Imaging characteristics of thoracic sarcomas - an illustration of interesting cases

Poster No.: C-1007
Congress: ECR 2013
Type: Educational Exhibit
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Keywords: Neoplasia, Education and training, Cancer, Diagnostic procedure, PET, MR, CT, Thorax, Oncology, Lung
DOI: 10.1594/ecr2013/C-1007

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Learning objectives

• To appreciate the wide spectrum of radiological appearances of thoracic sarcomas.

• To be able to differentiate thoracic sarcomas from primary lung cancer.

• To be able to offer a histological differential diagnosis based on certain imaging characteristics of thoracic sarcomas.

• To understand the key role the radiologist plays in diagnosis, evaluating disease extent and guiding management.
Background

Primary sarcomas of the thorax are an uncommon differential diagnosis for tumours involving the chest.

Primary thoracic sarcomas arise from soft tissues found in the mediastinum, lung, pleura and most commonly the chest wall.

The diagnosis can only be made after a process of exclusion of the more common malignancies such as primary lung cancer and metastasis.

The radiological differences are subtle and eventual diagnosis invariably lies in histological analysis. The principal role of radiology in the management of thoracic sarcomas is to evaluate the extent of disease and to provide guidance as to whether the tumour is resectable. However, imaging can also assist in narrowing the differential diagnosis by consideration of tumour location and other radiological features, which will be reviewed below.
Thoracic soft tissue sarcomas are often slow growing lesions and patients frequently present with chest wall pain. Plain radiographs commonly show a large solitary mass (Figure 1).

Although there is considerable overlap, it may be possible to favour a diagnosis of thoracic sarcoma over primary lung cancer or lung metastases, based on radiological appearances.

Primary thoracic sarcomas usually manifest as large, heterogeneous, poorly marginated and infiltrative masses (Figure 2) but can also appear as solitary pulmonary nodules or endobronchial tumours.

Metastases to the lungs is common with extra-thoracic sarcoma, in particular extremity sarcoma. Approximately 20% of patients with extremity sarcoma will have pulmonary metastasis at one point in their illness. Patients being considered for sarcoma metastatectomies may require sequential thoracic CTs to demonstrate that the number and growth rate of metastatic lesions is controlled.

The commonest sarcomas to metastasise to the lungs are osteosarcoma and Ewing's sarcoma\(^1\), which typically manifest as round smooth nodules. Their location is commonly subpleural or in the lung periphery and in the lower lobes (Figure 3). Metastases from osteosarcoma often demonstrate calcification (Figure 4). A lung primary can be differentiated, due to it usually appearing as a solitary mass, with a spiculated, ill defined contour.

Sarcoma metastases can appear very similar to metastases from other primaries such as testicular and renal cancer. If a patient has a known primary extrathoracic sarcoma, and lung nodules, as described above, are detected, they should be considered as being highly suspicious of metastases.

Imaging, in particular PET/CT, also has a role to play in assessing tumour response to chemo-radiotherapy (Figure 5). A well recognised post-treatment phenomenon to be aware of is that chemotherapy can sometimes lead to sarcoma metastases becoming cystic (Figure 6).
Resectability

The resectability of tumours which do not involve the chest wall, is generally determined by criteria similar to those used in cases of primary lung cancer and relates to the proximity of the lesion to the mediastinal structures (Figure 7).

For lesions involving the chest wall, important anatomical considerations are whether the tumour invades the brachial plexus\textsuperscript{2} or subclavian vessels (Figure 8).

For chest wall sarcomas it is important to evaluate whether the lesion invades the muscles of the chest wall. This has an important bearing on the need for chest wall reconstruction (Figures 9).

Similarly, lower thoracic sarcomas need to be scrutinized for sub-diaphragmatic invasion, for which MR can be useful (Figure 10).

Depicting invasion of the spinal canal with MR can also assist with surgical planning and the need for specialist spinal as well as thoracic surgical intervention (Figure 11).

