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Pictorial Review

# Imaging features of myxoid soft-tissue tumours

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Myxoid soft-tissue tumours are mesenchymal neoplasms, which are characterised by the production of abundant extracellular myxoid matrix. Imaging plays an important role in the diagnosis of these tumours as well as treatment planning. The imaging features as well as the clinical course for these lesions are highly variable, depending on both the anatomical location of the tumour and the histopathological subtype. This article, illustrated by histopathologically proven cases from our tertiary referral soft-tissue sarcoma centre, reviews the spectrum of imaging findings and characteristic signs seen with different types of benign and malignant myxoid soft-tissue neoplasms.

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## Introduction

Myxoid soft-tissue tumours are mesenchymal neoplasms, which are characterised by abundant extracellular myxoid matrix.[1] This myxoid matrix gives rise to high T2-weighted magnetic resonance imaging (MRI) signal, low computed tomography (CT) attenuation and low echogenicity; these imaging features are found to varying degrees in most myxoid tumours.

This article, illustrated by histopathologically proven cases from our tertiary referral soft-tissue sarcoma centre, reviews the spectrum of imaging findings and characteristic signs that can be seen with different types of benign and malignant myxoid soft-tissue neoplasms. This article follows conventional classification from the 2020 5th edition World Health Organization classification of bone and soft-tissue tumours. Electronic Supplementary Material figures are available in the online version of this review.

Although imaging features of different myxoid tumours overlap and are rarely characteristic, imaging plays an important role in the diagnosis of these tumours as well as treatment planning. Accurate anatomical information is vital for operative planning and differentiation of benign from aggressive lesions can help with risk stratification and to put biopsy results into clinical context.

All suspected myxoid soft-tissue tumours should be referred to a tertiary soft-tissue sarcoma centre where percutaneous imaged-guided biopsy for a definitive histological diagnosis. Biopsy of lesions with a high myxoid content can be challenging due to their gelatinous consistency.

## Benign lesions

### *Intramuscular myxoma*

Intramuscular myxoma is the most common myxoid soft-tissue tumour. These benign tumours consist of abundant myxoid stroma interspersed with cytologically bland spindle cells.[1] Intramuscular myxomas have a peak incidence between ages 40 to 70 years, with a roughly 2:1 female predilection. The large muscles of the thigh, shoulder girdle and

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buttock, in that order, are the most common sites. They present as slowly growing masses, which at the time of diagnosis are usually 5–15 cm in diameter.[2] Rarely (<15%), soft-tissue myxomas are intermuscular or subcutaneous.[3]

There is a recognised associated of intramuscular myxomas with polyostotic fibrous dysplasia, this is known as Mazabraud syndrome, the myxomas tend to occur near the fibrous dysplasia (Electronic Supplementary Material Fig. S1). The reported prevalence of Mazabraud syndrome in patients with fibrous dysplasia is 2% and the age at diagnosis is typically 30–40 years, younger than the typical age for presentation of myxoma in patients without fibrous dysplasia.[4,5]

The MRI appearances of myxomas although not pathognomonic are often typical. Intramuscular myxomas demonstrate fluid-like high signal on T2-weighted MRI images (Fig 1) with homogeneous low to intermediate signal on T1-weighted images (Fig 2; Electronic Supplementary Material Fig. S2). Contrast enhancement is almost always seen, usually mild to moderate and can be either diffuse or in a peripheral and septal pattern (Electronic Supplementary Material Fig. S3). Cystic spaces can be seen in approximately half of intramuscular myxomas. Fluid-sensitive sequences demonstrate high signal in the adjacent muscles in most cases, a finding caused by leak of myxomatous material due to the myxomas not being truly encapsulated. In 65–89% of cases, a thin rim of fat can be seen (Electronic Supplementary Material Fig. S3), most prominent at the superior and inferior poles of the lesion, represents fatty atrophy of adjacent muscle. The fatty pole can be seen with T1-weighted MRI or ultrasound and is often referred to as a "fat cap" (Fig 2).[6]

Ultrasound imaging demonstrates a well-defined hypoechoic to near anechoic mass with hyperechoic components in a "whorled" appearance with posterior acoustic enhancement (Fig 3). Myxomas typically demonstrate little to no internal vascularity, the presence of vascularity would be suspicious for a more aggressive lesion such as a myxofibrosarcoma (Figs 17 and 18).[2,7]

CT shows a well-defined intramuscular mass with attenuation between muscle and water with mild enhancement.[7]

