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Case Report

Uncommon but Aggressive: Imaging Diagnosis of a Large Primary Pulmonary Synovial Sarcoma

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Abstract

Primary pulmonary synovial sarcoma (PPSS) is an exceptionally rare and aggressive malignant tumor, accounting for less than 0.5% of all primary lung neoplasms. We report the case of a 32 year-old female presenting with progressive dyspnea and intense chest pain. Imaging revealed a large, well-defined pulmonary mass with heterogeneous enhancement and areas of necrosis on contrast-enhanced computed tomography (CT). No extra pulmonary primary lesion was identified. Histopathological analysis confirmed monophasic synovial sarcoma, supported by immunohistochemical positivity for epithelial membrane antigen (EMA) and cytokeratin, and detection of the SYT-SSX fusion transcript. This case highlights the critical role of imaging in the evaluation of unusual pulmonary masses and the importance of considering PPSS in the differential diagnosis, especially in younger patients. Early recognition is essential given its aggressive nature and poor prognosis.

Keywords: *Primary Pulmonary Synovial Sarcoma; Lung Mass; Computed Tomography, Rare Tumor*

Introduction

Pulmonary synovial sarcoma is an uncommon, high-grade malignancy originating from mesenchymal cells within the lung tissue, without evidence of a primary lesion elsewhere in the body. Representing a very small fraction of primary pulmonary tumors, it often poses a diagnostic dilemma due to its vague clinical presentation and non-specific imaging characteristics. In this report, we describe a unique case of an unusually large pulmonary synovial sarcoma identified through contrast-enhanced computed tomography, appearing as a well-circumscribed, heterogeneous mass containing necrotic components. The diagnosis was subsequently confirmed by histopathological analysis following a percutaneous transthoracic biopsy. This case underlines the need to include synovial sarcoma in the differential diagnosis of extensive intrathoracic masses and highlights the pivotal role of imaging in its detection and diagnostic workup.

Case Report

A 32-year-old female with no significant past medical history, was admitted to our department for evaluation of a left opaque hemi thorax. One month prior to admission, she had experienced left basithoracic pain. Clinical examination revealed signs of a left pleural effusion, while her general condition remained preserved. Chest radiography demonstrated a homogeneous opacity occupying the entire left hemi thorax (Figure 1). Imaging revealed a large pulmonary mass in the left lung, in close contact with the pleural and adjacent

vascular structures, producing a mass effect on the mediastinum and compressing the contralateral lung. In addition, a well-defined, dense contralateral pleural nodule is also noted (Figure 2).

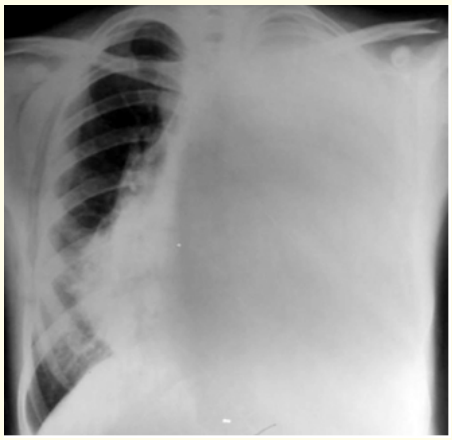

Figure 1: *Frontal chest X-ray showing a complete opacification of the left hemithorax with mediastinal shift, consistent with total left lung opacity.*

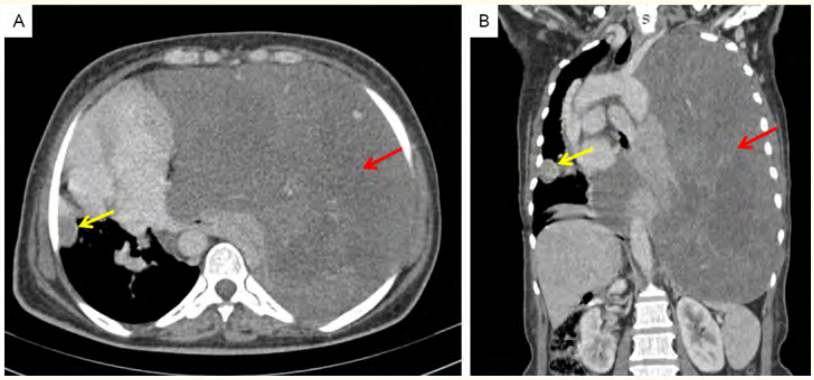

Figure 2: *(A, B) Axial and coronal contrast-enhanced CT scans of the chest (mediastinal window): a large hypodense pulmonary mass with heterogeneous enhancement and central necrotic areas is observed (red arrow), associated with marked cardiac and mediastinal shift and a contralateral pleural nodule (yellow arrow).*