Radiological correlates of histopathological types of thoracic sarcoma

The chest wall is the most common site for thoracic sarcomas and they can originate either in bone or soft tissue. The commonest osseous types are chondrosarcoma and Ewing’s and the most common soft tissue type is liposarcoma. Primary pulmonary sarcomas are rare and can mimic bronchogenic carcinomas so they are usually diagnosed only following a biopsy and exclusion of distant primary\textsuperscript{3}.

Sarcomas of the thorax, generally appear of heterogeneous appearance and enhancement on CT and MRI depending on the degree of necrosis, haemorrhage and cystic change. Clues to the histopathological origin of the tumour can sometimes be provided by the radiological appearances. A summary of the different features of thoracic sarcomas is summarised in the tables below.
<table>
<thead>
<tr>
<th>Type</th>
<th>Features</th>
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<tbody>
<tr>
<td>Osteosarcoma</td>
<td>Destructive bony tumour. Prominent matrix mineralisation.</td>
</tr>
<tr>
<td>Chondrosarcoma</td>
<td>Arising from chondral tissue. Cartilagenous matrix mineralisation (Figures 12 and 13).</td>
</tr>
<tr>
<td>Ewing's sarcoma</td>
<td>Large chest wall mass associated with bone destruction. No calcification. Extraosseous origin.</td>
</tr>
<tr>
<td>Synovial sarcoma</td>
<td>Arise from articular structures such as bursae and therefore found near joints (Figure 14).</td>
</tr>
<tr>
<td>Malignant schwannoma</td>
<td>Enlarging mass along the course of a nerve. Usually high signal on T2w (Figure 15).</td>
</tr>
<tr>
<td>Undifferentiated spindle cell sarcoma</td>
<td>Large mass with prominent haemorrhage and necrosis (Figure 16).</td>
</tr>
<tr>
<td>Liposarcoma</td>
<td>Adipose characteristics are maintained on CT and MRI unless poorly differentiated (Figure 17).</td>
</tr>
<tr>
<td>Malignant fibrous histiocytoma</td>
<td>Arises in muscle tissue. Matrix mineralisation is uncommon. Associated with previous radiation.</td>
</tr>
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**Table 1:** Features of primary thoracic sarcomas arising form the chest wall

**References:** Royal Brompton Hospital

<table>
<thead>
<tr>
<th>Type</th>
<th>Features</th>
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<tbody>
<tr>
<td>Leiomyosarcoma</td>
<td>Originates from smooth muscle cells usually in the pulmonary artery. Contrast enhancement can differentiate from filling defect.</td>
</tr>
<tr>
<td>Sarcomatoid mesothelioma</td>
<td>Pleural origin. Can present as diffuse pleural nodularity or solitary mass (Figure 18).</td>
</tr>
<tr>
<td>Angiosarcoma</td>
<td>Arises from vessels or the heart. Brisk contrast enhancement.</td>
</tr>
<tr>
<td>Rhabdomyosarcoma</td>
<td>Arise from muscle progenitor cells found in multiple tissues.</td>
</tr>
</tbody>
</table>
Table 2: Features of Primary thoracic sarcomas arising from an intra-thoracic location.

References: Royal Brompton Hospital
**Fig. 1:** Plain chest radiographs of primary chest wall sarcomas demonstrating; (a) Large left sided mass with osseous destruction of the posterior ribs and (b) posterior mediastinal mass with right sided rib scalloping, indicating the mass may have been present for some time.

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Fig. 2: Heavily calcified, irregular mass arising from the anterior chest wall with rib destruction. Pleural effusion suggests invasion of the pleura. The mass was a chondrosarcoma.

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Fig. 3: Axial CT of the chest demonstrating metastatic disease from a uterine leiomyosarcoma. Note the smooth, sharp margin of the lesions which are homogeneous and occur in the periphery of the lung parenchyma.

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Fig. 4: Axial CT scan demonstrating a calcified metastasis in the left lower lobe. The patient had a known lower limb osteosarcoma.

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**Fig. 5:** Coronal non contrast CT(a) showing a treated osteosarcoma of the right chest wall post chemotherapy and radiotherapy; Coronal PET(b) of the same lesion showing low grade uptake of FDG, suggesting partial treatment response. By comparison, on lung windows(c) coronal CT shows a calcified osteosarcoma lung metastasis with avid FDG uptake on PET(d).

