Characterizing fibrous soft-tissue tumors: Are we misdiagnosing a lot of these?

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Learning objectives

This educational exhibit aims to:

1. Review clinical features along with pathological and MR imaging characteristics of various benign, intermediate-grade and malignant fibrous soft-tissue tumors.
2. Illustrate the role and limitations of MRI in differentiating them from other similar appearing soft tissue lesions.
3. Update the reader with current modifications in WHO nomenclature for the group of fibrous malignancies in order to facilitate more uniformity in discussion with our oncology and pathology colleagues.

Background

Benign versus malignant

The issue whether MRI can differentiate benign soft tissue tumors from malignant ones remains controversial. It is generally considered that -

1. Malignant lesions are more heterogeneous on T2-weighted MR images due to internal hemorrhage and necrosis.
2. There is high specificity for malignancy when the tumor shows necrosis, involves bone and neurovascular structures and has a mean diameter of more than 66 mm as per one study.
3. The pseudocapsule of a malignant sarcoma can mimic well encapsulated appearance of a benign neoplasm.

The MRI findings of majority of soft tissue lesions remain non-specific with correct diagnosis reached in only 25% to 35% of cases at best. However, knowledge of tumor prevalence, presentation, patient age and lesion location along with MR characterization can significantly improve formulation of an educated differential diagnosis.

Introduction to fibrous soft tissue tumors

Fibrous soft tissue tumors which include sub categories of fibroblastic, myofibroblastic and fibrohistiocytic tumors are often confused with other soft tissue lesions due to their varied biological behavior and imaging appearances.
In 2002, WHO revised categorization of soft tissue tumors on the basis of their biological behaviour, and also revisited certain poorly defined terminologies.

Under this revised classification, two distinct varieties of intermediate grade neoplasms were identified and termed as 'locally aggressive' and 'rarely metastasizing' respectively. Notably amongst fibrous tumors, superficial and deep fibromatosis (desmoid tumors) fall under the former category while solitary fibrous tumor, inflammatory myofibroblastic tumor and plexiform fibrohistiocytic tumor fall under the latter category.

Revised terminology now describes myositis ossificans as fibroblastic and myofibroblastic lesion instead of a chondro-osseous lesion. Malignant fibrous histiocytoma has been replaced with undifferentiated pleomorphic sarcoma as the descriptor for tumors without a clear line of differentiation.

**Findings and procedure details**

We intend to review the clinical features along with pathological and MR imaging findings of commonly and not so commonly encountered fibrous soft tissue tumors in clinical practice.

**Benign Fibrous Tumours**

1. **Nodular fasciitis**

Nodular fasciitis is often confused with soft tissue sarcomas due to its rapid growth and variable MR appearance. It can occur superficially in the subcutaneous plane, along the fascia or located deep within the muscles. Histologically, nodular fasciitis may be myxoid, cellular or fibrous accounting for the variable T2 characteristics on MRI. The lesions with high cellular content or myxoid degeneration appear hyperintense on T2-weighted images (Fig. 1 on page 8) while those with highly collagenous contents are more mature and hypointense. These lesions show variable enhancement on post contrast images.

2. **Fibroma of the tendon sheath**
Fibroma of the tendon sheath is a rare slow growing fibrous tumor that most commonly occurs in the upper extremity of young men. Histologically, it represents the end stage of giant cell tumor of tendon sheath (GCTTS) and is hence found in close relation to a tendon. Due to collagenous and acellular nature, they are usually of low signal intensity on all MR sequences (Fig. 2 on page 9). T2 hyperintense signal intensity seen in several reported cases has been attributed to myxoid degeneration and increased cellularity. They can be differentiated from GCTTS by lack of blooming on gradient images.

3. Giant cell tumor of tendon sheath (GCTTS)

GCTTS, which histologically is composed of fibrous, inflammatory elements and hemosiderin, is more common than fibroma of the tendon sheath by a ratio of 2.7:1. Fibroma of the tendon sheath is less common in the lower extremities and hence a lower extremity lesion is more likely to be a GCTTS rather than the former. GCTTS has variable signal intensity on T1 and T2-weighted MR images due to above mentioned histological composition (Fig. 3 on page 9). They show variable, heterogeneous enhancement. Pressure erosion of underlying bone, cystic changes or calcifications are extremely uncommon.

