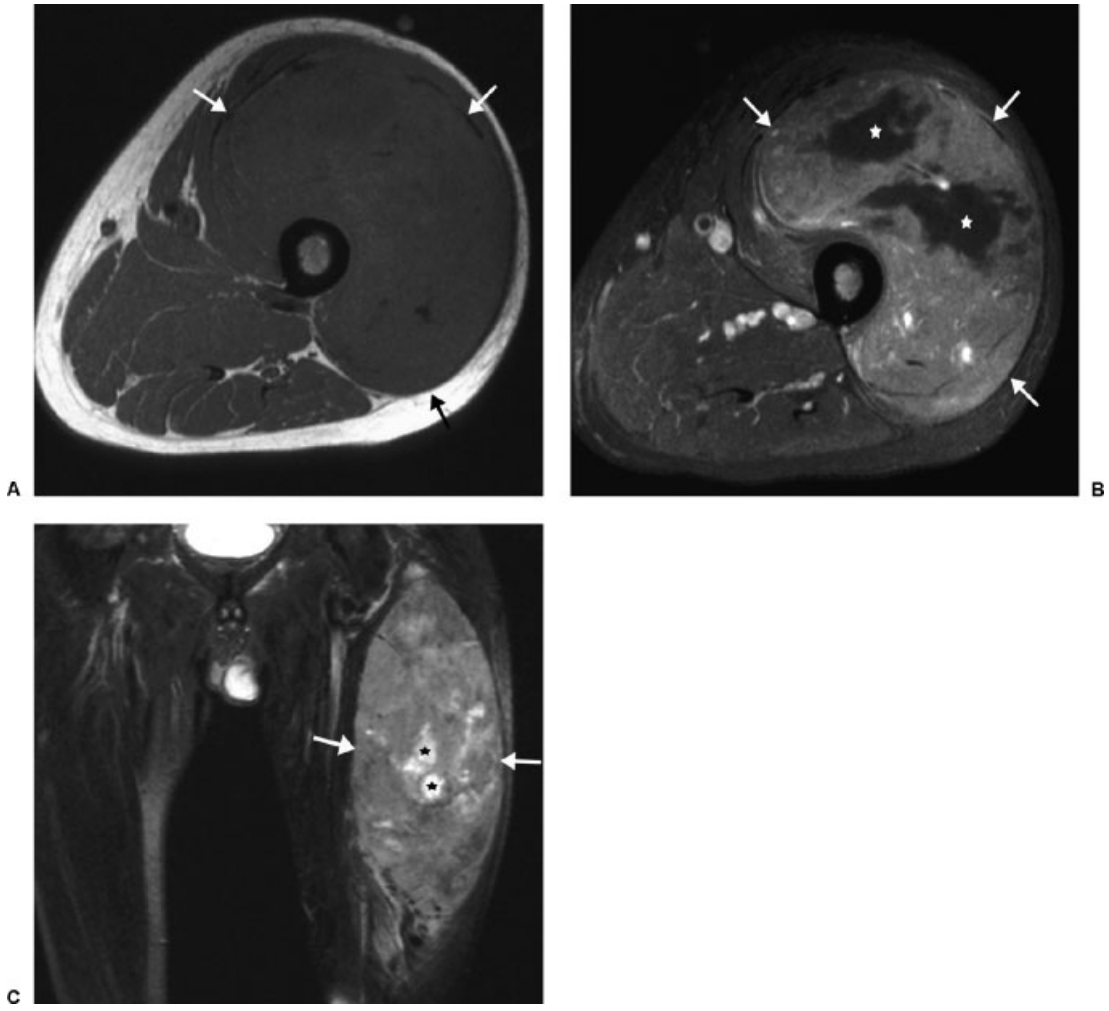

Figure 1 Pleomorphic malignant fibrous histiocytoma (undifferentiated high-grade pleomorphic sarcoma) in a 49-year-old man with a large thigh mass. (A, B) Axial T1-weighted (A, 500/20) and postcontrast fat-suppressed T1-weighted (B, 400/15) postcontrast with fat suppression MR images reveal a large intramuscular soft tissue mass (arrows) that is intermediate signal intensity on T1 weighting and has heterogeneous contrast enhancement with nonenhancing central areas of necrosis (asterisks). (C) Coronal fat-suppressed T2-weighted (4000/50) MR image reveals heterogeneous predominantly intermediate signal intensity soft tissue mass (arrows) with foci of high signal centrally corresponding to central necrosis (asterisks).

content of these lesions. Cross-sectional imaging frequently demonstrates low echogenicity on sonography, low attenuation or CT, low signal on T1-weighted MRI, and prominent high signal intensity on long TR MR images.[33,39] In our experience, this is more frequently a feature associated with myxoid MFH as compared with low-grade fibromyxoid sarcoma. However, these myxoid features are not unique to the fibrous malignancies and can also be seen with myxoid liposarcoma (see later discussion), neurogenic tumors, extraskeletal myxoid chondrosarcoma, and myxomas.

Hemorrhage is a frequent component of the fibrous malignancies involving the soft tissues and may be a predominant feature in a minority of cases (Fig. 2).[40] This can make distinction of hematoma versus hemorrhagic neoplasm (particularly MFH) a diagnostic dilemma with obvious clinical and medicolegal ramifications. Clinically, these patients may present with a "spontaneous" hematoma or hematoma out of proportion in size relative to the degree of trauma. This history alone should warrant concern for a hemorrhagic neoplasm, as opposed to a hematoma. On CT, the hemorrhagic regions are higher attenuation acutely but decreased attenuation chronically. MR similarly reveals time-dependent variation in signal intensity of hemorrhage owing to different component of hemoglobin and its breakdown products (oxyhemoglobin, deoxyhemoglobin, and methemoglobin). However, subacute blood most commonly reveals high signal intensity on both T1-weighted and T2-weighted MR images. Fluid levels may also be apparent on sonography, CT, and MR imaging. These features may significantly obscure the underlying neoplasm.

