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Review

Characterising benign fibrous soft-tissue tumours in adults: why is it so difficult and what do we need to know?

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Fibrous, myofibroblastic, and fibrohistiocytic soft-tissue tumours are amongst the most common benign soft-tissue lesions encountered in clinical practice. They demonstrate varied biological behaviour and imaging characteristics. Benign fibroblastic lesions, such as nodular fasciitis, are small, have a self-limited course, and rarely recur after excision, whereas deep fibromatosis and plexiform fibrohistiocytic tumours tend to exhibit more aggressive features and often have high recurrence rates. MRI with its superior tissue contrast, multiplanar imaging capability, and lack of ionising radiation is regarded as the preferred method of tumour evaluation, tissue characterisation, and assessment of treatment response. Histopathological features are depicted at MRI, reflecting the amount and distribution of the cellular and fibrous matrix. Cellular tumours tend to show higher T2 signal intensity and post-contrast enhancement as compared to tumours with greater collagenous content, which appear dark and show less enhancement. Awareness of MR characteristics, pathological behaviour, and common sites of occurrence of fibrous soft-tissue tumours will help radiologists to determine the appropriate differential diagnosis and guide patient management.

Introduction

Fibrous, myofibroblastic, and fibrohistiocytic soft-tissue tumours represent a large subset of mesenchymal tumours that are commonly encountered in adults during clinical practice with a varied range of anatomical locations, pathological features, and biological behaviour. Despite advances in imaging, characterisation of these soft-tissue lesions remains a challenging clinical problem.

The World Health Organisation (WHO) classification (2013) of fibroblastic and fibrohistiocytic tumours categorised these tumours into benign, intermediate (locally aggressive and rarely metastasising), and malignant. This review article will focus on the first two categories. These include benign fibroblastic lesions, such as nodular fasciitis, as well as intermediate lesions, such as deep fibromatosis and plexiform fibrohistiocytic tumours (PFTs), which tend to demonstrate more aggressive features. Notably amongst the intermediate-grade tumours, superficial and deep fibromatosis (desmoid tumours) fall under the “locally aggressive” subcategory whereas solitary fibrous tumours

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(SFTs), inflammatory myofibroblastic tumours, and PFTs are considered “rarely metastasising”. (Table 1)

Clinical evaluation and imaging approach

Careful clinical assessment of the region of concern should precede any imaging. The clinical history, age of the patient, and location of the fibrous mass are important pieces of information that when analysed with imaging findings can help to establish the diagnosis. Imaging characterisation of soft-tissue tumours in general should not be limited to MRI. Plain radiography helps to distinguish hard palpable lumps caused by underlying bony deformity, such as exostosis, and also enables assessment of soft-tissue calcifications, which can be specific for certain diagnoses such as myositis ossificans and calcifying fibrous tumours in the present context. Amongst soft-tissue sarcomas, synovial sarcomas show calcifications in a large proportion of cases. Specific aggressive features, such as bony erosion or destruction and joint involvement, can also be evaluated. Sonography is useful in determining the size and consistency of a superficial soft-tissue mass. High-resolution ultrasound allows solid structures to be distinguished from cystic ones and is useful in directing percutaneous biopsy. Colour Doppler ultrasound can also evaluate the vascularity of the lesion.

MRI with its superior tissue contrast, multiplanar imaging capability, and lack of ionising radiation is regarded as the preferred method of soft-tissue tumour evaluation, tissue characterisation, local staging, tumour extent, and surgical planning.[1] Multiplanar imaging is very helpful in determining the anatomical extent of the lesion and its relationship to adjacent structures.

Table 1
Overview of benign and intermediate fibroblastic and fibrohistiocytic soft-tissue tumours in adults.

Benign	Intermediate (locally aggressive)	Intermediate (rarely metastasising)
Nodular fasciitis	Superficial fibromatosis	Solitary fibrous tumor
Myositis ossificans	Desmoids-type fibromatosis	Inflammatory myofibroblastic tumor
Elastofibroma		Plexiform fibrohistiocytic tumor
Fibroma of tendon sheath		
Desmoplastic fibroblastoma		
Calcifying fibrous tumor		
Giant cell tumor of tendon sheath		
Other benign proliferations[a]		
Ischaemic fasciitis		
Mammary-type myofibroblastoma		
Angiomyofibroblastoma		
Cellular angiofibroma		
Nuchal-type fibroma		
Gardner fibroma		

[a] These lesions are rarely seen on imaging and will be excluded from this article discussion.

MRI technique

The present cases were examined using 1.5 or 3 T MRI systems. MRI images of the lesion should be acquired in at least two orthogonal planes, using T1-weighted and T2-weighted spin-echo sequences, the latter usually with fat saturation to make the lesion more conspicuous. A short-tau inversion time recovery (STIR) sequence may be added if adequate fat suppression is a concern, but in general, it should not replace T2-weighting, which has a better signal-to-noise ratio and higher resolution. Most fibrous tumours are heterogeneous and show mixed signal intensity on T2-weighting. T2 dark regions within the tumour usually represent collagenous bands, hyalinised regions or haemosiderin deposits for cases of desmoid tumours, fibroma of the tendon sheath (FTS), and giant cell tumour of the tendon sheath (GCTTS) respectively. Gradient-echo sequences are useful for identifying haemosiderin products, which are vital in making a diagnosis of GCTTS. The additional value of T1-weighted sequences with fat suppression has been described in characterising fibrous tumours by Gielen *et al.*[2] but has not been confirmed. However, post-contrast T1-weighted fat-suppressed images are useful in commenting upon vascularity and margins of fibrous tumours. The dark collagenous bands show no enhancement whereas the more cellular component comprising fibroblasts and other cells show enhancement. The enhancement seen in viable tumour also allows selection of biopsy site.

For surgical planning, it is crucial to determine the anatomical compartment of the lesion, whether it is intramuscular, subcutaneous, or intra-articular. Placement of a marker over the area of clinical concern is often useful to ensure that the area has been included within the field of view. MRI is also used to assess response to therapy and for recurrent disease surveillance.

Benign fibroblastic proliferations

Benign fibroblastic proliferations constitute a heterogeneous group of well-defined entities. Lesions such as nodular fasciitis are cellular, tend to grow rapidly, and can simulate sarcoma. Other fibroblastic proliferations, such as elastofibromas, FTS, and keloids, are more collagenous and less cellular. Although tumours under this category may have aggressive clinical presentation, they rarely recur after resection.

Nodular fasciitis

Nodular fasciitis also called pseudosarcomatous fasciitis or proliferative fasciitis is a benign soft-tissue lesion of proliferating fibroblastic–myofibroblastic cells. The lesion commonly presents as a fast-growing painful mass in the upper extremity of young adults (20–40 years age group), which may arouse suspicion of sarcoma.[3] Although the cause is unknown, it may be due to a reactive process triggered by local injury or inflammation. In a recent study from the Mayo Clinic, MYH9-USP6 gene fusion was