Imaging of fibrous lesions of the thorax: radiologic-pathologic correlation

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Learning objectives

1. To describe the imaging features of the main fibrous lesions of the thorax (chest wall, pleura, mediastinum, and lung parenchyma)

2. To review the correlated radiologic and pathologic findings of the main fibrous thoracic lesions
Background

Histologically, fibrous lesions of the thorax share the presence of fibroblasts and/or fibrosis. Most of these lesions are benign, have a mesenchymal origin, and are relatively rare.

In this paper we review the imaging features (conventional radiographs, CT, MRI, PET/CT...) of fibrous thoracic lesions and correlate them with their pathologic findings.

A definitive preoperative diagnosis of some of these entities may not be exclusively based on cytologic or histologic findings, but it may require taking into consideration the radiological and clinical context of the case.
CHEST WALL

**DORSAL ELASTOFIBROMA**

Dorsal elastofibroma (DE) is a non-encapsulated mass-like lesion, consisting of a benign proliferation of fibrous tissue and elastic fibers with fatty tissue. The most frequent location is the subscapular region (up to 99% of cases), between the muscles and the posterior chest wall. Other less frequent locations are chest wall, deltoid, and ischial tuberosities.

Its prevalence is around 2%, is usually more common in women, with a ratio of 8:1 (mean age: 60-70 years). Up to 50% of the patients are asymptomatic.

Histologically, the tumor consists of hypocellular masses with a mixture of eosinophilic collagen and elastic fibers. Fig. 1 on page 17

Chest radiography is usually normal in most cases. On ultrasound (US) it commonly appears as an echogenic mass with straight or curvilinear hypoechoic bands corresponding to fat.

CT usually shows a well-defined soft-tissue mass with thin fat attenuation bands within it. After injection of an iodinated contrast agent, the density of the mass remains virtually unchanged. Fig. 2 on page 17

MRI is the imaging test of choice: it typically reveals a well-demarcated subscapular mass showing the characteristic alternating pattern of fibrous tissue and fatty tissue. On T1- and T2-weighted images the fibrous component exhibits low signal intensity (nearly identical to that generated by the muscle), whereas the fatty component appears as linear streaks of fat signal intensity. STIR images show a poorly defined mass. After contrast administration there is usually no pathological uptake however, some areas may show subtle enhancement. The differential diagnosis must be made with other fibrous tissue-containing tumors: neurofibroma, cicatricial fibroma, and malignant fibrous histiocytoma.

On PET/CT imaging DE typically shows minimal or, less frequently, moderate FDG uptake. The characteristic CT features of DE on the CT component of the PET/CT helps to differentiate this entity from other, more aggressive, tumors.

**FIBROUS DYSPLASIA**
Fibrous dysplasia is a disease in which bone marrow is normal replaced by fibrous tissue. It is responsible for 30% of benign tumors of the chest wall.

Although most fibrous dysplasias are monostotic, approximately 20%-30% of them are polyostotic. In the chest wall, the ribs are most commonly affected. Fibrous dysplasia tends to be asymptomatic but can produce focal deformity or pathologic fractures. Up to 1% of the lesions of fibrous dysplasia may undergo degeneration into malignant fibrous histiocytoma, fibrosarcoma, osteosarcoma, chondrosarcoma.

Typical microscopic findings include irregular spindles of woven bone, usually nonmineralized, scattered throughout a fibrocellular matrix. Fig. 3 on page 18

Radiographs characteristically show unilateral fusiform enlargement and deformity with cortical thickening and increased trabeculation of one or more ribs. Amorphous or irregular calcification is often seen in the lesion on CT scans. Fig. 4 on page 19

On MRI, lesions typically display intermediate signal intensity on T1-weighted and proton density images. On T2-weighted images, the appearance of fibrous dysplasia varies from low to intermediate signal intensity depending on the presence of intra-lesional bony trabeculae. After the administration of intravenous contrast, most of the lesions enhance.

**MALIGNANT FIBROUS HISTIOCYTOMA**

Malignant fibrous histiocytoma (MFH) is an aggressive type of sarcoma that can arise both from soft tissues and bone.

