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Soft-Tissue Sarcomsa Fo Chest Wall

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I. Introduction

The chest wall consists of soft tissue, cartilage, and bone tissue. Tumors that arise from the soft tissue of the chest wall are less common than tumors in other parts of the body. Primary soft tissue tumors of chest wall include mainly adipocytic tumors, muscular tumors, vascular tumors, peripheral nerve sheath tumors, cutaneous lesions, fibroblastic-myofibroblastic tumors, and fibrohistiocytic tumors. More than 50% of the chest wall tumors are malignant [1]. CT and MRI play a crucial role in diagnosing chest wall tumors. Due to the rarity of these tumours, we analysed the imaging features of such tumours that we encountered in our institute over 1 year.

II. Material And Methods

This prospective observational study was conducted in the department of radiology and imageology at Nizam's Institute of Medical Sciences, Hyderabad over a period of 12 months. The institutional ethics committee permitted this project. The study included 30 patients of which 17 were males and rest were females between the age group of 5 to 70 years. All were referred to the Radiology department with clinical diagnosis of soft tissue chest wall lesion from August 2021 to September 2022. All the patients underwent CT Chest or MRI (CT in 15 cases, MRI in 14 cases, both CT and MRI in 1 case) and ultrasonography was done in 15 cases. Histopathological correlation was done for all patients. Written informed consent from the patients or patient's relatives was taken for inclusion in the study.

III. Results

Our study is a prospective study which included 30 patients in the age group of 5-70years with a mean age of 32years. The most common age group was 5-30 years (43.3%) followed by 31 to 50 years (40%). Majority of our patients were males, with male to female ratio of 1.3:1. There were 15 benign and 15 malignant primary soft tissue tumors of chest wall. Most common neoplasms were sarcomas (26.6%) followed by neurogenic tumors (23.3%), lymphoma (16.6%), vascular tumors (13.3%), lipoma (6.6%), PPNET (6.6%), Kikuchi's disease with secondary HLH (3.3%) and desmoid (3.3%). Most of the patients of sarcomas were in the age group of 30-40 years [25(83.3%)]. Of 30 cases evaluated for soft tissue chest wall lesions, the most common lesions encountered were Sarcomas (n=8, 26.6%). The details of imaging characteristics are depicted in table-1. The sarcomas proven were 3 cases of dermatofibrosarcoma protuberans, 2 cases of spindle cell sarcoma,1 case each of myofibroblastic type of spindle cell sarcoma, myxofibrosarcoma and myeloid sarcoma.

Table-1: Showing characteristics of 8 cases of sarcomas in our study

S.no	Age/sex	Site	Size (cm)	Imaging characteristics	Imaging Diagnosis	Final Diagnosis
1	33-M	Right deltoid	>5	CT-ill-defined iso to hypodense, homogenous lesion with no enhancement on post contrast.	Mesenchymal tumor	Myeloid sarcoma

				USG- heterogeneously hypoechoic lesion with increased vascularity		
2	34-F	Right anterior chest wall	<5	CT- irregular ill-defined isodense mass in subcutaneous planes with fingerlike projections at periphery and infiltrating skin. USG-hypoechoic lesion with increased vascularity	Fibromatosis	DFSP
3	36-M	Right anterior chest wall	>5	MRI-well-defined t1 hypointense, T2/STIR hyperintense lesion in subcutaneous plane abutting pectoralis muscle	Mesenchymal sarcoma	DFSP
4	47-M	Right anterior chest wall	<5	CT-well defined round isodense lesion in subcutaneous plane at thoracic inlet with minimal enhancement USG-hypoechoic lesion with internal vascularity	Neurogenic	DFSP
5	12-F	Left anterior chest wall	<5	MRI- T1 iso, T2/stir hyperintense lesion infiltrating skin and subcutaneous tissue with adjacent edema. USG-heterogenous hypoechoic lesion with anechoic cap around.	Mesenchymal sarcoma	Spindle cell sarcoma
6	22	Right scapular region	<5	MRI- small round t1/t2 hypointense, stir hyperintense lesion in subcutaneous plane	Mesenchymal sarcoma	Spindle cell mesenchymal tumor - myofibroblastic differentiation
7	41-M	Rt trapezius	>5	CT- large isodense homogenous lesion involving subcutaneous planes and trapezius and rt rhomboideus muscle. USG-highly vascular isoechoic lesion	Mesenchymal sarcoma	Spindle cell sarcoma
8	71-M	Left supraspinatus muscle	>5	CT- heterogenous mass with areas of iso and hypodensity and minimal heterogenous enhancement extending to skin and subcutaneous planes. USG- heterogenous hypoechoic lesion with internal vascularity	Fibromatosis	Myxofibrosarcoma