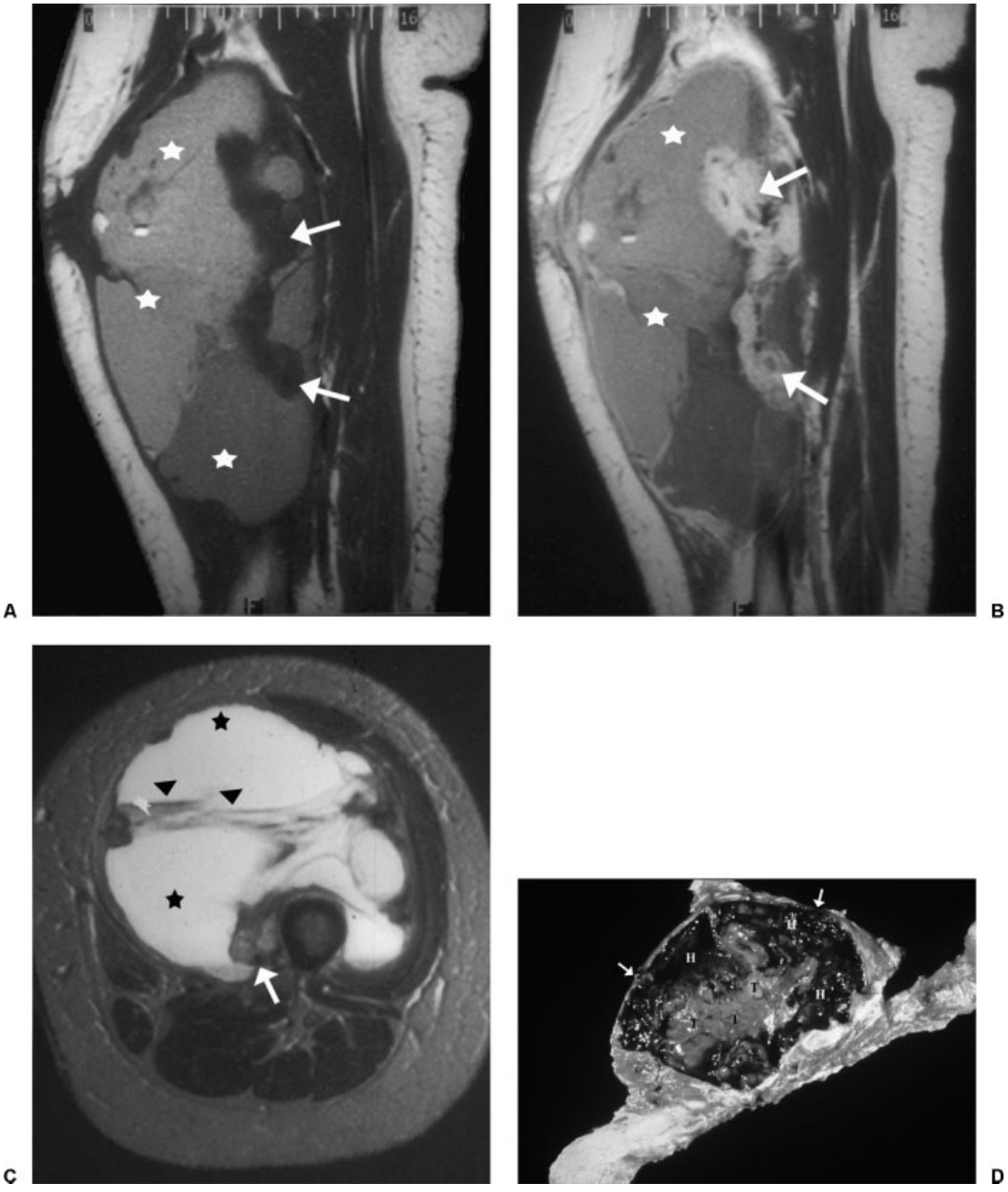

Figure 2 Pleomorphic malignant fibrous histiocytoma (undifferentiated high-grade pleomorphic sarcoma) presenting as a ''spontaneous hematoma'' in a 50-year-old man. (A–C) Multiple MR images including sagittal T1 weighting before (A, 500/20) and following intravenous contrast (B, 500/20) and axial T2 weighting (C, 2500/100) reveal the large mass that is predominantly composed of hemorrhage (asterisks show high signal intensity areas on short and long TR images) with fluid levels (arrowheads). Solid focus of viable malignant fibrous histiocytoma (viable tissue that should be biopsied) enhances after contrast and is intermediate in signal intensity on T2 weighting (arrows). (D) Photograph of sectioned gross specimen also demonstrates the hemorrhagic component (H) and viable portion of the tumor (T) and peripheral pseudocapsule (arrows) that contained the hemorrhage causing the well-defined margin and lack of surrounding edema on imaging.

Several imaging features are helpful in distinguishing a hemorrhagic neoplasm (such as an MFH) from a hematoma (Table 2).[32,33] Most importantly areas of solid viable tissue, not hemorrhagic foci, are often apparent and must be searched for diligently. These foci are most commonly peripheral in location (Fig. 2). Contrast enhancement patterns are also very useful in differentiation: Viable tumor nodules diffusely enhance, often in a peripheral nodular pattern (Fig. 2). Surrounding edema is frequently a prominent feature of

Table 2 Differentiation Neoplasm vs. Hematoma

Hematoma	Tumor
History of trauma or anticoagulants	Spontaneous
Surrounding edema	No surrounding edema, nodular wall
Thicker nonnodular wall	Solid nodular enhancing areas
Rim enhancement	No hemosiderin in wall
Hemosiderin in wall	

hematoma (particular in the early stages) as opposed to hemorrhagic neoplasm. The pseudocapsule surrounding neoplasm contains the hemorrhage, which limits significant adjacent edema (Fig. 2). Finally, the wall of a hematoma is usually thicker but not nodular, as compared with a hemorrhagic tumor. In addition, the wall of a chronic hematoma frequently contains hemosiderin, causing marked low signal intensity resulting from magnetic susceptibility artifact ("blooming") on gradient echo MR imaging sequences. This "blooming" artifact is often subtle but, in my experience, it is not seen in hemorrhagic neoplasms, aiding in distinction of these lesions. These differentiating features allow distinction of the vast majority of these cases. In the unusual situation that this differentiation cannot be confidently accomplished radiologically, follow-up imaging (to detect growth in a neoplasm versus stable size of hematoma) or, more commonly, biopsy may be necessary. Imaging is useful to direct biopsy to areas suggesting solid viable tissue more likely to harbor pathological diagnosis.

