The spectrum of imaging findings in primary chest wall tumors.

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Aims and objectives

1. Describe common primary chest wall tumors in terms of location, origin, tissue components, and clinical features.

2. Identify chest wall tumors on the basis of their chest X-ray (CXR) appearances and CT imaging.

3. Discuss imaging findings facilitating the differential diagnoses of bone and soft-tissue tumors.

Methods and materials

Tumors of the chest wall can be benign or malignant and can take origin from any of its components.
The majority of malignant chest wall tumors are metastatic lesions from other organs or they are the result of direct invasion of a tumor from the lung parenchyma.

Radiographic tools for imaging these lesions consist mainly of plain radiographs, CT, and MRI. Radiologic evaluation of these tumors is essential in determining resectability and planning reconstruction.

Results

Primary chest wall tumor is rare and represents about 5% of all thoracic neoplasms [1, 2]. It encompasses tumors of various origins, including bone and cartilage, soft tissue such as muscle, vessel, nerve, and even some hematologic diseases [1, 3, 4]. Only 8% of primary bone tumors occur in the chest wall. The clinical presentation of primary chest wall tumor is nonspecific. The most frequent symptoms in patients with primary chest wall

tumors were palpable mass and pain. Neurologic symptoms such as muscle weakness and brachial plexus neuropathy have also been reported [5, 6].

Imaging studies, including CXR and CT, are useful in determining the extent of tumor invasion, surgical treatment planning, and follow-up evaluation of patients with chest wall tumors. However, imaging features of chest wall tumors are nonspecific [7]. Soft tissue mass, bone destruction, calcification, and bone deformity are frequently described, but are not diagnostic of malignancy. It is sometimes difficult to make an accurate diagnosis before histologic examination.

I. Bone Tumors

Primary tumors of bony skeleton of chest wall are uncommon, comprising 5-10% of all bony tumors. Malignant neoplasms are significantly more common than benign ones are [8].

Approximately 95% of these primary bone tumors are located in ribs, with most of the remainder in the sternum [9]. The benign rib tumors include osteochondroma, chondroma, fibrous dysplasia, and histiocytosis X. Osteochondroma is the most common benign bone neoplasm. The neoplasm usually begins in childhood and continues to grow until skeletal maturity.

Chondrosarcoma is the most common primary malignant bone tumor of the chest wall and accounts for 33% of all primary rib tumors, with myeloma being the second most common. Ewing sarcoma is the most common primary bone tumor in children [10, 11]. Fibrous dysplasia is the most common benign tumorous condition of the osseous chest wall, accounting for approximately 30% of these tumors [12].

1. Malignant Bone Tumors

Traditionally, bone tumors that demonstrate extensive cortical destruction and extraosseous soft-tissue mass formation have been categorized as malignant. Bone destruction is better depicted with CT, whereas extraosseous mass formation is better visualized at MR imaging [13].

1.1 Chondrosarcoma Fig. 1 on page 7: is the most common malignant lesion arising from the bone, most frequently from the anterior portion of the ribs and less frequently from the sternum, scapula, or clavicle [14]. Chondrosarcomas frequently appear as a large, lobulated mass arising from a rib with scattered calcifications consistent with a bony matrix [15]. These lesions might be similar radiographically with their benign counterparts, enchondromas, osteochondromas, and osteoblastomas, therefore necessitating tissue biopsy for diagnosis. The size of the lesion can be used as a predictor of malignancy; lesions larger than 4 cm are considered to be malignant [15, 16, 17].

MR imaging sometimes allows further analysis of tissue contents. Chondrosarcomas may have high signal intensity on T2-weighted images, reflecting a hyaline cartilage matrix of uniform composition with high water content. Contrast enhancement with a peripheral and septal pattern is often present.Calcifications in tumors typically appear assignal voids with all MR pulse sequences [17].

1.2 Osteosarcomas :are true malignant bony primary tumors and usually arise from a rib. These tumors carry a worse prognosis and have similar radiologic findings to chondrosarcomas. In osteosarcoma, mineralized portions of tumors have low signal intensity on T1- and T2-weighted images, whereas nonmineralized areas and soft-tissue tumors have high signal intensity on T2-weighted images [17].

