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Case Report

Pathology Section

A Rare Case of Primary Pulmonary Synovial Sarcoma

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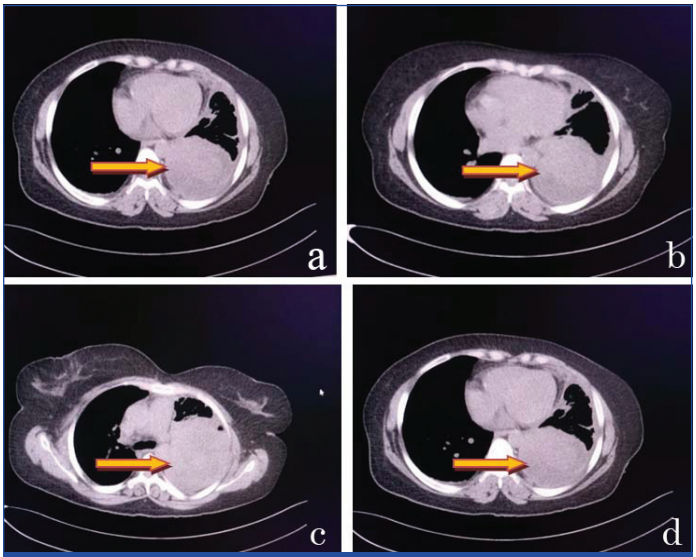
ABSTRACT

Synovial Sarcoma (SS) is a rare mesenchymal neoplasm arising commonly from the periarticular tissue. Primary SS involving the lung accounts for less than 0.5% of all lung tumours. SS are high grade aggressive tumours affecting the extremities of adolescents and young adults. Clinically, patient presents with cough, breathing difficulty, chest pain or haemoptysis and imaging studies may reveal a mass lesion but for definitive diagnosis, histopathology and immunohistochemical examination is needed. Hereby, authors report a case of 39-year-old female diagnosed with primary pulmonary SS with particular emphasis on the pathologic findings.

Keywords: Diagnosis, High grade, Histopathology, Mass lesion, Mesenchymal neoplasm

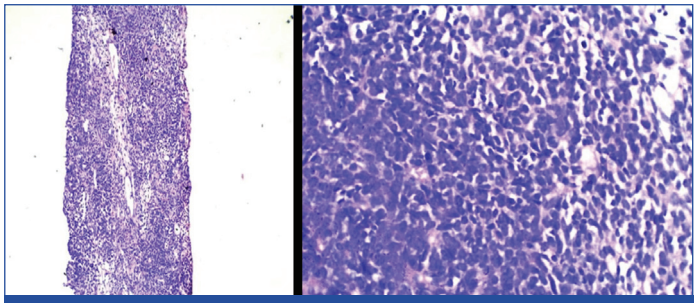
CASE REPORT

A 39-year-old female was admitted in Pulmonology Department, with complaints of dry cough for 1 month, left-sided chest pain and dyspnea on exertion for 1 week. Patient felt better when lying down on right side. She had no associated co-morbidities. Computed Tomography (CT) showed a huge multilobulated heterogeneously enhancing mass lesion measuring 13.8×8.6×6.7 cm with hypodense areas (likely necrotic areas) in left lungs involving upper, lingular and lower segments. Significant mass effect in the form of compression of left pulmonary artery, left main bronchus and left lower bronchus. Mass lesion was closely abutting the descending thoracic aorta and lower lobe pulmonary veins. Moderate left pleural effusion, adjoining lung collapse/consolidation [Table/Fig-1a-d]. Further CT guided biopsy and histopathological examination were done.



[Table/Fig-1]: a-d) CT images showing huge multilobulated heterogeneously enhancing mass lesion measuring 13.8×8.6×6.7 cm.

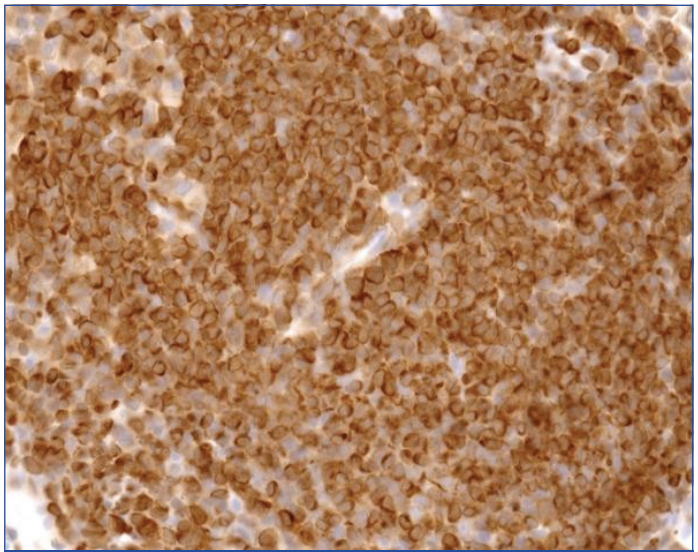
Intercostal Drain (ICD) tube was inserted and 2000 mL of haemorrhagic fluid was aspirated. CT guided biopsy of the lesion was done and sent for histopathological examination, which revealed, multiple linear soft tissue of pale brown in colour measuring 0.5-1.0 cm in length. Microscopic examination showed multiple linear fragments of neoplastic tissue composed of spindle cells only. The spindle cells were elongated in shape, having hyperchromatic nuclei with scanty eosinophilic cytoplasm and were arranged in fascicles and sheets [Table/Fig-2,3].