Contrast enhancement of the fibrous malignancies reveals several common patterns by CT or MR imaging. Diffuse, heterogeneous enhancement is the most frequently encountered pattern. Peripheral and nodular enhancement is also frequent. This enhancement pattern may be associated with central hemorrhage or necrosis (Fig. 1). In addition, the myxoid fibrous malignancies may also reveal peripheral nodular contrast enhancement.

Treatment of the fibrous malignancies is aggressive with wide surgical excision. High-grade lesions such as MFH are frequently treated with adjuvant radiation therapy and chemotherapy.[32,41,42] Despite this aggressive treatment, the overall prognosis for soft tissue MFH is unfavorable. Deep-seated MFH has a 43% rate of metastases. The overall 5-year survival rate for soft tissue MFH is 36 to 50%.[32,33,43] Distant metastases most frequently affect the lung (90%) and bone (8%).[32,43]

LIPOMATOUS MALIGNANCIES

Lipomatous malignancies currently include well-differentiated liposarcoma, dedifferentiated liposarcoma, myxoid liposarcoma, pleomorphic liposarcoma, and mixed-type liposarcoma. Previous nomenclature included a round cell variety of liposarcoma. However, the WHO has incorporated this lesion into the category of myxoid liposarcoma.[44,45]

Well-differentiated liposarcoma represents the most common type of liposarcoma, accounting for ~50% of lesions.[46–48] Evans and colleagues introduced the term *atypical lipomatous tumor* in 1979 as alternative nomenclature for well-differentiated liposarcoma, to reflect more accurately the lesion's lack of metastatic potential.[49,50] However, the designation of atypical lipomatous tumor has been variably applied and led to potential diagnostic confusion.[51] The WHO uses the terms *atypical lipomatous tumor* and *well-differentiated liposarcoma* synonymously because they are identical in morphology, karyotype, and biological behavior.[1] The WHO Committee on Classification of Soft Tissue Tumors further suggested that the choice in nomenclature "is best determined by the degree of reciprocal comprehension between the surgeon and pathologist to prevent either inadequate or excessive treatment." I personally restrict use of the term *atypical lipomatous tumor* to lesions in a subcutaneous location, reflecting their low morbidity and almost nonexistent potential for dedifferentiation (much less than 1%).[48,52] I strongly advocate the use of the term *well-differentiated liposarcoma* for lesions in all other locations, particularly in the retroperitoneum. That is because of the belief, in agreement with Weiss and Goldblum, that the description of atypical lipomatous tumor is not adequate to accurately emphasize the significant morbidity, risk of dedifferentiation, and mortality associated with these lesions.[52]

Similar to other soft tissue sarcomas, the typical clinical presentation of well-differentiated liposarcoma is that of a painless, slowly enlarging mass. Well-differentiated liposarcomas most frequently affect the deep soft tissue of the extremities (65 to 75% of cases). The retroperitoneum is the second most common location for this lesion, accounting for 20 to 33% of cases.[47,48]

Cross-sectional imaging of well-differentiated liposarcoma is frequently characteristic.[47,48,53–58] CT and MR imaging reveal a lesion composed of >50 to 75% adipose tissue with additional prominent nonlipomatous components in 91 to 96% of cases (Fig. 3).[48,59,60] The nonlipomatous components of the lesion commonly reveal thick (>2 mm) and numerous septa or focal globular or nodular regions (typically <2 cm in size) (Fig. 3). Well-differentiated liposarcoma can have an appearance similar to lipoma in 4 to 9% of cases.[48,59,60] Ohguri et al[60a] and Hosono et al[60b] have suggested that prominent contrast enhancement of the septa on MR imaging suggests well-differentiated liposarcoma as opposed to lipoma, which may aid in distinction in these cases. Lipomatous lesions in the retroperitoneal should always be considered a liposarcoma regardless of the intrinsic imaging appearance