1.3 Myeloma Fig. 2 on page 8: Multiple myeloma is the most common primary malignant neoplasm of the skeletal system. Radiologically, multiple destructive lytic lesions of the skeleton, as well as severe demineralization, characterize multiple myeloma. Almost 80% of patients with multiple myeloma will have radiological evidence of skeletal involvement on the skeletal survey most commonly effecting the following sites: vertebrae in 66%, ribs in 45%, skull in 40%, shoulder in 40%, pelvis in 30%, and long bones in 25% [18].

2. Benign Bone Tumors

The benign rib tumors include osteochondroma, chondroma, fibrous dysplasia, and histiocytosis X. Chondroblastoma, aneurysmal bone cyst (ABC), and giant cell tumor (GCT) are rare [19]. Osteochondroma, chondroma, and fibrous dysplasia constitute 60-70% of all benign chest wall tumors [20].

Radiographic evaluation with chest radiographs and CT scan is essential. A rapid increase in tumor size, cortical destruction, involvement of the surrounding tissues, and metastases suggest malignancy [21].

2.1 Fibrous dysplasia Fig. 3 on page 8: is the most common benign tumor arising from bone. These tumors account for approximately 30% of benign tumors of the chest wall [15, 16]. These fibrous tumors are slow-growing and are usually seen in the in the lateral or posterior arc of a rib. The tumor progresses by filling in the medullary cavity with fibrous

tissue, which can be demonstrated on CT or MRI [16]. The usual radiologic finding is an expanding lytic lesion in one of the ribs with a ground-glass appearance [15].

2.2 Chondromas Fig. 4 on page 9: Constitute 15% of benign neoplasms of the rib cage.usually arises from cartilaginous tissue at the costochondral junction[19]. CT reveals a slow-growing, lobulated, well-demarcated osteolytic lesion with or without bulging of cortical bone. The differentiation between chondroma and chondrosarcoma is impossible on clinical and radiographic findings.

2.3 Osteochondroma: Osteochondroma of the rib is exceedingly rare. They may present as a swelling in the chest wall or as an incidental finding on the chest radiograph [22]. Osteochondromas have a characteristic radiographic appearance, a sessile or stalked exostosis whose pedicle merges with the adjacent cortex, a peripheral rim of calcifications and stippled calcification within the tumour mass. MR imaging is the most accurate technique for demonstrating a cartilage cap and for evaluating changes in involved adjacent structures. Cartilage caps have high signal intensity on T2-weighted images, as do the cartilaginous tissues of other tumors, and cartilage cap thickness is an important indicator of osteochondroma-to-sarcoma transformation [13].

2.4 Langerhans cell histiocytosis of the rib: (LCH) in the ribs is very rare and difficult to diagnose using CT or PET. Tumor biopsy or resection is needed to diagnose LCH [23].

II. Soft-Tissue Tumors

In adults, the most common benign soft-tissue neoplasm is lipoma, and the most common malignant neoplasm is undifferentiated pleomorphic sarcoma (UPS), a term that is used interchangeably with *malignant fibrous histiocytoma*. In children, rhabdomyosarcoma and primitive neuroectodermal tumor (Askin tumor) are the most common malignant soft-tissue tumors [13].

1. Adipocytic Tumors

1.1 Lipoma: is the most common benign soft tissue lesion of the chest wall. These lesions can have intrathoracic and extrathoracic components, a dumbbell-shaped appearance, and tissue density consistent with fat, which makes these lesions easy to identify using CT or MRI [17]. On gadolinium-enhanced MR images, lipomas typically do not enhance. However, septa less than 2 mm in thickness are often visible and may enhance mildly [13].

