Rare mediastinal masses: Imaging aspects

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Authors: S. Hantous-Zannad¹, H. Mhalla¹, A. Zidi², H. Neji², I. Baccouche², K. Ben Miled M'rad¹;¹Tunis/TN, ²TN
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Learning objectives

- Illustrate a spectrum of rare mediastinal masses
- Describe specific imaging findings of those cases
- Outline which imaging modalities might be appropriate for their study
Background

The mediastinum includes structures bounded by the thoracic inlet, diaphragm, sternum, spine and pleura. Mediastinal masses can be located either in anterior, middle or posterior mediastinum. The CT and MRI characteristics and location within a particular mediastinal compartment can help determine the diagnosis. In fact, in adults, thymomas and lymphomas are the most common anterior lesions. Lymph node enlargement and bronchogenic cyst occur the most frequently in the middle mediastinum and neurogenic tumors and vascular masses in the posterior compartment. The etiology diagnosis of mediastinal masses can be approached regarding its location within a particular mediastinal compartment and its aspect on CT or MRI. In this exhibit we illustrated 11 cases of rare mediastinal masses confirmed by histological exams.
Six of the mediastinal masses described in our series were rare based on their histological findings (thymic cyst, thymolipoma, pleomorph carcinoma, angiomyxolipoma, synovial sarcoma and Abrikossof tumor). Four of them were unusual mediastinal locations of common chest or extra-thoracic masses (Hydatic cyst, paraganglioma, retrotracheal goiter and malignant sheath nerve tumor. Computed tomography (CT) approached the diagnosis in 4 cases regarding the location and the presence of characteristic cystic or fatty component: thymic cyst, thymolipoma, hydatic cyst, retrotracheal cystic goiter. Bone anomalies with posterior mediastinal mass suggested extramedullary hematopoiesis. CT scan was useful to predict local extension in the other cases: Abrikossof tumour, pleomorph carcinoma, malignant nervous tumour and synovial sarcoma. Magnetic resonance imaging allowed a better characterization of the inner components in synovial sarcoma. It helps suggesting the diagnosis when showing multilocular cystic aspect in hydatic cyst, signal of both fatty and soft tissue components in thymolipoma and when demonstrating an intensive enhancement with flow void aspect typical of paraganglioma.

**Thymic cyst:** A 56-year-old man. Incidental finding. Fig.1:

Thymic cysts are rare benign lesions of the mediastinum. Most of them are asymptomatic. These cysts are either of congenital or acquired origin resulting from involution changes of the gland. The diagnosis is controversial but a homogeneous hypodense anterior mediastinal mass is highly suggestive of thymic cyst [1]. Thymic cyst should be distinguished from cystic thymoma which can have an inner or peripheral enhancement.

**Thymolipoma:** A 54-year-old man complaining about right chest pain. Fig.2:

Thymolipoma is a very rare well encapsulated benign anterior mediastinal tumor. There is neither sex nor age predilection. It's often asymptomatic. Symptoms, when present, are generally attributed to mediastinal structures compression. Its etiology remains uncertain: true neoplasm of the thymus or hyperplasia of mediastinal fat engulfing thymic tissue. Typically, this tumor appears on chest radiograph as a large mediastinal opacity that can reach the diaphragm. In CT scan, it appears almost fatty with heterogeneous soft tissue density areas that represent thymic tissue. In MRI, it has both fat and soft tissue signal characteristics, which is in agreement with our findings [2].

**Retrotracheal goiter:** A 50-year old woman complaining about shortness of breath and dysphagia. Fig.3

The thyroid gland is located in the neck, even when it gets enlarged. The majority of mediastinal enlarged thyroids are immediately retrosternal and patients usually present with mediastinal organs compression symptoms. Goiters extending behind the trachea
are uncommonly reported [3]. Even when they occur, they are almost located on the right side of the mediastinum.

**Malignant nerve sheath tumor.** A 34-year-old woman complaining about chest pain. Fig. 4

Malignant nerve sheath tumors commonly involve the posterior mediastinum. In our case, this tumor originates from the vagus nerve. Clinicians and radiologists should be aware of the possibility of nerve sheath tumor even if they don't occur in the posterior mediastinum. The imaging criteria of malignant nerve sheath tumour are not specific enough to distinguish them from other malignant soft tissue tumor, so that neither CT nor MRI can establish definite diagnosis [4].

