

SOFT TISSUE SARCOMAS OF EXTREMITIES: A PICTORIAL REVIEW FROM TERTIARY CARE ONCOLOGY SETUP

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PJR October - December 2019; 29(4): 252-258

ABSTRACT

Radiologists are frequently encountered by soft-tissue sarcomas in their daily practice. Characterization and exact diagnosis of these soft-tissue sarcomas pose a great challenge, despite advances in imaging. By systematically using clinical history and imaging findings such as lesion location, mineralization on radiographs, and imaging characteristics on magnetic resonance images, one can narrow the differential diagnosis. Malignant neoplastic processes can mimic benign lesions and may be difficult to differentiate. Radiologists should have a thorough knowledge of the imaging characteristics of these lesions to advise on appropriate surgical management. Features that are more common in malignant lesions include large size (> 5 cm), deep site, inhomogeneous signal intensity, haemorrhage and necrosis, early and inhomogeneous contrast enhancement, irregular margins, surrounding soft tissue oedema and invasion of adjacent structures, including bone and neurovascular bundle. Advanced MRI techniques such as spectroscopy, perfusion and diffusion-weighted imaging may contribute to better soft tissue characterisation. If a lesion is lacking characteristic benign features then it should be regarded as indeterminate, and patient should undergo tissue diagnosis to exclude malignancy. Core biopsy is required as fine needle aspiration is inadequate. In this review article, we have tried to highlight imaging findings of soft tissue sarcomas mainly discussing the MRI imaging and their characteristic findings which can help a radiologist to narrow differential diagnosis before tissue diagnoses.

Key words: Soft tissue sarcoma, imaging, MRI, malignant

Introduction

Soft tissue masses of the extremities are relatively infrequent and may pose a diagnostic dilemma.¹

Plain radiography can be useful. It allows characterisation of mineralisation within a lesion and can be useful to assess for the presence of benign calcifications such as phleboliths. It is also useful to assess the extent of underlying bone and joint involvement, which can both suggest an etiology and have important implications for surgical treatment.^{2,3}

MRI provides excellent anatomical detail and allows for soft tissue characterisation which plays an

important role in formulating a differential diagnosis. It allows for local staging and description of the relationship of a lesion to adjacent anatomical structures (e.g. fascia, bone, muscle, neurovascular structures). Furthermore, advanced MRI techniques may aid in soft tissue characterisation. Lower apparent diffusion coefficient (ADC) scores on diffusion-weighted imaging (DWI) are generally seen in malignant lesions compared with benign lesions. Dynamic contrast-enhanced (DCE) perfusion imaging may aid in differentiating benign from malignant lesions based

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Submitted 26 November 2019, Accepted 10 December 2019

on patterns of contrast enhancement and washout. In addition, it can assess for viable, enhancing tumour to guide biopsy and may aid in early detection of residual/recurrent tumour post resection.^{4,5,6}

Our purpose is to make an overview of imaging findings of soft tissue sarcomas of limbs with emphasis on characteristic signs and correlate with histologic features.

Adult soft tissue sarcomas	• undifferentiated pleomorphic sarcoma (malignant fibrous histiocytoma): commonest type of soft tissue sarcoma
	• liposarcoma: second most common type
	• rhabdomyosarcoma
	• dermatofibrosarcoma
	• leiomyosarcoma
	• haemangiosarcoma
	• Kaposi sarcoma
	• lymphangiosarcoma
	• synovial sarcoma
	• malignant peripheral nerve sheath tumor
• extraskeletal chondrosarcoma	
• extraskeletal osteosarcoma	

List of adult soft tissue sarcomas

Soft tissue sarcomas:

Pleomorphic undifferentiated sarcoma (PUS):

Previously called as Malignant fibrous histiocytoma (MFH) and Fibrosarcoma. It is considered the most common type of soft tissue sarcoma and account for 25-40% soft tissue sarcomas. It occurs in adults in age range of 32-80 years with a slight male predilection. They are usually confined to the soft tissues and commonly seen in extremities. Plain radiographs will demonstrate a soft tissue mass with, curvilinear or punctate calcification in some cases. These are well circumscribed tumors appearing intermediate to low signal intensity on T1WS and high on T2WS however may show heterogenous signals on T1 and T2WS due to internal hemorrhage, calcification, necrosis, myxoid material. Prominent enhancement of solid components is seen. Better prognosis is associated with smaller tumors, superficially and distally located and histological grade.^{7,8,9}

(Fig. 1) shows 62 years male with Undifferentiated pleomorphic sarcoma. Patient presented with swelling in lower leg since 3 months.

