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Primary Pulmonary Synovial Sarcoma: Case Report with Unusual Histology and Diagnostic Pearls

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Abstract

Primary pulmonary synovial sarcoma is rare and poses a diagnostic challenge when unusual histologic features are present and t(x; 18) is negative. We present a case of a 49 year old female with left lung mass and left pleural nodule showing typical clinical and radiologic features of pulmonary synovial sarcoma with focal unusual histomorphology. While dense cellularity, interlacing fascicles, and hyalinized stroma were seen, this tumour showed unusual histologic features not usually seen with pulmonary synovial sarcoma but typical of other neoplasms, in particular, Verocay bodies, rhabdoid morphology, and vague rosette formation. Immunohistochemistry demonstrated Pan-Cytokeratin and CK7 immunoreactivity with focal S-100 expression, but were negative for muscle markers, other melanoma markers, and CD34. The tumour was negative for t(x; 18). In conclusion, awareness of focal unusual histology in otherwise typical pulmonary synovial sarcoma can prevent misdiagnosis, particularly when t(x; 18) is negative.

Keywords: Pulmonary; Pleura; Synovial sarcoma

Introduction

Primary pulmonary synovial sarcoma is an aggressive tumor sharing common histologic features with soft tissue synovial sarcoma [1-5]. Molecular testing for the pathognomonic t(x; 18) chromosomal translocation has enabled diagnostic confirmation in approximately 90% of cases [6]. In t(x; 18) negative cases, diagnosis must rely on histologic and immunophenotypic features. The differential diagnosis of primary pulmonary synovial sarcoma is particularly challenging when histologic features unusual to synovial sarcoma, but common to other neoplasms are focally present. This challenge is compounded with negative t(x; 18) findings. We present a case of primary pulmonary synovial sarcoma where unusual histology was present and t(x; 18) was negative. We offer diagnostic pearls to prevent misdiagnosis in such challenging cases.

Case Presentation

A 49 year old female presented with chest pain and shortness of breath with no significant medical or family history. Laboratory and cardiac work-up was normal and chest x-ray showed a left pulmonary mass. Computed tomography redemonstrated the pulmonary mass with likely metastatic left pleural nodule (Figure 1). Biopsy and surgical excision of tumour showed classic monophasic pulmonary synovial sarcoma histology with dense spindled cellularity, interlacing fascicles, and hyalinized stroma as diagnosed by a pulmonary pathologist, however with focal Verocay body formation, rhabdoid morphology and vague rosette formation (Figure 2-4). Immunohistochemistry showed pancytokeratin and CK7 immunoreactivity with focal S-100 expression, but were negative for muscle markers, other melanoma markers, and CD34. The SYT/SSX RNA fusion transcripts resulting from t(x; 18) (p11; q11) translocation were not detected using real-time reverse transcriptase-polymerase chain reaction. The patient showed no sign of widespread metastatic disease and was treated with complete resection of tumour and received adjuvant radiation treatment. The patient presented with recurrence and pericardial seeding eight months later and died of disease 10 months following diagnosis.

Discussion

Synovial sarcoma, although rare, is a primary pulmonary neoplasm with distinctive histology. The presence of focal unusual histologic findings characteristic of more common epithelial and mesenchymal tumors may lead to misdiagnosis. This is particularly problematic in small biopsies or in primary pulmonary synovial sarcomas that are negative for the pathognomonic t(x; 18) translocation. We present a case of primary pulmonary synovial sarcoma with unusual histologic

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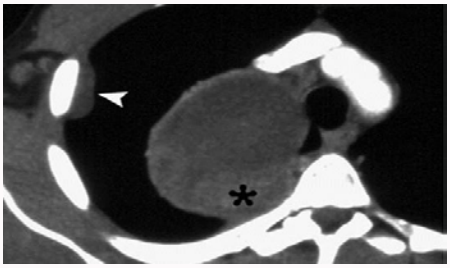

Figure 1: Synovial sarcoma in a 49 year-old female with dyspnea and pleuritic chest pain. Contrast-enhanced computed tomography scan demonstrates a predominantly cystic mass with an eccentrically thickened wall (asterisk) and peripheral rim enhancement. A metastatic nodule is seen (arrowhead) along the lateral pleural surface.