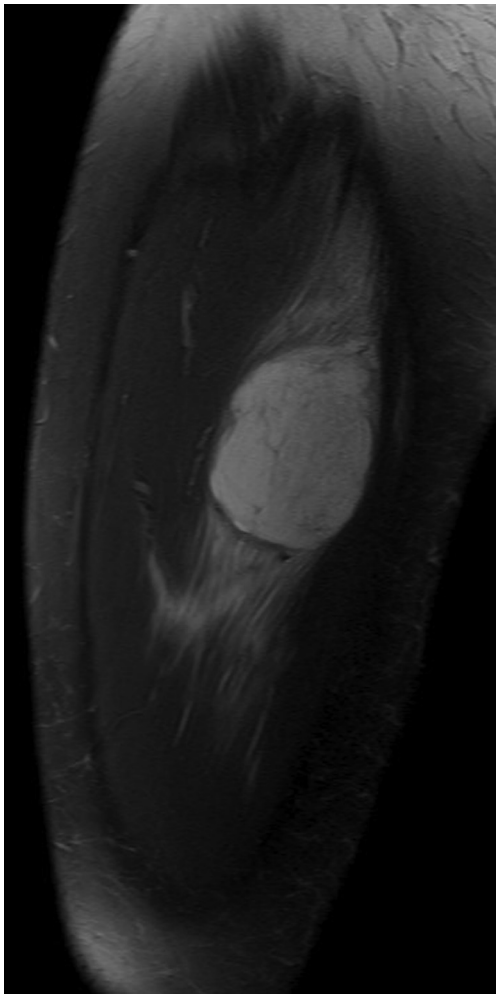

**Figure 1** Proton-density fat-saturated coronal MRI of an intramuscular myxoma in the right vastus lateralis muscle. Homogeneous high signal with high signal spreading superiorly and inferiorly within the muscle and bounded by the fascia in keeping with intramuscular leak of myxoid contents.

### *Deep (aggressive) angiomyxoma*

Deep angiomyxoma, also known as aggressive angiomyxoma, is a very rare tumour, which arises from the perineum or lower pelvis in women of childbearing age in over 90% of cases. These tumours are generally positive for oestrogen and progesterone receptors. Although benign, deep angiomyxoma has a high rate of local recurrence even with complete resection. Presentation is often non-specific and initial clinical impressions mimic that of a Bartholin gland cyst.[8]

The tumour is often large time of diagnosis, extending into the pelvis from the perineum. Deep angiomyxoma tends to grow around and displace rather than invade the pelvic viscera.[9] It appears markedly hypoechoic on ultrasound and hypoattenuating on CT due to its high myxoid content. It can be difficult to appreciate the deep extent of the tumour on ultrasound and given its propensity to arise superficially from the labia and perineum (Fig 4). On T2-weight MRI images, the tumour is usually high signal with "swirled" or "layered" bands of low T2 signal, which correspond to strands of fibroblastic tissue (Fig 5). A similar swirled or layered appearance is also seen on contrast-enhanced CT and MRI (Fig 6). The characteristic appearance and the anatomical relation of the perineal lesion extending into the pelvis is usually best appreciated on a sagittal T2-weighted MRI image.[10,11]

### *Juxta-articular myxoma*

Juxta-articular myxoma (JAM) is a rare variant of the intramuscular myxoma; it is similarly benign and shares

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**Figure 2** Coronal T1-weigthed MRI of an intramuscular myxoma in the right vastus lateralis muscle. Homogeneous T1-weighted signal with a prominent inferior triangle of high signal represents a "fat cap".

very similar imaging findings with the exception of its periarticular location. Over 80% of juxta-articular myxomas occur around the knee.[12] Juxta-articular myxomas, like intramuscular myxomas, demonstrate low echogenicity on ultrasound (Fig 7), low attenuation on CT and high T2-weighted MRI signal with variable contrast enhancement (Figs 8 and 9; Electronic Supplementary Material Fig. S4). Given the periarticular nature of JAM and its cyst-like appearance on imaging, it can be mistaken for a ganglion or synovial cyst, particularly if contrast-enhanced imaging is not performed.[13]

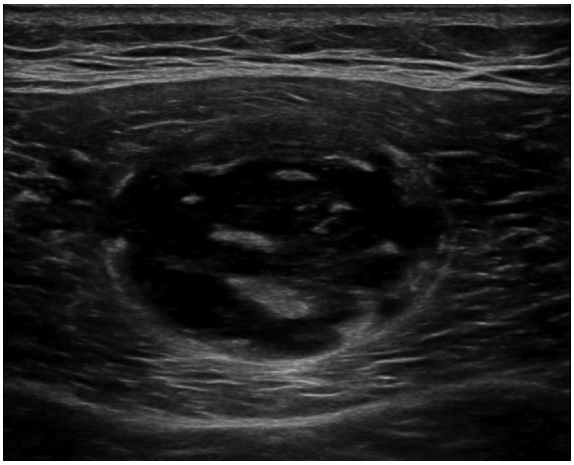

**Figure 3** Ultrasound of an intramuscular myxoma in the anterolateral right thigh. Hypoechoic lesion with swirled isoechoic contents, posterior acoustic enhancement and a hyperechoic fatty rim. Appearances are typical for intramuscular myxoma.