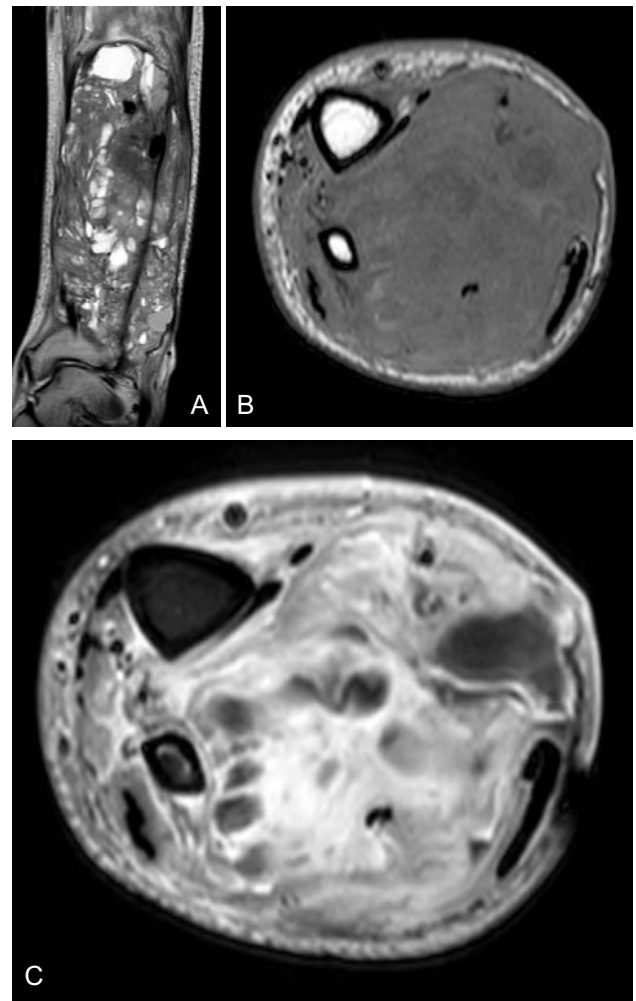


Figure 1a,b,c: showing coronal T2, axial T1 and post contrast T1WS. Large heterogeneously enhancing soft tissue lesion with internal cystic / necrotic areas predominantly involving posterior compartment muscles with encasement of the anterior as well as posterior neurovascular bundles. It shows heterogenous contrast enhancement with internal cystic areas.

Liposarcoma:

Liposarcomas are malignant tumors of fatty tissue and are the malignant counterpart of benign lipoma. They are usually seen in the extremities (75%), most commonly the thigh. MRI appearances are variable predominantly depending on the grade and amount of fatty tissue. Higher grade liposarcomas are often devoid of macroscopic fat and appearances are similar to other sarcomas.¹⁰

(Fig. 2) shows MRI of 37 years old male with swelling right medial thigh. Core biopsy showed Myxoid liposarcoma with more than 30% round cell component.

