Case Report

Primary pulmonary synovial sarcoma: A case report and review of literature

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Abstract

Primary pulmonary synovial sarcoma is a very rare, but highly aggressive tumor. Metastatic pulmonary sarcoma due to hematogenous dissemination is much more common. Hence why in any case of pulmonary sarcoma, whole body survey is necessary to exclude a primary tumor elsewhere. No clinical or radiological presentations are specific for pulmonary sarcoma hence; it is often confused with bronchogenic carcinoma. On the other hand, image-guided fine needle aspiration cytology (FNAC) is very much helpful in diagnosis of bronchogenic carcinoma, whereas, it may be inclusive in cases pulmonary sarcomas including primary synovial sarcoma, especially in cases of huge pulmonary masses. So why image-guided tru-cut core biopsy or open lung biopsy and their histopathological examination, supplemented by immunohistochemistry are preferable for the tissue diagnosis of pulmonary synovial sarcoma, although FNAC and immunocytochemistry may be used for the diagnosis. Surgical resection is treatment of choice, if it is not possible, palliative chemotherapy may be an option. Here, we report a rare case of primary synovial sarcoma which occupied almost whole of the right hemithorax in a 60-year-old male farmer.

Key words: Histopathology, immunohistochemistry, lung, primary synovial sarcoma, surgical resection

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Introduction

Synovial sarcoma is a rare soft tissue sarcoma accounting for 8% of all soft tissue tumors in the body.[1] It is not originating from the synovial tissue, but arising from pleuripotent mesenchymal tissue; hence, the term "synovial sarcoma" is a misnomer. It most commonly occurs in the extremities, especially in the close proximity of large joints, so it is mistakenly thought that it arises from synovium. Synovial sarcoma is also reported to occur in the lung, mediastinum, abdomen, head and neck, and heart.[2] Metastatic synovial sarcoma from extremities is the most common in pulmonary parenchyma and pleura.[3] Only 0.5% of all primary pulmonary malignancies is due to pulmonary sarcomas of which two are most common: Malignant fibros histiocytoma and synovial sarcoma.[4] Primary pulmonary synovial sarcoma is a highly aggressive lung tumor. It was first described by Zeren *et al*. in 1995.[5] Here, we report a rare case of primary pulmonary synovial sarcoma in a 60-year-old male farmer.

Case Report

A 60-year-old male farmer presented with right sided, dull aching chest pain, shortness of a breath, and dry cough for 1-month. There was no history of fever, wheeze, hemoptysis,

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anorexia, and significant weight loss. The severity of both the chest pain and dyspnea were gradually increasing. The pain was not relieved by simple analgesic and it disturbed his sleep at night. He was a smoker (15 cigarette/day for 15 years), but nonalcoholic.

On general survey, there was no anemia, clubbing, engorged neck vein, and palpable superficial lymph nodes. His temperature was 37°C, respiratory rate, 28 breaths/min, pulse rate, 96 beats/min, blood pressure, 110/70 mmHg, and on room air SpO_2 – 92%. Examination of respiratory system revealed reduced movement of right hemithorax, shifting of trachea and apical impulse to left, dull percussion note on the right side, diminished vesicular breath sound and vocal resonance on right side. Examination of other systems revealed no abnormality.

