Don’t forget chest wall tumors in the periphery

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Learning objectives

• Identify the imaging signs that are most useful for localizing and characterizing tumors of the chest wall.
• Describe the characteristic imaging findings in the most prevalent benign and malignant chest wall tumors.
Background

Primary chest wall tumors are a heterogenous group of neoplasms arising from bone, soft tissue, or cartilage of the chest wall. They are rare tumors, with an incidence of 2% of the population and represent approximately 5% of all thoracic neoplasms. Although primary chest wall tumors are diagnosed in every age group, they are more likely malignant in the extremes of age: in the young and the elderly.

Approximately 50% to 80% of chest wall tumors are malignant. Of these malignant tumors, approximately 55% arise from bone or cartilage and 45% from soft tissue. Overall 5-year survival after resection of primary chest wall neoplasms is approximately 60%. Recurrence can occur in up to 50% of patients, with a resultant 5-year survival of 17%.

Both can have vascular, muscle, nerve, cartilage, skin and fat origins. Among the malignant ones, there are still tumors that do not originate from any of these categories, such as Ewing sarcoma and synovial sarcoma. Other malignant tumors are more commonly found, such as myeloma multiple or solitary, soft tissue sarcoma, neuroblastoma, chondrosarcoma.

Among the benign ones, we can cite the example of the elastofibroma, neurofibroma, ganglioneuroma, schwanoma osteochondroma, haemangiopericytoma and lipoma.

CLINICAL PRESENTATION

Patients often present with a palpable enlarging mass. Less commonly (20%), asymptomatic patients are diagnosed due to an incidental finding on imaging as part of screening or for investigation of an unrelated condition.

When symptomatic, pain is the most frequent complaint. Approximately two-thirds of benign tumors will become painful, and nearly all malignant tumors will eventually cause pain, which can be a result of periosteal or neural invasion.

Pain is often vague and diffuse, affecting a particular region of the chest, and it is often attributed to a musculoskeletal cause, such as arthritis or recent trauma. Patients can also present with muscle weakness and atrophy of their upper extremities from compression of a tumor on the brachial plexus.

Due to the rarity of chest wall tumors, the time between onset of symptoms and diagnosis is often long.
Rapid increase in tumor size, involvement of surrounding tissues, and cortical destruction suggests malignancy, although they are not pathognomonic. Similarly, fixation to underlying tissues is not helpful in diagnosing malignant lesions, because it can be found in both malignant and benign conditions. There are no specific signs or symptoms that distinguish between benign and malignant lesions.

DIAGNOSIS

Imaging should be interpreted based on the location of the lesion and its size, the lesion’s effect on the bone, the bone’s response to the tumor, characteristics and composition of the tumor’s matrix and cortex, and any evidence of a soft tissue mass.

Chest x-rays (CXR), often the first imaging technique used in the diagnosis of chest wall tumors, can characterize the mass and demonstrate pulmonary. Additionally, some tumors are incidentally found on CXR. Although inadequate as the sole imaging modality in the current era of advanced imaging techniques, CXR can still provide useful information about tumor. Size, location, calcification, ossification, and bony involvement can all be ascertained.

CT is more sensitive than CXR in visualizing calcification and cortical destruction. It provides excellent information on size, location, bony involvement, and local infiltration of the tumor. Additionally, pulmonary metastases, if present, can be visualized. Tumor vascularity can also be ascertained when intravenous contrast is used. CT is also vital to operative planning and monitoring response to chemotherapy or radiation.