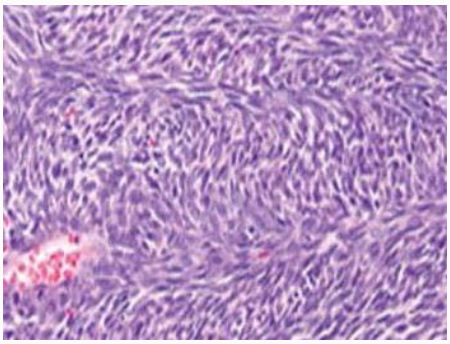

Figure 2: Typical histologic features of pulmonary synovial sarcoma characteristically composed of densely cellular interlacing fascicles.

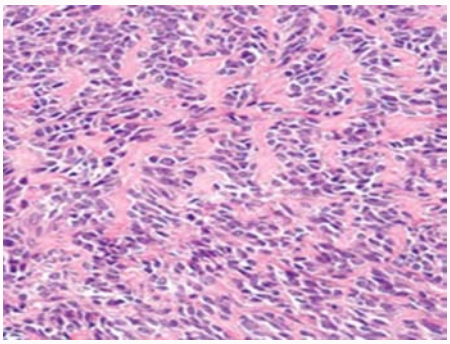

Figure 3: Unusual histologic features that may lead to misdiagnosis include Verocay body-like areas.

features, negative for t(x; 18). The focal unusual histology in primary pulmonary synovial sarcoma can erroneously suggest more common primary and metastatic pulmonary neoplasms. Verocay body-like areas in this case were similar to those seen in malignant peripheral nerve sheath tumor [7]. The stromal background of malignant peripheral nerve sheath tumor, however, typically lacks hyalinization and appears more basophilic. While focal immunoreactivity for S-100 can be present in both tumors, [7] primary pulmonary synovial sarcoma is often immunoreactive for cytokeratin 7, a finding not generally seen in malignant peripheral nerve sheath tumor [8]. Clinically, malignant peripheral nerve sheath tumors arise from nerve or neurofibroma and are associated with neurofibromatosis type I in approximately two-thirds of cases [9]. Primary pulmonary synovial sarcoma with focal vague rosette formation can lead to misdiagnosis as primitive neuroectodermal tumor [6,7]. Primary pulmonary synovial sarcoma may also be reminiscent of primitive neuroectodermal

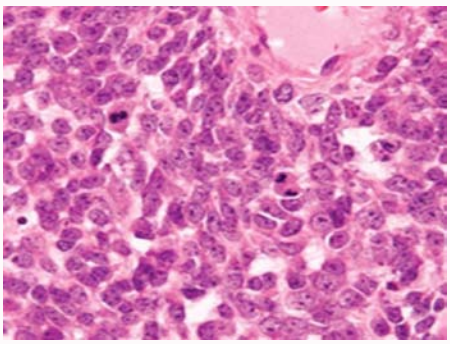

Figure 4: Rhabdoid morphology and vague rosette formation.

tumor when the former is poorly differentiated and displays round cell morphology. Unlike primary pulmonary synovial sarcoma, primitive neuroectodermal tumor typically has distinct cell borders, clear cytoplasm, scant stroma, and lacks hemangiopericytoma-like vasculature typically seen with primary pulmonary synovial sarcoma. Both tumors can express CD99, CD56, and cytokeratins, [7] although expression of cytokeratin 7 makes a diagnosis of primitive neuroectodermal tumor less likely [10]. Chromosomal translocation t (11; 22) is present in 85% of primitive neuroectodermal tumors [6]. We present a case of primary pulmonary synovial sarcoma with focal unusual histologic features that may erroneously suggest more common primary and metastatic pulmonary neoplasms. This unusual histology may be particularly challenging in small biopsies or when t(x; 18) is negative. Awareness of typical histology of pulmonary synovial sarcoma, their potential misleading unusual morphologic features, and prudent use of Immunohistochemistry will prevent misdiagnosis, even in t(x; 18)-negative cases.

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