## Malignant lesions

### *Myxoinflammatory fibroblastic sarcoma*

Myxoinflammatory fibroblastic sarcoma is a rare low-grade sarcoma characterised by a superficial mass in the extremities, usually involving the dermis and subcutis in young to middle-aged adults. It shows four elements on histopathological analysis: proliferative fibroblasts (spindle cells), myxoid matrix, associated inflammatory components, and Reed–Sternberg-like atypical giant cells.[14] Tumours may be well-defined or ill-defined and infiltrative, they are most commonly located along tendon

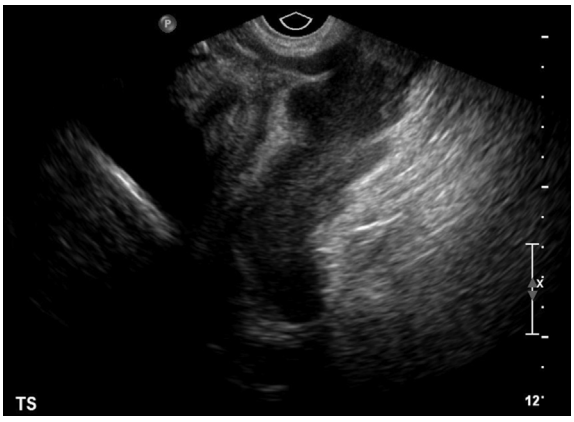


**Figure 4** Transvaginal ultrasound, transverse image taken with the probe superficially in vagina showing a deep angiomyxoma arising from the left labia and extending deep as well-defined hypoechoic lesion. This was fluctuant with dynamic compression and initially thought to represent a Bartholin gland cyst.

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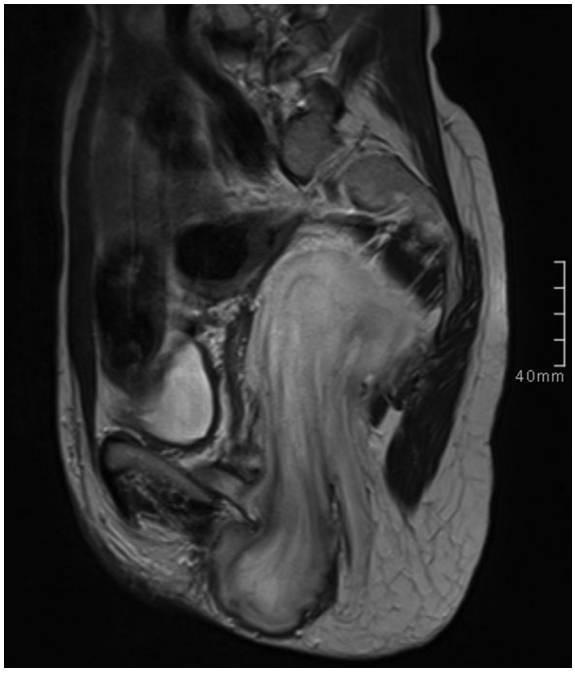


**Figure 5** MRI image of a pelvic deep angiomyxoma. Sagittal T2-weighted images how high signal mass arises from the perineum and extends deep and superiorly into the pelvis. The location and layered internal appearance to the lesion within bands of high and low signal are typical for a deep angiomyxoma.

sheaths. The imaging appearances of myxoinflammatory fibroblastic sarcoma is variable and dependant on the degree of infiltration. Ultrasound typically demonstrates a poorly marginated, very heterogeneous lesion with marked interval vascularity (Fig 10). High T2-weighted MRI signal typical of myxoid is normally seen but lesions can be variably homogeneous or heterogeneous on both

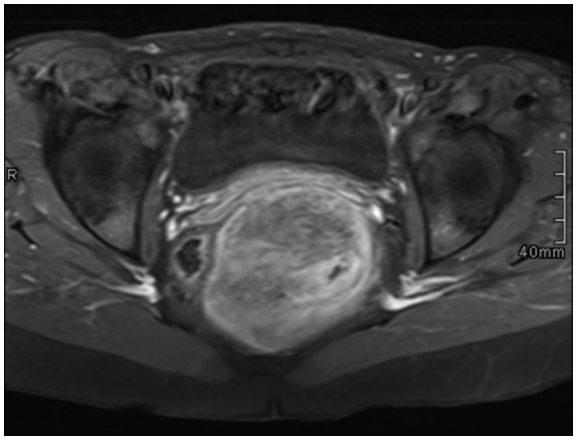


**Figure 6** MRI image of a pelvic deep angiomyxoma. Axial post-contrast fat-saturated T1-weighted images demonstrates avid heterogeneous enhancement. The mass is displacing the rectum to the right and the vagina anteriorly. There is no invasion of the rectum or vagina; the fat planes between the tumour and the adjacent structures are preserved.