Magnetic resonance (MR) imaging is the preferred modality for the evaluation of chest wall tumors. The superior spatial resolution afforded by MR imaging with the administration of contrast material often enables accurate characterization of the tumor tissue and extent, including differentiation from adjacent areas of inflammation. Meticulous attention to technique is necessary for optimal MR imaging. Standard spin-echo and fast spin-echo sequences are satisfactory for most evaluations, but the use of peripheral cardiac gating and respiratory compensation can reduce motion artifacts that often degrade MR images of the thorax. Prone positioning of the patient can lessen the occurrence of respiratory artifacts on images of anterior chest wall tumors. Surface coils are useful for obtaining detailed images of superficial chest wall lesions, whereas a dedicated torso coil should be used to optimize image quality for tumors with greater intrathoracic extent.
Imaging findings OR Procedure details

- MALIGNANT TUMORS

1. Ewing sarcoma family of tumors (malignant small round cell tumors)

The Ewing family of tumours includes Ewings tumor of bone, extra-osseous Ewings, primitive neuroectodermal tumours (PNETs) and Askin tumor. All belong to the group of "small round blue cell tumours of childhood" and account for 4% of childhood malignancy. Askin tumor is a sub-group of these tumors with an exclusively thoracopulmonary origin.

This family is the most common malignant tumor of the chest wall in children and young adults, frequently occurring in the 2nd decade of life. Most have a common genetic translocation (t11:22) and are believed to arise from primitive cells of the neural crest. They most commonly arise in the pelvis, humerus, and femur of young males, but approximately 15% arise in the chest wall.

Tumors usually occur in the rib, scapula, clavicle, or sternum but occasionally have an extraskeletal site of origin. Expansion of a chest wall tumor may cause the lung to collapse, or the neoplasm may invade the lung. Although tumors generally tend to displace adjacent soft-tissue structures rather than invade or encase them, large tumors may directly infiltrate the surrounding structures.

Imaging demonstrates a large, noncalcified, soft tissue mass associated with bone destruction. CXR demonstrates a characteristic "onion-peel" appearance caused by subperiosteal bone formation. Additionally, there is bony destruction with lytic and blastic lesions and periosteal elevation. CT scan typically shows a large ill-defined tumor with cystic degeneration with soft tissue extension, which may or may not be accompanied by calcification (Fig 1, 2 and 3). Tumors usually have increased signal intensity on T1-weighted MRI and intermediate intensity on T2-weighted images (Fig 4). However, larger tumors have heterogeneous intensity. Tumors show marked enhancement after intravenous administration of contrast material.

2. Synovial sarcoma

Synovial sarcomas are malignant soft-tissue tumours that occur primarily in young adults and are extremely rare in the chest wall. They constitute approximately 10% of all soft-tissue sarcomas, and arise de novo from mesenchymal tissue, which differentiates
sufficiently to have the histological appearance of synovium. The majority (80-95%) of tumours are reported in the extremities, with two-thirds being located in the lower limbs. Other sites of origin, although rare, include the head and neck, paravertebral region, chest and abdominal wall.

There is a wide age range at presentation, from childhood to late adulthood. Synovial sarcoma is most common in the 15- to 35-year-old age group. The patient typically presents with a slow-growing palpable mass, which may grow over weeks or months, thus simulating a benign lesion. These lesions are frequently painful.

Plain films can reveal a well-defined or lobulated soft tissue mass, with up to one-third of cases demonstrating punctate calcification, often in the periphery of the lesion.

CT scans typically show a soft-tissue mass with attenuation slightly higher than that of muscle and may show infiltration of adjacent structures (Fig 5 and 6). Cortical bone erosion or invasion is well depicted at CT, as are intratumoral calcifications, which are noted in 20%-30% of cases.

MRI is considered the modality of choice for the detection and staging of soft-tissue tumours. On T1-weighted MR images, most tumors show heterogeneous signal intensity that is predominantly equivalent to that of muscle. Small foci of high signal intensity on T1-weighted images, which are present in 45% of cases, indicate hemorrhage. Fluid-fluid levels can be striking and are seen in 15%-25% of patients. Findings of hemorrhage and fluid-fluid levels or high signal intensity on any MR image may be associated with a worse prognosis, because these tumors are usually large and extensively invasive of surrounding tissue. On T2-weighted images, marked heterogeneity is the rule, and various degrees of internal septation may be noted. These findings are especially characteristic of large tumors, 85% of which have heterogeneous signal intensity. A combination of three different signal intensity levels is present on T2-weighted images in 33% of cases: high signal intensity similar to that of fluid, intermediate signal intensity equal to or higher than that of fat, and low signal intensity close to that of fibrous tissue. This triple-signal-intensity pattern on T2-weighted images, when accompanied by small foci of high signal intensity on T1-weighted images, calcifications, and proximity to a joint, may indicate the diagnosis.