1.2 Liposarcoma: is the second most common type of soft-tissue sarcoma, accounting for 10%-35% of these lesions. The World Health Organization has categorized soft-tissue liposarcomas into five distinct histologic subtypes: well differentiated, dedifferentiated, myxoid, pleomorphic, and mixed type. Well-differentiated liposarcomas frequently demonstrate a diagnostic appearance on computed tomographic (CT) or magnetic resonance (MR) images, with a largely lipomatous mass (>75% of the lesion) and non

lipomatous components in thick septa or focal nodules. The CT or MR imaging finding of a nodular dominant focus (>1 cm in size) of nonlipomatous tissue in a well-differentiated liposarcoma suggests dedifferentiated liposarcoma, and biopsy should be directed at the nonadipose component [24].

2. Neurogenic tumors: are often benign and usually appear radiographically

to originate from intercostal nerve roots. MRI is extremely useful in identifying lesions that encroach on the neural foramen. The benign group includes schwannomas and neurofibromas. The target sign consists of relatively low signal intensity at the center of a lesion together with high signal intensity at the periphery.has been reported to be present at T2-weighted imaging in 50%-70% of neurofibromas and 0%-54% of schwannomas [25].

3. Vascular Tumors: include benign tumors (eg, hemangioma, lymphangioma), tumors showing intermediate biologic behavior (eg,

hemangioendothelioma) and malignant tumors (eg, angiosarcoma) [13].

3.1 Hemangiomas Fig. 5 on page 10: are soft tissue masses that are occasionally found in the chest wall and are identified radiographically by the presence of phleboliths and vascular enhancement [17].

3.2 Angiosarcoma: is the most aggressive type of soft-tissue tumor arising from vascular tissue,and it usually affects adults. Chronic lymphedema is well known to predispose angiosarcoma,although only approximately 10% of lesions demonstrate this association[13].

4. Fibroblastic-Myofibroblastic Tumors Elastofibroma Dorsi Fig. 6 on page 10 : is a pseudotumor of the soft tissue with a female predilection, reported to occur in deep dorsal regions between the thoracic wall and the lower third of the scapula. These lesions are often asymptomatic, and bilaterality is common. At MR imaging, elastofibroma dorsi typically manifests as a mass with a signal intensity similar to that of muscle on both T1- and T2-weighted images, with interspersed streaks of fat in a fascicular pattern [26].

5. Others malignant tumors:

5.1 Fibrohistiocytoma: is the most common malignant soft tissue tumor.The tumor characteristically occurs in patients over age 50 and is rare in children. It usually presents a painless, slowly growing mass. It may be more common after chest wall irradiation, and spreads widely along fascial planes or into muscle, which explains the high recurrence rate after resection [13].

5.2 Rhabdomyosarcoma is the second most common chest wall soft tissue malignancy. Wide resection followed by radiotherapy and chemotherapy results in a 5-year survival of about 70% [13].

Images for this section:

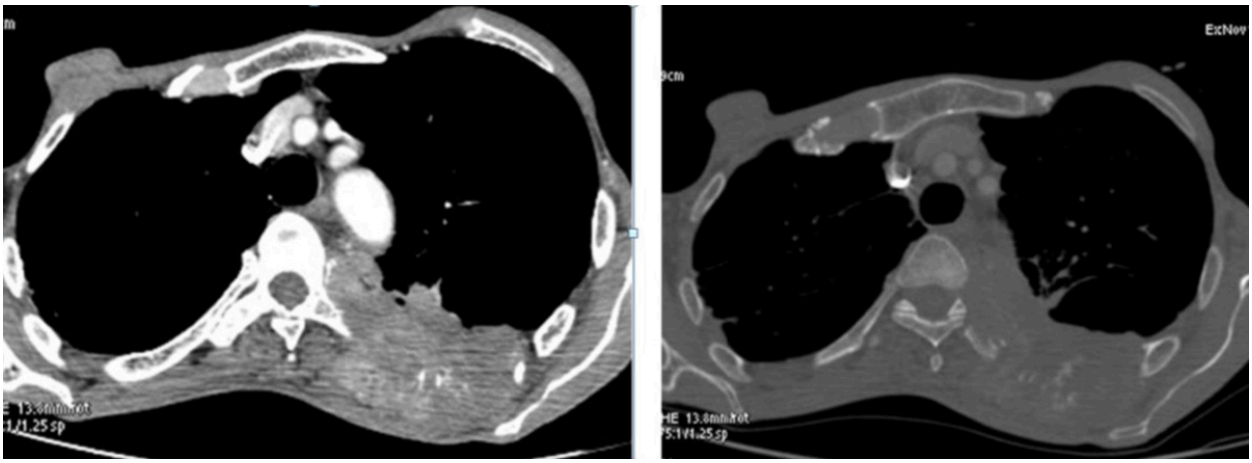

Fig. 1: CT imaging of a chondrosarcoma arising from the chest wall.