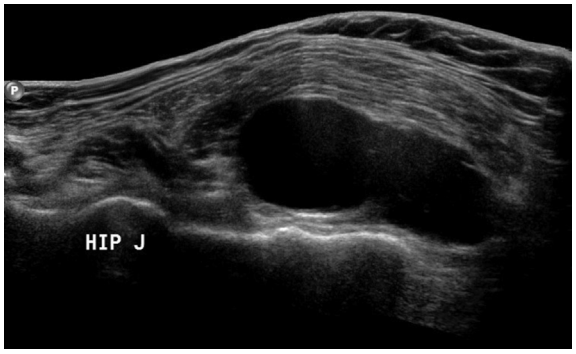


**Figure 7** Coronal panoramic ultrasound of a juxta-articular myxoma over the left hip just superficial to the proximal femur. The left gluteal muscles are displaced superficially by the lesion. This juxta-articular myxoma is well-defined with a bland hypoechoic internal structure.

T2-weighted and post-contrast MRI sequences (Fig 11; Electronic Supplementary Material Fig. S5). Due to the inflammatory nature of these lesions, surrounding oedema is seen on fluid sensitive MRI sequences. Comparison of post-contrast T1-weighted images with T2-weighted images can help to delineate tumour infiltration from oedema.[15,16,17] Demarcating the edge of the lesion and complete surgical excision can remain challenging, and it is therefore not surprising that the rate of local recurrence is between 28–51%. Distant metastases have been reported in approximately 1% of cases.[18]

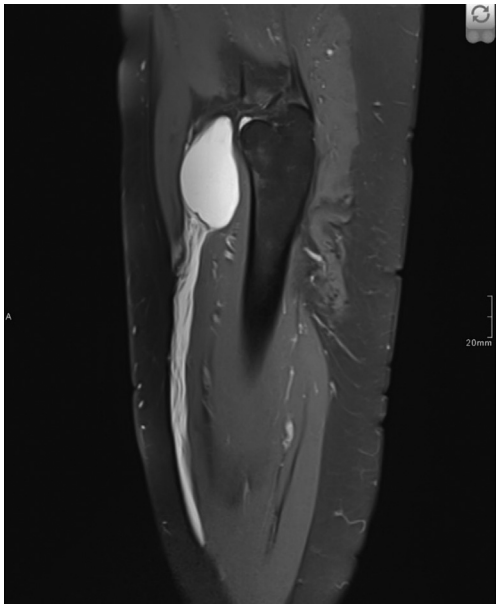


**Figure 8** Sagittal fat-saturated T2-weighted MRI of the left thigh showing the same juxta-articular myxoma as seen in Fig 7, which is well-defined with very high signal and thin low signal septa. There is high signal seen extending inferiorly along the fascial plane of the anterior compartment of the left thigh, which could be either oedema or leak of myxoid material. On this image, this lesion could equally be a large synovial cyst.

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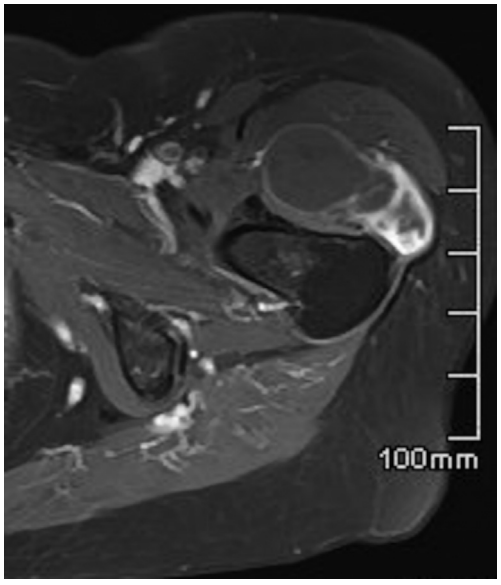


**Figure 9** Axial post-contrast fat-saturated T1-weighted MRI of the left hip showing the same juxta-articular myxoma as Figs 7 and 8. There is thin peripheral enhancement with thicker septal and lateral margin enhancement. This would be an unusual appearance for a synovial cyst, and therefore, this lesion therefore went on to percutaneous ultrasound-guided biopsy for a tissue diagnosis prior to surgical excision. See Electronic Supplementary Material Fig. S4 for corresponding pre-contrast fat-saturated image.

### *Myxoid liposarcoma*

Myxoid liposarcoma is a genetically distinct subtype of liposarcoma, characterised by a variable content of round cells. The higher the proportion of round cells, the more aggressive the lesion. It presents as a slow growing intramuscular mass in the lower limb in 75–80% of cases at a mean age of 48 years, a decade earlier than other variants of liposarcoma. Myxoid liposarcoma has a distinct pattern of non-pulmonary metastatic disease; frequently to the paraspinal soft tissues, retroperitoneum, and osseous metastases to the spine.[19]

MRI best demonstrates myxoliposarcoma, usually a multilobulate and well-defined lesion containing different constituent components: fat, myxoid and round-cell tissue. The fatty component of myxoid liposarcoma is typically <10% of total tumour volume and is seen as high T1 signal or low attenuation on CT. Although the proportion of fat is usually small it is seen on MRI in 90–95% of cases. Careful comparison of the T1-weighted images with fat-saturated T1-weighted images can demonstrate fat within internal septa of the lesion (Fig 12). A variable proportion of myxoid content with its high T2-weighted MRI signal and non-fatty, non-myxoid enhancing tissue representing round-cell tissue can be seen (Fig 13). Higher proportions of non-fatty, non-myxoid enhancing tissue are associated with poorer prognoses and more aggressive tumours.

