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Case Report

Primary pulmonary synovial sarcoma: a rare case report

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ABSTRACT

Primary lung sarcoma is an extremely rare tumor, accounting for less than 0.5% of all lung tumors. Histological subtypes are differentiated on the basis of immunohistochemical markers, such as vimentin, desmin, actin, CD99, and epithelial membrane antigen. A 50-year-old male presented with progressively increasing shortness of breath with cough for 2 months. 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. CT-guided Fine Needle Aspiration Cytology (FNAC) revealed spindle cell neoplasm. Histopathological examination revealed a spindle cell sarcoma. On immunohistochemistry the tumor cells expressed both epithelial membrane antigen and vimentin. Hence, final impression from immunohistochemistry was primary monophasic synovial sarcoma of lung.

Keywords: Synovial sarcoma, Lung, EMA

INTRODUCTION

Most lung tumors are malignant in origin and carcinoma by nature. Primary Pulmonary sarcoma is an extremely rare tumor, accounting for less than 0.5%[1,2] 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. The most common and important differential diagnosis is metastatis synovial sarcoma to the lung, which needs to be excluded with a thorough clinical and radiological examination. Such a rare case of primary pulmonary sarcoma diagnosed by routine haematoxylin and eosin staining of paraffin block sections and subsequent immunohistochemistry is being presented here.

CASE REPORT

A 50-year-old male presented with progressively increasing shortness of breath with dry cough for 2 months. Initially cough was non-productive, later it became productive with scanty white mucoid expectoration, but there was no history of haemoptysis. There was no history of past exposure to asbestos. On general examination, mild pallor was present, but there were no clubbing and palpable cervical and axillary lymph nodes. 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. 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 limits.

Complete haemogram and blood biochemistries were within normal limits. Left-sided homogenous opacity was seen on chest X-ray. Sputum for Acid Fast Bacilli (AFB) and malignant cells were negative. 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 (Figure 1). CT scan-guided Fine Needle Aspiration Cytology (FNAC) revealed a spindle cell neoplasm. On histopathological sections, it was shown that there were interweaving fascicles of densely packed elongated cells with plump nuclei, moderate degree of nuclear pleomorphism - suggestive of spindle cell sarcoma (Figure 2, 3). However, immunohistochemistry revealed that tumor cells expressed both epithelial membrane antigen, vimentin. Hence, final impression from immunohistochemistry was primary monophasic synovial sarcoma of lung.

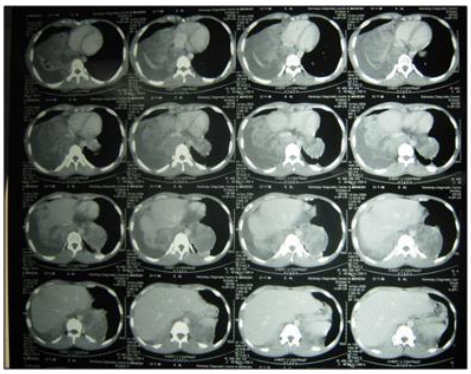

Figure 1: CECT thorax showing a large heterogenous mass in left lung.

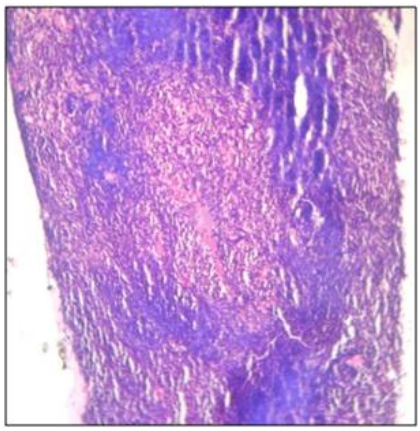

Figure 2: Photomicrograph showing densely packed spindle cells on low power (H&E stain, x100).

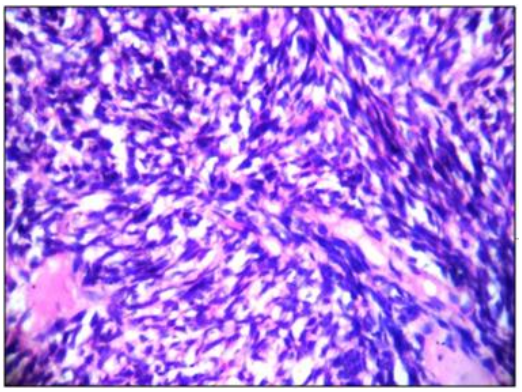

Figure 3: Photomicrograph showing interweaving fascicles of elongated cells. (H&E stain, x400).

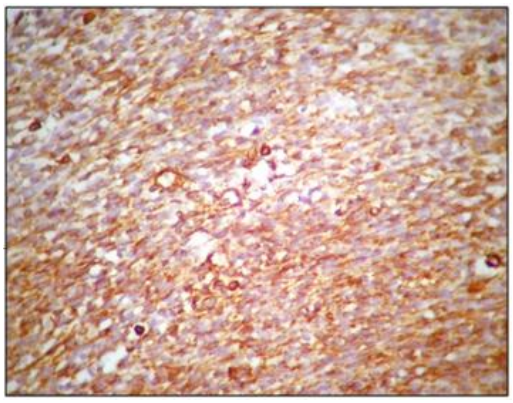

Figure 4: Photomicrograph showing presence of EMA positive spindle cells with membrane positivity (x400).

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] Primary pulmonary synovial sarcoma constitutes less than 0.5% of all pulmonary malignancies.[1,2] Histologic features of pulmonary synovial sarcoma are identical to its soft tissue counterparts.[1] 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 synovial 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. Monophasic synovial sarcoma, the most common pulmonary subtype is comprised solely of the spindle cell component.[2] 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, IHC is essential. Our case was characterized by the presence of spindle cell sarcoma on histopathological examination, the tumor cells being positive for epithelial membrane antigen, vimentin. Thus, diagnosis was primary monophasic synovial sarcoma of left lung. A similar case of primary pulmonary synovial sarcoma was reported by Roy et al.[3]

Synovial sarcomas express vimentin, epithelial membrane antigen and cytokeratin. CK7 & 19 are particularly useful because synovial sarcoma cells express these types of CKs and these are generally negative in other spindle cell sarcomas. Vimentin is usually expressed in the spindle cells of synovial sarcoma. Intranuclear and intracytoplasmic immunoreactivity for S-100 protein can be identified in upto 30% of the tumors. Bcl-2 and CD99 are frequently positive.[2,6]

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.[6,7,9] SYT-SSX 1 gene is associated with biphasic subtype and prognosis is bad, whereas monophasic subtype may have either 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.[3,6]

A total of 66% of primary pulmonary synovial sarcomas are centrally located and presents with post-obstructive pneumonia, atelectasis, and haemoptysis.[1] 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] Prognosis of primary pulmonary synovial sarcoma is poor.[2]

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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Ethical approval: Not required

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