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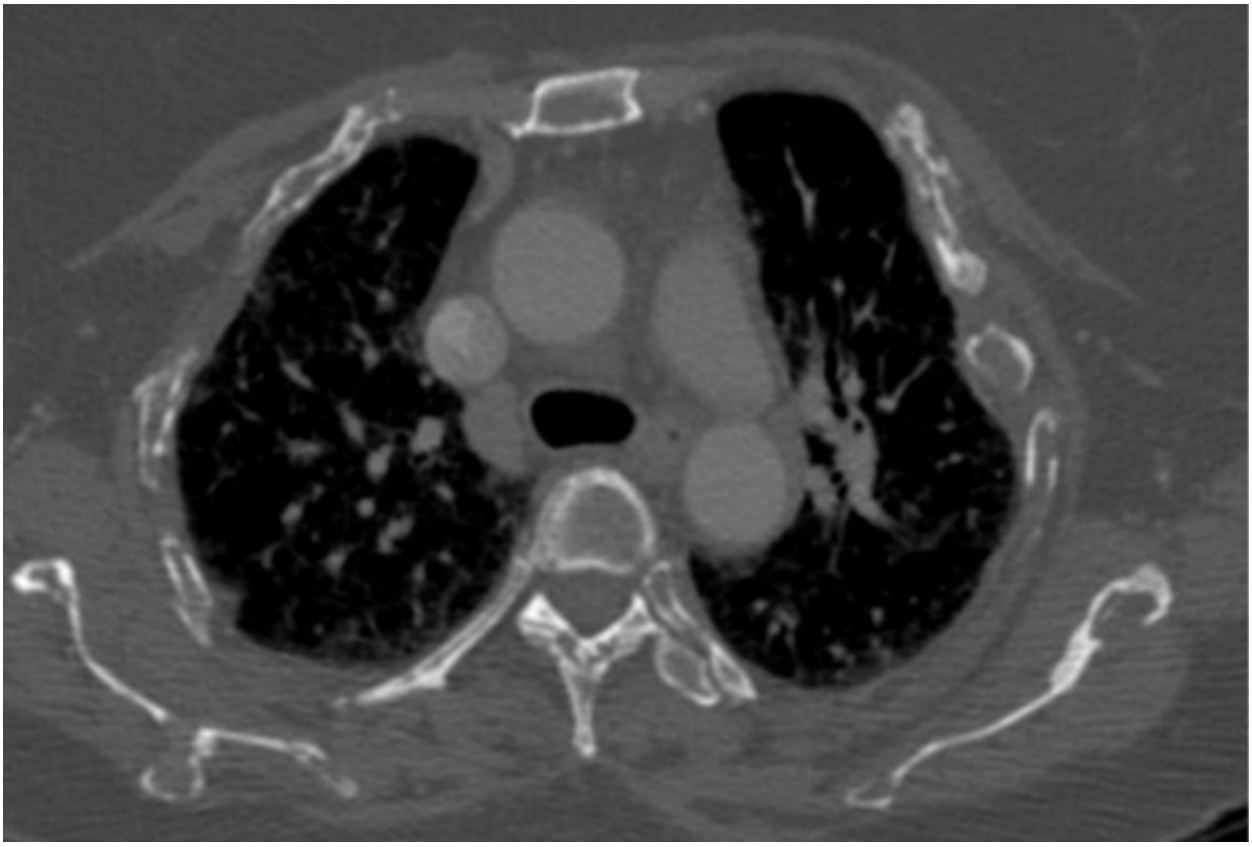

Fig. 2: CT imaging of multiple destructive lytic lesions of the skeleton in 50 years old women with myeloma.