**Paraganglioma.** A 21-year-old man complaining about hypertension and flushing. Fig 5

Mediastinal paraganglioma are rare highly vascularized tumors. They originate from chromaffin tissue located in the para aortic ganglia (middle mediastinum) and the paravertebral sympathetic chain ganglia (posterior mediastinum). The tumor tends to invade bordering structures and may also cause metastasis. Symptoms when present are related to cathecolamin hypersecretion or to a mass effect (dysphagia, shortness of breath and chest pain). MRI provides signal characteristics that may suggest the diagnosis. This is particularly important in case of non-functioning paraganglioma. In addition, MRI studies allow a precise evaluation of the extension of the tumor, which is necessary to plan an efficient surgical excision [5]

**Hydatic cyst.** 35-year-old man complaining about left chest pain. Fig.6:

The mediastinal location of hydatic cyst is rare (0.1%-0.5 % of all locations). The location in the posterior mediastinum results generally from an extension of vertebrae and ribs hydatidosis. In our case, there was no osseous lesion, which is an exceptional feature. CT and MRI allow a better study of spinal involvement when present [6].

**Hematopoiesis extramedullary:** A 20-year-old man. Suspicion of pulmonary embolism. Fig.7

Hematopoiesis is the formation and maturation of blood elements. In adults, it normally occurs in the marrow of epiphysis of long bones, ribs and vertebrae. When these sites are overpassed, blood formation process occurs in various extramedullary locations, particularly in the mediastinum. Symptoms are usually caused by the mass effect. In the thorax the most common imaging manifestations are paraspinal or rib well delimited expansion with mild homogeneous enhancement [7].
**Synovial sarcoma.** A 27 year-old-women complaining about general state alteration, breath shortness and weight loss evolving since one month. Fig.8

Soft tissue synovial sarcoma is far more common than pleuropulmonary synovial sarcoma and typically develops in para articular locations. It affects young and middle aged adults with no sex predilection. Thoracic location may involve the chest wall, the heart, the mediastinum, the pleura or the lung [8]. Computed tomographic images show a well-circumscribed heterogeneously enhanced lesion. Magnetic resonance imaging provides superior demonstration of nodular soft tissue and multilocular fluid-filled internal components in addition to peripheral rim enhancement after the intravenous administration of a gadolinium-based contrast material.

**Pleomorph carcinoma:** A 62-year-old man complaining about dysphagia and alteration of the general state. Fig.9

Pulmonary pleomorphic carcinoma is a rare epithelial tumor with aggressive clinical course. Its incidence ranges from 0.1 to 0.4% of all lung cancer [9]. CT features of a pleomorphic carcinoma of the lung show that these tumors are predominantly located at the lung periphery which is in disagreement with our findings [10]. We suggest in our case that the tumor had primarily developed in the trachea wall and then extended to the mediastinum.

**Angiomyxolipoma:** A 49-year-old woman complaining about an ill-defined right-sided posterior chest wall pain which had been evolving for a few weeks. Fig.10

Angiomyxolipoma is an extremely rare benign tumor. It is recognized as a variant of lipoma. This tumor occurs mainly in the subcutis. Only some cases of angiolipoma are found in the literature. On CT, the tumor is heterogeneous. The vascular component is predominant and shows an intense enhancement. This feature can suggest the diagnosis of angioma. The fatty component appears as small spots with low density. MRI shows that the fatty component has a high signal on T1-weighted images that is attenuated on sequences with fat signal saturation. It allows a better evaluation of the extent into the spinal canal and shows signs of spinal cord injury [11].

**Abrikossof:** A 43- year-old man complaining about dysphagia and shortness of breath. Fig.11

Abrikossof tumor, granular cell tumor or myoblastoma is a rare tumor originating from Shwann cells. It appears generally as unique benign lesion usually located to the tong, the skin or the genital organs. The lower part of the esophagus is the most frequent location in deep organs. Abrikossof tumor has no specific imaging features. It looks like the other mesenchymatous tumours of the esophagus. The diagnosis is always made by microscopic examination [12, 13].
Fig. 1: Thymic cyst: A 56-year-old man. Incidental finding (a) and (b) Chest radiograph shows a large homogeneous opacity with water tonality and smooth borders in the anterior mediastinum. Chest CT (c) axial slice before contrast and (d) axial slice after contrast demonstrates a retrosternal well circumscribed unilocular cystic mass without enhancement.

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Fig. 2: Thymolipoma: A 54-year-old woman complaining about right chest pain. (a) Chest radiograph showing a right-sided anterior mediastinal opacity with smooth borders. (b) CT axial slice before contrast and (c) CT axial slice after contrast shows a well circumscribed right cardiophrenic angle mass with fat area and soft tissue attenuation.