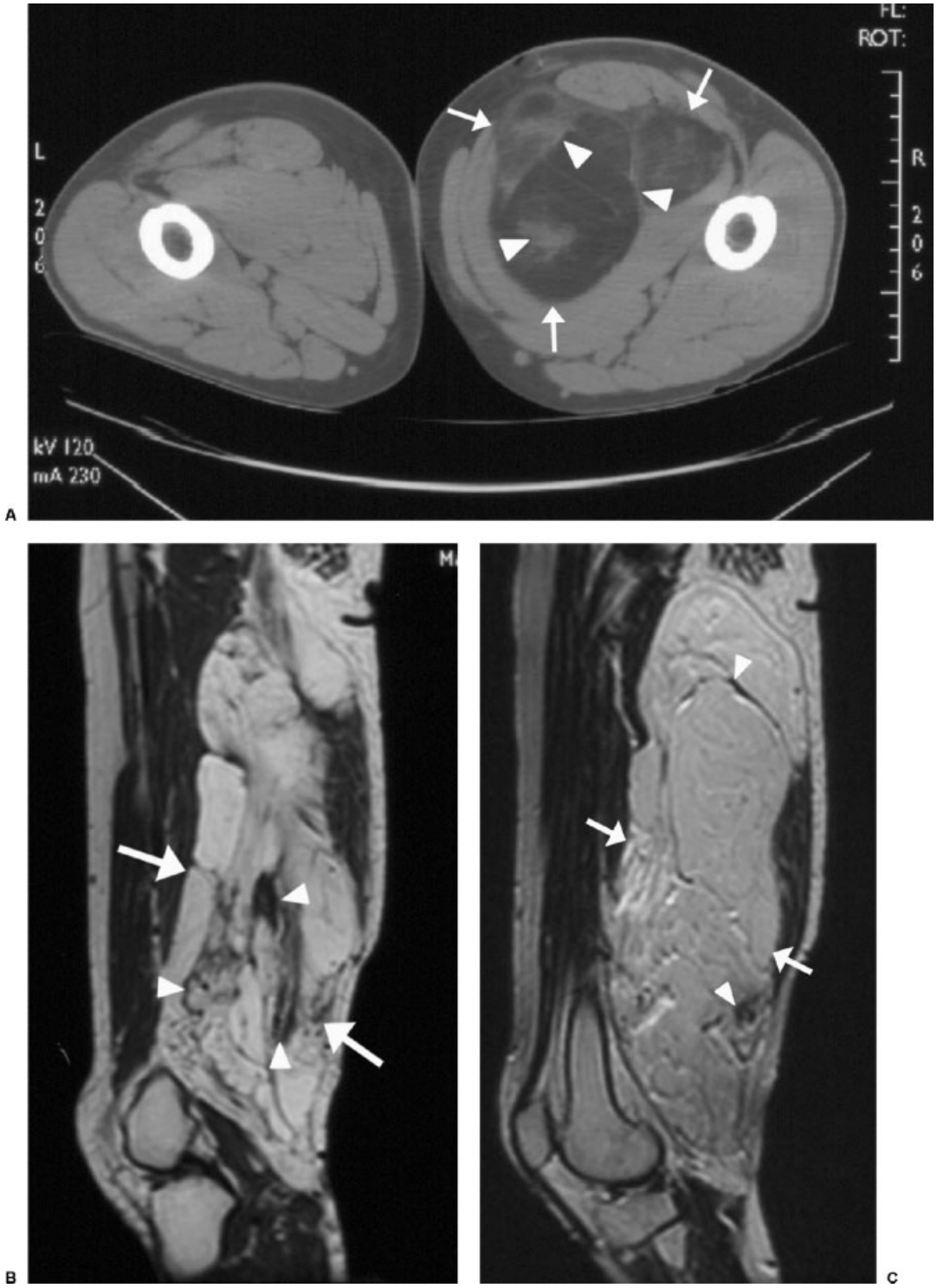

Figure 3 Well-differentiated liposarcoma of the distal thigh in a 65-year-old woman. (A–C) CT (A), sagittal T1-weighted (B, 650/14), and sagittal T2-weighted (C, 4500/98) MR images show a largely lipomatous soft tissue mass (arrows) with prominent nonlipomatous components seen as irregular thick septae and globular regions (arrowheads) typical of well-differentiated liposarcoma.

because lipomas are extraordinarily rare in this location. Biopsy of extremity lesions is often fraught with uncertainty, owing to the large size and heterogeneity of these lesions and possibility of not obtaining diagnostic tissue to allow differentiation from lipoma. Imaging may be helpful to direct biopsy to more nonlipomatous regions, allowing more confident pathological diagnosis.

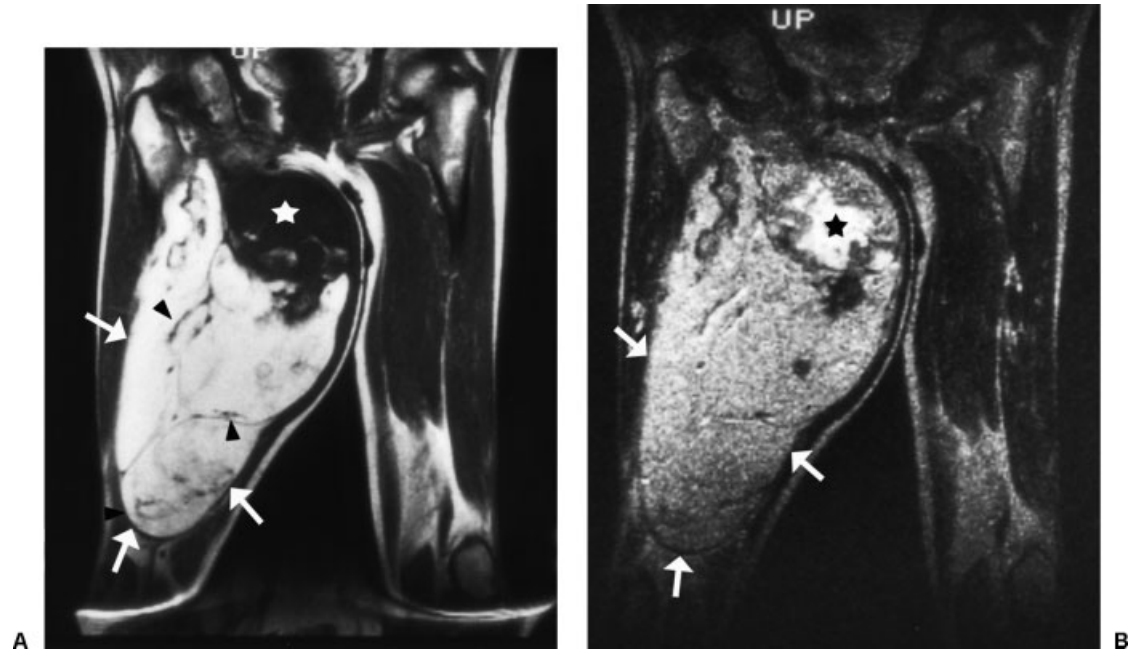

Figure 4 Dedifferentiated liposarcoma of the thigh in a 72-year-old man with a 10-year history of a slowly enlarging mass with more recent rapid growth. (A, B) Coronal T1-weighted (A, 500/16) and T2-weighted (B, 1600/120) MR images reveal a mass largely composed of tissue isointense to subcutaneous fat (arrows) but also containing thick septae (arrowheads). There is also a large nodular nonlipomatous component with nonspecific characteristics of low signal intensity on T1 weighting and heterogeneous intermediate to high signal intensity on T2 weighting superiorly (asterisks).