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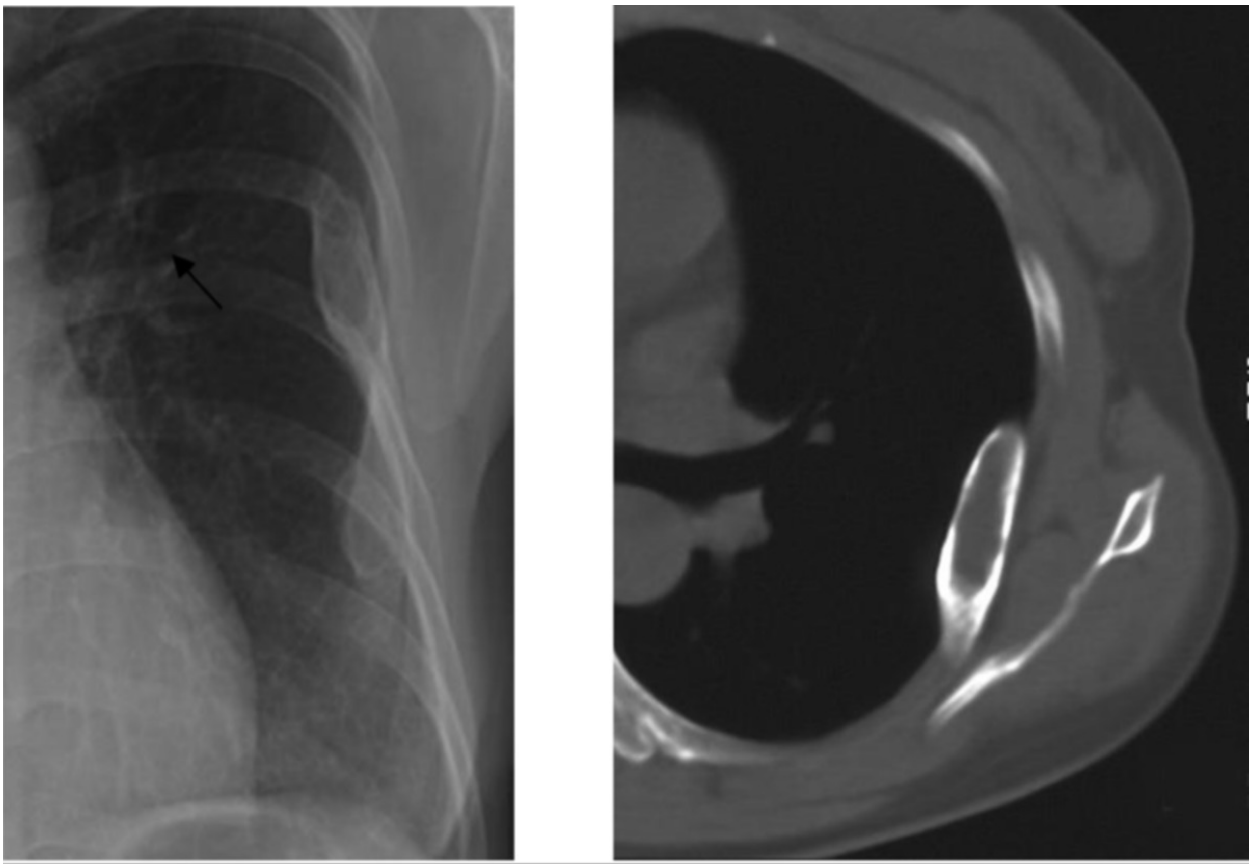

Fig. 3: Fibrous dysplasia of a rib in a 36-year old boy. Chest X ray and CT scan, bone window, shows a mass with expansile remodeling of the 7th rib.