3. Soft-tissue sarcoma

Soft-tissue sarcomas of the chest wall represent approximately 45% of primary malignant tumors of the chest wall and approximately 6% of soft-tissue sarcomas overall. Soft
tissue sarcomas of the chest come in a wide range of histologies, including malignant fibrous histiocytoma (MFH), angiosarcoma, leiomyosarcoma, spindle cell, liposarcoma, and undifferentiated sarcomas. These tumors can arise either de novo or in the setting of previous radiation therapy.

Most present with a painless chest wall mass (53%) and mean duration of symptoms is 12 months.

On CT, they usually have a heterogeneous appearance with varying levels of enhancement with intravenous contrast. Depending on the histology, sarcomas demonstrate varying degrees of calcification on CT scan (Fig 7 and 8). On T1-weighted MR images, the tumor has the same signal intensity as muscle and may be visible only because of its mass effect. On T2-weighted MR images, the tumor shows heterogeneous high signal intensity. Heterogeneous strong enhancement is observed after the administration of contrast material.

4. Solitary plasmacytoma

Solitary plasmacytoma is an isolated form of multiple myeloma, a tumor arising from plasma cells. It is considered one of the few "medical tumors" of the chest wall. Two-thirds of patients progress to develop generalized myeloma within 3 years of diagnosis, with poor prognosis, but the remainder of the patients achieve permanent cure.

Solitary myeloma is diagnosed in patients at a mean age of about 50 years, in contrast to multiple myeloma, in which the age range at manifestation is 50-70 years. Patients typically present with pain without a mass.

Solitary myeloma of bone manifests radiologically as a multicystic expansile mass (Fig 9) or purely osteolytic focus without expansion. Extraosseous solitary myeloma, which manifests as a nonspecific soft-tissue mass, progresses less frequently to multiple myeloma. At MR imaging, tumors show low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Contrast-enhanced MR imaging may be an effective means of monitoring the response to therapy.

5. Neuroblastoma
Neuroblastoma is a tumor of early childhood with the majority of tumours occurring before 5 years of age. Tumors arise in sympathetic nervous tissue anywhere in the body and usually present with a mass or bone pain. Neuroblastoma primarily involving the chest wall is very uncommon and usually represents a rib metastasis that has grown in situ.

Due to their neural origin, tumors show invasion through neural foramina giving a "dumbbell" appearance and may cause spinal cord compression (Fig 10, 11 and 12). Tumors show high signal on T2W with heterogeneous but marked contrast uptake. Central necrosis and pleural effusions may be present in large lesions.

6. Chondrosarcoma

Chondrosarcomas, the most common malignant primary tumors of the chest wall, usually occur in the anterior part of the wall, arising from the costochondral arches or sternum. Nineteen percent of chondrosarcomas occur in the ribs. Most of these are primary lesions, but 10% arise from preexisting benign tumors, such as chondromas, exostoses, or osteochondromas. Additionally, they can be associated with trauma.

Two peak periods of prevalence have been identified—the first, at less than 20 years of age, and the second, at more than 50 years of age. These tumors occur twice as often in men as in women, and they most frequently are found along the upper five ribs, adjacent to costal cartilage.

Bone destruction, irregular contours, and intratumoral mineralization are characteristic but variable features detected on chest radiographs.