Most of the patients of sarcomas were in age group of 30-40 years, youngest patient was 12-year-old patient of spindle cell sarcoma and oldest was 71 years, a case of myxofibrosarcoma. 6 cases were males and 2 were females. 4 patients had lesions larger than 5cms (largest dimension) at the time of presentation and 4 patients had sizes less than 5cms. Two cases of DFSP were misdiagnosed as Neurogenic tumor, fibromatosis on CT, a case of Myxofibrosarcoma was misdiagnosed as fibromatosis on CT. Most of the cases were given a diagnosis of mesenchymal tumor.

IV. Discussion

Sarcomas accounts for less than 1% of all malignant mesenchymal tumors [2,3]. According to WHO there are more than 100 different varieties. Undifferentiated soft tissue sarcoma constitutes a major group. Spindle cell sarcoma is one of the rare types of undifferentiated soft tissue sarcoma.

Dermatofibrosarcoma protuberans [DFSP]

DFSP arise from dermis and infiltrates into subcutaneous tissue. Rarely does it arise from subcutaneous tissue. Trunk, scalp, breast, extremities are common sites. Tumor size varies from 1 cms to several cms with asymmetrical extension into subcutaneous tissue, sometimes penetrates the fascial plane. Rarely does it arise from muscle.

Of 3 cases of DFSP, 1 patient (36yr old) had large (>5cm) well defined lesion on MR imaging with low signal on T1-weighted images and higher signal than fat on T2-weighted sequences in subcutaneous plane at right upper chest wall abutting pectoralis muscle which was typical of DFSP similar to that described by Xincheng et al [4] (Figure-1). In their study, CT findings of 16 cases (17 tumors) with DFSP confirmed by pathological findings were, 15 cases appeared as solitary iso-hypodense, ovoid, or round mass at the cutaneous and subcutaneous tissue, 12 cases demonstrated isodense masses on unenhanced CT images. The mean diameter of tumors was 4.0 cm,

and the depth was 1.7 cm. The margin was well defined (n = 15 [88.2%]) or ill-defined (n = 2 [11.8%]). Fifteen tumors revealed moderate or marked homogeneous (n = 12 [80%], smaller lesion, diameter <5 cm) or heterogeneous (n = 3 [20%], larger lesion, diameter ≥5 cm) enhancement on enhanced CT indicating intratumoral necrotic and cystic degeneration areas. One of three cases of DFSP has small nodular lesion which was misdiagnosed as neurogenic tumor (Figure-2). In a study by Diago A et al **[5]** 30 patients with DFSP that underwent ultrasound examination classic finger-like projections were observed in 73.3% of cases. A posterior hyperechoic area extending deep into the subcutaneous tissue correlated with the honeycomb DFSP pattern and was observed in 53.3% of patients. Concordance between ultrasound and histopathologic depth measurements was excellent. In our study one case had classic finger-like projection but was misdiagnosed as fibromatosis on CT (Figure-3). In a study by Ryan K.L. Lee et al **[6]** retrospectively analysed 8 patients of dermatofibrosarcoma protuberans to evaluate the +ultrasound appearances of dermatofibrosarcoma protuberans (DFSP). Most (78%) tumors were in the subcutaneous region. The shape of the tumor was round in 67% (6/9) and ovoid in 33% (3/9) of cases. The margin was poorly defined in one (11%) and well-defined or circumscribed in eight (89%) tumors. All the tumors showed a mildly lobulated border and had a heterogeneously hypoechoic matrix, often with rounded, ovoid, or occasionally linear discrete hypoechoic areas. Most (67%) tumors showed moderate vascularity on colour Doppler imaging. This vascularity tended to be more profound peripherally rather than centrally, similar findings were observed in our study (Figure-3).