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Fig. 6: Patient, treated for left sided pneumothorax, with bilateral lower lobe leiomyosarcoma metastases on chemotherapy. CT(a) demonstrates the cystic lesion in the right lower lobe. Plain radiograph(b) shows the cyst which has ruptured and contains a fluid level.

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Fig. 7: Axial T2w MRI showing a right anterior chest wall mass that is seen to invade not only the mediastinal fat but also the right atrium. Histological analysis showed synovial sarcoma.

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Fig. 8: Osteosarcoma invading the left lung apex and pleura but sparing the subclavian artery and the brachial plexus. Heavily calcified mass with destruction of the first rib. Calcification is demonstrated as areas of signal void on MRI. Sagital T2w(a) demonstrates the separation of the left subclavian artery(characterised by signal void(arrowhead)), from the mass, better than CT(b). (d)Note the interscalene fat pad in the normal position on the right(arrow), but displaced on the left(arrow). If this fat pad is obliterated, then invasion of the brachial plexus should be suspected[4]. This is easier demonstrated on MRI than CT(c).

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**Fig. 9:** Axial contrast enhanced CT(a) and T1w(b) of an undifferentiated spindle cell sarcoma. The mass is seen to invade the chest wall, necessitating reconstruction surgery, but the pectoralis muscle is spared. Note the fat plane between the mass and the muscle on CT and MRI(arrows).

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Fig. 10: Axial T2w MRI(a) and contrast enhanced CT(b) of a chondrosarcoma returning high T2w signal, arising from the right costal cartilage. Note the clearer depiction of the plane between the mass and the liver on MRI which demonstrates lack of liver invasion.

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Fig. 11: Chondrosarcoma. Axial T1w(a) and T2w(b) MRI demonstrates widening of the right neural foramen and clear invasion of the spinal canal. Note the scalloping that the mass is causing to the vertebral body.

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Fig. 12: Chondrosarcoma. Axial, contrast enhanced CT scan showing a heavily calcified chest wall mass causing cortical destruction of the adjacent rib.

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Fig. 13: Chondrosarcoma with mediastinal and intraspinal extension. Axial, non contrast (5mm maximum intensity projection) CT demonstrating typical "arc like" calcification of chondrosarcoma.

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Fig. 14: Synovial sarcoma. Axial, contrast enhanced CT scan showing a chest wall mass without bony destruction or calcification arising in close proximity to the costovertebral articulation.

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Fig. 15: Malignant schwannoma. Large mass arising from the rib usually high signal in T2w images (b) and low signal on T1w (a) MRI imaging due to myxoid tissue composition. Often demonstrates dots of low signal (arrows) on T2w imaging which represents collagen and condensed Schwann cells.

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Fig. 16: Undifferentiated spindle cell sarcoma. Coronal, contrast enhanced CT scan showing a large, extra pulmonary, left sided thoracic mass.

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Fig. 17: Liposarcoma. (a), Axial post gadolinium T1w fat suppressed images showing a posterior chest wall mass with signal suppression and enhancing septations. (b) T2w axial imaging showing high signal return from the mass. (c) Axial, contrast enhanced CT scan showing the low attenuation fatty mass with septations. Biopsy showed well differentiated liposarcoma. The presence of thick septae, nodules and large mass size favours liposarcoma over lipoma. Note the intercostal chest wall invasion(arrow).

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Fig. 18: Axial post contrast, lung window, CT scan slice showing a soft tissue mass arising from the pleura. Biopsy showed sarcomatoid mesothelioma. These tumours can mimic malignant mesothelioma or present as a solitary mass.

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Conclusion

- Thoracic sarcomas encompass a wide range of tumour subtypes and subsequently have a wide range of clinical presentations and imaging manifestations.

- Occasionally they can mimic other malignancies and pose an even greater diagnostic challenge.

- Key interpretation steps include ascertaining whether the tumour arises from the chest wall and what its composition is - features that could help to determine a histological subtype.

- The radiologist plays an important role, not only in diagnosis but to guide management.
References

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