

Pulmonary Sarcoma, an Unusual Lung Mass: A Report of Two Cases and Review of Literature

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Abstract

Case Report

Primary pulmonary sarcoma is a rare malignant tumor, accounting for less than 0.5% of all lung tumours. Here we describe two cases, a 72 year old gentleman who presented with a lung mass and a 66 year old man who presented with paraparesis and lung mass. Chest X-ray and Thoracic computed tomography showed a 4.5 x 4 cm mass in the lower lobe of right lung in our first patient and a 10 x 8 cm pleural based mass in right lower lung in our second patient. Histopathology was suggestive of synovial sarcoma in the first patient and sclerosing spindle cell sarcoma in the second. First patient underwent resection of the tumor and adjuvant radiation and had progression after 10 months. Second one received palliative radiation to bone metastasis and is on chemotherapy.

Keywords: pulmonary sarcoma, lung mass, synovial sarcoma, resection, chemotherapy, radiation.

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INTRODUCTION

The most common primary lung tumours are Non-small cell cancer, Small cell lung cancer and Carcinoid. Sarcomas in lung are usually metastatic lesions and primary sarcoma of the lung is rare. Here we report two cases of primary pulmonary sarcoma diagnosed by computed tomography (CT) and guided biopsy.

CASE REPORT

A seventy two year old goldsmith presented with complaints of progressively increasing cough over the past two months with occasional episodes of hemoptysis and pleuritic pain. He did not give any history of fever, dyspnea or hoarseness of voice. He was a nonsmoker. He gave a past history of thoracotomy done forty eight years ago following accidental aspiration of gold piece. On general examination his performance status was good and he did not have clubbing, lymphadenopathy or any mass. Scar of thoracotomy was present in right lower chest. There was hyper resonance and decreased breath sounds in right inframammary region. Other systems were within normal limits. The hemogram and serum biochemistries were normal. Radiograph of the chest showed an opacity in the right lower lobe (figure 1 a). Contrast enhanced CT scan of thorax showed a heterogeneously enhancing mass in subpleural region of

posterior basal segment of right lower lobe of 45 x 40 mm with moderate hydropneumothorax (figure 1b). Positron emission tomography showed uptake in lower lobe of right lung with SUV of 4.34 (figure 1 c). Bone scan was normal. He underwent CT guided biopsy from the lesion. Histological examination showed spindly cells arranged in fascicles along with intervening small nests of cells with moderate cytoplasm and vesicular nucleus. Spindly cells were positive for bcl2 and the nests of cells were positive for cytokeratin.

Immunostaining for p40, chromogranin and synaptophysin were negative (figure 2). A diagnosis of synovial sarcoma biphasic type was made. He underwent right lower lobectomy. Tumour size was 4.5 x 4 x 1cm with resected margins free of tumour and adjacent pleura showing nodules of tumour deposit. He received adjuvant radiation 66 Gy in 33 fractions. Adjuvant chemotherapy was not given in view of advanced age. CT evaluation done ten months later showed radiological progression with enhancing nodular thickening involving pleura and conglomerate retrocaval deposits and he succumbed to his illness after six months.

The second patient was a sixty six year old man who presented with low backache and weakness of both lower limbs of one month duration. He also complained of severe pain in the hip region and

paresthesia of both lower limbs. He was a smoker with 80 pack years. CT showed a pleural based mass of 10 x 8 cm in lower lobe of right lung abutting posterior chest wall and lower thoracic vertebrae, a subpleural nodule in right middle lobe and mild pleural effusion. Multiple metastases in bone with a cortical break in left acetabulum and ilium and metastasis in both lobes of liver were also seen. Open lung biopsy showed hypocellular /sclerosed areas with cellular areas in between. The cellular areas are composed of fascicles and loosely arranged spindly cells with pleomorphic

oval nuclei and moderate amount of clear cytoplasm, 5-6/hpf mitosis and scattered multinucleate giant cells. Tumour cells were positive for CD34, desmin, SMA, CK and negative for S100, HMB45, calretinin, thrombomodulin, WT1, CD31 and myogenin. A diagnosis of sclerosing spindle cell sarcoma is made. Patient received palliative radiation to left iliac bone and ischium (30 Gy in 10 fractions) followed by palliative chemotherapy with ifosfamide and doxorubicin.