On ultrasound lesions are vascular, heterogeneous with larger regions of fat appearing as low echogenicity foci (Fig 14). Whole-body MRI with diffusion-weighted imaging is a useful technique to evaluate for distant soft-tissue

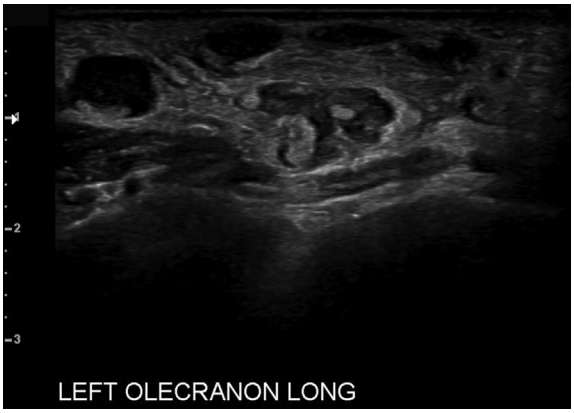


**Figure 10** Sagittal ultrasound image of an myxoinflammatory fibroblastic sarcoma posterior to the left olecranon showing a poorly defined, heterogeneous lesion in the subcutaneous tissues.

metastases and has become the modality of choice for evaluation of metastases.[20,21,22] MRI is favoured over CT, positron-emission tomography (PET)-CT or bone scintigraphy because approximately 50% of bone metastases are negative on all three of the former modalities but positive on MRI.[23]

### *Myxofibrosarcoma*

Myxofibrosarcoma typically presents as a mass in the extremities (75% of cases) in the sixth decade of life.

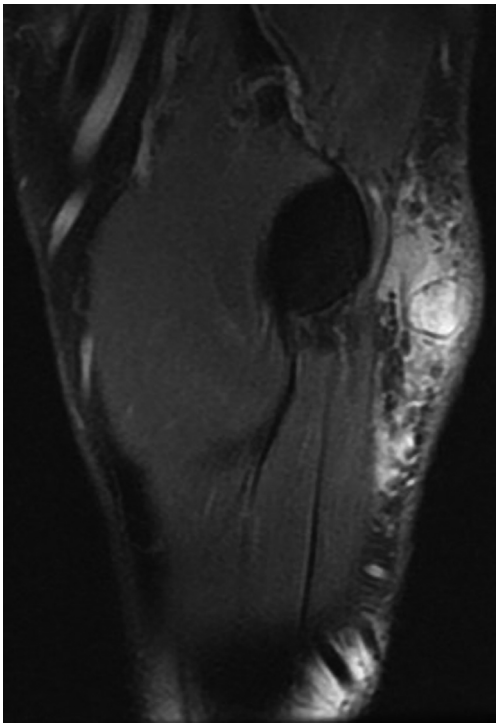

**Figure 11** Sagittal fat-saturated T2-weighted MRI of the left elbow in the same patient as Fig 12. The tumour is ill-defined with heterogeneous high T2 signal and infiltrative spread along the fascial planes. See Electronic Supplementary Material Fig. S5 for axial images.

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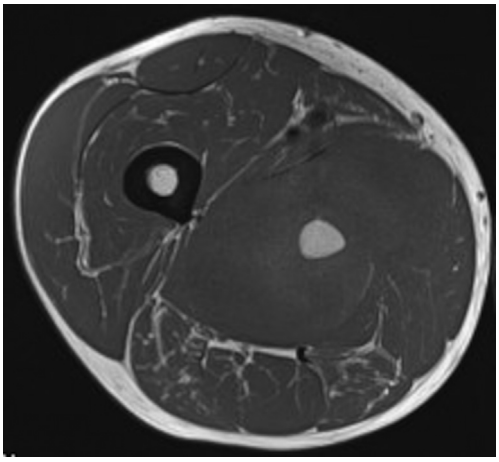

**Figure 12** Axial T1-weighted MRI of the right thigh with a myxoliposarcoma deep in the posterior compartment of the thigh. The lesion is difficult to differentiate from the adjacent muscles as the bulk of the lesion has isointense signal to muscle. There is a round focal area of high T1 signal fat centrally in the lesion.