The local recurrence rate of extremity and retroperitoneal well-differentiated liposarcoma is 43% and 91%, respectively.[61,63] A mortality rate of 33% has been reported for retroperitoneal lesions and 14% for groin lesions.[61–63] These figures further emphasize the shortcomings in the use of the term *atypical lipomatous tumor*. Radiation therapy may be used as an adjuvant to surgical resection to reduce the incidence of local recurrence, particularly for retroperitoneal and mediastinal lesions. In my opinion, I would strongly advocate against its use in the vast majority of extremity lesions because of potential complications of radiation. This must be compared and contrasted in the clinical context to the nonmetastasizing behavior of this lesion.

Dedifferentiated liposarcoma arises within a well-differentiated lesion, representing a biphasic tumor. The nonadipose component is typically a high-grade cellular sarcoma (often MFH or fibrosarcoma). Dedifferentiation occurs in ~10% of well-differentiated liposarcomas, and the incidence depends on lesion location.[64] The estimated risk of dedifferentiation in retroperitoneal lesions is 15% and much higher than other locations.[64] It has been suggested that this higher incidence of dedifferentiation in retroperitoneal tumors is related to the delayed diagnosis at this site and larger size at detection as compared with extremity lesions.[64] The estimated risk of dedifferentiation for deep-seated extremity lesions is ~5%.[64]

Imaging features correlate with the biphasic nature of this lesion. The largest component has features of well-differentiated liposarcoma, as previously described. However, in addition, there is a focal nodular nonlipomatous component that is typically larger than 2 to 3 cm in size (Fig. 4).[48] This nonadipose focus represents the region of dedifferentiation and usually shows nonspecific intrinsic imaging characteristic of a solid mass on cross-sectional imaging (Fig. 4). The CT attenuation of this region is similar to muscle. On MR imaging, there is low to intermediate signal intensity on T1 weighting and intermediate to high signal intensity on T2 weighting. Diffuse contrast enhancement is typically seen on MR in the focus of dedifferentiation as well. Other causes of focal nonlipomatous regions within a well-differentiated liposarcoma include collagenized areas, metaplastic mineralization, and fat necrosis. In my experience, these areas are usually smaller than 1 to 2 cm, and radiography or CT can detect areas of calcification optimally. Peripheral rim enhancement on postcontrast MR may help identify fat necrosis and allow distinction from a region of dedifferentiation.

It is vital for radiologists to recognize that any prominent solid nodular focus (>2 cm) within a well-differentiated liposarcoma should be biopsied because of the concern for dedifferentiation and its implication on treatment and prognosis. In fact, a strong argument can be made that this distinction is more important for patient management than the distinction of lipoma versus well-differentiated liposarcoma of the extremities.

Dedifferentiated liposarcoma treatment is much more aggressive than well-differentiated lesions, with

wide surgical excision (if possible) as well as adjunct radiation therapy and chemotherapy. Despite this aggressive therapy, the overall local recurrence rate is 41%, and in retroperitoneal lesions it is 100%.[47,48,64] The overall mortality rate for dedifferentiated liposarcoma is 28 to 30% and nearly 100% in retroperitoneal lesions. Distant metastases to the lungs, liver, and bone occur in 15 to 20% of cases.[47,48,65]

Myxoid liposarcoma represents 20 to 50% of all liposarcomas and is the second most common subtype of these lesions.[31,48,66] The WHO has coalesced myxoid and round cell subtypes as myxoid liposarcoma in recognition of the continuum of these lesions. However, despite this nomenclature, detection of areas suspicious for containing significant round cell components (> 5%) is important because of treatment and prognostic implications.

Myxoid liposarcomas most commonly affect the lower extremity (75 to 80% of lesions) and occur in patients approximately a decade younger (fourth and fifth) than other liposarcomas.[47,48] The clinical presentation is that of a painless, enlarging soft tissue mass that may be quite large (> 15 cm).

Cross-sectional imaging of myxoid liposarcomas typically reveals a large intramuscular (70 to 80%) soft tissue mass.[48] The high water content of the myxoid component predominates by volume and causes hypoechogenicity on sonography, low attenuation on CT, low signal intensity on T1 weighting, and very high signal intensity on T2-weighted MR images (Fig. 5).[67,68] However, the myxoid components are nonspecific as to diagnosis because many other neoplasms have this consistency.[69] The pathognomonic feature by imaging is the additional detection of an adipose component (usually < 5 to 10% of the tumor volume) within the myxoid background (Fig. 5). This is best accomplished by MR imaging as compared with CT and/or sonography.[70] The literature suggests that the detection of fat is seen in 42 to 78% of cases of myxoid liposarcoma (Fig. 5).[47,48,71] However, in my experience, the incidence of identifying this pathognomonic appearance is much higher (~90 to 95%).[48,72] Detection of the adipose component can be quite subtle, particularly when very small in volume. Careful attention is needed in comparing T1-weighted and T2-weighted MR images in the same plane (axial plane is usually optimal), and correlation to fat-suppressed or fat-saturated sequences aids in detection of these foci of fat.

Myxoid liposarcomas may simulate a cyst on CT and noncontrast MR images in 5 to 22% of cases, without evidence of an adipose component (Fig. 6). These imaging features may simulate ("cyst mimickers") ganglion, synovial cyst, bursa, liquefied hematoma, or abscess. The lesion location is usually the clue that the lesion is a "cyst mimicker" requiring additional imaging, as opposed to the expected position of a ganglion, synovial cyst, or bursa (Fig. 6). Lack of surrounding edema and thick walls as well as absence of correlative appropriate clinical history typically allows distinction from liquefied hematoma or abscess. Myxoma is an additional consideration, although these lesions are usually small and intermuscular.[73,74] Sonography or post-contrast MR imaging provides convincing evidence of the solid nature of myxoid liposarcoma. Sonography reveals a hypoechoic (but not anechoic) mass.[39] MR imaging demonstrates either diffuse or nodular enhancement, not a thin rim and septal pattern as expected for a truly cystic mass (Fig. 6).

