Primary pleuropulmonary synovial sarcoma

A 34-year-old male smoker with unremarkable past medical history presented with 5 weeks history of shortness of breath, chest pain and nonproductive cough.

Imaging Findings:
Posterior and lateral chest X-ray demonstrated a left-sided opacity in the anterior mediastinum associated with contralateral tracheal deviation.
Axial contrast-enhanced chest computed tomography showed a pleural-based large and well-defined mass with heterogeneous enhancement, nodular soft tissue components mixed with areas of low attenuation surrounded by atelecstatic lung tissue in the left hemithorax associated with contralateral shifting of the mediastinum.
T2-weighted MRI images better delineated the internal heterogeneity of the mass with nodular soft tissue and multilocular fluid-filled internal components.
No lymph node involvement or distant metastatic disease was noted.

Discussion:
Synovial sarcoma is a rare mesenchymal tumour (2.5%–10% of all soft-tissue sarcomas) characteristically seen in adolescents and young adults. [1]
More than 90% of synovial sarcomas affect the extremities, arising especially near large joints and particularly near the knee, but other locations have been described (lung, pleura and mediastinum among others). [2]
Pulmonary synovial sarcoma represents 0.1%–0.5% of all lung neoplasms. [3]
Pleuropulmonary synovial sarcoma (PPSS) does not arise from the synovial membranes but receives its name due to its similarity with synovial tissue tumours under the light microscopy [4] and a specific chromosomal translocation specific to synovial sarcoma.
Macrosopically PPSS are composed of a variety of solid, cystic and necrotic components and microscopically of a proliferation of oval to spindle-shaped tumour cells. Histologic subtypes include monophasic (the most frequent), biphasic and poorly differentiated (with poorer prognosis). [5]
Pleuropulmonary synovial sarcoma has a more aggressive clinical behaviour than soft tissue synovial sarcoma with an overall 5-year survival rate of 50%. [6]
Patients can present with cough, shortness of breath, haemoptysis and chest wall pain [7].

Its typical presentation on chest radiographs is a well-marginated mass with uniform opacity based either in the lung or in the pleura, sometimes with associated ipsilateral pleural effusion. CT characteristically shows a well-defined soft-tissue mass with heterogeneous enhancement and areas of fluid indicating necrosis or haemorrhage, sometimes associated with an ipsilateral pleural effusion but without lymphadenopathy, a finding which points more towards bronchogenic carcinoma. Even if calcification is seen in about 30% of para-articular tumours, PPSS usually lacks tumour calcifications. [7] MRI is helpful to better differentiate nodular soft tissue from multilocular fluid-filled internal components and to determine the extent of tumour invasion. The characteristic “triple sign” seen on MRI imaging (gray, dark and bright) represents tumour, haemorrhage and necrosis. [8]

Due to the rarity of PPSS, there are no guidelines on its optimal treatment. The current standard is surgery followed by chemotherapy or radiation, the latter being used after wide excision in high-grade (G2-3), deep and >5 cm lesions and in cases where R0 resections have not been achieved [9].

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Primary pleuropulmonary synovial sarcoma, Primary and metastatic lung neoplasms, Localised fibrous tumours of the lung and pleura, Malignant mesothelioma, Pleuropulmonary blastoma and other rare parenchymal sarcomas

**Final Diagnosis:** Primary pleuropulmonary synovial sarcoma

**References:**


Figure 1

Description: Posterior (a) and lateral (b) chest X-ray demonstrated a left-sided opacity (asterisk) in the anterior mediastinum associated with contralateral tracheal deviation. Origin: Department of Radiology, Hospital HM Montepríncipe, Madrid, 2019
Description: Posterior (a) and lateral (b) chest X-ray demonstrated a left-sided opacity (asterisk) in the anterior mediastinum associated with contralateral tracheal deviation. Origin: Department of Radiology, Hospital HM Montepríncipe, Madrid, 2019
Description: Contrast-enhanced axial CT scan shows a pleural-based large and well-defined mass with heterogeneous enhancement, nodular soft-tissue components mixed with areas of low attenuation (asterisk) surrounded by atelectatic lung tissue (arrow) in the left hemithorax associated with contralateral shifting of the mediastinum. Origin: Department of Radiology, Hospital HM Montepríncipe, Madrid, 2019
Description: Axial T2-weighted MRI image: T2-weighted MRI image better delineates the extent of the mass and its internal heterogeneity with nodular soft tissue and multilocular fluid-filled internal components (asterisks). Origin: Department of Radiology, Hospital HM Montepríncipe, Madrid, 2019
**Description:** Sagittal T2-weighted MRI image: T2-weighted MRI image better delineates the extent of the mass and its internal heterogeneity with nodular soft tissue and multilocular fluid-filled internal components (asterisks). **Origin:** Department of Radiology, Hospital HM Montepríncipe, Madrid, 2019
Description: Coronal T2-weighted MRI image: T2-weighted MRI image better delineates the extent of the mass and its internal heterogeneity with nodular soft tissue and multilocular fluid-filled internal components (asterisks). Origin: Department of Radiology, Hospital HM Montepríncipe, Madrid, 2019