MFHs occur principally as a mass of the extremities, abdominal cavity, or retroperitoneum in adults. However, they only rarely occur in the chest wall. It’s the most common sarcoma of adulthood (20-30%), with a peak incidence is in the fifth decade of life. This tumor can be associated with previous bone lesions, including Paget disease and bone infarcts, and is the most common sarcoma to develop after irradiation.

Initially described as a tumor of histiocyte origin, currently the histogenesis of this entity is not clear. MFH manifests a broad range of histologic appearances with four subtypes (storiform-pleomorphic, myxoid, giant cell, and inflammatory). The storiform-pleomorphic subtype is the most common form, accounting seen as a soft-tissue heterogeneous mass that exhibits intense enhancement following contrast for more than two-thirds of all cases. Fig. 5 on page 20

On CT studies, MFH is usually administration Myxoid MFH, the second most common subtype, typically shows a characteristic CT appearance of central low attenuation (due
to the myxoid matrix at the center of the lesion) and nodular peripheral enhancement of the more cellular tumor components.

MRI is the imaging method of choice for MFH because of its ability to provide superior contrast between tumor and muscle, and excellent definition of surrounding anatomy. On T1- and T2-weighted MR images, MFH may appear either homogeneous or heterogeneous. On T1-weighted images, most tumors have a signal intensity equal to that of muscle, whereas and on T2-weighted images, MFHs usually exhibit a signal intensity equal to or greater than that of fat. MFH usually enhances intensely and heterogeneously after contrast administration.

PLEURA

SOLITARY FIBROUS TUMOR OF THE PLEURA

Solitary fibrous tumor of the pleura (SFTP) is an uncommon neoplasm of mesenchymal origin that can be benign or malignant. This tumor is more frequent in persons between the fifth and sixth decades of life and there is no significant sex predilection. The majority of SFTP are benign, but up to 15-20% have a malignant behaviour.

Histologically, SFTP are composed of spindle cells within a background of collagen stroma, often in a whorled pattern or patternless. These tumors are highly vascular and have a propensity to undergo myxoid degeneration. The diagnosis is confirmed by characteristic positive immunohistochemical staining. Fig. 7 on page 22

SFTP represent less than 5% of pleural masses, and malignancy is present in up to 35% of cases. Most of these tumors originate from the visceral pleura (70%). These lesions are often pedunculated with a fibrovascular stalk.

SFTP is usually an incidental finding at chest radiography or CT. CT can better depict the pleural origin of the lesions than radiography. The appearance in these studies is nonspecific and prompts further investigation.

On chest radiographs, SFTP appear as peripheral, smooth, well-demarcated soft-tissue density masses. Pedunculated tumors can present as mobile masses at serial examinations.

SFTP are often located in the periphery of the thorax or positioned along a fissure. SFTP can be hypodense or hyperdense with respect to muscle. The attenuation depends on the collagen content, (hyperdense lesions usually have abundant collagen). Calcifications and pleural effusions are uncommon findings in SFTP. The presence or absence of
calcifications is not a helpful distinguishing feature between benign and malignant SFTP. Fig. 8 on page 23

At MRI, SFTP are usually isointense on T1-weighted images and show a variable signal intensity on T2-weighted images, having what has been described as a "black and white" mixed pattern. The presence of rounded or linear low-intensity foci on both T1- and T2-weighted images is attributable to the collagen content, low cellularity, and associated reduced proton mobility. These are vascular tumors that are vigorously enhancing. This combination of features produces a "chocolate chip cookie" appearance.

At PET (PET/CT) benign tumors typically exhibit low-grade and relatively uniform FDG activity, whereas malignant SFTP tend to be strongly hypermetabolic and more heterogeneous

The imaging differential diagnosis of SFTP includes mesothelioma, lymphoma and metastasis.

**DESMOID TUMOR (FIBROMATOSIS)**

Desmoid tumors (DT), also called deep or aggressive fibromatosis, are uncommon mesenchymal neoplasms with a fibrotic bandlike consistency.

The incidence of these rare tumors has a slight female preponderance and peak incidence in the third and fourth decades. Trauma, prior surgery, pregnancy, and oral contraceptive use are considered as risk factors. Estrogen has been implicated as a growth factor.

DT are locally aggressive lesions without potential for distant metastases.