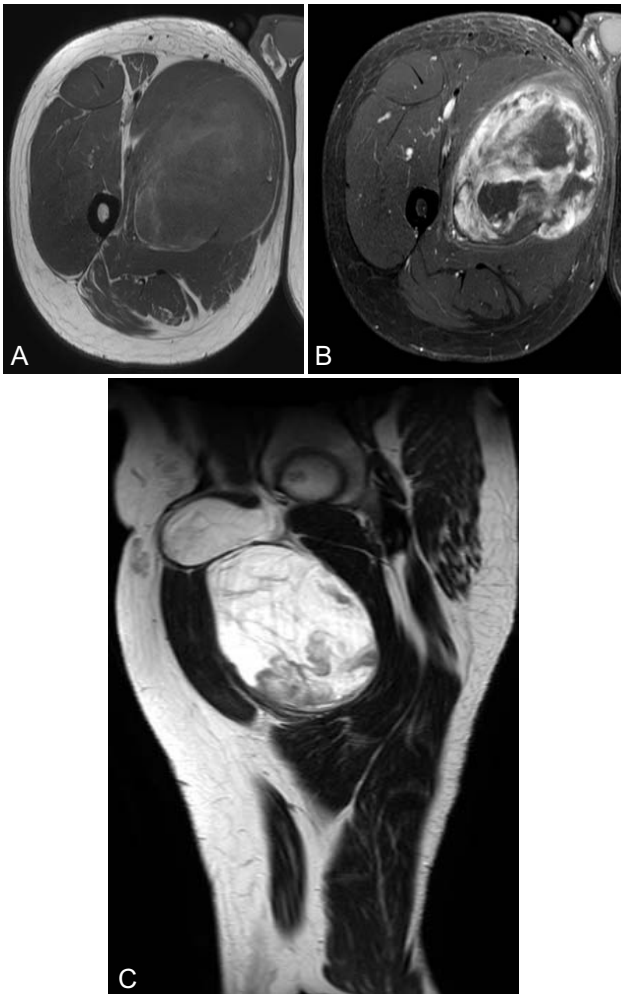


Figure 2a,b,c: axial T1, Post contrast T1WS and sagittal T2WS showing large lobulated heterogeneously enhancing soft tissue tumour within the proximal adductor compartment of the right thigh showing subtle T1 hyperintense signals representing fat component.

Synovial sarcoma:

Synovial sarcomas are relatively common and most commonly involving the soft tissue surrounding the knees. Radiographs may be normal however in 30% may show a mass with dystrophic calcifications, smooth pressure erosions or direct bony invasion. These may be T1 hypo to slightly hyperintense to muscle or could be heterogenous. T2 signals are mostly hyperintense or can be heterogenous due to internal necrosis and cystic degeneration. Due to the high tendency of lesions to bleed, there might be areas of fluid-fluid levels. Enhancement is usually prominent and can be diffuse (40%) heterogeneous (40%) or peripheral (20%).^{8,11}

(Fig. 3) shows post contrast MRI of 18 years old female with biopsy proven synovial sarcoma.