4. Desmoplastic Fibroblastoma (Collagenous fibroma)

Desmoplastic fibroblastoma is commonly seen in middle-aged and elderly men. They are usually located in the subcutaneous plane although fascial and muscular extension is common. This nature of involvement and MR signal intensity (Fig. 4 on page 11) is often confused with a desmoid tumor, which is an intermediate grade locally aggressive recurring tumor. However unlike desmoid tumors, desmoplastic fibroblastomas do not show their characteristic low signal band-like morphology and are less infiltrative even when large.

5. Calcifying fibrous tumor

It is a rare, tumor-like lesion histologically characterized by fibrotic proliferation, infiltration of inflammatory cells, and dystrophic or psammomatous calcifications. MR appearance mimics that of other fibrous lesions or pigmented villonodular synovitis (PVNS) when it is found close to a joint. The lesion is usually well-circumscribed and appears hypointense on both T1 and T2-weighted images due to predominance of hyalinized collagenous tissue (Fig. 5 on page 12). It lacks hemosiderin which helps in differentiating it from PVNS. Post-contrast images may reveal mild to moderate enhancement. Plain films or CT scan usually demonstrate calcification.
6. Myositis ossificans

Heterotopic ossificans refers to the ectopic formation of bone, commonly after an inciting trauma or neurological event (particularly stroke). MR appearance changes with the age of the lesion. In early stages, the lesion shows heterogeneous high T2 signal with an indistinct low signal-intensity rim representing calcification. After several weeks to months of progressive maturation, the lesion reveals high T1 and T2 signal intensity which is in keeping with ossification (Fig. 6 on page 10). Late lesions typically do not enhance, or may enhance minimally. In the more mature lesions, presence of high T1 and T2 signal with suppression of marrow fat on fat saturated images mimics a lipomatous lesion. Conventional radiography and computed tomography are useful in the evaluation of this osseous nature of the more mature lesions.

Intermediate grade "locally aggressive" Tumors

1. Superficial fibromatosis

They are group of fibroblastic proliferations that arise from fascia (Fig. 7 on page 14) or aponeuroses at the palmar, plantar, penile (Peyronie disease) and knuckle pad locations. Plantar fibromatosis (Ledderhose disease) usually effects the non-weight bearing medial portion of the plantar aponeurosis (Fig. 8 on page 15). Linear extension of the lesion along the aponeurosis (fascial tail sign) is frequent and best seen after contrast administration. Known association exists with diabetes, epilepsy and alcohol induced liver disease.

2. Desmoid-type Fibromatosis (extra-abdominal fibromatosis)

Desmoid tumors most frequently present as a firm soft tissue mass in young females. Early stage lesions are often cellular and appear heterogeneously hyperintense on T2-weighted MR images in the background of low signal collagenous areas. In particular, presence of low signal band-like morphology is helpful in making the diagnosis (Fig. 9 on page 16) while other features such as poor margination, heterogeneous signal and neurovascular or skeletal involvement are often confused with soft tissue malignancies. Moderate to marked heterogeneous enhancement is usually noted after contrast administration. Recurrence is seen in approximately 50 % cases.

Intermediate grade "rarely metastasizing" Tumors
1. Extrapleural solitary fibrous tumor

Histologically confused with hemangiopericytomas, these usually present as a slow growing mass in a middle-aged adult. Large tumors may also cause paraneoplastic hypoglycemia. On MRI, these are of intermediate signal intensity on T1W and of high signal intensity on T2W images. This non specific MR signal intensity in the presence of prominent perilesional vessels and marked post contrast enhancement is more specific (Fig. 10 on page 17). A known fat containing variant can be confused with a liposarcoma on MRI.