Microscopically, DT consist of poorly defined fascicles of uniform spindle cells and fibroblasts in dense collagen stroma. Necrosis is rare. The microscopic features are similar to, or undistinguishable from, fibromas or fibrosarcomas. Fig. 9 on page 24

CT usually shows a soft-tissue mass of variable attenuation and enhancement. Heterogeneous attenuation may be seen because of necrosis or degeneration. Fig. 10 on page 25

MRI, with its excellent soft-tissue contrast, is best suited for optimum evaluation of extraabdominal desmoids because it allows accurate depiction of their relationship with adjacent structures. Desmoids may have heterogeneous signal and inhomogeneous enhancement because of variable distribution of spindle cells, collagen, and myxoid matrix. On T2-weighted and proton density images, signal intensity is usually intermediate, between skeletal muscles and subcutaneous fat, at times with the presence of hypointense bands corresponding to collagen bundles.
The differential diagnosis of DT includes other neoplasms, such as lymphoma, pleomorphic sarcoma, and fibrosarcoma.

**DESMOPLASTIC MESOTHELIOMA**

Malignant mesothelioma is a rare but aggressive (usually fatal) neoplasm. These tumors have assumed importance during the past decades because of their increased incidence, especially among persons with previous exposure to asbestos. 90% of mesotheliomas are associated with asbestos exposure.

Malignant mesotheliomas are divided into three histological subtypes: epithelial (55%), sarcomatous, and mixed or biphasic. Desmoplastic malignant mesothelioma is a subgroup of malignant mesothelioma, in which 50% or more of the tumor is fibrous and has low cell content. Fig. 11 on page 26

Chest radiography is of limited utility and non-specific, demonstrating a pleural opacity which may extend around and encase the lung. Reduction in volume of the affected hemithorax is common resulting in shift of the mediastinum towards the lesion. Rib destruction may be evident. Mediastinal lymph node enlargement and pleural effusion may also be seen.

CT is the primary modality used for the study of pleural mesothelioma. The findings that suggest mesothelioma are: unilateral pleural effusion, soft tissue nodular pleural thickening and interlobar fissure thickening. Growth typically leads to tumoral encasement of the lung with a rindlike appearance. Calcified pleural plaques are found in 20% of cases and may become engulfed by the primary tumor. Sarcomatoid variants may demonstrate osteosarcoma or chondrosarcomatous components which may also be calcified. There is contraction of the affected hemithorax with associated ipsilateral mediastinal shift, narrowed intercostal spaces and elevation of the ipsilateral hemidiaphragm. Malignant mesothelioma is usually a locally aggressive tumor with frequent invasion of the chest wall (obliteration of extrapleural fat planes, invasion of intercostal muscles, displacement of ribs or bone destruction), mediastinum (heart, pericardial thickening or pericardial effusion, esophagus and trachea) and diaphragm. Malignant mesothelioma frequently metastasizes to the contralateral lung and local lymph nodes. Fig. 12 on page 27

In patients with potentially resectable disease, MRI can provide additional staging information. Relative to muscle, mesothelioma is typically iso-or slightly hyperintense on T1-weighted images and moderately hyperintense on T2-weighted images. Mesothelioma enhances following gadolinium administration.

PET (PET/CT) is becoming useful in two clinical settings: for differentiating between benign and malignant asbestos-related pleural thickening, and for assessing for nodal
metastases. In addition there appears to be a correlation between the degree of FDG uptake and the biological aggressiveness of the tumor.

The imaging differential diagnosis includes: loculated pleural effusion, benign asbestos-related pleural disease, pleural metastases, peripheral bronchogenic carcinoma, solitary fibrous tumor of the pleura, and pleural fibrosis/fibrothorax.

**DESMOPLASTIC SMALL ROUND CELL TUMOR OF THE PLEURA**

Desmoplastic small round cell tumor (DSRCT) of the pleura is a rare aggressive malignancy with a predominance of male patients and increased frequency during the second and third decades of life. The progenitor cell may arise from the mesothelium or from the submesothelial or subserosal mesenchyme with potential for multilineage differentiation.

The majority of cases occur in the abdomen. The pleura is the most frequent extrabdominal location of DSRCT.

