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Lung: Case Report

Primary Pulmonary Synovial Sarcoma With Extensive Myxoid Change Masquerading as a Lung Hydatid Cyst

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Synovial sarcoma is a rare malignant mesenchymal neoplasm primarily affecting adolescents and young adults, and it typically arises from deep soft tissues near large joints. Although commonly found in extremities, it can occur in various anatomic locations. We present a rare case of a 29-year-old man with primary pulmonary synovial sarcoma manifesting as a cystic mass masquerading as a lung hydatid cyst. Histopathologic examination, immunohistochemistry, and molecular analysis aided in accurate diagnosis. This case underscores the importance of considering synovial sarcoma in the differential diagnosis of pulmonary cysts and highlights the crucial role of pathologic examination in guiding treatment decisions.

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Synovial sarcoma is a malignant mesenchymal neoplasm constituting approximately 5% to 10% of all soft tissue sarcomas. It has a definite male predilection, and it predominantly affects adolescents and young adults ranging in age from 10 to 40 years and is rarely seen beyond 50 years.[1] Synovial sarcoma usually arises from the deeper soft tissue of extremities, especially in close vicinity to large joints such as the knee.[2] However, it has been infrequently reported in the retroperitoneum, mediastinum, oropharynx, tonsil, central nervous system, bone, male and female genital tract, and visceral organs such as kidney, heart, pancreas, pleura, and lung.[3,4]

Although metastasis to the lung from extrapulmonary sarcomas is commonly encountered, primary pulmonary sarcomas are extremely rare, comprising a mere 0.1% to 0.5% of all malignant lung diseases.[4] We describe the case of a 29-year-old man with primary pulmonary synovial sarcoma showing extensive myxoid stroma, masquerading as a lung hydatid cyst. This case is remarkable not only because of the rare primary site of occurrence of synovial sarcoma but also because of the unusual clinical, radiologic, and histopathologic presentation.

A 29-year-old man presented with an insidious onset of cough and shortness of breath for the previous 2 months. There was no history of fever, evening rise of temperature, significant weight loss, chest pain, palpitations, or sputum production. A chest roentgenogram showed a large cystic mass in the right lower lobe of the lung. Computed tomography (CT) revealed a large cyst with internal septations, anterolaterally abutting the costal margin (Figure a). Cystectomy was performed with closure of the remaining cyst cavity with normal saline solution, and the cyst was sent for histopathologic examination. Sections examined revealed an infiltrative, moderately cellular tumor composed of spindle cells with eosinophilic cytoplasm, arranged in vague fascicles and a herringbone pattern with abundant myxoid stroma. The cells had ovoid to elongated nuclei with regular contours, finely dispersed, homogenous chromatin, and inconspicuous nucleoli. No epithelial components, necrosis, calcification, metaplastic elements, or staghorn vasculature were seen. The mitotic count was 4 to 5/10 high-power fields (Figures b-d).

On immunohistochemistry, the cells showed diffuse strong immunoreactivity for B-cell leukemia/lymphoma 2 protein, vimentin, neural cell adhesion molecule (CD56), calretinin, and cluster of differentiation 99 (CD99) and focal positivity for epithelial membrane antigen. marker of proliferation Kiel 67 labeling index was 40%

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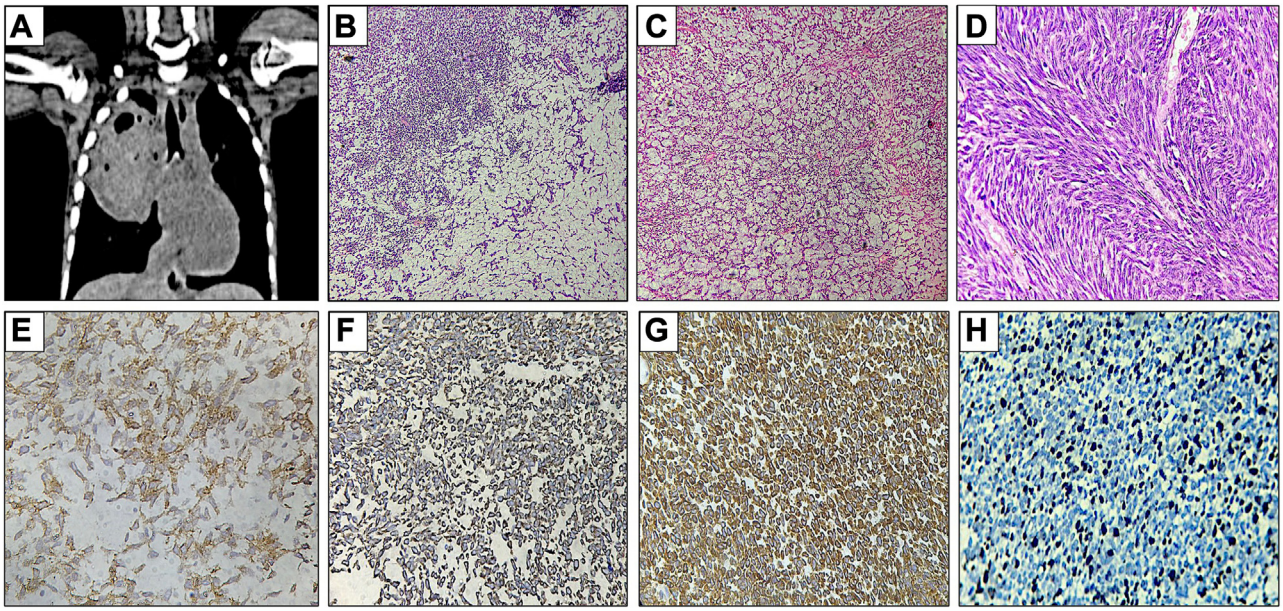

FIGURE Radiologic and histopathologic photomicrographs from the patient. (a) Contrast-enhanced computed tomography revealed a large cyst with internal septations. (b–d) Hematoxylin and eosin–stained sections revealed an infiltrative, moderately cellular tumor composed of spindle cells with eosinophilic cytoplasm, arranged in vague fascicles and a herringbone pattern with abundant myxoid stroma. (Original magnification ×100 and ×400.) (e–h) The cells exhibited positive immunostaining for (e) epithelial membrane antigen, (f) vimentin, and (g) B-cell leukemia/lymphoma 2 protein, with (h) a marker of proliferation Kiel 67 proliferation index of 40%. (Original magnification ×400.)

