Primary leiomyosarcoma of the mediastinum

A 58 year-old woman was admitted to our hospital for dyspnoea and chest tightness.

Chest radiograph showed a large homogenous opacity, with well-defined contours, in the right superior mediastinum (Fig 1).

To further investigate the mass chest CT was performed. CT showed a large soft tissue mass, 14x11x14 cm, with multilobulated edges located at the right superior mediastinum. The mass displaced the mediastinal structures without evidence of cleavage plane with the mediastinal structures. Contrast-enhanced CT showed heterogeneous enhancement of the lesion due to areas of necrosis; the mass appeared to be highly vascularised with evidence of neoangiogenesis (Fig. 2). It infiltrated the superior vena cava and the pleural wall, and many collateral vessels were observed (Fig. 3).

The patient underwent mediastinoscopy to perform biopsy. The histological examination classified the lesion as a richly cellular leiomyosarcoma with areas of necrosis and high grade of mitosis (50 mitosis/10HPF).

Due to the local spreading of the disease the patient underwent radiotherapy and chemotherapy with doxorubicin.

Discussion:

The primary tumours of the mediastinum include: thymomas, lymphomas, sarcomas, germ cell tumours and vascular tumours.

Primary sarcomas of the thorax are rare; they are classified according to their histological features and constitute a large group of tumours that occur in the lung, mediastinum, pleura and chest wall. Angiosarcoma, leiomyosarcoma, rhabdomyosarcoma, and mesothelioma (sarcomatoid variant) are the most commonly encountered histopathological types.

Leiomyosarcomas are extremely rare tumours, which develop from smooth muscle, usually in the esophagus and large vessels. In very rare cases, leiomyosarcomas develop from small vessels in the soft tissue of the mediastinum. Approximately 5-10 % of all tissue sarcomas are leiomyosarcomas and they affect predominantly women during the 5th and 6th decade of life. Clinical symptoms of mediastinal leiomyosarcoma such as dysphagia and dyspnoea are related to its large size and the subsequent compression of mediastinal structures.

CT is the study of choice in the evaluation of a mediastinal mass because it can show characteristic features of
malignancy, it can evaluate the local extension, the spreading of the disease and the relationship between the mass and the surrounding structures. Although primary thoracic sarcomas commonly manifest as large, heterogeneous masses, they have a wide spectrum of radiological manifestations, including solitary pulmonary nodules, central endobronchial tumours and intraluminal masses within the pulmonary arteries. Leiomyosarcoma of the mediastinum may manifest at CT as a large necrotic multilobular mass which dislocates and invades the surroundings organs as in the case reported [3].

Treatment of localised leiomyosarcoma is based on surgical excision, either alone or in combination with radiotherapy of the mediastinum. In the case of locally advanced disease or metastatic disease surgery is not recommend and chemotherap - generally doxorubicin - is the treatment of choice with or without radiotherapy [4]. In conclusion, mediastinal leiomyosarcoma is a rare tumor, which can be suspected when the CT excludes the presence of a primary tumour of the lung or metastatic disease. It appears as a wide necrotic mass with high tendency to invade the surrounding organs, is highly aggressive, and histological examination is required to reach a reliable diagnosis.

**Differential Diagnosis List:** Primary leiomyosarcoma of the mediastinum, Other mediastinal tumours, Metastatic disease, Primary tumours of the lung

**Final Diagnosis:** Primary leiomyosarcoma of the mediastinum

**References:**


Figure 1

Description: Contrast enhanced CT scans at two levels (a,b) detect a mediastinal mass at the right superior mediastinum. The mass shows heterogeneous enhancement with necrotic areas. There is contralateral shift of mediastinum to the left. Origin:
Figure 2

Description: Multiplanar reconstruction in coronal (a) and sagittal (b) planes confirms invasion of the superior vena cava and shows the local spreading of the disease. Origin:
Description: Chest radiograph in two projections (a,b) shows a wide mass in the right superior mediastinum. Origin:
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