[Table/Fig-2]: Photomicrograph showing linear fragment of neoplastic spindle cells (Haematoxylin and Eosin, 10X magnification).
[Table/Fig-3]: Photomicrograph showing spindle cells arranged in sheets with hyperchromatic nuclei and scanty cytoplasm (Haematoxylin and Eosin, 40X magnification). (Images from left to right)

Areas of necrosis were also seen. Mitosis was less frequent 0-1 per 10 on High Power Field (HPF). A provisional diagnosis of spindle cell lesion was made and Immunohistochemical examination (IHC) was suggested to rule out high grade sarcoma.

The IHC showed diffuse positivity for BCL-2 (B-Cell Lymphoma-2) as in [Table/Fig-4], Transducin-Like Enhancer of split-1 (TLE-1), Cluster Differentiation-99 (CD-99); focally positive for Creatine kinase (CK) and Synaptophysin; while negative for Thyroid Transcription Factor-1 (TTF-1), Protein 40 (p40), Smooth Muscle Antigen (SMA), Soluble-100 protein (S-100 protein), Cluster Differentiation 34 (CD 34), Cluster



[Table/Fig-4]: Photomicrograph showing immunohistochemical stain for BCL-2 positive.

Differentiation 31 (CD 31), Erythroblast transformation specific-Related Gene (ERG) and Chromogranin. The MIB-1 labelling index was approximately 15% in areas of highest proliferative activity. Final diagnosis was given as malignant spindle cell tumour consistent with primary pulmonary Synovial Sarcoma (SS), favouring monophasic spindle cell subtype. National Fedration of French Cancer Centres (Fédération Nationale des Centres de Lutte Contre Le Cancer, FNCLCC) grade-favour grade 2. Patient was referred to Oncology Centre for further management and follow-up.

DISCUSSION

The SS has been reported most commonly in the periarticular tissue. As described by Rajeev LK et al., SS arising as a primary tumour in the lung is very rare, most common are metastatic carcinoma from other organs and accounts for less than 0.5% of all primary lung tumours [1]. SS is a misnomer because it resembles synovial cells on microscopic examination but does not arise from the synovium [2]. Recent literature by Singhal S et al., has proved that these tumours are derived from the pluripotent mesenchymal cells that have the capability of aberrant epithelial differentiation [2,3]. Primary pulmonary sarcomas commonly seen are fibrosarcomas and leiomyosarcomas [4].

Patient generally present in the young and middle aged adults with chest pain, breathlessness, cough and sometimes haemoptysis and mostly non smokers [1,2,4]. In present case, the patient was a middle-aged adult woman with complaints of left-sided chest pain and breathing difficulty on exertion. A definitive diagnosis needs a coordinated approach by compiling the clinical details, imaging study findings, histopathological and immunohistochemical staining to exclude other possible primary and metastatic tumours of the lung [1,5].

Primary Pulmonary SS are divided into four histological subtypes based on the prominence of either spindle cells or epithelioid cells: monophasic fibrous, monophasic epithelial, biphasic and poorly differentiated. Biphasic subtype can be easily diagnosed with the presence of both spindle and epithelial cells, but monophasic subtype is difficult to diagnose and the most common subtype which needs immunohistochemistry for definitive diagnosis. Immunoreactivity for cytokeratin, vimentin and epithelial membrane antigen along with negativity for CD34 are sensitive markers for monophasic SS [1,2,5]. In the present case, the tumour showed focal cytokeratin and synaptophysin positivity; diffuse positivity for BCL-2, TLE-1 and MIC-2. CD31 was negative excluding lymphoma; CD34 was negative which excluded solitary fibrous tumours; S-100 negative excluded malignant peripheral nerve sheath tumour and SMA negative excluded leiomyosarcomas. Diffuse positivity for BCL-2 was the clinching point which gave a diagnosis of primary pulmonary SS favouring monophasic spindle cell subtype; and Positron Emission Tomography (PET) CT did not reveal any tumour at other sites of the body. The differential diagnosis of monophasic SS includes fibrosarcomas, leiomyosarcomas, haemangiopericytoma, peripheral nerve sheath tumours, intrapulmonary solitary fibrous tumours and squamous cell carcinoma - spindle cell variant [1,3,6].

Singhal S et al., suggested that cytogenetic testing increases the diagnostic specificity of these tumours but are not essential [2]. Reciprocal translocation in the chromosome (X;18)(p11.2;q11.2) leads to fusion of the SYT gene on chromosome 18 to either SSX 1 and SSX2 in the xp11 region; molecular testing to detect SYT-SSX fusion gene can be done through Fluorescent In-Situ Hybridisation (FISH). Reverse Transcriptase-Polymerase Chain Reaction (RT-PCR) can be used to differentiate between biphasic and monophasic subtypes [2,6,7,8,9] Kasthuri S et al., suggest that the treatment includes surgical resection (lobectomy or pneumonectomy) which should be followed by adjunctive chemotherapy or radiotherapy. Prognosis depends on the age of the patient, size of tumour, number of mitotic figures (>10/HPF) and extent of tumour necrosis [1,3,5,7,10].

Since, the patient was referred to Oncology Centres for further surgical and oncology management, follow-up could not be done.

CONCLUSION(S)

Primary Pulmonary SS is a very rare yet aggressive tumour and has very poor prognosis. Definitive diagnosis is obtained through clinical, radiological, histopathological, immunohistochemical and molecular investigations; which help to exclude the possibilities of other primary or metastatic malignancies. Adequate surgical resection with negative margins along with adjuvant chemotherapy and radiotherapy, if needed, is the present treatment modality. Very close follow-up of the patient is needed due to the high recurrence rate of these tumours.

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