Myeloid Sarcoma

Myeloid sarcoma is extra-medullary myeloid cell tumor or chloroma originating from extramedullary tumor originating from immature granulocyte or monocyte.it often invades bone, soft tissue, lymph nodes, skin, and PNS. It is seen in 2-14 % of AML patient [7]

We had 1 case of myeloid sarcoma at right shoulder with CT imaging findings of large ill-defined iso-hypodense, homogenous lesion with no enhancement. Ultrasound showed heterogeneously hypoechoic lesion with increased vascularity. Numerous studies have noted myeloid sarcomas may involve muscles, but a parameningeal location appears to be a preferred site for these masses. Lesions are slightly hypodense to muscle at CT and isointense to bone marrow on both T1- and T2- weighted MR images; they enhance homogeneously after injection of contrast medium. They have micronodulations. These imaging patterns are immensely helpful in differentiating granulocytic sarcoma from synovitis, hematoma, or abscess. Since there was no enhancement in a case of granulocytic sarcoma, we thought to be mesenchymal sarcoma, and exact diagnosis was not offered (Figure-4). Early diagnosis is important as treatment can be offered. Guermazi et al reported breast masses in myeloid sarcoma were irregular in shape, homogenous, hypoechoic solid with ovoid shaped circumscribed masses with posterior shadowing [8].

Myxofibrosarcoma

Myxofibrosarcomas occur usually in 6th decade in extremities, trunk, or head in subcutaneous plane as multinodular growth. It may present as cutaneous lesion also with infiltrative margins and grows along facial plane. These lesions can multiply after surgical excision despite negative margins after wide excision of the tumour. In a study conducted by Brendan Waters et al [9]**.** 33 recurrent low-grade myxofibrosarcomas 40% were in upper extremities. 40% in lower extremities and 20% in chest wall. Median size of lesion was 6.5 cm in their series, presenting as mass like or as multinodular and superficial spreading type. Our case was ill-defined nodular (Figure 5).

In this study, authors describe the margins as tail -like with well defined, tail like and infiltrating or tail-like well-defined and infiltrating. Lesions wereT1 hypointense to muscle, T2 hyperintense to muscle and enhancement greater than muscle. On CT the masses showed variable enhancement. Our case the enhancement was less than that of muscle. Ten patients with myxofibrosarcoma in whom MRI with contrast, with or without CT were analysed to determine and show patterns of recurrent low-grade myxofibrosarcoma. At imaging, the largest recurrences were mass-like in four patients (40%), were multinodular in two (20%), and had a superficial spreading configuration in one (10%); recurrences had both mass-like and multinodular configurations in three patients. Tumor margins were well defined in four patients (40%). In six patients (60%), the largest recurrence showed more than one type of margin: tail-like and infiltrative in three (30%), tail-like and well-defined in two (20%); and tail-like, well-defined, and infiltrative in one (10%). Perilesional edema pattern was seen in three (30%) patients. In two of the five patients who underwent contrast-enhanced CT, the largest recurrence enhanced more than muscle in the periphery of the lesion, but it was less than muscle in the centre. In two other patients, the entire lesion enhanced more than muscle; and in the fifth patient, the lesion showed CT attenuation slightly less than that of muscle.