The main histological features are the desmoplastic fibrous stroma, the sheets of relatively undifferentiated small cells (*"small blue round cell tumor), and divergent differentiation by immunohistochemistry. Fig. 13 on page 28

Imaging features: DSRCT of the pleura usually presents as multiple pleural soft-tissue masses. Areas of central low attenuation within tumors at CT suggest hemorrhage or necrosis. The degree of tumor enhancement with intravenous contrast is variable. Calcification is seen in 20% of cases. Fig. 14 on page 29

The radiological differential diagnosis includes: pleural metastases, malignant mesothelioma, desmoid tumor/fibromatosis.

**PLEURAL PLAQUES AND DIFFUSE PLEURAL THICKENING (BENIGN ASBESTOS-RELATED PLEURAL DISEASE)**

The most common manifestation of asbestos exposure is pleural plaques, which are discrete areas of fibrosis that usually arise from the parietal pleura but may also arise from visceral pleura.

Pleural plaques (PP) are strongly associated with inhalation exposure to asbestos. There is usually an extremely long latency period between the exposure to asbestos and the development of pleural plaques (20-30 years after exposure). PP are more frequent in men and are usually asymptomatic and incidentally found on chest radiographs.
Histologically, the plaques are relatively acellular, with a "basket-weave" appearance of collagen bundles. Although asbestos fibers/fibres are sometimes seen, asbestos bodies are usually absent. **Fig. 15 on page 30**

The typical distribution of plaques is the posterolateral chest wall (between the seventh and tenth ribs), lateral chest wall (between the sixth and the ninth ribs), the dome of the diaphragm (pathognomonic) and the mediastinal pleura. The apices and costophrenic angles are typically spared. The size and number of plaques are variable. Calcification is present in 15-20% of cases.

Visceral pleural plaques are associated with abnormality in the adjacent lung parenchyma, including short interstitial lines of fibrosis that radiate from the plaque ("hairy plaques") or parenchymal opacities. Visceral plaques have a predilection for the interlobar fissures.

CT is the modality of choice for assessment of PP, since it may identify plaques in any thoracic region, whether calcified or not. **Fig. 16 on page 31**

The differential diagnosis for PP includes adipose tissue, rib fracture, "rib companion shadows", and other pleural masses such as metastases and mesothelioma.

Diffuse pleural thickening (DPT) is less specific for asbestos exposure because other causes of exudative effusions can lead to it. It results from thickening and fibrosis of the visceral pleura, which leads to fusion with the parietal pleura, and is preceded by benign pleural effusion.

Histologically DPT is similar to pleural plaques, except that fusion of the pleural layers is suggestive of more intense inflammation. The underlying process is thought to be inflammation and fibrosis of lymphatic vessels and may be a direct extension of lung fibrosis.

The CT criteria for diagnosis DPT are: a continuous sheet of pleural thickening more than 5 cm wide, more than 8 cm in craniocaudal extent, and more than 3 mm thick.

Differentiation of DPT from PP can be difficult. Apart from the appearances mentioned above, DPT has ill-defined, irregular margins from all angles, whereas PP are well defined and do not tend to extend for more than four interspaces unless they are confluent. DPT involves the interlobar fissures (visceral pleura), whereas PP normally do not.

The differential diagnosis for diffuse pleural thickening includes organizing effusion, chronic infection, connective tissue diseases, talcosis, pleural metastases, and mesothelioma.
INFLAMMATORY MYOFIBROBLASTIC TUMOR OF THE LUNG (INFLAMMATORY PSEUDOTUMOR OF THE LUNG)

Inflammatory myofibroblastic tumor (IMFT) of the lung is a rare lesion accounting for only 0.7% of lung tumors. This entity is also known as inflammatory pseudotumor or plasma cell granuloma. It is a relatively uncommon non-neoplastic tumor-like process with an unidentified etiology that usually occurs in children and young adults. Inflammatory pseudotumor is a rare disease in adults.

