***Case report***

**PRIMARY SYNOVIAL SARCOMA (PPSS) - A RARE CASE REPORT ON DIAGNOSIS AND MANAGEMENT.**

**ABSTRACT**

Primary pulmonary synovial sarcoma is a rare and distinct entity, comprising 0.5% of all primary lung malignancies. It typically manifests clinically with cough, chest pain, shortness of breath, or hemoptysis, and a mass lesion may be visible on X-ray or CT scan. Diagnosis is established through histopathology and immunohistochemistry. Here, we present a case of a 71-year-old male who exhibited cough with mucoid expectoration for 1 month, accompanied by breathing difficulties, weight loss, and evening rise of temperature, and the histopathological report revealed primary pulmonary synovial sarcoma of the lung.

KEYWORDS : Primary Pulmonary synovial Sarcoma, Synovial Sarcoma, Surgical Resection, Chemotherapy, Histopathology

**INTRODUCTION**

Synovial sarcoma is an uncommon cancer originating from mesenchymal cells, accounting for approximately 8% of all sarcomas and 0.5% of all malignancies [1]. The lung is an atypical location for primary synovial sarcoma, with metastatic disease from other organs being far more prevalent. The optimal management of primary pulmonary synovial sarcoma (PPSS) remains challenging due to its rarity, resulting in diverse and non-uniform treatment strategies. Optimal treatment includes surgery, chemotherapy, and radiotherapy . PPSS tends to exhibit aggressive local behavior with infrequent distant metastasis. The prognosis is guarded, with a 5-year mortality rate of around 50%, which can be risk aggressive for tumors exceeding 5 cm in size and high mitotic activity [2]. Our case study outlines the diagnostic methodology and therapeutic regimen for a 71-year-old male with an unremarkable medical history, wherein primary pulmonary synovial sarcoma was diagnosed using CT thorax, CT-guided biopsy, histopathological analysis, and immunohistochemistry (IHC). The patient underwent three cycles of chemotherapy.

**CASE DESCRIPTION**

A 71year old male, who presented with c/o cough with mucoid expectoration for 1 month - associated with breathing difficulty , history of weight loss and evening rise of temperature.The patient's medical history was notable for a long-standing smoking habit, with no documented evidence of previous respiratory infections, including tuberculosis. Physical examination revealed a well-nourished individual without signs of anemia, jaundice, or peripheral cyanosis. Respiratory assessment indicated decreased air entry into the right side of the lungs.

Computed tomography (CT) scans of the thorax, with and without contrast, demonstrated a large, lobulated soft tissue mass measuring approximately 8.3 x 8 cm, with extensive involvement of the hilar region and significant encasement of the bronchus intermedius and lower lobe airways. The mass is obliterating superior venacava . Nodular soft tissue noted in the right bronchial lumen. The mass appeared to infiltrate the pericardium at that level and left atrium with enhancing soft tissue component between the parietal and visceral pericardium adjacent to the mass with minimal pericardial effusion. Passive collapse of middle lobe noted with wedge shaped area of consolidation with air bronchogram right lower lobe. Centrilobular emphysematous changes seen in both lungs predominately involving right upper lobe. Interlobular septal thickening noted at right upper lobe with minimal peribronchovascular consolidation. Panlobular emphysematous changes noted left lower lobe. Minimal to moderate right pleural effusion and thin pleural effusion noted.Subcentimetric enlarged lymph node noted right upper paratracheal, both lower paratracheal and subscranial region. CT appearance consistent with advanced central bronchogenic carcinoma right middle lobe region infiltrating right hilar structure with endobronchial tumor extension and pericardial infiltation and small mediastinal lymphadenopathy. Right endobronchial mass biopsy suggest diagnosis of synovial sarcoma.Bronchoalveolar lavage from right bronchus for cytology showed scattered bronchial epithelial cells, lymphocytes, neutrophils and alveolar macrophages. Bronchial brushings for cytology positive for malignant cells. CT guided biopsy specimen lung shows necrotic material with scattered atypical cells.