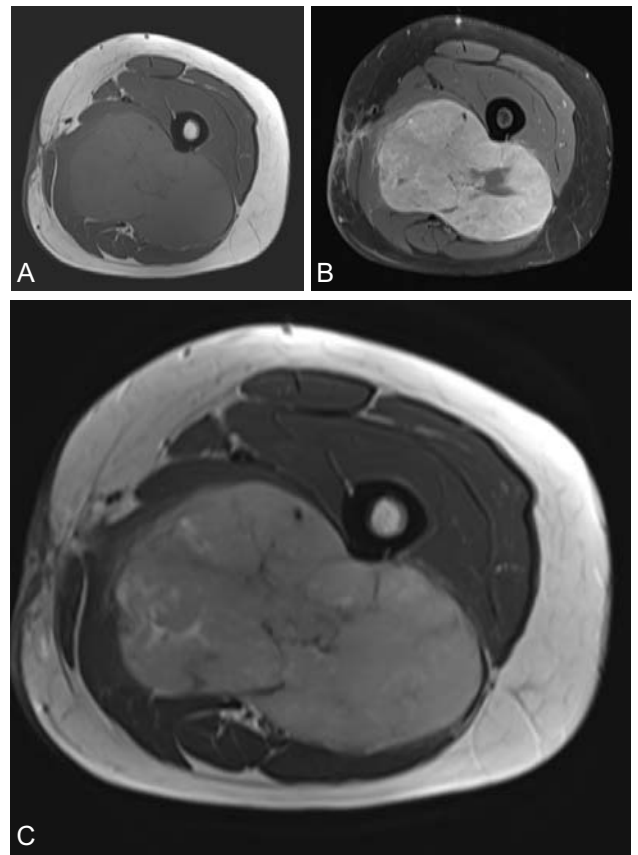


Figure 3a,b,c: axial T1, CE T1WS and T2WS. Large enhancing tumor in posterior compartment of the left thigh. Iso to hypointense to muscles on T1WS and hyperintense on T2WS, showing avid contrast enhancement. Post biopsy changes are seen along medial aspect. The superficial femoral neurovascular bundle and sciatic nerve are draped over the lesion.

Rhabdomyosarcoma:

Rhabdomyosarcoma is a primitive malignant soft tissue sarcoma with a skeletal muscle phenotype. Embryonal variety tends to occur in the first decade of life, whereas alveolar type is more common in adolescents and young adults. Metastases occur to the lymph nodes, lungs and bones are seen. These are large necrotic lesions with a lobulated margin and lymphovascular invasion. MRI demonstrates a lesion that is iso- to hyperintense to muscle on T1WS, heterogeneously hyperintense on T2W with moderate to marked enhancement with contrast. Prominent serpentine flow voids are often a feature of these highly vascular lesions, as is internal hemorrhage.^{6,12}

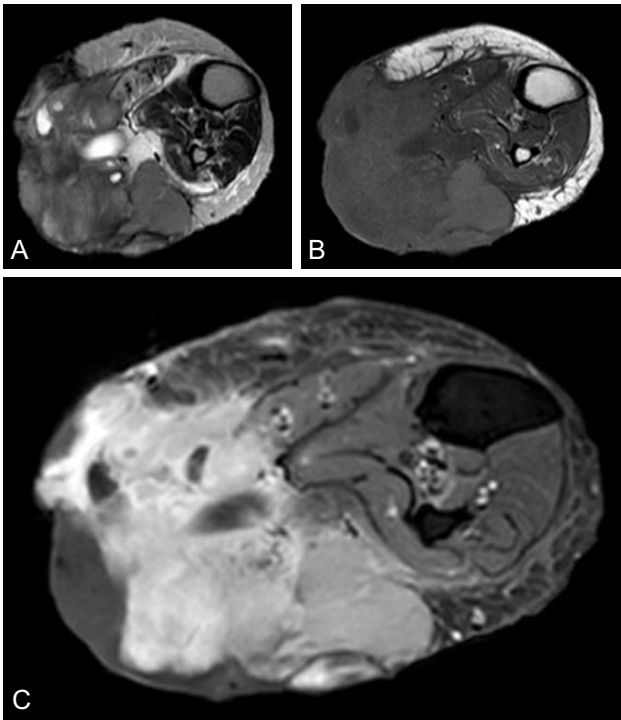


Figure 4a,b,c: axial T2, T1 and CE T1WS. Large exophytic mass in 69 years old female appearing heterogeneous on T2, showing intense postcontrast enhancement with internal areas of necrosis. This was biopsy proven pleomorphic rhabdomyosarcoma

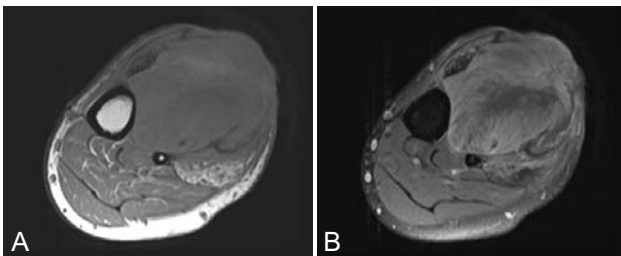


Figure 5a,b: T1 and CE T1WS. 27 years old male with embryonal rhabdomyosarcoma of left lower limb presented with swelling for the past one year. Anterolateral aspect of upper leg shows soft tissue mass with mild post contrast enhancement. Neurovascular bundle is draped along the mass. No cortical destruction.

