## Case Report

# PRIMARY INTRATHORACIC PLEUROPULMONARY SYNOVIAL SARCOMA WITH RIB METASTASIS: A RARE ENTITY

Palwasha Gul[1], Pari Gul[2]

**ABSTRACT**

Primary synovial sarcoma is common in limbs, in para particular location. Lungs are commonly involved by metastatic sarcoma due to hematogenous spread whereas primary pulmonary synovial sarcoma in the lungs is an infrequent entity but highly aggressive. Symptoms and imaging findings are non-specific to make definite diagnosis, therefore it is easily confused with other pathologies. The key to correct diagnosis is histopathology.

Primary pleuropulmonary synovial sarcoma is extremely rare. It is important to excluded other sites of primary synovial sarcoma. We report 50 years old female who presented with cough and dyspnea. CT scan showed enhancing circumscribed mass in left hemithorax which proved to be synovial sarcoma on core biopsy.

**Key Words:** Synovial sarcoma, Lung, Metastasis

**doi: https://doi.org/10.51127/JAMDCV3I3CR01**

**How to cite this:**
Gul P, Gul P. Primary intrathoracic pleuropulmonary synovial sarcoma with rib metastasis: a rare entity. JAMDC. 2021; 3(3): 134-136
doi: https://doi.org/10.51127/JAMDCV3I3CR01

## INTRODUCTION

Synovial sarcoma (SS) constitutes 8% of all soft tissue tumors in the body. The term "synovial sarcoma" is a misnomer, as it arises from pluripotent mesenchymal tissue and not the synovial tissue as the name suggests. Its common presentation is in limbs, in para particular location, so it is mistakenly thought to originate from synovium. Common location is close to the large joints in 90% of the cases, however in 10% of the cases it can be seen at other locations such as lung, mediastinum, abdomen, head and neck and heart.[1, 2]

Primary pulmonary sarcomas in the lungs constitutes only 0.5% of all primary lung malignancies with only a few case reports in the literature. Primary SS in pleuropulmonary and mediastinal location has more aggressive course as compared to SS of the extremities.

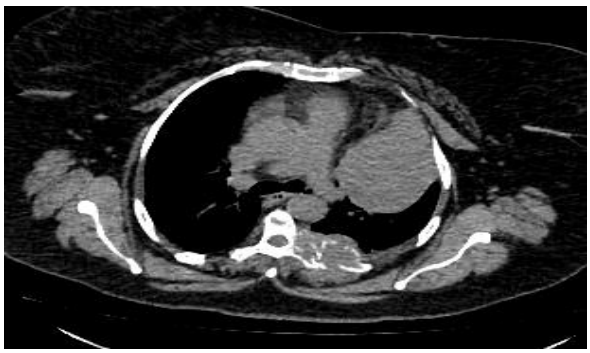
[1]Senior Registrar, Radiology Department, BMCH, Quetta.
[2]Assistant Professor, Radiology Department, BMCH, Quetta.

Multi-disciplinary approach is needed for correct diagnosis which includes clinical/ imaging findings, tissue sampling and immunohistochemical investigations to exclude alternate neoplasia and metastatic sarcoma.[2]

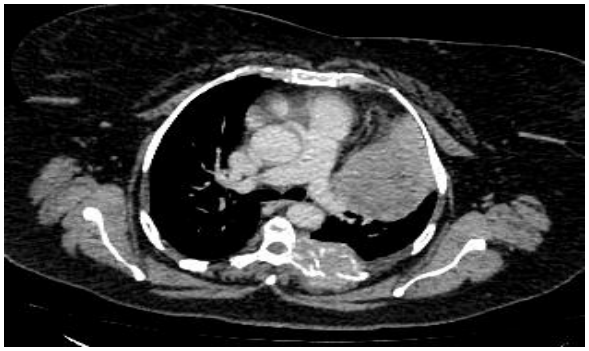
## CASE REPORT

We present a case of 50 years old married female. She presented with cough and dyspnea. Her x-ray chest showed an opacity in left upper lobe. Primary lung malignancy was concern of pulmonologist and radiologist therefore contrast enhanced CT was performed on 128 slicer Toshiba prime aquiline. CT scan showed circumscribed enhancing mass with smooth margins (Fig 1a, b, c). It showed broad interface with adjacent pleura. The mass was abutting left pulmonary artery and left sided cardiac chambers. Partial encasement of left upper lobe bronchus was also noted. No mediastinal lymph nodes were seen. There was destructive mass with soft tissue component in left posterior 5$^{th}$ rib (Fig 1 a, b). There was minimal left pleural effusion.