In view of the large tumor size and hilar vessel involvement, the patient was not a candidate for right pneumonectomy at that time. The patient underwent a multi-agent chemotherapy regimen consisting of Ifosfamide and Doxorubicin as the primary treatment approach. Chemotherapy was administered for three cycles, with Doxorubicin at a dose of 70 mg and Ifosfamide at a dose of 2000 mg, along with Mesna, following a three-weekly protocol but we could not save the patient.

**DISCUSSION**

Synovial sarcoma is an uncommon malignant tumor of mesenchymal origin, representing approximately 8% of all soft tissue sarcomas [1].Primary occurrence in the lung is particularly rare, with primary pulmonary synovial sarcoma (PPSS) comprising only about 0.5% of all pulmonary cancers [3]. Lung involvement is more frequently seen as a site of metastasis from other primary tumors rather than as a primary origin. Due to its rarity, the management of PPSS lacks standardized treatment protocols. Common presenting symptoms include cough, chest pain, dyspnea, and hemoptysis. While PPSS shares histological features with synovial sarcomas of soft tissue, distinguishing it clinically and radiologically from other primary or metastatic lung neoplasms remains challenging [4]

Diagnosing primary pulmonary synovial sarcoma (PPSS) involves evaluating clinical symptoms, imaging studies, tissue pathology, and immunohistochemical analysis, primarily to differentiate it from metastatic sarcomas and other primary lung tumors4 . Immunohistochemistry helps identify the sarcoma subtype, with typical markers such as vimentin, BCL-2, and epithelial membrane antigen (EMA) frequently being positive, while others like S-100 protein, CD34, desmin, actin, and CD99 are usually negative or variable [5] . In our case, the tumor cells expressed vimentin, CD99, BCL-2, TLE-1, and showed focal positivity for PAN-CK, whereas EMA was absent. Despite the use of multiple markers, none are entirely specific to PPSS, which complicates its definitive identification.

In the study conducted by Rahmaniar D et al, a synovial sarcoma located in the lower-left lung lobe was successfully removed via wedge resection without lymphadenectomy. Histology confirmed a spindle cell tumor positive for EMA and CD99. The patient received three cycles of doxorubicin at a dose of 25mg/m2 and ifosfamide at 3000 mg/m2 accompained by mesna, following a three week protocol. A chest CT after treatment showed complete response, and follow-up over time revealed no recurrence. In this case, early-stage diagnosis and prompt treatment likely contributed to the favorable outcome, with imaging studies and follow-up confirming a complete response and no recurrence [4]

There is no standardized treatment protocol for primary pulmonary synovial sarcoma (PPSS); however, surgical management remains the mainstay of therapy. Although there are no expert guidelines or consensus on the ideal surgical approach, complete tumor excision aiming for R0 resection margins is typically pursued to reduce the risk of local recurrence. Depending on the tumor size and location, procedures such as lobectomy or pneumonectomy are commonly performed. In selected cases, partial resection or lumpectomy combined with mediastinal lymph node dissection may be appropriate [6].  Surgery is generally not advised for tumors that are extensively invasive or have already metastasized. Systemic chemotherapy using a combination of doxorubicin and ifosfamide is considered standard, with reported response rates ranging from 25% to 60%. This drug combination has demonstrated clinical benefit in treating various types of sarcomas [4]

**CONCLUSION**

Our case highlights the challenges in diagnosing and managing Primary Pulmonary Synovial Sarcoma (PPSS), a rare and aggressive malignancy. Early diagnosis and treatment are important for improving prognosis. The diagnosis of primary pulmonary synovial sarcoma is based on clinical symptoms, radiological examination, pathology, and immunohistochemistry . Treatment is not standardized; however, surgical intervention plays a predominant role and is the primary strategy. But in our case there is an involvement of large sized tumour and hilar vessel involvement, therefore surgical intervention was not preferred and chemotherapy regimen was initiated but could not save the patient.

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