Spindle Cell Sarcoma

Spindle cell sarcoma is rare subtype of undifferentiated / unclassified soft tissue that lack a specific line of differentiation. Histomorphologically they may be pleomorphic, round cell, and spindle cell variants. Spindle cell sarcoma occurs in any age and sex. Usually, they present late, and diagnosis is delayed. Metastases and recurrence are common. There were 3 cases of spindle cell sarcoma in our series and all the cases were given a diagnosis of mesenchymal tumor and on HPE the diagnosis is spindle cell neoplasm (Figures-6,7,8). Two cases were large than 5cm and one case was small in size. These Tumors are usually sharply mariginated, heterogenous in tissue attenuation or in signal intensity. Calcification may be observed, but necrosis /cavitation are rare. Tumors are highly vascular with evidence of feeding artery may be visualised in CT MR angiography. They show avid enhancement on CT or MRI. Haemorrhage is not uncommon. Mostly these tumors are difficult to differentiate from other sarcomas [10]. One of our cases had well defined margin and other two had ill-defined border, it was heterogenous in signal intensity in one.

V. Conclusion

Precise diagnosis of various soft tissue tumours based on imaging is difficult. Myeloid sarcoma, myxofibrosarcoma, and spindle cell sarcoma are rare tumours. Imaging diagnosis of DFSP can be to some extent. History, imaging, and biopsy are necessary to reach a final diagnosis in suspected soft-tissue sarcomas of the chest wall.

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Legends

Figure- 1: 36/M -Dermatofibrosarcoma of chest wall appearing heterogenous on T1W and T2W images involving skin and subcutaneous tissue. The gross appearance is similar to the MRI images.

Figure-2: Case of dermatofibrosarcoma recurrence in 47/M. Small isodense nodule on CT appearing well defined hypoechoic on USG.

Figure- 3: 34/F case of DFSP of chest wall showing lobulated isodense mass in subcutaneous plane infiltrating pectoralis muscle on CT and lesion is hypoechoic on USG having internal vascularity. It was diagnosed as deep fibromatosis on imaging and final diagnosis was on HPE

Figure- 4: 33/M -case of myeloid leukaemia having a granulocytic sarcoma deposit in right deltoid. On CT there is focal bulge in deltoid with loss of architecture. It is isodense to muscle; but on USG the mass appears to be mildly hypoechoic with internal vascularity.

Figure- 5: 71/M Case of myxofibrosarcoma. CECT reveals Ill-defined heterogeneously enhancing hypodense lesion in the subcutaneous plane infiltrating infraspinatus and teres minor muscles on dorsal aspect of left scapular region – s/o Recurrence. On USG the lesion is heteroechoic and vascular on doppler.

Figure- 6: 12/F -well-defined heteroechoic SOL with vascularity increased on USG and appear hyperintense on T2W and STIR images. SPINDLE CELL TUMOR

Figure- 7: 22/M- Spindle cell neoplasm with myofibroblastic differentiation in right posterior chest wall. T1 isointense, T2 hyperintense lesion on MRI and it is homogenously isoechoic with mildly vascular.

Figure- 8: 41/M- Soft tissue sarcoma of trapezius muscle appears homogenous on NECT an highly vascular on USG

Figure-1

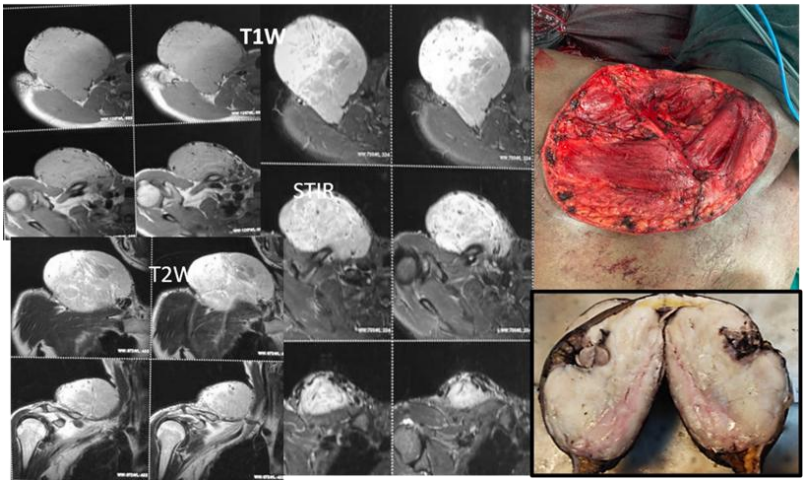
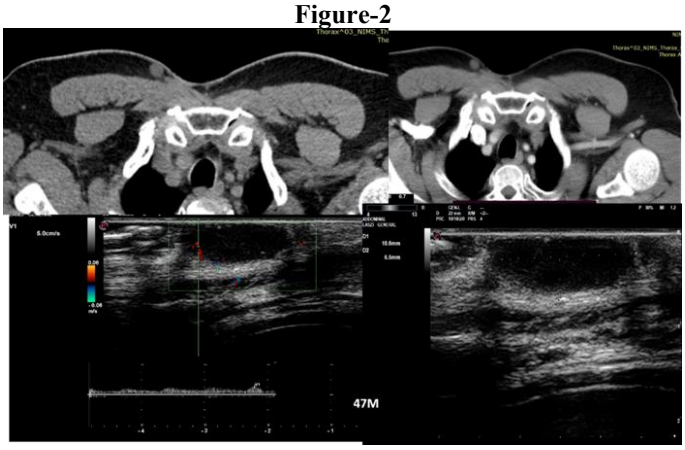