Abdominal ultrasound and bronchoscopy were unremarkable. A CT-guided transthoracic biopsy of the pleural masse revealed a poorly differentiated malignant proliferation composed of spindle and globular cells arranged in fascicles and sheets, with moderately eosinophilic cytoplasm and hyperchromatic nuclei showing moderate cytonuclear atypia. Immunohistochemical staining was positive for vimentin and negative for PS100 and bcl2, consistent with a diagnosis of primary pleural synovial sarcoma (Figure 3). Chemotherapy was planned; however, the clinical course was rapidly unfavorable, and the patient died before receiving the first cycle.

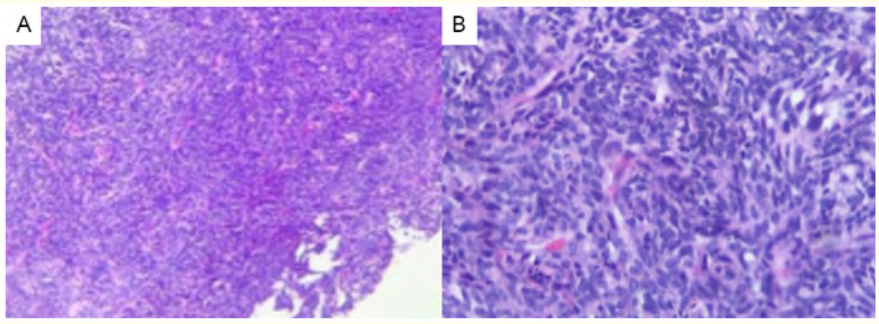

Figure 3: *4× and 20× H&E stained sections of pleural synovial sarcoma. (A) At low magnification, the lesion demonstrates alveolar lung tissue infiltrated by sarcoma. (B) At higher magnification, the tumor is composed of densely packed spindle cells arranged in an interlacing, herringbone-like pattern.*

Discussion

Synovial sarcoma (SS) is a rare and aggressive malignant tumor that typically affects young adults. It originates from pluripotent mesenchymal stem cells rather than true synovial tissue, although its name derives from the histological resemblance to synovial structures and its frequent periarticular location. SS accounts for about 2.5% - 10% of all sarcomas, with a slight male predominance (sex ratio ≈1.5). While most cases arise in the extremities and peri-articular soft tissues, thoracic involvement is uncommon (≈8%), and primary pulmonary forms are exceedingly rare, representing less than 0.1% of lung cancers [1,2].

Primary pulmonary synovial sarcoma (PPSS) is an exceptionally rare entity, and its diagnosis is challenging due to the frequent occurrence of pulmonary metastases from synovial sarcomas of other origins. Establishing PPSS therefore requires careful exclusion of secondary disease through a comprehensive correlation of clinical, radiological, histopathological, immunohistochemical, and cytogenetic findings [1,5]. The clinical presentation is most often dominated by respiratory symptoms such as cough, hemoptysis, dyspnea, or nonspecific chest discomfort. In peripheral locations, PPSS may remain asymptomatic until invasion of adjacent structures, including the pleura or chest wall, which can result in chest pain. Less commonly, the tumor may be discovered incidentally or through evaluation of secondary manifestations [1,3,6]. Imaging findings are nonspecific and often mimic other pulmonary neoplasms: chest radiography typically demonstrates a well-circumscribed opacity, while computed tomography reveals a sharply demarcated, heterogeneously enhancing mass, occasionally containing necrotic or calcified areas.