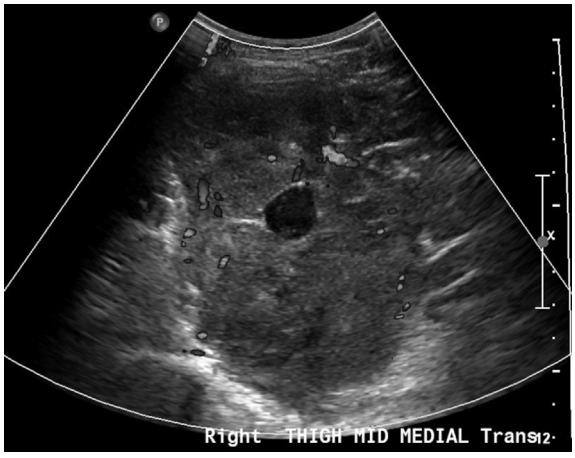


**Figure 14** Axial ultrasound image with colour Doppler of the same myxoliposarcoma from the right thigh. The lesion is moderately well defined with mildly heterogeneous echotexture. The lesion is mostly isoechoic to muscle but with a well-defined central focus of hypoechoic fat. There is internal vascularity.

Myxofibrosarcoma tends to have ill-defined margins with infiltrative spread along fascial and vascular planes. These tumours can arise from the subcutaneous tissue, dermis, and intermuscular or intramuscular planes.[24] The prognosis is relatively poor; local recurrence rates are 50–70% due to difficultly in achieving complete local resection with, low-grade lesions trend to recure as high-grade disease with a greater predilection for metastasis. The five-year survival is 60–70%.[25]

These lesions appear heterogeneous on T1-and T2-weighted MRI images with heterogeneous contrast enhancement (Fig 15; Electronic Supplementary Material Fig. S6). Areas of intralesional haemorrhage can mimic the fat of a myxoliposarcoma but haemorrhage will retain its high signal on fat-suppression. Marked perilesional oedema is common and similar to myxoinflammatory fibroblastic sarcoma, comparison of post-contrast and T2-weighted imaging can help delineate tumour from oedema and aid the surgeon in planning the extent of resection required. An enhancing "tail" of tissue extending from the bulk of the lesion along fascial planes is a characteristic sign seen with myxofibrosarcoma. It is this spread along the fascia which is thought to be the reason for such high rates of local recurrence (>50%; Fig 16).

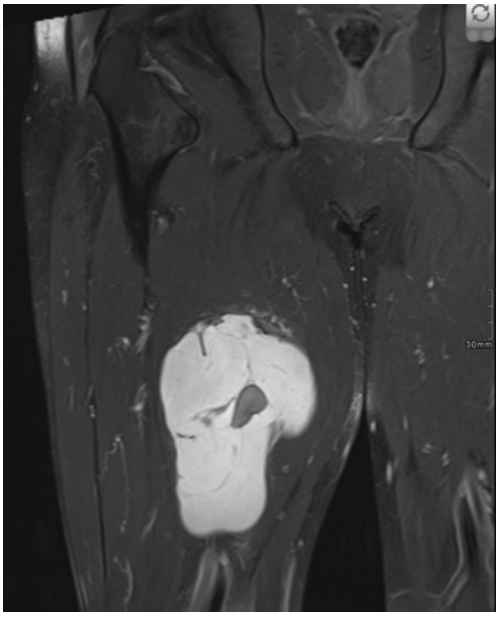

**Figure 13** Coronal fat-saturated T2-weighted MRI image of the same myxoliposarcoma in the right thigh. The myxoliposarcoma is well defined, demonstrating high signal with internal septa. The signal from the central focal area of fat has been nulled by the fat saturation.

On ultrasound myxofibrosarcoma is usually heterogeneous with marked internal vascularity. The infiltrative nature of these tumours can be difficult to appreciate with ultrasound and when myxofibrosarcoma is towards the low-grade end of its spectrum it can look similar to an intramuscular myxoma (Fig 17).[26] The imaging appearances can also overlap with myxoliposarcoma, extraskeletal myxoid chondrosarcoma and myxoinflammatory fibroblastic sarcoma.

### *Low-grade fibromyxoid sarcoma*

Low-grade fibromyxoid sarcoma (LGFMS) and myxofibrosarcoma are distinct entities with different histopathological characteristics despite their similar names. LGFMS has a relatively indolent clinical course but approximately 1% of case will present with metastatic disease, most commonly pulmonary metastases.

LGFMS consists of collagen and myxoid components in variable proportions. The collagen or fibrous components will have low signal on all MRI sequences and the myxoid component will have typical high T2 signal. Post-contrast enhancement is variable. In contrast to the

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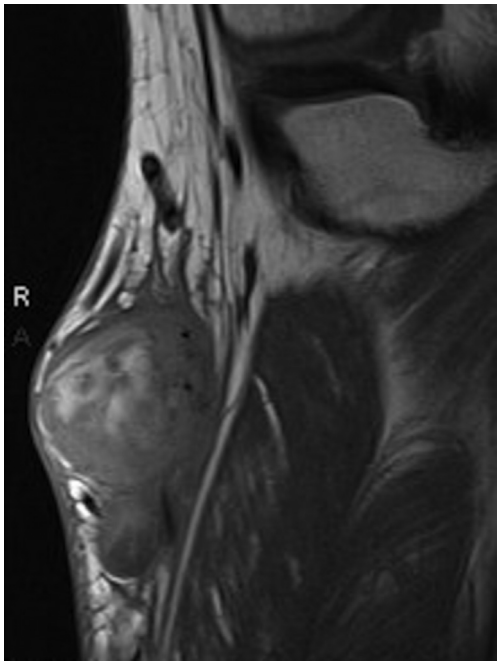