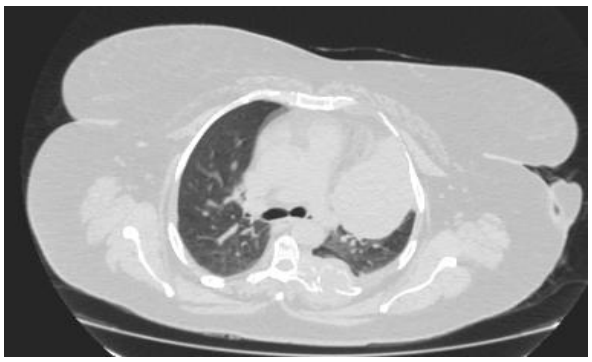
Radiology report gave possibility of lung mass with osseous metastasis and CT guided biopsy of the main mass and rib lesion was planned. In the meantime, clinical examination of the periphery and imaging of the abdomen and pelvis was also performed which was negative for primary malignancy / distant metastasis. Biopsy of lung mass was consistent with synovial sarcoma whereas rib lesion was consistent with its metastasis. Patient was referred to oncologist and thoracic surgeon for further management



a



b



c

**Fig-1:** Plain (a), post contrast (b) and lung window (c) from Contrast enhanced CT chest showing pleuropulmonary enhancing circumscribed mass in left upper lobe abutting left main pulmonary artery. Left 5th rib shows destructive lesion with enhancing soft tissue component

## DISCUSSION

Primary pulmonary SS is a rare entity. The average age of onset for pleuropulmonary location is 38.5 years. Both genders are equally affected with similar predilection for right and left lungs.[2,3] Our patient was 50 years old female with mass in left upper lung. The secondary synovial sarcoma is frequent in lungs however primary is a rare entity. The primary tumor is usually seen in the soft tissues. Literature states that primary synovial sarcoma of the lungs mostly presents as a large pleural-based, inhomogeneous lesion.[1] Pleural effusion is reported, while mediastinal lymphadenopathy was infrequent.[2] In most cases of primary synovial sarcomas of the lung, the primary presentation is with chest pain, hemoptysis, cough, shortness of breath or ipsilateral pleural effusion. Our patient presented with cough and dyspnea. Streak of pleural effusion was also noted in our case on left side ipsilateral to the mass.

Essay and colleagues were first to suggest the term “pleuropulmonary” to describe the anatomic subtype of primary SS originating from either the lung or the pleura, as there were difficulties in defining the exact anatomic location in most cases. There is no large data defining the precise number of pleuropulmonary SS cases worldwide.[4]

Surgical resection is the treatment of choice, although chemotherapy and radiotherapy can also be used as an option. There is no standardized treatment for primary lung SS.[2]

The prognosis for patients with primary lung synovial sarcoma is poor, with an overall 5-year survival rate of 50 percent. Factors predicting a worse prognosis with synovial sarcomas include tumor size (>5 cm), male patients, older age (>20 years), extensive tumor necrosis, high grade, large number of mitotic figures (>10 per 10 hpf), neurovascular invasion, and recently, the *SYT-SSX1* variant.[5]

## CONCLUSION

Primary synovial sarcomas although rare but can be encountered in clinical practice. It cannot be diagnosed or predicted radiologically as primary synovial sarcomas doesn't give any specific findings on imaging. However, imaging can play important role in assessing its size/ morphology and its metastasis. The key to correct diagnosis is histopathology as in our case.

**Disclaimer:** none

**Ethical review committee**: approved by hospital ethical committee (BMCH, Quetta).

## AUTHOR'S CONTRIBUTION

PG: Data collection, Data analysis and Drafting
PG: Review critically

## REFERENCES

1. Bhattacharya D, Datta S, Das A, Halder KC, Chattopadhyay S. Primary pulmonary synovial sarcoma: a case report and review of literature. Int J Appl Basic Med Res. 2016 Jan 1;6(1):63-5. doi: 10.4103/2229-516X.174019
2. Mrabet FZ, Ouazzani HE, Akkari LE, Hammi S, Bourkadi JE, Zouaidia F. Primary pleuropulmonary synovial sarcoma: a case. Case Rep Pulmonol. 2018 Apr 4;2018:5190271. doi: 10.1155/2018/5190271
3. Rajeev LK, Patidar R, Babu G, Babu MS, Lokesh KN, Okaly GV. A rare case of primary synovial sarcoma of lung. Lung India. 2017 Nov 1;34(6):545-7. doi: 10.4103/lungindia.lungindia_7_17
4. Podbielski FJ, Sambo TE, Salamat A, Blecha MJ, Connolly MM. Primary pulmonary synovial sarcoma. PLEURA. 2016 Feb 29;3:1-2. doi: 10.1177/2373997516632755
5. Chang CC, Chang PY. Primary pulmonary synovial sarcoma. J Cancer Res Pract. 2018 Mar 1;5(1):24-6. doi: 10.1016/j.jcrpr.2017.09.002