Round cell components have features that may be detected on MR imaging, in my experience (Fig. 7).[48,75] These foci are often nodular, with nonspecific features of solid but nonmyxoid elements (Fig. 7). On CT, the attenuation of the round cell areas is similar to muscle, and on MR intermediate signal intensity on T1 weighting and T2 weighting (Fig. 7). Identification of foci with this appearance in the background of a myxoid liposarcoma should alert the radiologist to biopsy these regions, to aid in appropriately aggressive preoperative treatment.

Myxoid liposarcomas are intermediate-grade lesions treated by wide surgical excision; adjuvant chemotherapy may also be employed. Myxoid liposarcomas metastasize in 23% of cases with an overall 5-year survival rate of 47 to 77%.[48,51] However, the incidence of metastases incrementally increases with larger percentages of round cell components from 35% (5 to 10% round cell component) to 56% (> 25% round cell component), emphasizing the clinical importance of identifying these regions.[48,76] Metastases from myxoid liposarcoma demonstrates a predilection from nonpulmonary locations (94%), particularly the pleura, pericardium, peritoneum, chest wall, and retroperitoneum.[48] Metastatic lesions may maintain their high water content at imaging evaluation.

Pleomorphic liposarcoma represents 5 to 15% of all liposarcomas.[48,77] This lesion typically affects patients > 50 years of age.[48,77] The lower extremity is the most frequently involved site (accounting for 50 to 60% of cases), with the upper extremity affected in 20% of patients.[48]

Cross-sectional imaging of pleomorphic liposarcoma reveals a large heterogeneous intramuscular mass. Areas of necrosis and hemorrhage are common. Areas of adipose tissue are less frequently detected by MR imaging (62 to 75%) as compared with other subtypes of liposarcoma.[48,55] This is a reflection of the higher degree of anaplasia of these high-grade neoplasms. Signal intensity of viable tumor areas on T2 weighting are usually intermediate to high signal but not as markedly increased as in myxoid liposarcoma. Biopsy at least partially directed at the adipose-containing areas, if present, can aid the pathologist in preoperative diagnosis of these lesions.

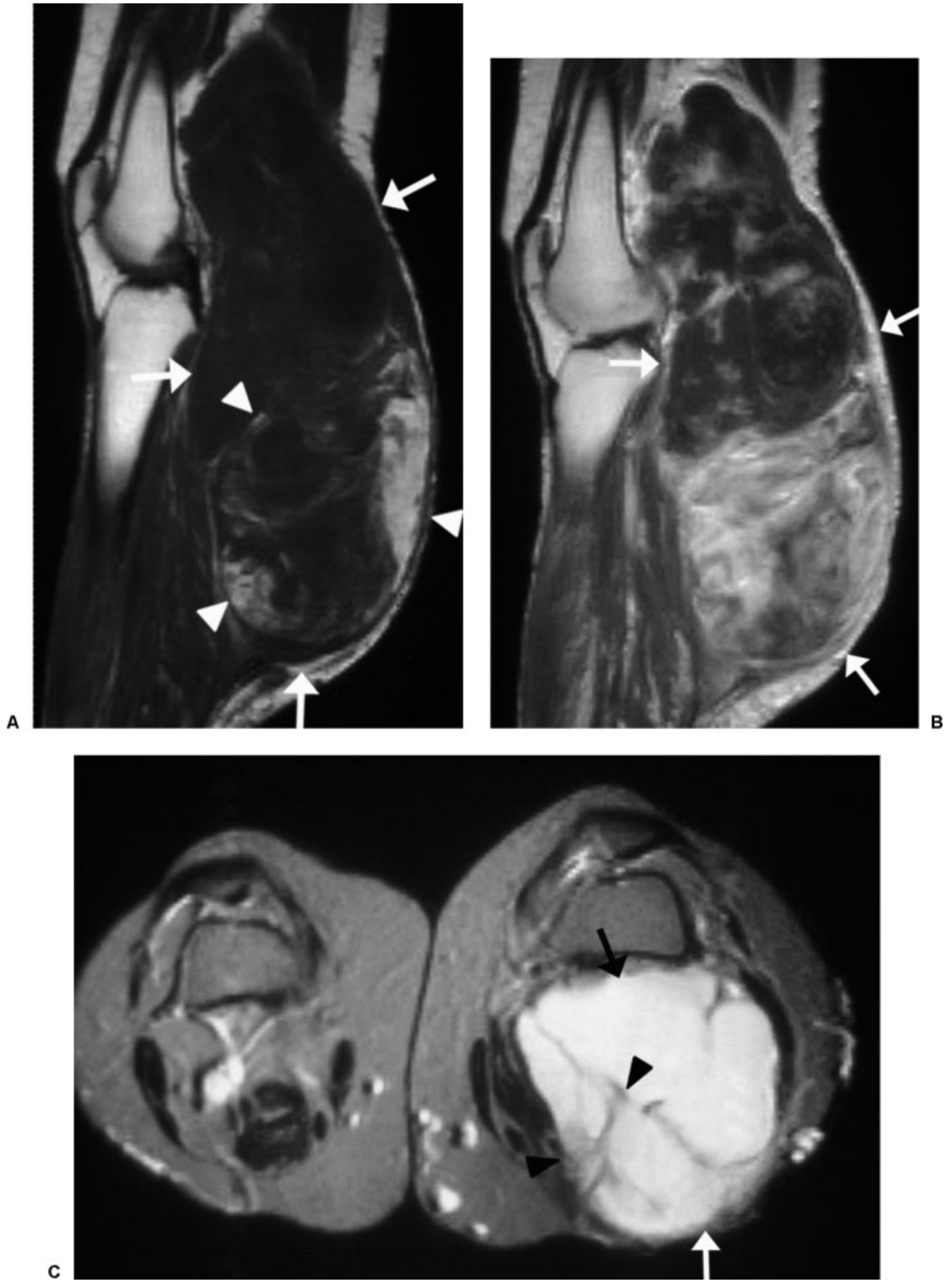

Figure 5 Myxoid liposarcoma of the popliteal region in a 60-year-old woman with a painless, slowly enlarging mass. (A–C) Sagittal T1-weighted (A, 500/20) MR images both (A) before and after (B) intravenous contrast and axial T2-weighted (C, 2500/90) MR images show a large heterogeneous intermuscular popliteal mass (arrows). The predominant signal intensity is that of a high water content mass with low signal on T1 weighting and high signal on T2 weighting. However, focal areas in the septae and several small nodular regions (arrowheads) (< 10% of the tumor volume) are isointense to subcutaneous fat. Following contrast administration there is thick and nodular peripheral and septal enhancement most prominent inferiorly.