2. Plexiform Fibrohistiocytic tumor

These present as poorly demarcated, slow growing, superficial masses in the extremities of adolescent and young adults. MRI appearance is described in Fig. 11 on page 18. Multinucleated giant cells are present on histology; the fibroblastic subtype resembles fibromatosis. It rarely presents with systemic metastases or oncologic osteomalacia as in our case.

3. Inflammatory myofibroblastic tumor (pseudotumor)

Histologically they are composed of cells associated with both acute and chronic inflammation including lymphocytes and plasma cells, myofibroblastic spindle cells and collagen (fibrous reaction). Depending upon the predominance of particular type of cells, varied signal intensity is seen on T2 weighted MR images (Fig. 12 on page 13). As in other organs, these pseudotumors are great mimics of other neoplasms on CT and MRI. CT may show presence and type of calcification and bone reaction. It is observed that they are usually located close to neurovascular structures. Wide resection should be considered since there is high risk of local recurrence with possibility of malignant transformation.

Fibrous Malignancies

Malignant fibrous histiocytomas/ Undifferentiated pleomorphic sarcoma

Past research has revealed lack of true histiocytic differentiation in malignant fibrous histiocytomas (MFH) and this led WHO to modify the terminology of its various pathological subtypes to to undifferentiated pleomorphic sarcoma, undifferentiated high-grade pleomorphic sarcoma (formerly pleomorphic MFH), undifferentiated pleomorphic sarcoma with giant cells for MFH (giant cell type), undifferentiated pleomorphic sarcoma
with prominent inflammation (formerly inflammatory MFH) and myxofibrosarcoma (formerly myxoid MFH).

Undifferentiated pleomorphic sarcomas usually occur in deep soft tissues of extremities between 4th to 7th decades. They are also the most common post-radiation sarcomas.

Histologically, they are composed of anaplastic spindle cells with varying degree of differentiation, characteristically arranged in a fascicular or so-called herringbone pattern. On gross pathology, hemorrhage and necrosis can be seen in high grade tumours.

**Fibrosarcoma and other fibrous malignancies**

Term fibrosarcoma is also becoming increasing obsolete and is now reserved for describing tumors such as dermatofibrosarcoma protuberans or sclerosing epithelioid fibrosarcoma. Other true histological varieties of fibrous malignancies include the relatively common (adult and infantile fibrosarcoma) and some extremely rare variants (low-grade fibromyxoid sarcoma, myxoinflammatory fibroblastic sarcoma, low-grade myofibroblastic sarcoma and inflammatory myofibroblastic tumor).

Most cases of previously diagnosed fibrosarcomas may in fact be synovial sarcomas, malignant peripheral nerve sheath tumor or other pathological types.

**Imaging findings**

Although these fibrous malignancies are pathologically distinct, imaging findings of these lesions largely overlap. On MR imaging, these tumors are commonly of heterogeneous signal intensity which is representative of their intrinsic tissue characteristics (Fig. 13 on page 19 and Fig. 14 on page 20), necrosis, hemorrhage and calcifications. Hemorrhage and calcification cause blooming on gradient images. These tumors at least in the early stages are well circumscribed due to a pseudocapsule, emphasizing that definition of tumor margin should not be used to determine the benignity of a soft tissue neoplasm.

Fibrous malignancies typically enhance in a diffuse and heterogeneous pattern after administration of intravenous contrast. Peripheral and nodular enhancement may be seen in lesions with central necrosis, hemorrhage or in myxoid tumours.

Myxoid features are are not unique to fibrous malignancy and are also seen in myxoid liposarcoma, neurogenic neoplasms, soft tissue myxoid chondrosarcoma and myxoma.
Low grade myxofibrosarcomas amongst fibrous malignancies need a special mention due to their propensity to spread along fascial and vascular planes, far beyond the site of primary tumor. On MRI, these tumors very frequently show a tail-like margin and superficial spreading configuration, as is also seen in our case (Fig. 14 on page 20). Low grade myxofibrosarcoma tends to become progressively higher in grade at recurrence. Knowledge of these unusual characteristics helps in surgical planning.