Dermatofibrosarcoma protuberans(DFSP):

Seen in adolescents, it is a rare slow growing superficial mass which may ulcerate. It demonstrates iso to slightly hypointense signal on T1W and high signal on both conventional T2W and fat suppressed T2W sequences with intense enhancement. Increased T2 signal may be seen spreading from the lesion within the adjacent skin. Dermatofibrosarcoma protuberans (DFSP) has a low metastatic potential but local recurrence after surgical resection may occur.^{13,14}

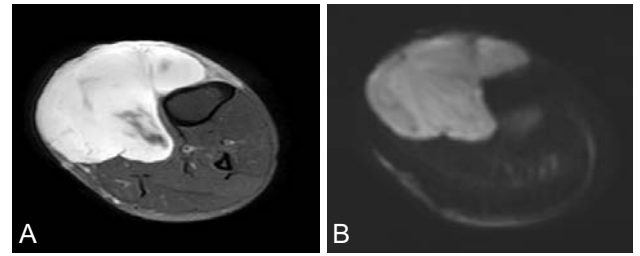


Figure 6a,b: CE T1 and DWI. Diffusion restricting avidly enhancing mass involving subcutaneous region along medial aspect of proximal left tibia. No muscular invasion / neurovascular encasement noted. This was biopsy proven recurrent DFSP in 55 years male.

Angiosarcoma:

Angiosarcoma is a rare mesenchymal malignant neoplasm of the vascular or lymphatic endothelium, accounting for 2% of all soft-tissue sarcomas. Brightly enhancing soft tissue mass, often hyperintense on T2WI, with prominent flow voids, It has tendency to metastasize to liver, lungs and lymph nodes.¹⁵ (Fig. 7) shows 81 years old male presented with swelling since 3 months. HP showed spindle cell angiosarcoma. Patient had no distant metastasis.

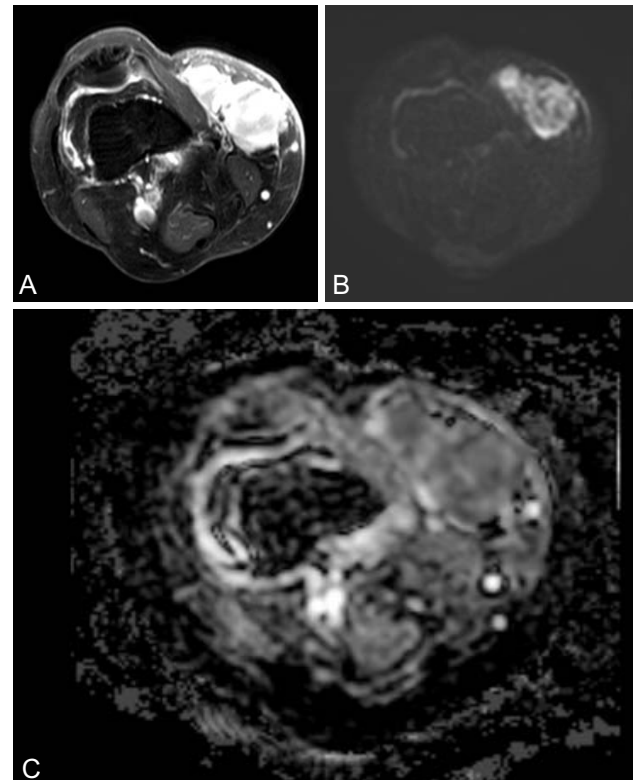


Figure 7a,b,c: CE T1WS, DWI and ADC mapping shows large subcutaneous lesion, showing avid post contrast enhancement. There is intense internal DWI restriction. no muscular involvement was seen.