Early-stage lesions demonstrate a thickened cortex whereas higher grade lesions usually have complete cortical destruction, along with a soft tissue mass. Alternatively, the growth of a benign cartilage tumor or expansion of their cartilage cap may suggest malignant degeneration.

CT is more sensitive than radiography and MR imaging for delineation of chondroid matrix calcifications. The better-differentiated chondrosarcomas appear on CT scans as well-defined, densely calcified soft-tissue masses, with foci of dense chondroid matrix calcification (Fig 13). T1-weighted MR images show lobulated masses with signal intensity similar to that of muscle, and T2-weighted MR images show masses with signal intensity equal to or greater than that of fat. Enhancement after administration of intravenous contrast material typically is heterogeneous, especially at the periphery.
7. Elastofibroma

Elastofibroma dorsi are muscular tumours of the posterior chest wall. They characteristically occur at the inferior angle of the scapula and have a reported prevalence of 2% in the elderly population. The lesions are periscapular in 99% of reported cases and are reportedly bilateral in 10-66% of cases. Synchronous lesions in the infraolecranon region are also common.

Although the cause of these lesions is not clear, Jarvi and others thought of elastofibroma as a pseudotumor, reactive in nature, and attributable to mechanical friction of the scapula against the ribs during heavy manual labor. Elastofibroma is thought to be more common on the right side, presumably related to the side of the patient's dominant handedness, and is also thought to be more common in manual laborers. Elastofibroma is several times more common in women than in men.

On CT, elastofibroma is typically a poorly defined, inhomogeneous soft-tissue mass with attenuation similar to that of skeletal muscle, containing linear streaks of fat attenuation (Fig 14 and 15). On MR imaging, the appearance is similar-soft tissue with signal intensity similar to that of skeletal muscle on both T1- and T2-weighted images, interlaced with streaks of tissue with the signal intensity of fat. This appearance corresponds to areas of dense fibrous connective tissue interlaced with areas of fat. Heterogeneous enhancement following gadolinium administration can be seen. Deformation of adjacent bony structures has also been reported.

8. Benign Peripheral Nerve Tumors

a) Schwannomas

Chest wall schwannomas arise from spinal nerve roots and intercostal nerves and typically occur in patients between 20 and 50 years of age.
Radiographs do not usually depict small schwannomas, but bone erosion or scalloping can occasionally be seen.

Nonenhanced CT scans of schwannoma typically show a well-circumscribed homogeneous mass with attenuation slightly less than or equal to that of muscle (Fig 16). On CT scans acquired after contrast material administration, the attenuation of the mass is equal to or slightly greater than that of muscle, and any cystic or necrotic areas in the mass appear nonenhanced.

The signal intensity of schwannoma on T1-weighted MR images is equal to or slightly greater than that of muscle and on T2-weighted images is markedly greater, with increased contrast between the high-signal-intensity nerve sheath tumor, intermediate-signal-intensity fat, and low-signal-intensity muscle. The nerve from which the tumor originated can often be seen along one side of the mass. Small tumors tend to enhance brightly and uniformly after intravenous administration of contrast material, whereas the enhancement pattern of larger lesions may be more heterogeneous because of central cystic change. The presence of bone erosion without destruction indicates the benign nature and slow growth rate of this lesion.

b) Ganglioneuromas

Ganglioneuromas originate from the sympathetic ganglia in the chest wall. Although this tumor most often arises de novo in young adults, it also may occur as a maturation of neuroblastoma. The mass is composed of mature ganglion cells, Schwann cells, and nerve fibers, and it is often large and encapsulated, with delicate trabeculation.

Ganglioneuromas usually manifest radiologically as ovoid, sharply marginated paravertebral masses (Fig 17 and 18). Calcification occurs in 25% of cases. The tumor has either homogeneous or heterogeneous attenuation on CT images and homogeneous intermediate signal intensity on both T1- and T2-weighted MR images. Curvilinear bands of low signal intensity are seen on both T1- and T2-weighted images, giving the lesion a whorled appearance.

c) Neurofibroma
Neurofibromas originate from peripheral nerves and in up to 60% can be associated with neurofibromatosis type I, multiple plexiform neurofibromas, or multiple endocrine neoplasia. They are slow-growing masses, often occurring between the ages of 20 and 30 years.