Images for this section:

Fig. 1: Nodular Fasciitis in a 39 year old female who presented with a rapidly enlarging left wrist lump. MR T1 axial image (a) shows a well circumscribed lobulated mass of intermediate signal intensity along the volar aspect of wrist. It lies in close relation to the flexor carpi ulnaris tendon sheath (orange arrow). The mass also abuts the flexor retinaculum (blue arrow) though there is no infiltration of underlying flexor tendons. It shows heterogeneous high signal on T2 (F/S) axial image (b). Intense enhancement is seen on T1 post contrast image with an irregular non enhancing area within it (c). No susceptibility is detected on gradient-echo sequence (d) which helps in differentiating it from giant cell tumor of the tendon sheath.
Fig. 3: Giant cell tumor of flexor tendon sheath in a 45 year-old-man who presented with a non tender lump for 1 year. MR images reveal a lobulated soft tissue lesion wrapping the flexor tendon. It is of intermediate signal intensity on T1W image (a). It is iso to slightly hyperintense on T2W fat saturated image (b), demonstrates small foci of susceptibility on gradient image (c) (red arrow) and shows avid, heterogenous enhancement on post contrast image (d).
**Fig. 2:** Fibroma of tendon sheath in a 63 year old male with a right upper limb lump. Colour doppler ultrasound study (a) shows a well-defined isoechoic ovoid-shape mass in the subcutaneous fat, indenting the underlying muscle without invading it. The mass shows minimal internal vascularity. T1 axial (b) and corresponding T2 (F/S) (c) images show a homogenously hypointense lesion (arrow) in the deep fascial plane of the arm which indents upon the triceps muscle. There is no evidence of intramuscular invasion. On T1 post contrast image (d), the lesion shows minimal enhancement.
**Fig. 6:** Late stage myositis ossificans in a 55 year-old- paraplegic female who presented with tender palpable mass in the right thigh. Axial T1W (a), T2W fat saturation (b) and post contrast (c) images reveal a non enhancing lobulated fat-based lesion in the anterior aspect of right thigh. Plain radiographs (d) demonstrate extensive ossification around the right hip joint.
Fig. 4: Desmoplastic fibroblastoma in a 57 year old female with a painless lump over the first webspace of hand for about 3 years. Located within the belly of the first dorsal interosseous muscle, the lesion demonstrates intermediate to low signal intensity on T1 axial (a) and diffusely hypointense signal intensity on T2 (F/S) (b) images. On T1 post contrast image (c), the lesion shows heterogeneous enhancement. It abuts the second metacarpal and second dorsal interosseous muscle and is inseparable from the extensor tendon of index finger. No invasion of the adjacent bone is evident.
Fig. 5: Calcifying fibrous tumor in a 28 year old male who presented with a progressively increasing lump around the left knee. MRI shows a well defined ovoid subcutaneous lesion anteromedially in the left knee which appears hypointense on T1 axial (a) and T2 (F/S) (b) images. Mild contrast enhancement is seen on T1 post contrast image (c). The lesion abuts the medial patellar retinaculum. No deep infiltration is seen. (d) Radiograph of the knee shows calcification within this mass (orange arrow).
**Fig. 12:** Benign inflammatory fibrosclerotic lesion (pseudotumor) in a 48 year old female who presented with a mobile, non tender lump over the palmar aspect of left hand. T1 axial image (a) shows an intermediate signal intensity lesion overlying the 4th metacarpal. On T2 (F/S) axial image (b) the lesion reveals hypo to intermediate signal intensity. The lesion is closely related to the palmar interosseous muscle of fourth webspace but remains separate from the flexor tendon sheath by a clear fat plane. On colour doppler ultrasound study (c), the lesion appears iso to hypoechoic and demonstrates vascularity. The flexor tendon is seen deep to it.
**Fig. 7:** Superficial fibromatosis in a 45 year old patient who presented with a painful lump in the left thigh for about 1 year. T1 axial image (a) shows an intermediate signal intensity subcutaneous lesion (orange arrow) in the posterolateral aspect of the thigh with low to intermediate signal on corresponding T2 (F/S) axial image (b) and homogeneous enhancement on T1 fat saturated post contrast sequence (c). It is closely related to the deep fascia between the iliotibial band and the posterior compartment of the thigh with no deep extension seen.