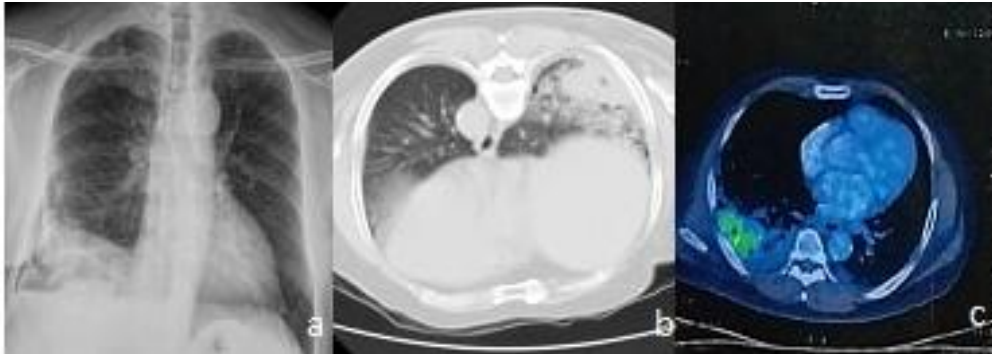


Fig-1a: Chest X ray showing right lower zone opacity. **1b:** CT axial view showing heterogeneously enhancing right lower lobe mass lesion. **1 c:** PET CT image showing FDG uptake in mass lesion

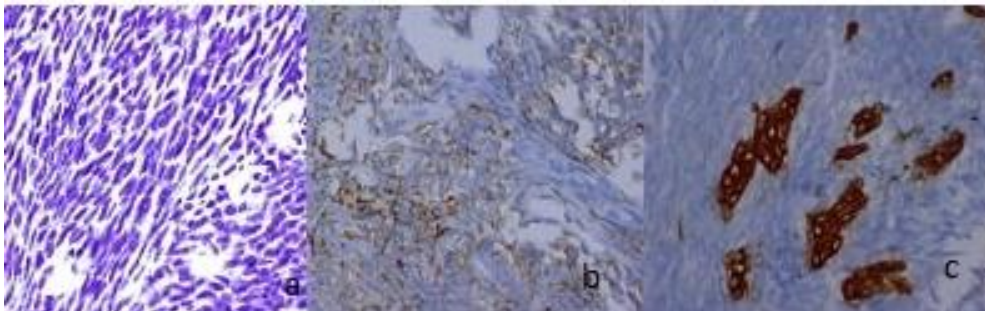


Fig-2 a: Cellular neoplasm composed predominantly of spindly cells with hyperchromatic nucleus (H E X 400). **2 b:** Atypical cells showing positivity for bcl2 (IHC x 400). **2 c:** Atypical cells showing positivity for CK (IHC x 400)

DISCUSSION

Synovial sarcoma is a malignancy of soft tissues. They commonly affect the extremities and are seen very rarely in the head and neck, mediastinum, heart, esophagus, lungs, abdominal wall and mesentery [1].

Primary lung sarcoma is an extremely rare clinically aggressive tumor which constitutes between 0.1% to 0.5% of all lung tumors [2]. The usual location is central and is commonly seen as a postobstructive pneumonia or atelectasis. Clinically they present with cough or hemoptysis. Peripheral tumors as in our case are less common and are usually asymptomatic, but can infiltrate adjacent structures or metastasize to distant organs [3]. Morphologically distinct variants of synovial sarcoma include monophasic fibrous (spindle), monophasic epithelial, and biphasic and poorly differentiated [4]. Biphasic subtype is diagnosed on the basis of the presence of both epithelial and spindle cells.

Molecular diagnosis is through cytogenetic study by Reverse transcriptase polymerase chain reaction method to detect reciprocal translocation (X;18) (p11.2;q11.2). The expression of SYT-SSX1 variants has been associated with worse behavior [5]. Other adverse prognostic factors are male gender, tumor size, necrosis, higher histological grade, mitotic rate and neurovascular invasion [6]. Despite its high sensitivity, molecular testing is not mandatory for the diagnosis of synovial sarcoma. In our case, diagnosis of synovial sarcoma was made based on the histological and immunohistochemical findings.

Lung is the most common site of metastasis in sarcoma, with approximately half of sarcoma patients developing lung lesions at some point in their lifetime. Workup to rule out any other primary site should be done before concluding a diagnosis of primary lung sarcoma.

A study of 12 patients with primary pulmonary sarcoma over a 28 year period who were treated with resection followed by adjuvant chemo/radiation was reported. Median overall survival was 48 months and 5 year overall survival was 38%[7]. Another case of primary fusiform sarcoma of lung in a 50 year old smoker whose tumour was unresectable was also reported. He died after 2 cycles of chemotherapy [8]. Two cases of primary lung sarcoma one treated with chemotherapy and other with surgery followed by chemotherapy which recurred after 5 months was also reported [9]. Treatment and prognosis of primary pulmonary sarcoma is similar to other soft tissue sarcomas. Surgical resection with negative margins is the primary goal, followed by adjunctive chemo and/or radiotherapy. Most common chemotherapeutic agents used are ifosfamide and doxorubicin. Primary pulmonary sarcoma has an aggressive clinical course and they often recur. The median survival is 48 months and five year survival ranges from 38 to 48%[10]. Our first patient also recurred soon after treatment and second one presented with metastasis.

CONCLUSION

Primary pulmonary synovial sarcoma is a very rare and aggressive entity. It is usually treated with surgery followed by adjuvant chemotherapy. The outcome is generally poor.

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