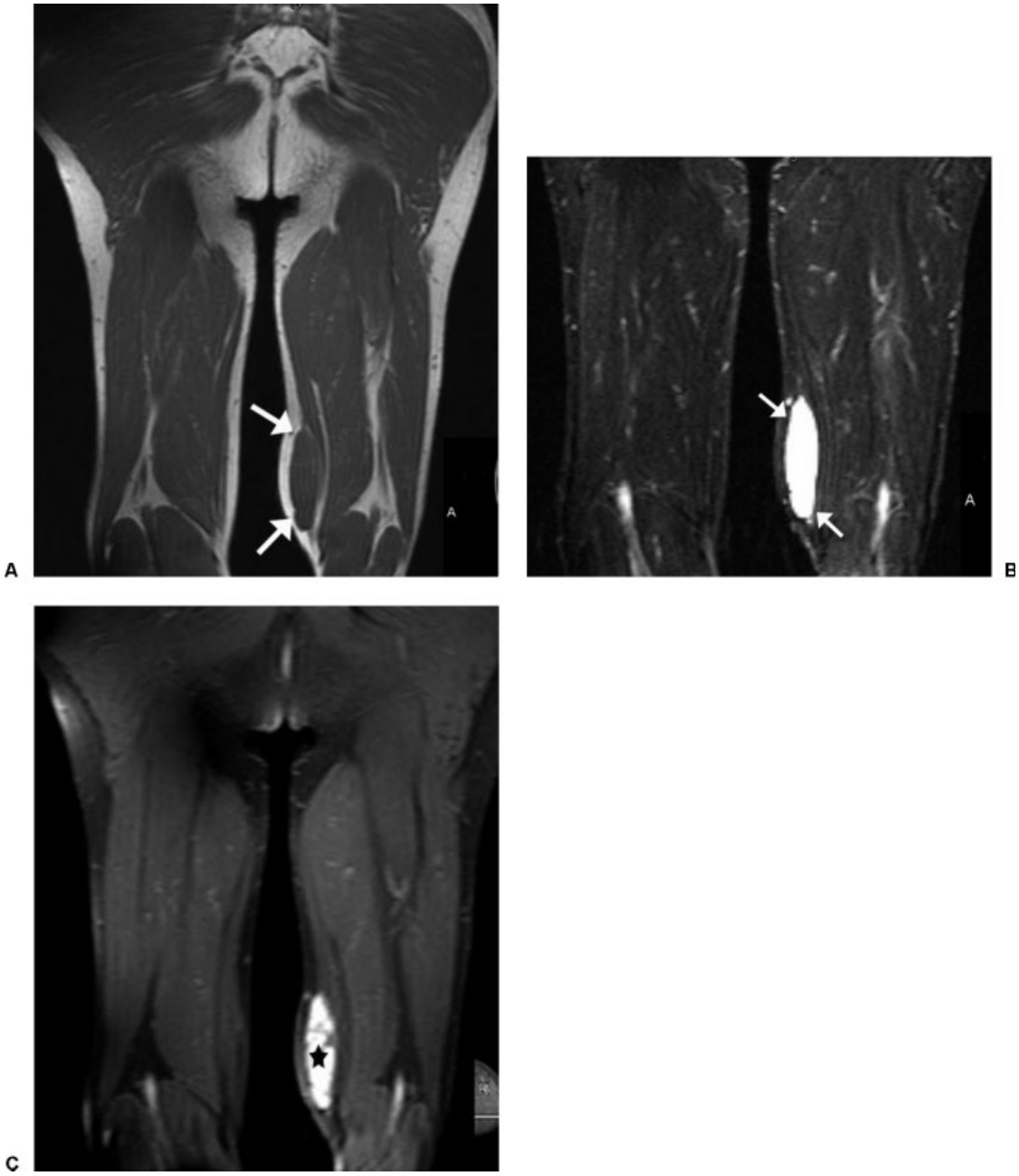

Figure 6 Myxoid liposarcoma simulating a cyst in a 48-year-old woman with a slowly enlarging painless mass in the distal thigh. (A, B) Coronal T1-weighted (A, 480/17) and T2-weighted (B, 4500/100) MR images show a relatively well-defined homogeneous intermuscular mass (arrows) with low to intermediate signal intensity on T1 weighting and homogeneous high-signal intensity on T2 weighting. No fat is apparent. Although characteristics simulate a cyst, the location is markedly atypical, requiring further imaging to exude a myxoid tumor masquerading as a cyst (cyst mimicker) (C) The coronal T1-weighted fat suppressed MR image following contrast (C, 500/20) demonstrates the solid noncystic consistency of the mass with prominent diffuse enhancement (asterisk).

As expected for a high-grade sarcoma, the prognosis of pleomorphic liposarcoma is poor. Multimodality therapy to include aggressive surgical resection, chemotherapy, and radiation therapy is employed in treatment and has improved 5-year survival from 21% reported in 1962 to 63% in 2001.[48,78–80] Similar to other subtypes of liposarcoma, metastases most frequently affect the lung.