**Fig. 8:** Plantar fibromatosis in a 40 years old male. Short axis images of right foot reveal a T1 isointense (a), T2 intermediate to hyperintense signal intensity (b) subcutaneous lesion stuck to the medial cord of the plantar fascia. The lesion shows avid, mildly heterogeneous post contrast enhancement (c). It demonstrates areas of T1 and T2 hypointensities within which are suggestive of the fibrous nature of tumor. There is no infiltration of the underlying plantar muscles. On ultrasound (d), the same nodule appears hypo to isoechoic and causes focal fusiform bulge of the plantar fascia. The patient had similar appearing nodules in the other foot as well.
**Fig. 9:** Desmoid-type fibromatosis in a 47 year old female who presented with painful posterior thigh swelling for 2 years which was progressively increasing in size. T1W axial image (a) shows an intermediate signal intensity mass (arrows) in the lateral aspect of the posterior compartment with heterogenously hyperintense signal intensity on T2W axial image (b). There are thick band-like low signal intensity areas within this mass which are suggestive of this neoplasm. T1 post contrast image (c) demonstrates avid enhancement of the mass barring that of the low signal intensity collagenous areas.
Fig. 10: Solitary fibrous tumour in a 32 year old male who presented with a lump on the anteromedial side of the left thigh. T1 axial image (a) shows a lobulated subcutaneous T1 isointense lesion. On corresponding T2 (F/S) axial image (b), the lesion demonstrates heterogenous increased signal intensity with avid enhancement seen on T1 post contrast fat saturated image (c). Prominent perilesional vessels are seen (orange arrows) which are helpful in making the diagnosis.
**Fig. 11:** Plexiform fibrohistiocytic tumor in a 25 year old male who presented with hypophosphotemia, osteomalacia and a lump in his sole. T1W (a) and T2W fat saturated (b) images show a lobulated, infiltrating low signal intensity lesion in the subcutaneous plane of the sole which shows minimal enhancement on T1W fat saturated post contrast image (c).
Fig. 13: Low grade fibrosarcoma in a 49 year old female with rapidly increasing wrist lump. MRI reveals a relatively well circumscribed ovoid mass in the ulnar aspect of the right distal forearm, located primarily in the subcutaneous plane. It lies in close proximity to the flexor carpi ulnaris tendon (blue arrow), with no clear fat plane seen between the two. This lesion is predominantly of low signal intensity on T1W (a) and T2W (b) images with heterogenously bright T2 signal areas (orange arrow). On T1 fat saturated post contrast image (c), the lesion appears very vascular and shows mildly heterogeneous enhancement. Dark linear strands are also seen on T1, T2W and post contrast images which suggest fibrous nature of this lesion. No blooming was seen on the gradient images (not shown). On ultrasound (d), the lesion appears isoechoic. The flexor carpi ulnaris tendon lies deep to it (arrow).
**Fig. 14:** Recurrent Myxofibrosarcoma in a 80 year old man with a recurrent soft tissue mass (arrows) in the expected location of the biceps muscle. The mass is of intermediate signal intensity on T1W image (a) and of high signal intensity on T2W fat-saturated image (b). The mass appears to have slightly irregular margins with tail-like configuration (orange arrow). T1 post contrast image (c) shows avid enhancement. The tumour abuts underlying brachialis muscle and is closely related to the neurovascular structures (brachial vessels, median nerve and basilic vein) (blue oval). The patient later underwent right scapulothoracic amputation.
Conclusion

Although histology is required in many cases, the radiologist by systematically using clinical history, lesion location and signal intensity characteristics on MR images can develop appropriate differential diagnoses. MR imaging is also useful to determine the origin of these lesions, in defining their extent and relation to adjacent structures, and in follow-up after therapy.

Musculoskeletal radiologists who are involved in evaluation of these fibrous tumours need to be aware of the modifications of the WHO nomenclature for the group of fibrous malignancies, so that it allows more uniformity in discussion with our oncology and pathology colleagues.

Personal information

References