(Figures e-h). The cells were negative for pancytokeratin, thyroid transcription factor 1, napsin A, CD34, S100 protein, myogenin, MyoD1, desmin, smooth muscle actin, Wilms tumor antigen 1, and CD45. Fluorescence in situ hybridization for t(X;18): synovial sarcoma 18: synovial sarcoma X1 gene fusion revealed a split signal, consistent with synovial sarcoma 18 (18q11.2) rearrangement. Thus, a final impression of synovial sarcoma, monophasic variant with extensive myxoid change, was suggested.

Subsequent positron emission tomography combined with CT showed no evidence of any other metabolically avid lesion elsewhere in the body. The postoperative period was uneventful. Currently, the patient is under follow-up for 2 years with no signs of recurrence or metastasis.

COMMENT

Synovial sarcoma is a malignant soft tissue spindle cell sarcoma that can exhibit variable epithelial differentiation. It constitutes about 5% to 10% of all mesenchymal malignant diseases and shows a slight male predilection (1.2:1).[5]

Although synovial sarcoma is most commonly found in the extremities (85%-90% of cases), it has been reported at almost every anatomic location in the body. Metastasis, particularly in the lung, occurs in approximately 50% of cases, although primary pulmonary synovial sarcoma is extremely rare.[5-7]

Histopathologically, synovial sarcoma is characterized by 2 major variants: monophasic synovial sarcoma, composed entirely of spindle cells; and biphasic synovial sarcoma, which includes both spindle cells and epithelial components. These subtypes can show dedifferentiation, thus posing diagnostic challenges, especially when mimicking other myxoid sarcomas.[8]

Radiologically, synovial sarcoma typically manifests as solid, multilobulated masses with calcifications, often near joint spaces. However, cystic presentations are rare and can lead to diagnostic dilemmas by masquerading as benign lesions.[8]

Immunohistochemistry plays a crucial role in diagnosing synovial sarcoma, with markers such as B-cell leukemia/lymphoma 2 protein, vimentin, CD56, calretinin, CD99, and epithelial membrane antigen aiding in its identification. Molecularly, the characteristic t(X;18)(p11;q11) fusion is found in 90% of cases, detectable by fluorescence in situ hybridization or real-time reverse transcriptase-polymerase chain reaction.[5]

In evaluating a cystic lung lesion, it is essential to consider a differential diagnosis to ensure accurate diagnosis and management. Radiologic features such as daughter cysts and a characteristic "water lily sign" can aid in differentiation from hydatid cysts. Bronchogenic cysts, typically congenital, necessitate imaging modalities such as CT or magnetic resonance imaging to delineate their relationship with adjacent structures and bronchial tree communication. Pulmonary sequestrations require thorough imaging evaluation to identify anomalous arterial supply and confirm noncommunication with the bronchial tree. Additionally, the patient's age and clinical history aid in distinguishing pulmonary lymphangiomas, often seen in pediatric populations. For cystic neoplasms, comprehensive evaluation, including imaging characteristics, the presence of solid components, and assessment of primary malignant diseases elsewhere in the body, is crucial. Moreover, histopathologic examination, immunohistochemistry, and molecular studies may be necessary for definitive diagnosis, particularly in cases with atypical presentations or inconclusive imaging findings.

Treatment involves complete surgical excision with negative margins, often supplemented by radiotherapy and chemotherapy, especially in patients with unresectable tumors. Emerging therapies such as pazopanib, a receptor tyrosine kinase inhibitor, show promise in improving overall survival.[5]

Primary pulmonary synovial sarcoma carries a considerable risk of local recurrence after surgical resection, with reported rates varying on the basis of factors such as tumor size, grade, and adequacy of surgical margins. Close follow-up with imaging studies is necessary to monitor for any signs of recurrent disease. Despite the primary site of this tumor in the lung, metastases can occur both locally within the thorax and to distant sites such as bones and lymph nodes. Vigilant surveillance for metastatic disease is essential because early detection may affect treatment strategies and prognosis. The prognosis of primary pulmonary synovial sarcoma can vary widely depending on various factors, including tumor size, histologic grade, the presence of necrosis, and response to treatment. Generally, primary pulmonary synovial sarcoma has been associated with poorer survival outcomes compared with its soft tissue counterpart, likely a reflection of challenges in early detection, delayed diagnosis, and limited treatment options for lung-based malignant diseases. Given the rarity and heterogeneity of primary pulmonary synovial sarcoma, a personalized approach to management is required. This includes careful consideration of factors such as tumor characteristics, patient comorbidities, and response to initial therapy in determining subsequent treatment strategies. Multidisciplinary collaboration among oncologists, thoracic surgeons, radiologists, and pathologists is essential to optimize treatment outcomes and enhance survival in patients with primary pulmonary synovial sarcoma.

This case highlights the importance of considering primary pulmonary synovial sarcoma in the differential diagnosis of lung cysts, despite its rarity, and emphasizes the critical role of accurate pathologic diagnosis in guiding appropriate treatment and follow-up.

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PATIENT CONSENT

Obtained.

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