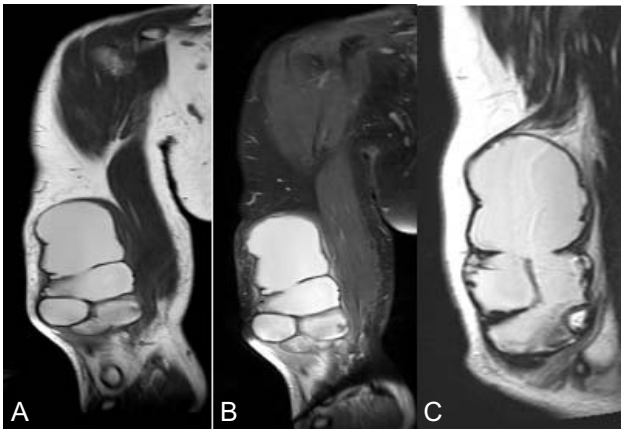


Figure 8a,b,c: T1, CE T1WS, T2WS. 81 years old male with epitheloid angiosarcoma. lobulated intramuscular multiloculated cystic mass within the anterolateral compartment of the upper limb with involvement of radial nerve. The internal contents of the lesion appeared T1, T2 and STIR hyperintense, suggestive of internal proteinaceous or hemorrhagic contents.

Extraskelatal chondrosarcoma:

Extraskelatal chondrosarcomas make up only 2% of soft-tissue sarcomas. They tend to be of higher grade than conventional intramedullary chondrosarcomas, with the majority being of the myxoid (most common) or mesenchymal varieties. Low to intermediate signal on T1WS and very high intensity on T2WS. Most demonstrate heterogeneous moderate to intense contrast enhancement on CE T1WS.¹⁶

(Fig. 9) shows 34 years old female presented with pain and swelling since 3 weeks. Core biopsy showed mesenchymal chondrosarcoma of the soft tissues.

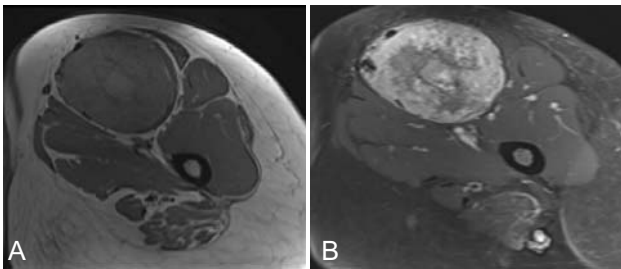


Figure 9a,b: T1 and post contrast T1WS shows well circumscribed lesion showing post-contrast enhancement with central necrosis.

Leiomyosarcoma:

Leiomyosarcoma has male predominance and most commonly occurs in the retroperitoneum, closely followed by the lower extremities. The age range is 35 to 79 years. In the extremities, they typically present as a solitary painful nodule. On MRI, they appear as

a nonadipose soft tissue neoplasm with areas of necrosis.¹⁷

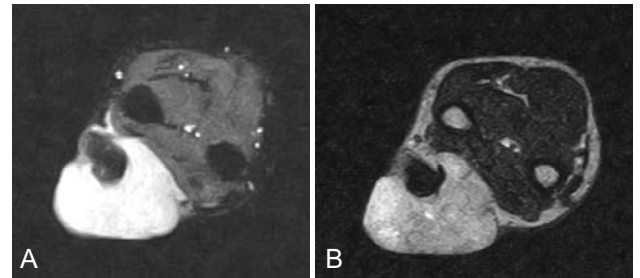


Figure 10a,b: shows CE T1WS and T2WS. Lobulated superficial soft tissue mass posteromedial aspect of the forearm. This shows post contrast enhancement with an eccentric focus which appears of low signal intensity in all the sequences and is likely to represent either calcification or chronic haematoma. Excision biopsy was consistent with Malignant spindle cell sarcoma favoring leiomyosarcoma.

Round cell sarcoma:

Undifferentiated small round cell sarcomas, comprise a rare subset of highly aggressive sarcomas. Diagnosis requires radiology and histopathology. These exhibit high rates of metastasis to lung and brain, often with a poor prognosis, and median survival is < 2 years.¹⁸

(Fig. 11) shows 54 years female presented with left hand swelling since 1 year. HP showed Undifferentiated round cell sarcoma.

