A Primary Synovial Sarcoma of Lung

Roy PP, Das A¹, Sarkar A¹, Dwari AK², Datta S¹

Department of Pulmonary Medicine, Midnapore Medical College, Midnapore, Paschim Midnapore, ¹Medical College, Kolkata, ²Bankura Sammilani Medical College, Bankura, West Bengal, India

Abstract

Primary pulmonary synovial sarcoma is an extremely rare tumor. The diagnosis is established only after sarcoma like primary lung malignancies and metastatic sarcoma have been excluded. It has four subtypes: monophasic fibrous, monophasic epithelial, biphasic, and poorly differentiated subtypes. We report a case of a 55-year-old man, who complained of left-sided chest pain and shortness of breath, had a large heterogeneous mass, occupying most of left hemithorax, associated with ipsilateral pleural effusion, seen on contrast enhanced computed tomogram of thorax. Computed tomography guided tru-cut biopsy revealed spindle cell sarcoma. On immunohistochemistry, tumor cells expressed epithelial membrane antigen, CD99, bcl-2 and Calponin and were immunonegative for cytokeratin. So, final diagnosis was primary pulmonary synovial sarcoma. Primary pulmonary synovial sarcoma is a rarely reported case of malignant neoplasm of lung. Histopathology, immunohistochemistry, and cytogenetics, if possible, are essential for confirmation of its diagnosis.

Key words: Immunohistochemistry, lung mass, primary pulmonary synovial sarcoma, tru-cut biopsy,

Address for correspondence: Dr. Anirban Das, Assistant Professor, Peon Para, Bhatchala, P.O. Sripalli, Burdwan – 713 103, West Bengal, India.
E-mail: dranirbandas_chest@rediffmail.com

Introduction

Most lung tumors are malignant in origin and carcinoma by nature. Primary lung sarcoma is an extremely rare tumor, accounting for less than 0.5% of all lung tumors. The variety of soft tissue sarcomas reflects the range of the mesenchymal tissues present in the lung. Three most common sarcomas include leiomyosarcoma, malignant fibrous histiocytoma, and synovial sarcoma.¹

Histological subtypes are differentiated on the basis of immunohistochemical markers, such as vimentin, desmin, actin, CD99, and epithelial membrane antigen. As most of the mesenchymal malignant tumors have a benign counterpart and some epithelial tumors have sarcomatoid differentiation (renal cell carcinoma, melanoma), specific histopathological diagnosis including evaluation of the grade of the lesion is very important. Metastases from extrapulmonary sarcomas are undoubtedly more common than primary pulmonary sarcomas.

Therefore, they must be considered before the diagnosis of primary lung sarcoma is accepted. Such a rare case of primary pulmonary sarcoma diagnosed by computed tomography (CT) guided tru-cut biopsy and subsequent immunohistochemistry is being presented here.

Case Report

A 55-year-old male presented with left-sided pleuritic chest pain for 2 months and progressively increasing shortness of breath with dry cough for 1.5 months. Chest pain was not relieved by simple analgesic. Shortness of breath was not associated with wheeze. Initially cough was nonproductive; later it became productive with scanty white mucoid expectoration, but there was no history of hemoptysis. He smoked 25 bidis daily for last 32 years, i.e., he was exposed to 8 pack year smoking. But there was no history of past exposure to asbestos. On general survey, mild pallor was present, but there were no clubbing and palpable cervical and axillary lymph node. His respiratory rate was 24 breaths/min, pulse rate 108 beats/min, and blood pressure 120/80 mm Hg.
Examination of respiratory system revealed decreased movement of the left side of the chest wall with ipsilateral fullness. Trachea was shifted to right. Vocal fremitus was diminished and percussion note was dull over all areas of left side. Vesicular breath sound was diminished and vocal resonance was decreased on the left side. Examination of abdomen did not reveal any lymphadenopathy, ascites, and hepatosplenomegaly. Other systems were within normal limit.

