Management of Primary Pulmonary Synovial Sarcoma: A Case Report from Northern Pakistan and Review of Literature

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Abstract

Primary pulmonary synovial sarcoma is a rare lung tumor and has mesenchymal origin. It is difficult to diagnose clinically and should be differentiated from other primary and metastatic lung tumors. The definitive diagnosis should be made via careful histopathological evaluation along with immunohistochemistry. Cytogenetic studies detect SYT-SSX-1 variant which has got diagnostic and prognostic value. The disease has poor outcome and complete cure is difficult to achieve. We hereby report a case of 40 years old female who was diagnosed and treated for metastatic primary pulmonary synovial sarcoma at Atomic Energy Cancer Hospital, NORI.

Keywords: Primary pulmonary synovial sarcoma; Lung; Immunohistochemistry

Introduction

Primary pulmonary synovial sarcoma (PPSS) is an extremely unusual malignancy associated with aggressive behavior and poor clinical outcome [1]. The tumor originates from mesenchymal cells [1]. Synovial sarcomas make up to 8% of soft tissue sarcomas most commonly seen in extremities [1]. Pulmonary sarcomas are even rare making up to just 0.1-0.5% of primary lung malignancies [2]. The differential diagnosis includes metastatic sarcomas and another primary lung cancer [3]. Primary pulmonary synovial sarcoma behaves like soft tissue synovial sarcoma [4]. PPSS may arise from lung parenchyma, pleura, chest wall and mediastinum [5].

The molecular analysis shows characteristic chromosomal translocation t (x; 18) which confirms diagnosis in up to 90% patients and is also a poor prognostic feature [6]. In the rest of 10% cases diagnosis is established through histopathology along with immunohistochemistry [6]. The disease has predilection for young male patients [5]. Our patient was 40-year-old female who presented with cough associated with hemoptysis. She was diagnosed with Stage IV PPSE and received palliative chemotherapy. Only few such cases have been reported in the literature so far and no case has been previously reported from Northern Pakistan.

Case Report

A 40-year-old female patient, non-smoker and non-addict, known diabetic and hypertensive controlled on oral medication and with past surgical history of cholecystectomy for gallstones and hysterectomy for fibroid uterus presented with complaint of dry cough for 4 months. It was later associated with hemoptysis. On examination, patient had a good performance status. Chest examination showed decreased bilateral air entry in lungs, bilateral breast examination was unremarkable and there was no axillary and cervical lymphadenopathy. Rest of the systemic examination was normal.

Figure 1: CT scan chest (lung window) showing multiple soft tissue density intrapulmonary and pleural based nodules in bilateral lung fields.

She was investigated, and CT scan chest and abdomen was done that showed bilateral innumerable nodular opacities both intrapulmonary as well as pleura based, of variable sizes (Figure 1). Largest intrapulmonary mass in right upper lobe measured 6 x 5.1 cm (AP x TR) and large pleura-based mass along right lung
measured 9 x 4.8cm (AP x TR) (Figure 2). Liver showed a large cyst in segment IV measuring 5.3 x 5.4cm (AP x TR) (Figure 3). There was no evidence of any breast mass lesion and axillary lymphadenopathy. No focal osseous lesion was seen.

PET scan was done that showed hypermetabolic soft tissue lesions involving upper lobes of both lungs, the largest one occupying right lung with SUV of 6.8. Hypermetabolic posterior pericardiac lymph nodes were observed. There were numerous pleural and parenchymal deposits in bilateral lungs. There was no evidence of hypermetabolic hepatic and osseous metastasis.

Patient underwent right lung upper lobe wedge resection. The specimen consisted of two grey white linear cores, largest core measuring 0.8 x 0.1cm and smaller core measured 0.3 x 0.1cm. Microscopic examination revealed spindle shaped neoplastic cells arranged in short fascicles with interspersed blood vessels. Unremarkable lung parenchyma was identified. Foci of entrapped pulmonary glandular stricture were also seen close to the edge of the lesion. The mitotic activity was low with less than 4 mitosis/10 HPF. No pleomorphism was seen. Immunohistochemistry revealed that tumor cells were immunopositive for WT-1, TLE-1, Bcl-2 and CD-99 while EMA, Cytokeratin AE1/AE3, Cytokeratin 7, Cytokeratin 19, GATA-3, CD34, STAT-6, S-100 and Sox-10 were negative. The diagnosis of Monophasic Synovial Sarcoma of lung was made.

The case was discussed in multidisciplinary tumor board and the patient was planned for palliative chemotherapy. Echocardiography was advised to the patient that was normal with ejection fraction of 65%. The patient was started on combination chemotherapy with Ifosphamide and Doxorubicin combination along with Mesna in 3 weekly protocol. Patient received 6 cycles of chemotherapy with remarkable subjective and radiological response. The patient was kept on close follow up. She remained asymptomatic for 6 months after which she presented again with complaint of right hypochondrial pain.

CT scan repeated showed a large pleura based heterogeneously enhancing soft tissue density mass lesion along right para-vertebral region extending superiorly from T6 vertebral body to T9-10 intervertebral disc level inferiorly, adjacent to superior segment of right lower lobe measuring 71 x 61mm associated with loculated pleural effusion tracking in to right oblique fissure with associated fissural thickening (Figure 4). Multiple intrapulmonary and pleura based soft tissue density nodules of varying sizes were noted in bilateral lung fields with some of them showing cavitation. Hepatic cyst in the liver was static. Traction bronchiectatic changes were noted in anterior segment of right upper lobe (Figure 5).

Repeat echocardiography for ejection fraction was normal. Patient was counseled thoroughly about the advanced nature of the disease and poor prognosis and was planned for second line chemotherapy.

**Discussion**

Thoracic primary synovial sarcomas are very rare neoplasms mostly originating from lung, heart, mediastinum and esophagus [1]. The incidence of primary pulmonary synovial sarcomas is very low with male predominance [5]. WHO classifies the disease as mesenchymal tumor with most common sites of origin being...
The primary pulmonary synovial sarcoma is an extremely rare malignancy and is a diagnostic as well as a therapeutic challenge. Radiologists and histopathologists should report carefully keeping rare disease presentations in mind so that correct diagnosis can be made. Such cases should be discussed in multidisciplinary tumor boards for most appropriate management plan.

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