Complete hemogram and blood biochemistry were within normal limits. Sputum for acid fast bacilli, Gram-stain, and pyogenic culture were negative. Chest X-ray – posteroanterior view showed right sided homogenous opacity with central mediastinum. Contrast enhanced computed tomography (CECT) scan of thorax revealed huge right sided pleural based heterogeneous intraparenchymal mass occupying almost whole of the right hemithorax with contralateral shifting of the mediastinum [Figure 1]. Computed tomography (CT)-guided fine needle aspiration cytology (FNAC) showed spindle cell neoplasm. CT-guided tru-cut biopsy revealed a cellular spindle cell tumor in long fascicles. The cells had plump, hyperchromatic nuclei, indistinct cytoplasm and conspicuous mitotic figures – suggestive of solitary fibros tumor, sarcomatoid pleural mesothelioma, or sarcomatoid carcinoma of lung [Figure 2]. On immunohistochemistry, spindle-shaped tumor cells were strongly positive for bcl-2 [Figure 3], but negative for CD34, cytokeratin (CK), epithelial membrane antigen (EMA), calretinin, and WT-1. CECT brain and ultrasonography of abdomen revealed no abnormality. Radionuclide bone scan did not detect any metastatic bony lesion. Hence, the diagnosis was right sided primary pulmonary monophasic synovial sarcoma. As the tumor encroaching the mediastinal vascular structures, surgical resection of this huge tumor could not be done. Cytotoxic chemotherapy comprising of ifosfamide and doxorubicin was given, but the patient died after completion of first cycle chemotherapy.

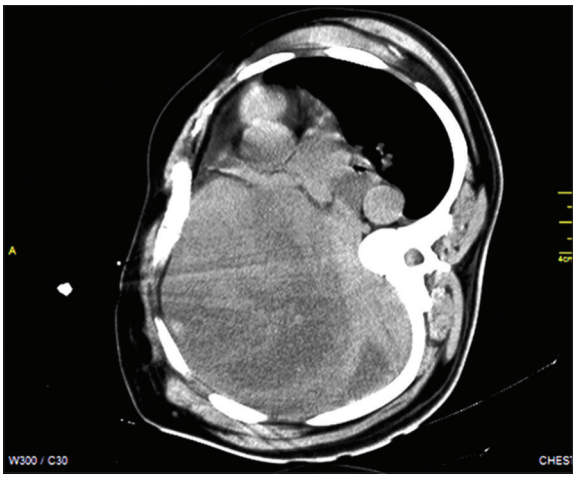

Figure 1: Contrast-enhanced computed tomography scan of thorax showing huge right sided pleural based heterogeneous intraparenchymal mass occupying almost whole of the right hemithorax with contralateral shifting of the mediastinum

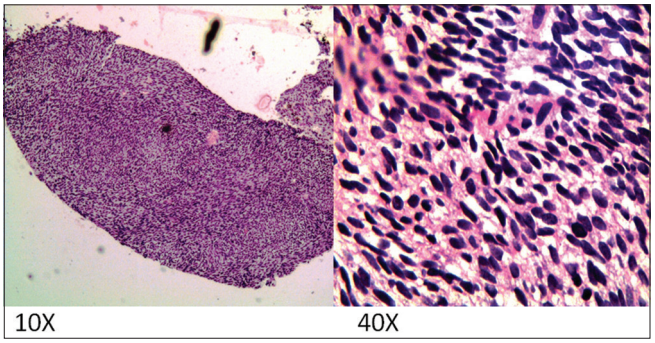

Figure 2: Photomicrograph of histopathology of computed tomography-guided tru-cut biopsy of right sided lung mass showing sarcomatoid malignant neoplasm (H and E, ×10 and ×40)

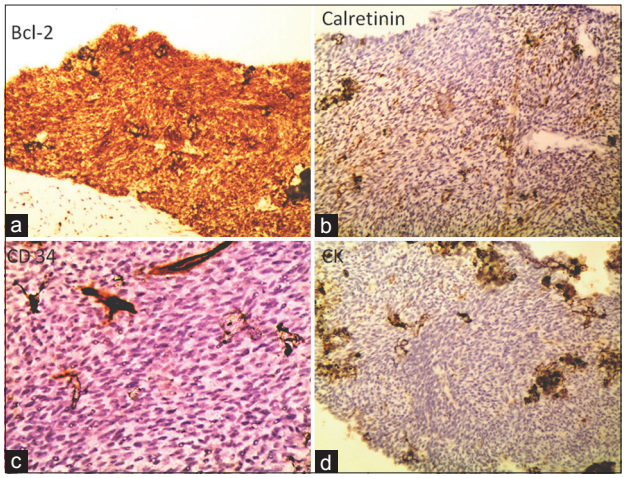

Figure 3: Photomicrograph of immunohistochemistry of computed tomography-guided tru-cut biopsy tissue showing (a) bcl-2 positivity, (b) Calretinin negativity, (c) CD34 negativity and (d) cytokeratin negativity of spindle-shaped tumor cells