**Figure 15** Coronal pre-contrast T1-weighted MRI of a myxofibrosarcoma in the medial aspect of the left. The tumour has heterogeneous T1 signal with foci of internal high signal in keeping with haemorrhage. See Electronic Supplementary Material Fig. S6 contrast-enhanced imaging where non-haemorrhagic components of the tumour show avid enhancement.

myxofibrosarcoma, LGFMS are less heterogeneous, with little surrounding oedema and no fascial extension.

In most cases, LGFMS appears similar to many of the other myxoid soft tissue lesions but where the portion of fibrous tissue is high, with predominantly low T2 signal, the differential diagnosis would include fibrosarcoma (Fig 18; Electronic Supplementary Material Fig. S7).[27,28,29]

*Extraskeletal myxoid chondrosarcoma*

Extraskeletal myxoid chondrosarcoma is an intermediate to high-grade tumour characterised by malignant chondroblasts in a myxoid matrix. Extraskeletal myxoid chondrosarcoma is most common in the lower extremities/hip region (63% of lesions) in older males (63% male predominance).[30,31]

Imaging features are non-specific. Extraskeletal myxoid chondrosarcoma are usually well-defined lobulate lesions in the deep tissue. Heterogeneous signal is shown on both T1-and T2-weighted images as well as post-contrast images due to mixed haemorrhage, cystic areas and necrosis being common (Electronic Supplementary Material Fig. S8). The presence of an ossified internal chondroid matrix is rarely reported.[32,33]

*Ossifying fibromyxoid tumour*

Ossifying fibromyxoid tumour (OFT) is a rare intermediate grade tumour which most frequently affects middle

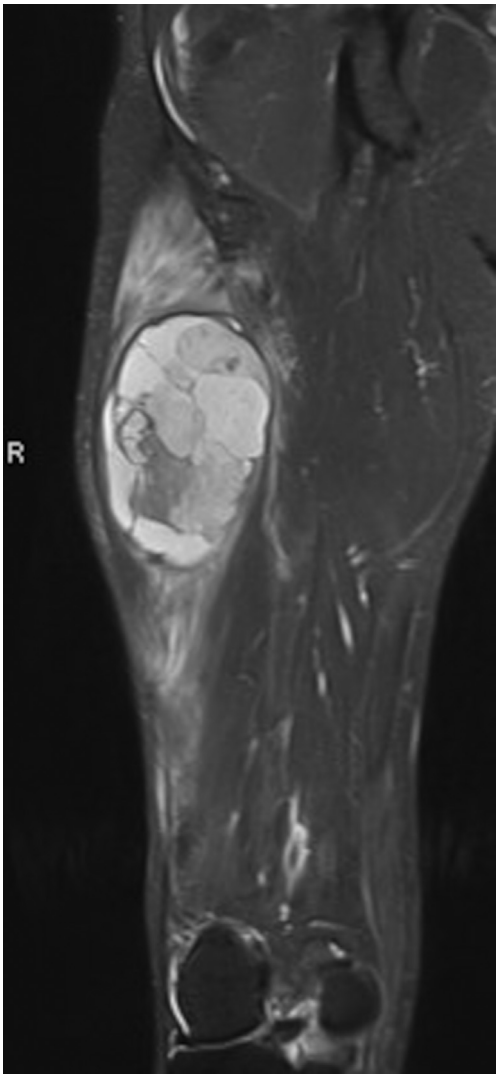


**Figure 16** Coronal fat-saturated T2-weighted MRI of another myxofibrosarcoma in the right thigh demonstrating high signal with internal septa and intramuscular oedema spreading inferior and superior to the tumour. This tumour has imaging features which are similar to the intramuscular myxoma in Fig 1. This example shows the overlap in imaging features between a benign myxoma and the malignant myxofibrosarcoma. Some clues to this lesion being more aggressive than a benign myxoma are the presence of thick internal septa and a solid enhancing soft-tissue component (not shown in this image).

aged adults.[34] Most OFT display benign histopathological features and a benign clinic course with complete local excision proving curative.[35] Ossification with an incomplete shell of metaplastic lamellar bone is seen in 70% of lesions.[36] This peripheral rim of ossification can be seen on plain radiograph but is best demonstrated on CT.[37] Thirty per cent of lesions are non-ossifying.