Immunohistochemistry plays a key role in establishing the diagnosis by excluding other entities. Most cases demonstrate epithelial marker expression, particularly epithelial membrane antigen (EMA), along with cytokeratins. CD99 positivity is observed in approximately 60% of cases, and S100 protein in about 30%, with occasional reactivity for calretinin. Molecular confirmation can be obtained through RT-PCR or FISH, which typically identify the characteristic t(X;18) translocation involving the SSX1 or SSX2 genes on chromosome X (Xp11) [4,9]. A complete staging workup, including cerebral and abdominopelvic imaging, is recommended to exclude secondary lesions and confirm the pulmonary origin of the tumor. Involvement of mediastinal lymph nodes is uncommon.

The prognosis of PPSS parallels that of synovial sarcomas arising in other locations. Key factors linked to poorer outcomes include age above 20, male sex, tumors larger than 5 cm, extensive necrosis, high mitotic counts, neurovascular invasion, incomplete surgical resection, and the SYT-SSX1 fusion variant. The most important determinant remains achieving complete tumor resection with negative margins. The 5-year survival rate is around 50% [11].

Adjuvant radiotherapy is recommended for tumors larger than 5 cm, given that synovial sarcoma is classified as a high-grade malignancy; it improves local tumor control. In contrast to soft tissue synovial sarcoma, for which chemosensitivity is established, the role of chemotherapy in PPSS remains unclear. However, some studies suggest that ifosfamide may be beneficial in selected cases, particularly in unresectable or metastatic disease, where it has been associated with improved survival [9,10].

Conclusion

Primary pulmonary synovial sarcoma (PPSS) is a rare and highly aggressive tumor, usually presenting with respiratory symptoms and showing nonspecific radiological features. Histologically, the two most frequent subtypes are monophasic and biphasic. Management is challenging, particularly when the tumor arises in the chest wall, due to its rarity and broad differential diagnosis. No standardized treatment guidelines exist because of the limited data available, but the therapeutic approach typically combines surgical resection with radiotherapy, which improves local control. Neoadjuvant chemotherapy may also facilitate resection in selected patients, while systemic therapy, often ifosfamide-based, is considered in unresectable or metastatic disease. Despite multimodal treatment, the overall prognosis remains poor.

Bibliography

1. Pandey L., *et al*. "Primary synovial sarcoma of the lung: a rare presentation, diagnostic dilemma and review of literature". *BMJ Case Reports* 13.11 (2020): e237678.
2. Gupta A., *et al*. "Pulmonary synovial sarcoma". *Respiratory Medicine Case Reports* 25 (2018): 309-310.
3. Ammar A., *et al*. "Une localisation primitive inhabituelle du synovialosarcome: le poumon". *Revue des Maladies Respiratoires* 23.3 (2006): 285-287.
4. Dermawan JKT and Policarpio-Nicolas MLC. "Cytological findings of monophasic synovial sarcoma presenting as a lung mass: report of a case and review of the literature". *Diagnostic Cytopathology* 47.9 (2019): 948-955.
5. Eilber FC and Dry SM. "Diagnosis and management of synovial sarcoma". *Journal of Surgical Oncology* 97.4 (2008): 314-320.
6. Satoh H., *et al*. "Primary synovial sarcoma of the chest wall". *Journal of Thoracic Oncology* 2.11 (2007): 1060.
7. Eisenberg RB and Horn RC. "Synovial sarcoma of the chest wall report of a case". *Annals of Surgery* 131.2 (1950): 281-286.
8. Bui-Mansfield LT., *et al*. "Radiologic-pathologic conference of Keller Army Community Hospital at West Point, the United States Military Academy: synovial sarcoma of the chest wall". *American Journal of Roentgenology* 179.4 (2002): 880.
9. Mbatchou Ngahane BH., *et al*. "Évaluation des facteurs pronostiques du synovialosarcome thoracique". *Revue des Maladies Respiratoires* 27.1 (2010): 93-97.
10. Bhattacharya D., *et al*. "Primary pulmonary synovial sarcoma: a case report and review of literature". *International Journal of Applied and Basic Medical Research* 6.1 (2016): 63-65.
11. Thway K and Fisher C. "Synovial sarcoma: defining features and diagnostic evolution". *Annals of Diagnostic Pathology* 18.6 (2014): 369-380.

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