Rare case: Monophasic pleuropulmonary synovial sarcoma

Dear Editor,

Synovial sarcoma accounts for 7%-10% of all soft-tissue sarcomas. The most common sites of origin are the thigh, knee, ankle, foot, and upper extremities.[1] Primary synovial sarcoma arising in lung is very rarely seen in clinical practice.[2,3] The use of molecular techniques in diagnosis has recently increased the number of published reports of synovial sarcoma at unusual sites such as pleura, prostate, peritoneum, and lungs.[2] Pleuropulmonary synovial sarcoma represents small subset of pulmonary sarcoma with similar histological, morphological, and chromosomal translocation as their soft-tissue counterparts.[4,5]

Classically, synovial sarcoma has a biphasic pattern and is composed of sheets of spindle cells and sharply segregated epithelial cells forming gland-like areas.[6] Diagnosis of monophasic synovial sarcoma, particularly at unusual location like mediastinum, is a challenge as it is often misdiagnosed as primary pulmonary spindle cell sarcoma/carcinoma or metastatic carcinoma.[4] Further work up including immunohistochemistry, Polymerase Chain Reaction, Fluorescence *in situ* hybridization (IHC, PCR, and FISH) are required for confirmation of diagnosis. T (x; 18) (p11.2; q11.2) is highly specific gene mutation for synovial sarcoma.[7] Using clinical, histopathological, and immunohistochemical findings, the diagnosis may be made with a fair amount of certainty and in developing countries due to financial constraints, molecular diagnosis may not be possible all the time.[8] This is particularly important for cases like ours from developing countries where financial and infrastructural constraints are major drawbacks.

A 40-year-old female presented with complaints of cough, chest pain, and shortness of breath with progressive weight loss since past one year. The past medical history was unremarkable, with no evidence of tuberculosis. The radiological studies X-ray (PA view) showed a large round opacity in lower zone of right lung with small nodules in middle and lower zones of left lung. The ultrasonography of thorax showed a mass lesion in right lower lobe of lung. Bronchoscopy revealed a right-sided growth involving middle and lower pulmonary lobes. No abnormality was seen on left side on bronchoscopy. The computed tomography (CT) of the thorax showed a well-defined mass measuring 8 × 7 × 6 cm in the right lung infiltrating the diaphragm with small nodules of 1-2 cm in the left upper and lower lung lobes. The origin of the mass from lung substance or pleura could not be identified with certainty [Figure 1].

CT guide fine needle aspiration cytology was performed, but unfortunately the material aspirated was inadequate for reporting. Bronchoalveolar lavage was reported as negative for malignant cells.

Finally, right side partial lobectomy was performed. On histopathology, we received three soft-tissue pieces. The largest one measuring 19 × 15 × 10 cm consisted of part of lung tissue with tumor. A well-circumscribed tumor measuring 19 × 9 × 9 cm was seen. The cut surface of tumor was fleshy, homogenous gray white with areas of calcification. The other two pieces comprised of lung tissue measuring 7 × 6 × 2 cm and tumor measuring 6 × 6 × 4 cm.

Microscopy examination [Figure 2] showed a spindle cell tumor with varied pattern of cellularity having areas of high cellularity mixed with low cellularity areas. The tumor cells were arranged in random, fasciculate and at places storiform pattern. Cytologically, the cells had varying degree of anaplasia with fusiform to plump cells having hyperchromatic nuclei and mild-to-moderate pink eosinophilic cytoplasm. Necrosis was minimal. There were occasional mitotic figures seen. Metastatic calcification was also noted. It was interpreted as low-grade spindle cell neoplasm of uncertain histogenesis. Based on the location

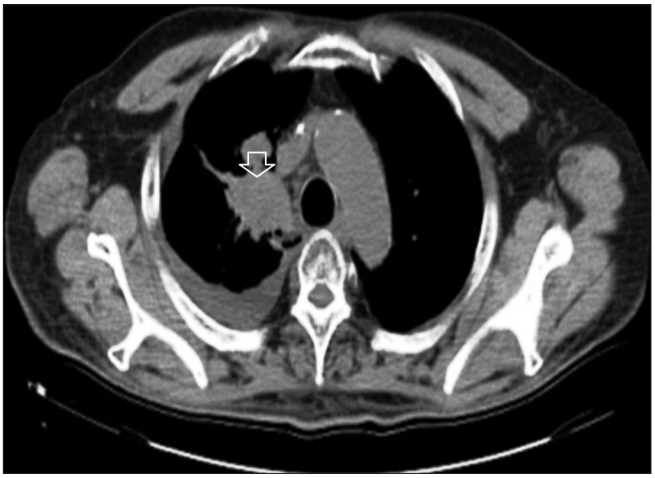

Figure 1: CT Chest showing right side lung mass lesion extending into pleura with multiple small nodules on left side

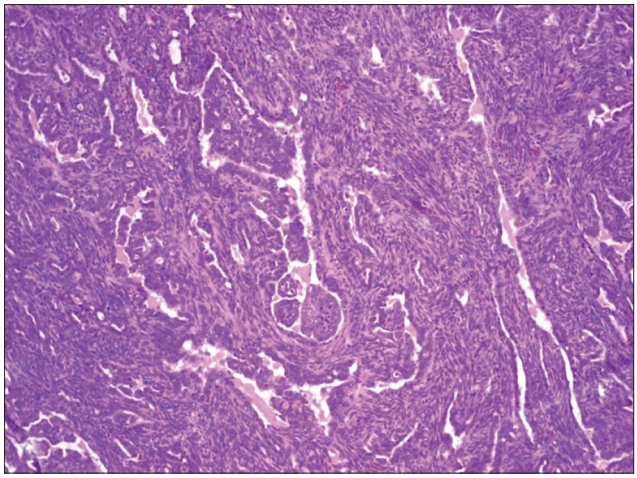

Figure 2: Hematoxylin and Eosin stained section from tumor showing randomly arranged spindle cells with areas of low and high cellularity

of tumor in mediastinum, a differential diagnosis of solitary fibrous tumor and low-grade mesothelioma was suggested. The tumor on immunohistochemical analysis was EMA, cytokeratin5/6, mic-2, bci-2, and calponin positive. TTF-1, calretinin, WT-1, and CD-34 were negative. The tumor was diagnosed as synovial sarcoma.

Unfortunately, the molecular genetic work up could not be performed in our case due to financial and infrastructural constraints. The mainstay of treatment is surgery with postsurgery chemotherapy. Free surgical margins increase the survival of patients and prevent local recurrence. The overall prognosis is poor in pleuropulmonary synovial sarcoma.[9] Tumor size more than 5 cm, male gender, advanced age >20 years, high mitotic activity >10 mitosis/10 HPF, presence of tumor necrosis, and SYT-SSX1 gene variant are the poor prognostic factors.[10]

To conclude, pleuropulmonary synovial sarcoma is a rare tumor. The diagnosis of this tumor requires meticulous clinicopathological correlation, immunohistochemistry, and molecular techniques Familiarity with this entity is essential as it carries poor prognosis.

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