Discussion

Primary pulmonary synovial sarcoma is a very rare, but highly aggressive malignant neoplasm. It is most commonly seen in adolescents and young adults. Males are most common affected.[6] In most cases of primary synovial sarcomas of the lung, the patients present with chest pain, cough, shortness of breath, hemoptysis, or ipsilateral pleural effusion.[7] In most cases, a huge pleural-based, heterogeneous, intrathoracic mass

is seen at presentation. Necrosis and hemorrhage are almost always present within the surgically resected mass which is greyish-white or yellowish on gross appearance.

Unlike the other pulmonary sarcomas, histologically it is mainly composed of two morphologically different types of cells: Epithelial cells or fibroblast-like spindle cells. Histopathologically, it is classified into four types: Biphasic, monophasic fibros type, monophasic epithelial type, and poorly differentiated type.[8] Biphasic synovial sarcoma is most common type and considered as the classic type.[9] It is composed of both epithelial cells and spindle cells. The epithelial cells are characterized by large, round or oval vesicular nuclei and abundant pale-staining cytoplasm with distinct cell border. They form solid cords, or nests, or they border pseudoglandular, cleft-like, or cyst-like spaces. The cleft-like spaces resemble normal synovium. Epithelial cells are surrounded by the spindle-shaped cells forming solid, compact sheets. The spindle cells are of uniform appearance with oval, dark-staining nuclei and scanty amount of indistinct cytoplasm. Mitotic figures occur in both epithelial and spindle-shaped cells. Monophasic fibros variant is characterized by predominance of spindle cells with immunopositivity for CK and EMA with only a minute focus of epithelial rests.[8] Monophasic epithelial type exhibits predominance of epithelial cells with formation of gland-like structures. It is difficult to differentiate it from metastatic and primary carcinomas of lung. Diagnosis of poorly differentiated synovial sarcoma is very difficult as it is a crossover of all other variants. Microscopically, it is composed of solidly packed oval or spindle-shaped cells of small size, intermediate appearance between epithelial and spindle cells, with little differentiation, simulating small cell carcinoma of lung.[9] It is associated with a most aggressive course and a worst prognosis among all four variants.[10]

FNAC of lung mass may be inconclusive, as in our case. Hence, image-guided needle biopsy or open lung biopsy or thoracoscopic biopsy is required to make the diagnosis. On immunohistochemistry, it is shown that both the epithelial and spindle cell elements of synovial sarcoma are positive for CK, EMA, bcl-2, and vimentin.[11] A synovial sarcoma may show reactivity for calretinin and S-100 protein. CD99 is positive in 50–100% of synovial sarcomas.[12]

The prognosis of patients with primary pulmonary synovial sarcoma is very poor, as it is a very aggressive tumor with a 5-year survival rate of 50%.[13] Poor prognostic factors are: Size >5 cm, male sex, age >20 years, extensive tumor necrosis, large number of mitotic figures (>10/10 high-powered fields), neurovascular invasion, and SYT-SSX1 variant.[14] The treatment of choice is a complete surgical resection, although there is no standardized therapy.[15]

Conclusion

All lung tumors are not bronchogenic carcinoma, although rare, synovial sarcomas may occur in very few numbers of patients. Clinically they cannot be differentiated from other tumors. Cytology may be inconclusive, especially in huge pulmonary sarcomas and histopathology is required to establish the diagnosis and for cytogenetic study. Immunohistochemistry is preferable for detection of the tumor subtype of this rare tumor entity.

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Conflicts of interest
There are no conflicts of interest.

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