Mixed-type liposarcoma represents 5 to 12% of all liposarcomas.[48] As implied by its designation, this lesion represents a combination of the other subtypes. This combination is most frequently of myxoid and

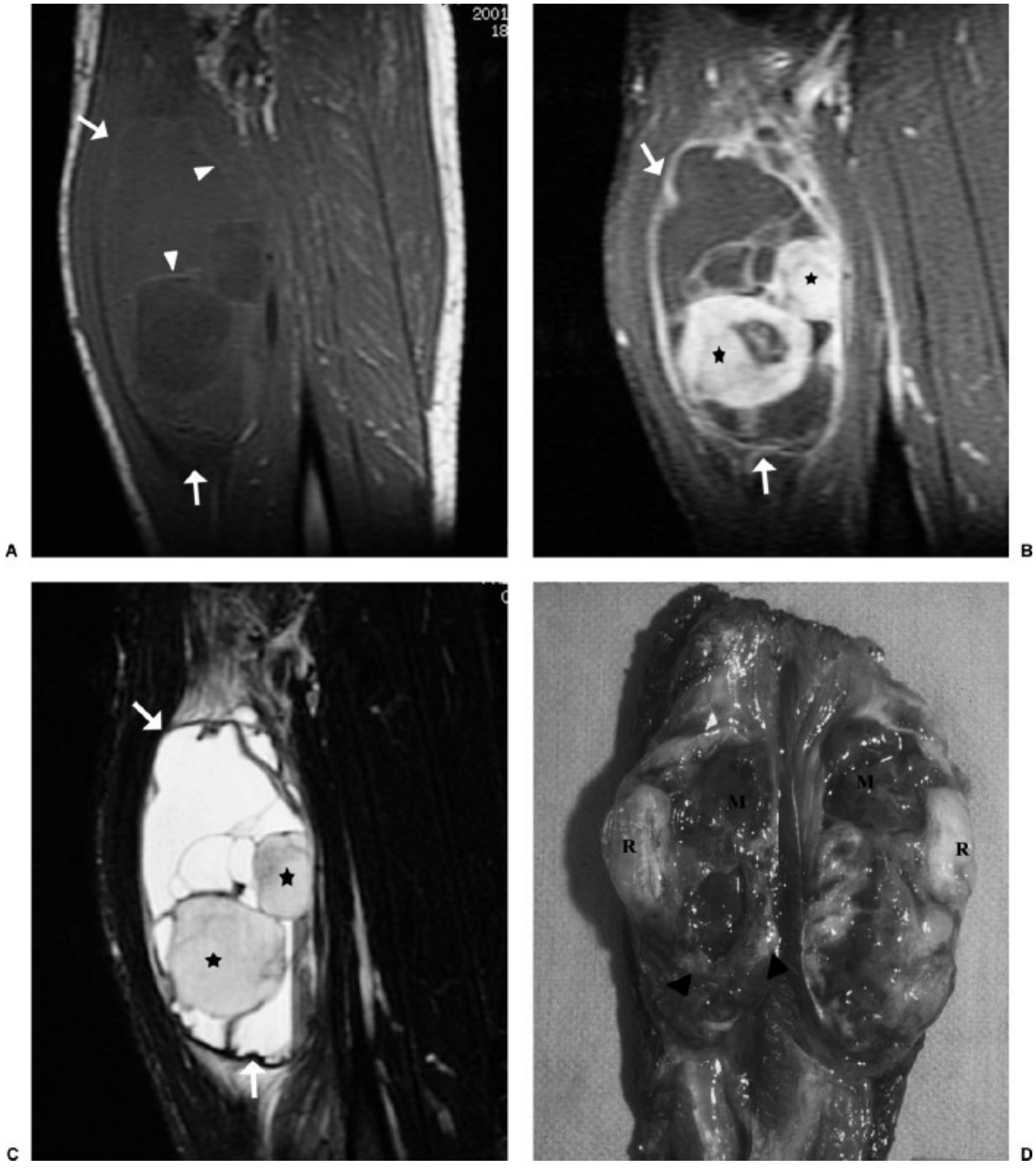

Figure 7 Myxoid liposarcoma with prominent round cell component in the calf of a 40-year-old man with a slowly enlarging mass. (A–C) Sagittal T1-weighted (616/8) MR images before (B) and after (B, 516/8; also with fat suppression) intravenous contrast and T2-weighted (C, 7176/91) fat-suppressed MR image reveal a large soft tissue mass (arrows) to contain a subtle area of adipose tissue isointense to subcutaneous fat (arrowheads). The T2-weighted MR image (B) reveals markedly high signal intensity in the majority of the mass, and in correlation to the subtle fat on T1 weighting the findings are those of a myxoid liposarcoma. However, there are several prominent nodules that are diffusely enhancing and only intermediate signal intensity on the long TR image (asterisks) representing the round cell component. (D) Long axis sectioned gross specimen demonstrates similar features of a heterogeneous multinodular partially myxoid mass (M) with a prominent round cell component (R) and subtle adipose tissue (arrowheads).

well-differentiated subtypes of liposarcoma or myxoid and pleomorphic. In my experience, imaging may give a more accurate representation of the overall morphology, with distinct separate areas of each subtype, as opposed to histological evaluation of biopsy material from small areas of these often large lesions. The intrinsic imaging characteristics of these lesions are, as previously described, for the various subtypes of liposarcoma. Mixed-type liposarcoma typically affects older patients. The retroperitoneum and abdominal cavity are the most common locations of these lesions. The mediastinum and deep soft tissue of the extremity are less frequently affected.

CONCLUSIONS

The working group of the WHO for classification of tumors of soft tissue and bone convened in 2002. This meeting resulted in modification of nomenclature for soft tissue neoplasms, particularly in the group of fibrous and lipomatous malignancies. I have reviewed these modifications and the causes for these changes, primarily consisting of suggesting the replacement of the term *MFH* with *undifferentiated high-grade pleomorphic sarcoma* and combining myxoid and round cell liposarcoma. I have also emphasized the radiological appearances of these lesions. In my opinion, it is important for radiologists involved in the evaluation of these lesions to have an understanding of the current nomenclature. This allows more uniformity in discussion with our orthopedic oncological and pathological colleagues to strengthen our team approach in the diagnosis and treatment of these patients.

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DISCLAIMER

The opinions or assertions contained herein are the private views of the author and are not to be construed as official or as reflecting the views of the Departments of the Army, Navy, or Defense.

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