Pathologically, IMFT are characterized by the presence of fibrous, hyalinized bands interrupted by irregular patterned sheets of mononuclear cellularity. These mononuclear cells include lymphocytes, histiocytes, and notable numbers of polyclonal plasma cells. Fig. 17 on page 32

Radiologically, IMFT typically presents on chest radiography as a solitary, peripheral mass, usually >3 cm in size. There is a predilection for the lower lobes, and they are predominantly sharply circumscribed. Rarely, IMFT may calcify, cavitate, invade the mediastinum or hilum, or present with pleural effusion. Five percent of cases are multiple, and 10% are endobronchial pure lesions. On CT, these masses can be more heterogeneous than homogenous. Although few lesions have been studied with chest MRI, the T1-signal is heterogeneous and slightly greater than that of skeletal muscle. T2-signal is typically high, and heterogeneous contrast enhancement following gadolinium administration is seen. Fig. 18 on page 33

SCLEROSING HEMANGIOMA

Sclerosing hemangioma (SH) is a benign neoplasm of the lung, characterized by the presence of vascular proliferation with a marked tendency for sclerosis. Multiple theories have been proposed for its histogenesis including mesothelial, mesenchymal, epithelial, and neuroendocrine origin. However, SH seems to originate from pneumocytes type II. This tumor usually occurs in middle-aged young women. In most cases SH is an incidental imaging finding (70-78%) in an asymptomatic patient; however, some patients report a history of hemoptysis (2/3 of symptomatic patients), chest pain and history of previous pneumonia.

Histologically, the tumor is composed of two cell types: polygonal (also called round or interstitial or lesional) cells and the cuboidal (also called lining or surface) cells. These
cells are arranged in varying patterns such as in sheets or papillae, hemangiomatous or sclerotic patterns. Fig. 19 on page 34

On chest radiographs, SH typically presents as a peripheral, solitary, well-defined, homogeneous nodule or mass. Its location is usually subpleural, preferably in the lower lobes. On CT scan, SH also presents as a well-circumscribed subpleural mass that exhibits enhancement following contrast administration. On MRI, T1-weighted high signal intensity areas correspond to regions including abundant clear cells, whereas the T2-weighted low signal intensity areas usually correspond to the fibrotic or hemorrhagic regions within the tumors. The areas of high signal intensity on T2-weighted images, as well as the areas demonstrating contrast enhancement, correspond to the hemangiomatous portions of the tumor. Fig. 20 on page 35

**ROUNDED ATELECTASIS**

Rounded atelectasis (RA) is a radiological entity which was first described in 1928. It usually presents as a rounded opacity in a peripheral location and may mimic lung cancer on imaging.

Typically, RA is seen with benign pleural disease caused by asbestos exposure. Many other causes have been reported, including tuberculosis, trauma, pulmonary infarction, cardiac failure, uremia, Dressler's syndrome...

Histologically, the pleural fibrosis is superficial to the elastic and interstitial layer of the pleura, which is of normal thickness but characterized by prominent wrinkling and infolding. Fig. 21 on page 36

Chest radiograph typically shows a rounded peripheral mass with curvilinear opacities extending from the lung hilum to the pleural surface. CT usually shows a characteristic curving ("comet-tail sign") of bronchi and vessels entering a peripheral rounded or oval-shaped mass.

On CT RA appears as a rounded peripheral mass abutting a thickened pleura. Its diameter lies between 3.5 and 7 cm and its density is close to that of soft tissue. Homogeneous enhancement after contrast administration is usually seen, but is very variable. Fig. 22 on page 37

Calcifications and extraparietal pleural fat accumulation may be found within the thickened neighboring pleura or within the mass itself.

MRI may be used in some circumstances where it is difficult to differentiate between RA and lung cancer. On MRI RA typically exhibits a signal intensity that is higher than
that of muscle and lower than that of fat on T1-weighted images, while on T2-weighted images the signal is similar to or lower than that of fat and enhances after gadolinium administration.

PET/CT may be helpful for differentiating RA from lung cancer in some patients. FDG-PET/CT typically shows a metabolically inactive peripheral lung lesion in association with the pleural thickening and the "comet tail" sign.

When the imaging findings are equivocal, percutaneous needle biopsy of the mass can be valuable for clarification.

**SILICOSIS**

This type of occupational lung disease results from repeated inhalation of silica dust. The risk has been associated with certain occupations (mining, quarrying, tunneling).