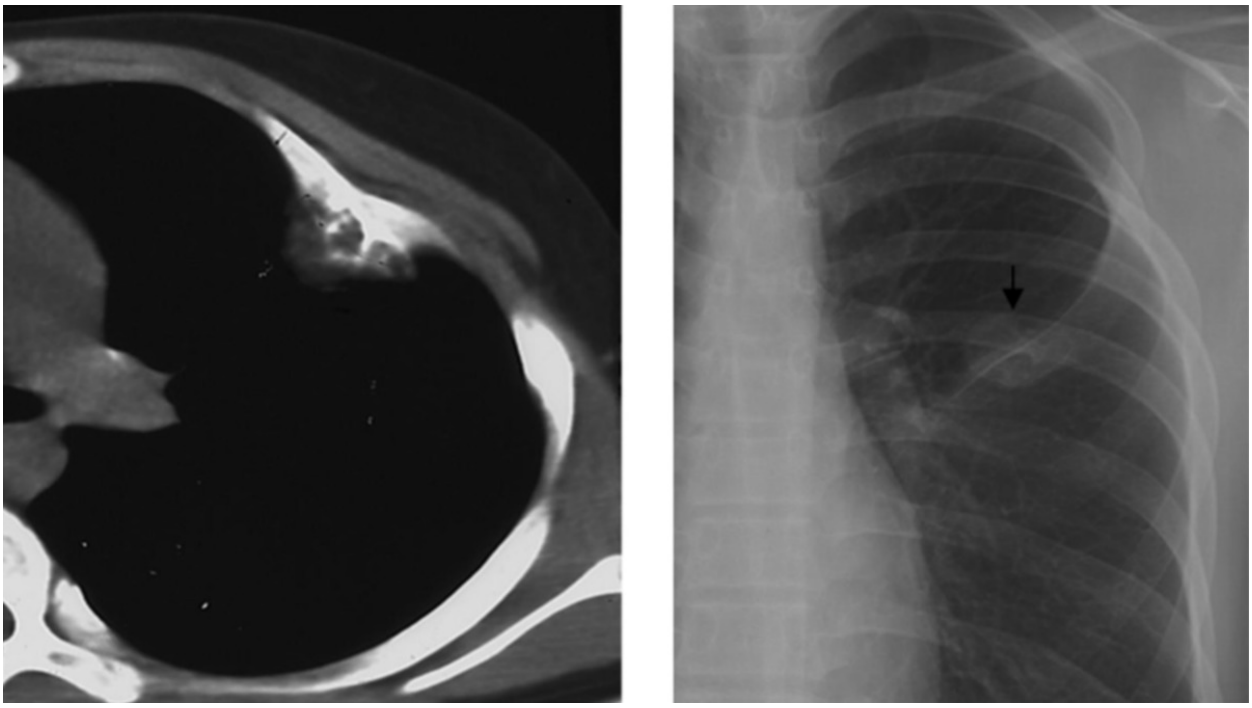

Fig. 4: Enchondroma of a rib in a 25-year old woman. Chest X ray and Contrast- CT scan shows a rib mass with cortical expansion at the costochondral junction. Note the punctate calcification, suggesting a chondroid matrix.