Neurofibromas have a low muscle-like attenuation on non-contrast CT and show heterogeneous enhancement post contrast. They may demonstrate cystic degeneration, with a central zone that is more cellular and a peripheral more stromal zone, creating a target appearance on both T2-weighted images and gadolinium-enhanced MRI. These tumors can grow into the spinal canal in a dumbbell fashion.

9. Osteochondroma

Osteochondroma is a benign tumor consisting of bone and cartilage, but composes less than 10% of primary chest wall tumors. It is the most common benign bone tumor and accounts for nearly 50% of all benign rib tumors. Young men are most commonly affected, and men are affected 3 times more often than women.

The mass originates from the cortex of the rib, usually on the metaphysis. In the chest, osteochondromas are most commonly found at the costochondral junction. When the tumor grows outward, it presents as a painless mass, but is typically asymptomatic when it grows inward. Osteochondroma can result in complications, including fractures, bone deformities, and nerve compression. Multiple lesions can indicate familial osteochondromatosis. Malignant transformation can occur and should be suspected if there is pain, erosion of the bone, irregular calcifications, or thickening of the cartilage cap.

Most are round and measure less than 9 cm. The tumors are characteristically pedunculated osseous protuberances arising from the surface of the parent bone (Fig 19 and 20). Imaging is usually adequate for diagnosis, demonstrating punctate or flocculent calcifications with a mineralized hyaline cartilage cap best seen on CT (Fig 21, 22 and 23). The cortex and medullary space blend into the underlying bone, which is how a definitive diagnosis is made on CT or MRI.

10. Haemangiopericytoma
Haemangiopericytoma is a soft tissue tumor that rarely occurs in children. Two forms are described: an infantile form that has a relatively benign course and an adult form that is more aggressive. The infant form may be multifocal and may regress spontaneously. The adult form tends to metastasize and recur locally.

CT and MRI show a mass with vascular invasion and necrosis but appearances are non-specific (Fig 24, 25 and 26).

11. Lipoma

Lipomas are well-circumscribed encapsulated masses composed of adipocytes that differ very little from normal fatty tissue. They typically occur in patients who are 50-70 years of age, and they are most frequent in the obese. Most lipomas that originate in the chest wall are deep lipomas, which tend to be larger and less well circumscribed than superficial lesions.

On CT and MR images (Fig 27), lipomas generally appear to be internally homogeneous and do not enhance after intravenous contrast material administration. However, multiple thin septa often are present that appear slightly enhanced on CT scans and have low signal intensity on fat-suppressed T1-weighted MR images. The key to diagnosis is by the identification of fat within the lesion either by CT (attenuation of -100UH) or MRI using fat suppression (FS) techniques.

They can be difficult to differentiate from low-grade liposarcomas on imaging, but liposarcomas have heterogeneous enhancement and tend to be larger. Imaging features that favour a diagnosis of liposarcoma include size greater than 10 cm, thick internal septations, nodular non-adipose areas and lesions with less than 75% fat signal characteristics.
Fig. 0: PNET in a 14-year-old man. CT scan shows a large ill-defined and heterogeneous mass, associated a bone destruction.

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Fig. 0: PNET in a 14-year-old man. CT scan shows a large ill-defined mass fills the thoracic cavity and displaces the aortic arch.
**Fig. 0:** PNET in a 14-year-old man. Coronal reformatted CT scan shows a large ill-defined mass fills the thoracic cavity and displaces the mediastinum. There is a metastatic nodule at right upper lobe.