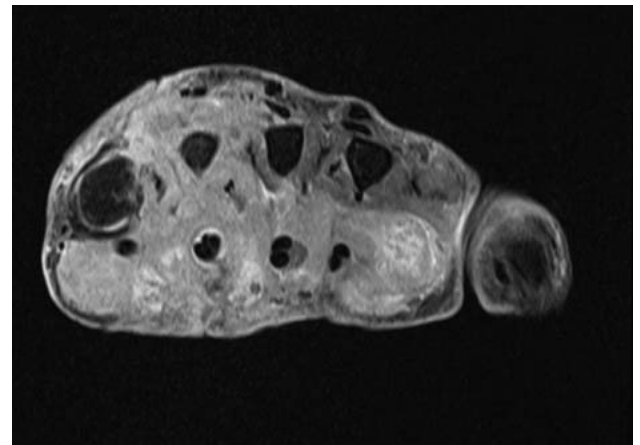


Figure 11: CE T1WS. Enhancing tumour encasing the long flexor tendons with extension through third and fourth spaces into the dorsum of the hand where it is inseparable from the extensor tendons as well.

Malignant peripheral nerve sheath tumors:

Malignant PNSTs account for 6% of soft-tissue sarcomas and are associated with type 1 neurofibromatosis in 50% of cases. On T1WI, it has low signal

and on T2WI, it has high signal. Malignant peripheral nerve sheath tumor enhances brightly on postcontrast images and often has a central dark area secondary to necrosis and hemorrhage. These tumors are continuous with the involved nerve.¹⁷

(Fig. 12) shows enlarging distal thigh mass since 5 months in 20 years old male.

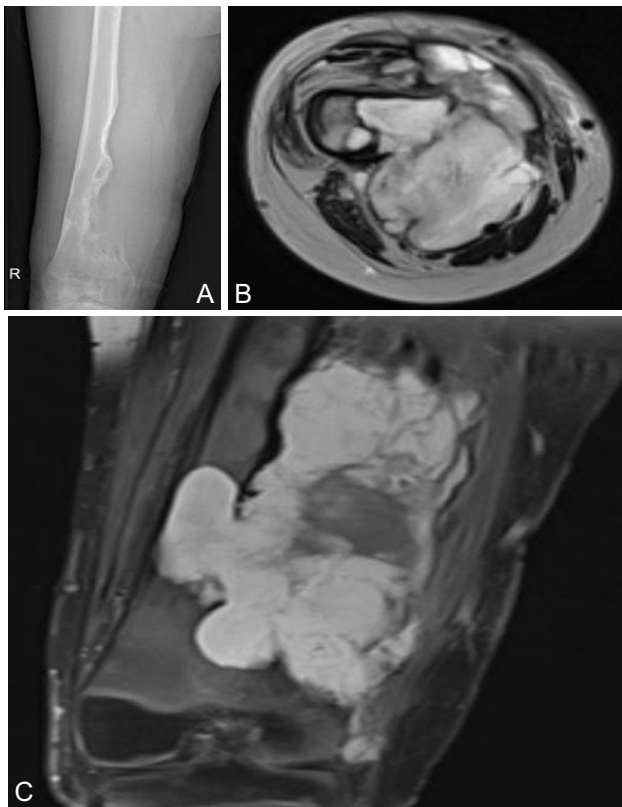


Fig 12a,b & c: X ray femur shows large cortical defects and erosions in the distal femoral metadiaphysis, with wide zone of transition. Large lobulated T2 hyperintense tumor (Fig. b) in the posteromedial soft tissues causing scalloping and invasion into the bone. Avid post contrast enhancement seen (Fig. c).

Conclusion

MRI is the imaging modality of choice when dealing with soft tissue lesions of extremities. Certain soft tissue tumours are identifiably benign because of their signal characteristics, morphology or location. However there is huge overlap between benign and malignant soft tissue tumors in which case high level of suspicion for malignancy is suggested. Histopathological diagnosis remains the gold standard for final diagnosis.

Soft tissue sarcomas are generally a diagnostic dilemma needing the complimentary use of both radiology and pathology for their accurate diagnosis.

Funding: None

Conflict of interest: None declared

Ethical approval: The study was approved by institutional review board.

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