Histopathologically, silicotic nodules are composed of mature collagen in the central portion, with a peripheral zone of particle-laden macrophages that appears markedly different from the surrounding lung parenchyma. A number of birefringent silicate crystals 1-3 µm in length usually can be identified at the cellular level with the use of polarized light microscopy. Fig. 23 on page 38

Radiographic manifestations of acute silicosis consist of bilateral perihilar consolidation and/or ground-glass opacities. CT findings include centrilobular nodular ground-glass opacities and multifocal patchy ground-glass opacities, sometimes adopting a "crazy paving" pattern.

Radiographic and CT findings of simple chronic silicosis consist of multiple well-defined nodular opacities that range from 2 to 10 mm in diameter, predominantly in the upper lobes and posterior segments of the lung. At thin section CT, nodules are usually observed in centrilobular, paraseptal, and subpleural regions and have a perilymphatic distribution. Calcification of hilar and mediastinal lymph nodes is common and typically occurs at the periphery of the node ("eggshell" calcification pattern), and is highly suggestive of silicosis. Imaging findings of complicated silicosis ("progressive massive fibrosis") consist of large symmetric bilateral opacities with irregular/spiculated margins, predominantly in the upper and middle zones of both lungs. Fig. 24 on page 39

**PULMONARY HYALINIZING GRANULOMA**

Pulmonary hyalinizing granuloma (PHG) is a relatively new entity recently described characterized by fibrosing lesions of the lung, which have central whorled deposits of lamellar collagen. Its true incidence is unknown, but it is considered considered a rare disease of unknown origin. It has been associated with other entities such as mediastinal
or retroperitoneal fibrosis and infectious processes (mainly tuberculosis). The mean age at presentation is 44 years, with a range from 19 to 77 years. There does not appear to be a sex or racial predilection. Patients are typically mildly symptomatic at presentation or are completely asymptomatic.

Histologically, PHG consists of whorled bundles of hyalinized collagen lamellae arranged haphazardly and concentrically around blood vessels. These are usually accompanied by perivascular collections of lymphocytes, plasma cells, and/or macrophages. Fig. 25 on page 40

Chest radiographs usually demonstrate multiple, frequently bilateral, ill-defined pulmonary nodules or masses, which are of variable size. Solitary lesions are rare. Fig. 26 on page 41

CT findings typically shows randomly distributed nodules and masses with well-defined borders, some with and some without calcium. Although not commonly seen, the calcification is usually focal, central, and irregular. Differential diagnosis must be made with pulmonary metastases that may calcify (osteosarcoma and chondrosarcoma metastases, metastases from mucin-producing adenocarcinomas, and metastases from thyroid and choriocarcinoma).

**CARCINOSARCOMA**

Carcinosarcoma of the lung is an exceedingly rare neoplasm composed of a mixture of carcinoma and sarcoma elements that accounts for less than 1% of all malignant lung tumors. The average age of presentation of carcinosarcomas is 60 years with men to women ratio of 4:1; more than 90% of these patients have a history of heavy smoking.

Lung carcinosarcomas are composed of a mixture of carcinoma and sarcoma elements. The carcinomatous component is most commonly squamous cell carcinoma, followed by adenocarcinoma and large cell carcinoma. The sarcomatous component commonly comprises the bulk of the tumor and shows poorly differentiated spindle cell features. Foci of differentiated sarcomatous elements such as chondrosarcoma and osteosarcoma may be seen. The sarcomatous component probably arises from a previously existing carcinoma through mesenchymal metaplasia or some other divergent tissue differentiation process. Fig. 27 on page 42

The imaging findings are nonspecific, but typically consist of a solitary huge mass or extensive opacity due to associated obstructive pneumonitis and atelectasis. Fig. 28 on page 43
Fibrosing mediastinitis (FM) is a rare non-malignant disorder caused by proliferation of acellular collagen and fibrous tissue within the mediastinum. FM has been associated with other idiopathic fibrosing entities such as retroperitoneal fibrosis or Riedel's thyroiditis, as well as with autoimmune diseases.

Histologically, FM is characterized by the presence of abundant, paucicellular fibrous tissue infiltrating and obliterating adipose tissue. The fibrous tissue can contain patchy infiltrates of mononuclear cells. Because fibrosing mediastinitis is associated with a multiplicity of clinical syndromes and diseases, great care must be exercised when one evaluates open or needle biopsy specimens that demonstrate fibrosis.