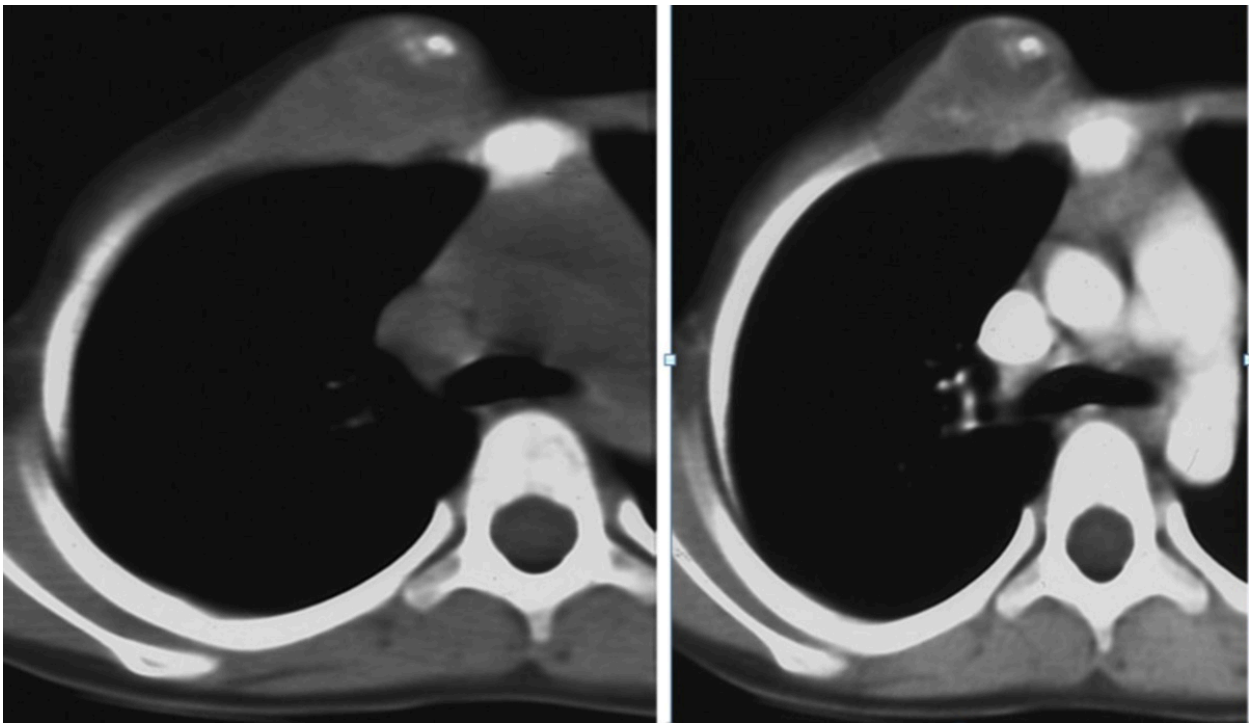

Fig. 5: CT imaging of a cavernous hemangioma containing a phlebolith.

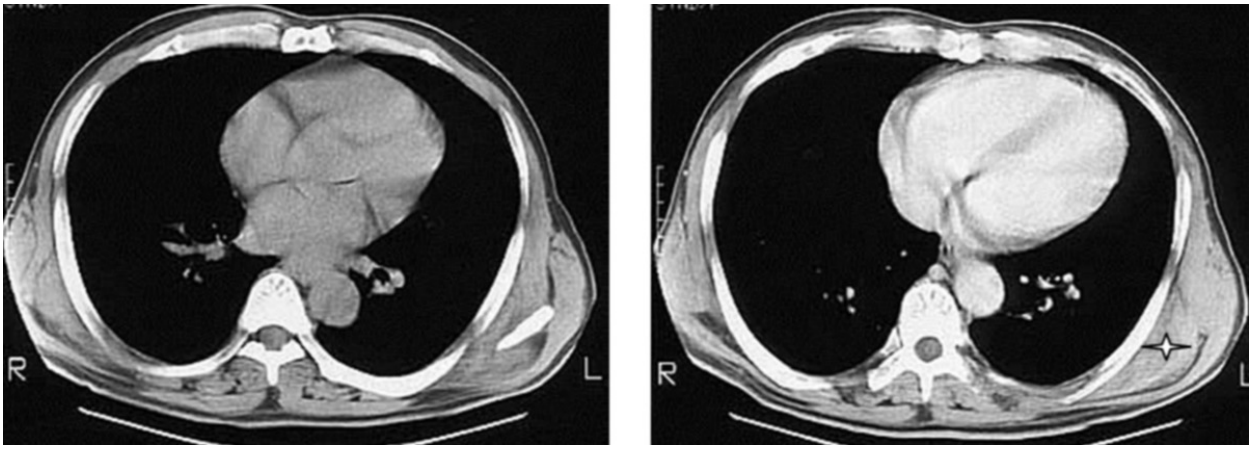

Fig. 6: Elastofibroma in a 56 -year old woman. (a) Axial CT-enhanced scan shows a crescent shaped soft-tissue lesion with diffuse streaky enhancement (arrows) in the deep dorsal region between the thoracic wall and the lower third of the scapula.

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Conclusion

Chest Wall Tumors is a difficult disease to characterize radiographically because of its diffuse nature and propensity to infiltrate between tissue planes. Surgical biopsy remains the gold standard for diagnosis.

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