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Fig. 0: PNET in a 14-year-old man. Coronal reformatted image T2-weighted shows a large ill-defined mass fills the thoracic cavity.
**Fig. 0:** Synovial sarcoma in a 71-year-old woman. CT scan shows soft-tissue mass with attenuation slightly higher than that of muscle with infiltration of left hemithorax.
Fig. 0: Synovial sarcoma in a 71-year-old woman. Coronal reformatted CT scan shows soft-tissue mass with attenuation slightly higher than that of muscle with infiltration of left hemithorax.

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Fig. 0: Soft-tissue sarcoma in a 42-year-old woman. CT scan shows a large heterogeneously enhancing mass between the subcutaneous tissues and the deep layer of fat and muscle.

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Fig. 0: Solitary plasmacytoma in a 71-year-old woman. Contrast-enhanced CT scan at the level of the aortic root shows a soft-tissue mass that originates from a right rib.

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**Fig. 0:** Neuroblastoma in a 5-year-old man. CXR shows a left paravertebral mass at level of carina.

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**Fig. 0:** Neuroblastoma in a 5-year-old man. Contrast-enhanced coronal reformatted CT scan shows a left ill-defined paravertebral mass.

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Fig. 0: Neuroblastoma in a 5-year-old man. Contrast-enhanced CT scan at the level of the aortic arch shows a heterogeneous left paravertebral mass.

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**Fig. 0:** Chondrosarcomas in a 32-year-old woman. CT scans shows a well-defined, soft-tissue masses, with a subtle foci of dense chondoid matrix calcification.

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**Fig. 0:** Elastofibroma in a 63-year-old woman. CT scan shows a bilateral poorly defined, inhomogeneous soft-tissue mass with attenuation similar to that of skeletal muscle, containing linear streaks of fat attenuation at the inferior angle of the scapula.
Fig. 0: Elastofibroma in a 63-year-old woman. Coronal reformatted CT scan shows a bilateral poorly defined, inhomogeneous soft-tissue mass with attenuation similar to that of skeletal muscle, containing linear streaks of fat attenuation at the inferior angle of the scapula.

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Fig. 0: Schwannoma in a 35-year-old man. CT scan shows a well-circumscribed homogeneous mass with attenuation slightly less than or equal to that of muscle.
Fig. 0: Ganglioneuroma in a 4-year-old man. T2-weighted MR images shows a sharply marginated paravertebral masses with a intermediate signal intensity.

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Fig. 0: Ganglioneuroma in a 4-year-old man. Coronal reformatted T2-weighted MR images shows a sharply marginated paravertebral masses with a intermediate signal intensity.

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Fig. 0: Osteochondroma in a 27-year-old woman. CXR shows a extrapulmonar mass demonstrating calcification in the apex of left lung.

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**Fig. 0:** Osteochondroma in a 27-year-old woman. Nonenhanced CT scan shows a sessile osseous protuberances demonstrating punctate or flocculent calcifications with a mineralized hyaline cartilage cap in the second left rib.

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Fig. 0: Osteochondroma in a 14-year-old woman. Nonenhanced CT scan shows a pedunculated osseous protuberances arising from the surface of rib toward the heart.

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Fig. 0: Haemangiopericytoma in a 56-year-old woman. Contrast-enhanced CT scan shows a soft tissue mass invading a rib.

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Fig. 0: Haemangiopericytoma in a 56-year-old woman. Contrast-enhanced CT scan shows a soft tissue mass with necrosis.

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Fig. 0: Haemangiopericytoma in a 56-year-old woman. Contrast-enhanced CT scan shows a soft tissue mass invading a rib.

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Fig. 0: Lipoma in a 64-year-old woman. Nonenhanced CT scan at the level of the left inferior pulmonary vein shows a well-defined mass with the same attenuation as fat in the left part of the chest wall.

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Conclusion

Although the radiologic manifestations of benign and malignant chest wall tumors frequently overlap, differences in characteristic location and appearance occasionally allow a differential diagnosis to be made with confidence.
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References