FM usually manifests on chest radiographs as non-specific widening of the mediastinum, with distortion and obliteration of normally recognizable mediastinal interfaces or lines. The middle mediastinum is affected more often, particularly the subcarinal and right paratracheal regions. The right side of the mediastinum is more commonly involved than the left. The anterior and posterior mediastinum are much less frequently involved. Less frequently, a focal hilar mass is observed. Calcification is seen in up to 86% of patients.

FM typically manifests on CT as an infiltrative mass of soft-tissue attenuation that obliterates normal mediastinal fat planes and encases or invades adjacent structures. Two patterns of FM have been identified on CT scans: a focal pattern and a diffuse pattern. The focal pattern (80%) manifests as a mass of soft-tissue attenuation that is frequently calcified (60%) and is usually located in the right paratracheal, subcarinal regions or in the hila. Contrast-enhanced CT is useful in cases of suspected superior vena cava, pulmonary arteries and veins obstruction. CT is also useful for assessing the site, length and severity of airway stenosis.

On MRI, FM typically manifests on T1-weighted images as a heterogeneous, infiltrative mass of intermediate signal intensity. Its appearance on T2-weighted is more variable: regions of both increased and markedly decreased signal intensity are frequently seen in the same lesion. Areas of decrease signal intensity are thought to indicate the presence of calcification or fibrous tissue, and areas of increased signal intensity may indicate more active inflammation. The differential diagnosis of FM at CT/MRI includes other infiltrative lesions of the mediastinum, such as lung cancer, metastatic carcinoma, lymphoma, mediastinal sarcoma or, in rare cases, mediastinal desmoid tumors.
Riedel’s thyroiditis (RT) is an extremely rare form of chronic thyroiditis, characterized by a fibroinflammatory process which partially destroys the thyroid, often involving surrounding tissues. The fibrotic process invades adjacent structures of the neck and extends beyond the thyroid capsule.

RT is thought to be a manifestation of the systemic disorder multifocal fibrosclerosis (such as retroperitoneal fibrosis, mediastinal fibrosis and sclerosing cholangitis). It is most often seen in women and the mean age of diagnosis is 40 years.

Histologically, RT is characterized by fibrotic changes in the thyroid gland. However, these changes cannot be reliably distinguished from the fibrotic changes that are often associated with anaplastic thyroid carcinoma. The involved portion is typically described as stony or woody, and is relatively avascular (“cuts like cartilage”), and is often white or pale gray. Fig. 31 on page 46

Enlargement of the affected gland and compression or invasion of adjacent structures, such as strap muscles, trachea, esophagus or carotid, may be observed on CT or MRI. These studies cannot reliably distinguish between RT and invasive thyroid malignancy.

CT shows affected areas of the thyroid to be hypodense. The area is usually isodense with the neck muscles. There is decreased enhancement after the administration of iodinated contrast, especially if extensive fibrosis is present. Fig. 32 on page 47

On MRI, the affected thyroid gland is typically hypointense on T1 and T2-weighted images with decreased enhancement with gadolinium contrast use.

PET shows an increased uptake of FDG in the involved thyroid and a decrease in activity in response to successful corticosteroid therapy, so it may be used to monitor the effectiveness of this treatment.
Fig. 1: Edematous eosinophilic collagen and elastic fibers interwoven aspect rosary, degenerated discs or globules fragmented sawn linear arrangement.

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Fig. 2: CT shows a well-defined soft-tissue mass with thin fat attenuation bands within it.

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**Fig. 3:** Classic microscopic appearance of fibrous dysplasia consisting of small, principally nonmineralized, trabeculae of woven bone (arrow) in bland cellular and collagenous matrix. (H and E)

Fig. 4: Expansive bone lesion in the seventh rib, without cortical fracture or soft tissue mass, consisting of a thickening segmental and circumferential cortical bone, with a cystic central component. The thickened cortical bone has a "ground glass" attenuation.

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**Fig. 5:** Perivascular granuloma with a central and a peripheral ring, a peripheral reactive growth with a central scar. This central area is wrapped with a layer of chronic inflammatory cells (fibroblasts and lymphocytes).