Areas of ossification appear as low signal intensity on all MRI sequences. The internal appearance of OFT is non-specific with mixed high and low T2 signal. OFT that are more extensively ossified can show high T1-weight fatty marrow signal centrally in ossified components and

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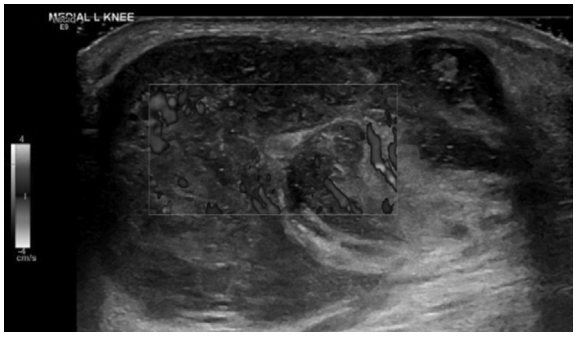


**Figure 17** Axial ultrasound image of the medial aspect of a left knee with a myxofibrosarcoma. The tumour appears well defined, moderately heterogeneous, and hypoechoic. There is marked internal vascularity.

osteoblastic activity on bone scintigraphy.[38] The ultrasound appearances are non-specific, usually demonstrating a well-circumscribed low-echogenicity mass, occasionally with a very bright rim that shadows posteriorly if there is significant marginal calcification (Fig 19). Differential diagnoses on imaging would include ossifying haematoma, myositis ossificans, periosteal osteosarcoma, extraskeletal chondrosarcoma, and other ossifying soft-tissue sarcomas or metastases.

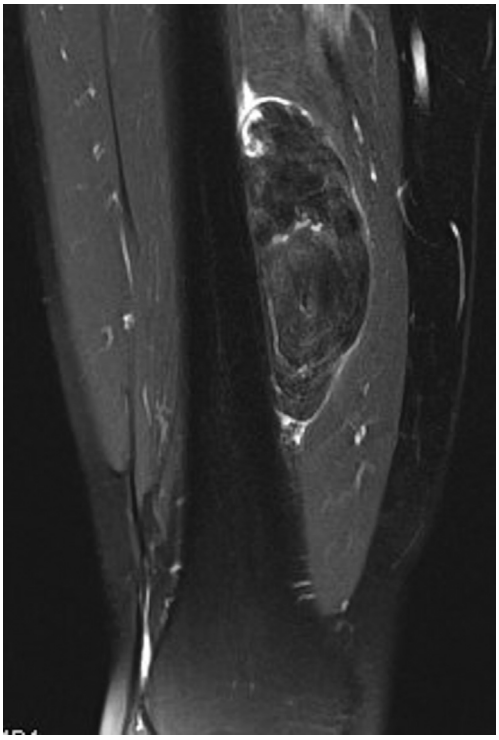

**Figure 18** Coronal fat-saturated T2-weighted MRI of the right thigh of a low-grade fibromyxoid sarcoma. The lesion demonstrates heterogeneous low T2-weighted signal with sparse foci of high signal. The predominantly low signal on all sequences is typical of a fibrous composition with the high signal foci represents a small myxoid component. These appearances are typical but not pathognomonic of a low-grade fibromyxoid sarcoma. See Electronic Supplementary Material Fig. S7 for the corresponding T1 image.

**Figure 19** Colour Doppler ultrasound image of OFT in the right deltoid muscle. The 2 cm lesion is well defined with mixed but predominantly low echogenicity. Low-grade internal Doppler flow is present, which led to management with excision biopsy. This OFT does not demonstrate peripheral high echogenicity with posterior shadowing which would be expected in cases with dense peripheral calcification.

## Conclusion

Soft-tissue myxoid tumours are an uncommon, heterogeneous group that share a common myxoid extracellular matrix which gives rise to high T2-weighted MRI signal, low CT attenuation and low echogenicity; these imaging features are found to varying degrees in most myxoid tumours.

Although imaging features of different myxoid tumours overlap and are rarely characteristic, there are certain discriminating features. Myxomas are usually well defined and intramuscular with a whorled internal appearance and may show a fatty cap. Deep angiomyxoma affects the perineum in women of childbearing age, presenting as large mass extending into the pelvis. Juxta-articular myxoma are seen in close proximity to large joints and can mimic synovial cysts. Myxoinflammatory fibroblastic sarcoma presents in young adults affecting the extremities in an infiltrative pattern. Myxofibrosarcoma also presents as an infiltrative mass in the extremities but typically in older adults. Myxoid liposarcoma is the most common malignant myxoid tumour and internal fat signal is seen on MRI. The presence of incomplete peripheral ossification suggests ossifying fibromyxoid and a calcified internal chondroid matrix suggests extraskeletal myxoid chondrosarcoma.

Ultimately, in clinical practice, suspected myxoid tumours will undergo percutaneous imaged guided biopsy in a tertiary soft-tissue sarcoma centre for a definitive histological diagnosis. Nevertheless, the radiologist plays an important role in providing accurate anatomical descriptions and in assessing whether lesions are likely benign or aggressive. Biopsy of myxoid lesions tends to yield poor tissue cores if the myxoid content of the lesion is high due to its gelatinous consistency.

## Conflict of interest

The authors declare no conflict of interest.

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## Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.crad.2023.05.005.

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