Complete hemogram and blood biochemistries were within normal limit. Left-sided homogenous opacity along with contralateral mediastinal shifting was seen on chest X-ray. Sputum for acid fast bacilli (AFB) and malignant cell were negative. A total of 500 mL of pleural fluid was aspirated from the left side and pleural fluid analysis revealed lymphocyte predominant, exudative, hemorrhagic pleural effusion with adenosine deaminase value, 17.5 U/L. Papanicolaou stain of pleural fluid revealed no malignant cell. On contrast enhanced computed tomography (CECT) of thorax a large heterogeneous mass with multiple areas of necrosis, occupying almost whole of left hemithorax was seen along with left-sided pleural effusion [Figure 1]. CT-guided fine needle aspiration cytology (FNAC) revealed spindle cell neoplasm. On histopathological section of CT-guided true-cut biopsy, it was shown that there were sheets of spindle cells with plump nuclei, moderate degree of nuclear polymorphism, and mitotic Figures in more than one high power field – suggestive of spindle cell sarcoma [Figure 2a]. However, immunohistochemistry revealed that tumor cells expressed epithelial membrane antigen, CD99, bcl-2 and calponin and were immunonegative for cytokeratin [Figure 2b]. Hence, final impression from immunohistochemistry was primary synovial sarcoma of lung.

**Discussion**

Synovial sarcoma is a rare mesenchymal tumor, accounting for 10% of all soft tissue tumors.[2] It occurs most commonly in adolescents and young adults, in soft tissues of the extremities, but lung is also involved. [2] It is a highly aggressive malignant neoplasm, with a slight male predilection, and is not related to cigarette smoking. The diagnosis of primary pulmonary synovial sarcoma requires clinical, radiological, pathological, and immunohistochemical investigations to exclude alternative primary tumours and metastatic sarcoma.

Primary pulmonary synovial sarcomas are of four subtypes – monophasic fibrous (spindle), monophasic epithelial, biphasic, and poorly differentiated, monophasic subtype being most common.[3,4] Diagnosis of biphasic subtype is easy as both, epithelial and spindle cell components are present. Close differential diagnoses of monophasic subtype are fibrosarcoma, hemangiopericytoma, leiomyosarcoma and spindle cell variant of squamous cell carcinoma, as all are spindle cell neoplasms. Hence, to differentiate monophasic subtype of synovial cell sarcoma from others, immunohistochemistry is essential. Our case was characterized by the presence of spindle cell sarcoma on histopathological examination, the tumor cells being negative for cytokeratin, but positive for epithelial membrane antigen, CD99, bcl-2 and calponin. Thus, diagnosis was primary synovial sarcoma of left lung. A similar case of primary pulmonary synovial sarcoma was reported by Mermigkis et al.[5]

A total of 66% of primary pulmonary synovial sarcomas are centrally located and presents with post-obstructive pneumonia, atelectasis, and hemoptysis.[6] Peripheral tumors are less common and usually asymptomatic, but may infiltrate adjacent pleura, thoracic wall, and mediastinum, or metastasize to hilar or mediastinal lymph nodes, adrenal, brain, and spinal cord.[1] In our case, adjacent pleura was infiltrated by the tumour, resulting in ipsilateral pleural effusion, but there was
no distant metastasis. Prognosis of primary pulmonary synovial sarcoma is poor.

Cytogenetic study by reverse transcriptase–polymerase chain reaction (RT-PCR) helps to differentiate monophasic and biphasic form. Synovial sarcoma is characterized by a reciprocal chromosomal translocation (X;18) (p11.2; q11.2) which results from fusion of SYT gene on chromosome 18 to either of two genes, SSX 1 and SSX 2 on chromosome X.[6] SYT–SSX 1 gene is associated with biphasic subtype and prognosis is bad, whereas monophasic subtype may have one of two fusion transcripts, SYT–SSX 1 or SYT–SSX 2. All tumors with SYT–SSX 2 gene show monophasic morphology. Despite its high sensitivity, molecular testing is not required, if the diagnosis of synovial sarcoma is certain or probable on the basis of clinical, histological, and immunohistochemical evaluations.[6]

The present treatment includes surgical resection, followed by adjunctive chemo- or radiotherapy. Extensive clinical examination, followed by full body CT scan was done to exclude primary synovial sarcoma located peripherally and distant metastases. Hence surgical excision was planned, but the patient refused to continue treatment further and was lost to follow-up.

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