Figure-2



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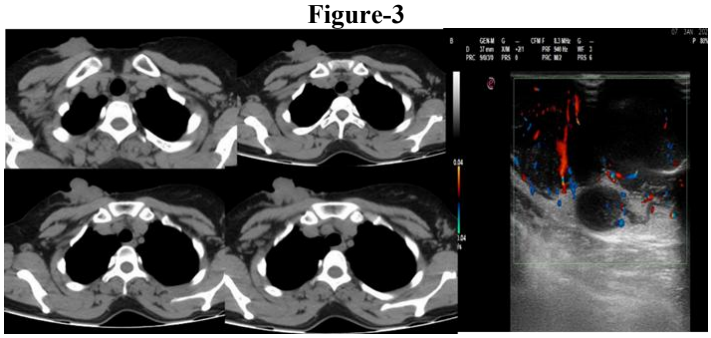

Figure-4

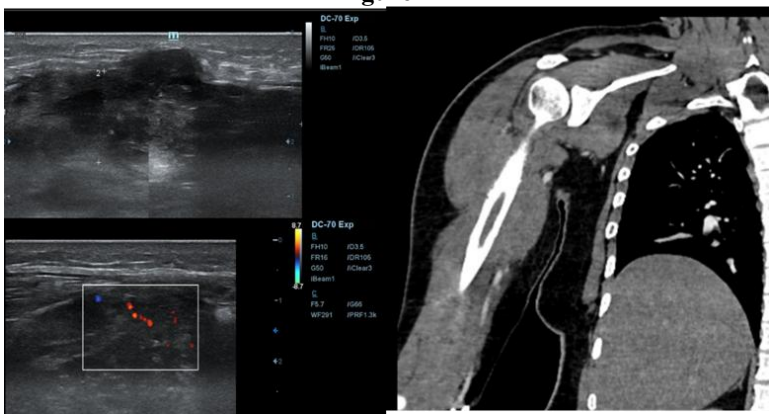