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Fig. 3: Fig.3: Retrotracheal goiter: A woman complaining about shortness of breath and dysphagia (a), (b) and (c) CT axial slices shows a retrotracheal thyroid cystic goiter extending in the middle mediastinum behind the trachea.

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**Fig. 4:** Fig.4: Malignant nerve sheath tumor. A 34-year-old woman complaining about chest pain. (a) and (b) Chest radiograph shows a right mediastinal opacity with smooth borders and water-like tonality displacing the azygoesophageal line reflecting its middle mediastinum location. (c) CT axial slice after contrast shows a soft tissue heterogeneous mass with irregular necrotic central area located in the middle mediastinum and extending to the costovertebral gutter. (d) MRI axial T1 weighted after gadolinium and (e) T2 weighted images shows a well defined right mass with smooth borders, heterogeneous enhancement and necrotic central area with low intensity signal in T1 and high intensity signal in T2 images.

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**Fig. 5:** Paraganglioma. A 21-year-old man complaining about hypertension and flushing. (a) and (b) Chest radiograph shows a posterior left opacity with homogeneous tonality and smooth borders. (c) MRI: coronal T1 weighted before gadolinium, (d) coronal, and (e) sagital T1 weighted after gadolinium confirms the tumor with intermediate signal intensity on T1 weighted image, high signal intensity in T2 weighted image and intense heterogeneous enhancement with small areas of signal void.

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Fig. 10: Angiomyxolipoma: A 49-year-old woman complaining about an ill-defined right-sided posterior chest wall pain which had been evolving for a few weeks. Chest radiograph identified a homogeneous right opacity with smooth borders making an obtuse angle with mediastinum in its lower part reflecting its mediastinal origin. This opacity extended above the clavicle on the postero-anterior film. CT scan (c) and (d) axial, (e) coronal and (f) sagittal mediastinal window post contrast shows a heterogeneous right posterior mediastinal mass located in the costovertebral gutter with a vascular blush and small fat areas. The mass extends into the vertebral canal through an intervertebral foramen and squeezed the thoracic spinal cord.

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Fig. 9: Pleomorph carcinoma: A 62 years-old man complaining about dysphagia and alteration of the general state. CT scan (a) axial mediastinal window post contrast, (b) axial lung window and (c) coronal mediastinal window post contrast shows a well circumscribed left mediastinal mass that is very close to the esophagus and the trachea with heterogeneous enhancement and necrosis areas. This mass is associated to left mediastinal node enlargement and culminal nodule.

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Fig. 8: Fig.8: Synovial sarcoma. A 27 year-old-women complaining about general state alteration, breath shortness and weight loss evolving since one month. (a) Chest radiograph demonstrates a left-sided mediastinal opacity and pleural effusion. (b) CT coronal reconstruction shows a well circumscribed heterogeneous low enhanced mass surrounded by collapsed lung and associated with an ipsilateral pleural effusion. MRI (c) axial T1 weighted before contrast, (d) axial T2 weighted and (e) axial T1 weighted after contrast provides a better demonstration of nodular soft tissue and multilocular fluid components. There is a heterogeneous and intense enhancement after contrast.

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Fig. 7: Extramedullary Hematopoiesis: A 20 years old man. Suspicion of pulmonary embolism (a) Chest radiograph shows two opacities displacing the left paraspinal line reflecting its posterior location (b) CT axial slice post contrast and (c) bone windows demonstrates a homogeneous enhanced paraspinal mass with bone anomalies suggesting the diagnosis of extramedullary hematopoiesis.

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Fig. 6: Hydatic cyst. A 35 years old man complaining about left chest pain. (a) and (b) Chest radiograph shows posterior mediastinal opacity without disruption of the left margin of the heart. (c) CT axial slice after contrast shows a well defined multiloculated cystic mass of the posterior mediastinum adjacent to collapsed lung tissue. (d) MRI axial T2 weighted and (e) coronal T1 weighted after gadolinium helps confirm the multilocular cystic aspect of the posterior mediastinal mass and doesn't show any involvement of the spine and the ribs.

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**Fig. 11:** Abrikossof tumor: A 43-year-old man complaining about dysphagia and shortness of breath. CT scan (a) axial and (b) sagital mediastinal window post contrast shows a homogeneous left-sided middle mediastinal mass. This mass invades the trachea and the oesophagus.

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Conclusion

Conventional chest radiography detects mediastinal masses and helps defining further appropriate imaging techniques regarding the mediastinal compartment involved and the potential association with other chest lesions. CT and MRI can suggest the diagnosis preoperatively even in rare mediastinal conditions. Definite diagnosis is histological.
References