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Fig. 6: CT demonstrated the presence of a relatively well defined, ovoid shaped mass at the anterior chest wall with bony infiltration. The mass showed heterogeneous weak enhancement (arrow).

**Fig. 7:** Spindle cells of variable distribution in short ill-defined fascicles (pattern, no pattern), with areas of hyalinization.

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**Fig. 8:** CT demonstrates a homogeneous well-defined, non invasive, lobular, soft-tissue mass adjacent to the chest wall.

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Fig. 9: Spindle cells with uniform appearance surrounded and separated by collagen bundles.

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**Fig. 10:** Chest CT demonstrates a mass extending from the lateral chest wall nearly to the mediastinum. On pathologic review, the tumor biopsy was characterized as desmoid tumor.

Fig. 11: Proliferation of bland-appearing spindle cells with haphazard growth pattern.

**Fig. 12:** CT shows a multi-lobed mass located in the anterior mediastinum, which extends to the chest wall and pleural masses.

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**Fig. 13:** Core biopsy shows well-defined nests of small round blue tumor cells separated by abundant desmoplastic stroma (hematoxylin-eosin).

**Fig. 14:** Voluminous pleural soft-tissue mass with areas of central low attenuation within tumor.

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Fig. 15: Photograph shows multiple raised pearly plaques that arise from the parietal pleura.

Fig. 16: CT shows multiple calcified pleural plaques in both hemithorax, related to chronic exposure to asbestos.

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Fig. 17: Monoblastic cell proliferation with increased inflammatory infiltrates including many plasma cells in a fibrotic stroma.

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Fig. 18: Tomography scan of the chest shows a spiculated opacity in the left lower lobe, suggestive of malignancy. The wedge resection of this lesion showed only focal organizing pneumonia without any evidence of malignancy.

Fig. 19: Pseudovascular formations, with pleomorphic cells, low mitotic activity and pseudopapillary pattern within sclerotic and angiomatous areas.

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**Fig. 20:** Peripheral spiculated pulmonary nodule with pleural retraction and eccentric calcification.

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**Fig. 21**: Superficial pleura uniformly thickened by fibrosis, while the elastic layer of the pleura shows duplication with prominent wrinkling.

Fig. 22: Axial CT scan shows an ovoid mass, pleural thickening, and linear comet tail of rounded atelectasis.

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Fig. 23: Hyaline nodules and areas of fibrosis with silicon particle-laden histiocytes.

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Fig. 24: Mediastinal window illustrating a conglomerated mass with punctiform calcifications in the posterior region of the right lung, and adjacent pleural thickening. Also note egg-shell-type calcifications on the periphery of the lymph nodes.

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**Fig. 25:** Perivascular granuloma with a central scar which is wrapped with a layer of chronic inflammatory cells (fibroblasts and lymphocytes).

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**Fig. 26:** Multiple pulmonary hyalinizing granulomas.

Fig. 27: Presence of two types of elements, pleomorphic cells, and other spindle cell of mesenchymal origin, sarcomatoid.

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Fig. 28: CT scan with contrast of the chest showing large left upper lobe lung mass involving the pleural surface.

© Olobatoke et al Pulmonary carcinosarcoma initially presenting as invasive aspergillosis: a case report of previously unreported combination. Diagnostic Pathology 2010 5:11.
Fig. 29: Photograph showing numerous inflammatory cells interspersed with collagen bundles consistent with mediastinal fibrosis.

Fig. 30: Soft tissue lesion that infiltrates the right inferior pulmonary vein with associated ipsilateral pleural effusion.

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**Fig. 31:** Riedel thyroiditis. A lymphocytic infiltration is evident within a background of fibrosis and cell atrophy.

Fig. 32: Computed tomography (CT) scan of the neck and the thoracic outlet. This shows a large thyroid mass predominantly affecting the right lobe of the thyroid gland. The trachea has been pushed over to the left.

Conclusion

The importance of correctly identifying and characterizing these tumoral and pseudotumoral (usually benign) thoracic lesions is that they can sometimes mimic malignancy. A correct preoperative diagnosis is, quite frequently, difficult to achieve.


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