Figure-5

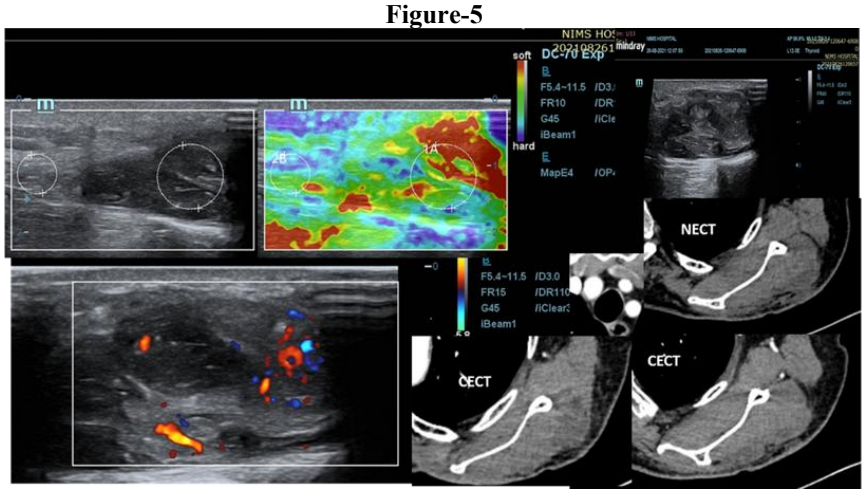

Figure-6

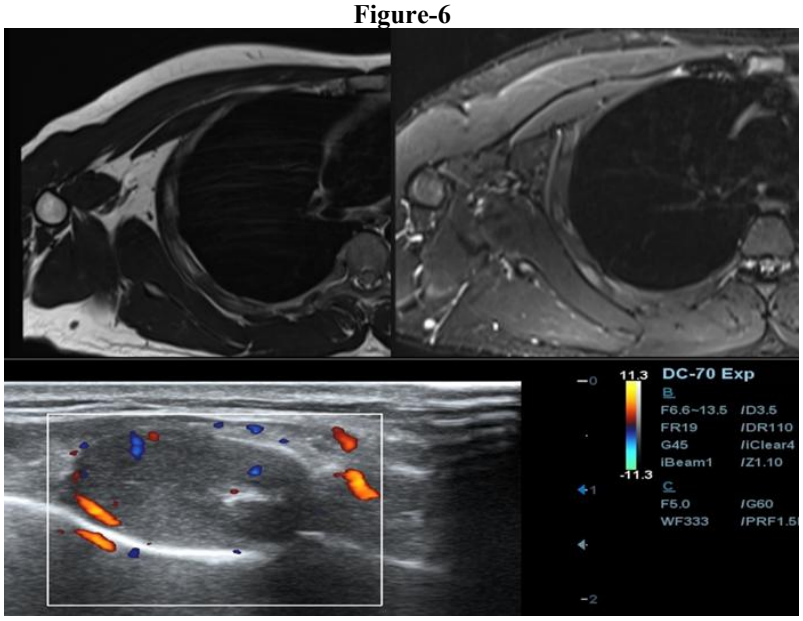

Figure-7

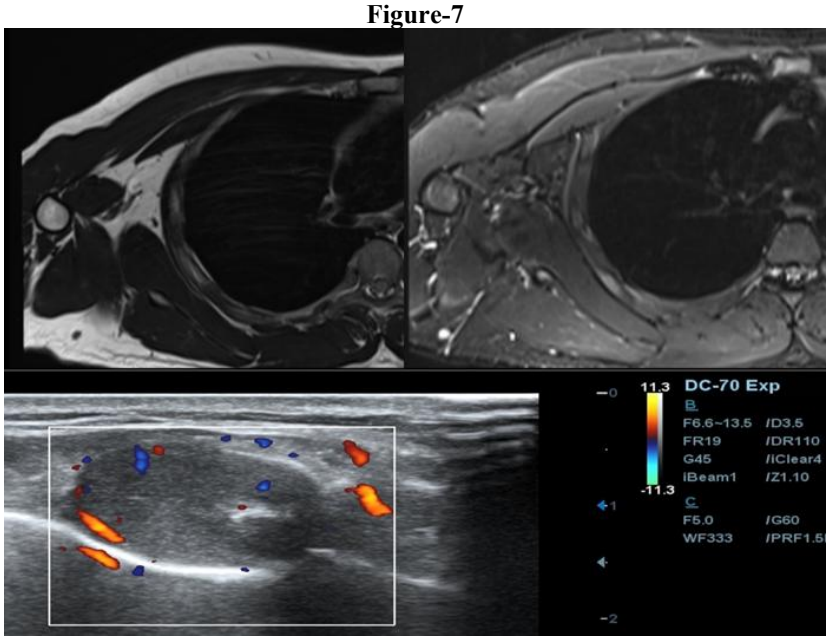

Figure-8