

Primary Synovial Sarcoma of Mediastinum- Imaging Features In Two Cases

DrN.L.N.Moorthy¹, DrMeenakshi Swain², DrS.Jayabhaskar Reddy³,
DrS.Padmaja⁴

¹Dr N.L.N.MOORTHY Professor Radiodiagnosis

²Dr MEENAKSHI SWAIN Senior Consultant Histopathologist

³Dr S.JAYA BHASKAR REDDY Professor pathology

⁴Dr S.PADMAJA Assistant Professor Radiodiagnosis

Department of Radio diagnosis, Apollo institute of medical science and research

Apollo health city, jubilee hills HYDERABAD 500096Telangana INDIA

Corresponding Author: DrS.Padmaja

Abstract: Synovial sarcoma also called malignant synovioma is a type of soft tissue sarcoma occurring near joint capsule or tendon sheath in young adults. Histologically the tumor is characterized by the presence of small uniform spindle cells and epithelial cells giving it a biphasic appearance. The diagnosis is mainly based on the above pattern of histopathology along with the presence of translocation $t(X:18)(p11.2;q11.2)$. Synovial sarcoma involving other sites like mediastinum is extremely rare. We present the imaging features in two cases of histologically proven synovial sarcoma in young individuals presenting as huge intrathoracic masses.

Key Words; synovial sarcoma- mediastinum- soft tissue sarcoma

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I. Introduction:

Though labelled as synovial sarcoma, the tumor does not arise from synovium, but arises from pluripotential mesenchymal cells that are present in tendons, tendon sheath, joint surfaces. They arise mostly around joints. The rare and uncommon sites of involvement of synovial sarcoma are trunk, retroperitoneum, mediastinum and intestines, lungs, pleura, chest wall etc¹. Synovial sarcoma is classified into three subtypes a) monophasic b) poorly differentiated c) biphasic types. The ideal method to differentiate the subtypes is by morphology and immunohistochemistry. Synovial sarcomas are positive for epithelial membrane antigen (EMA), bcl-2, CD 99, S-100 and vimentin. They are negative for myoD1, and myogenin. Majority of synovial sarcomas show translocation involving SYT gene on chromosome 18q11 and SSX1 or SSX2 gene on chromosome X². In the thoracic cavity synovial sarcoma has been reported to arise from lungs, pleura, chest wall and mediastinum. Less than 20% arise in sites other than musculoskeletal system. The other soft tissue sarcomas that infrequently involve the mediastinum include rhabdomyosarcoma, sarcomatoid mesothelioma, leiomyosarcoma, angiosarcoma, malignant peripheral nerve sheath tumor, hemangiopericytoma etc. Synovial sarcomas are highly aggressive tumors with metastases to lungs being common.

II. Case Report:

Case 1: A 30 year old male patient presented with progressive shortness of breath and right sided chest pain of two months duration. He had nonproductive cough without hemoptysis. There was no history of fever or past history of pulmonary tuberculosis or any other systemic disorder. Chest radiograph revealed homogenous opacity in right hemithorax with mediastinal shift. Routine laboratory investigations were normal. CT chest showed a large 18.3 x 17.2 cm well defined solid heterogeneously enhancing mass in the right hemithorax. The mass displaced heart and insinuated to left side compressing right and left atrium. Right lung was displaced cranially and partially collapsed. There was minimal right side pleural effusion. Multiple subcentimeter prevascular lymph nodes were present. The left lung was normal. (fig 1 A,B,C). CT abdomen and neck were normal. PET-CT after injecting 11.5mCi of 18F FDG showed a solid mass with increase uptake (SUVmax- 8.6) along with cystic areas. The mediastinal lymph nodes showed increased uptake (SUVmax -2.8). The provisional diagnosis on CT imaging was large mediastinal tumor with lymphadenopathy. CT guided biopsy was performed. Case 2: A 50 year old male patient presented with chest pain, dry cough, shortness of breath since 3 months. There was no fever or any significant past history. Chest radiograph showed dense

opacity in right upper zone. Chest CT scan revealed enhancing mediastinal mass in right upper chest. MRI with contrast showed heterogeneously enhancing mixed signal intensity mass in right upper thorax. (fig 2 A,B). The histopathology from the mass in both patients showed sheets and ill defined fascicles of spindled tumor cells in a scant collagen stroma (fig 3 A). The cells have spindle to elongate hyperchromatic nuclei . Focally tumor cells are arranged radially around the blood vessels. Foci of necrosis were seen (fig 3 B). Immunohistochemistry report: CD34,S 100,Desmin, Calretinin , Pancytokeratin were negative in the tumor cells. CD99 and Bcl2 were positive in the tumor cells. TLE-1 was positive in tumor cells. MIB -1 index was about 25%.(fig 4). The above findings were consistent with synovial sarcoma.

III. Discussion:

Compared to primary pleural tumors, metastatic invasion of pleura is more common. The most common primary pleural mass is mesothelioma. The others being localized solitary fibrous tumor. pleural liposarcoma. As per WHO classification pleural tumors are divided into mesothelial, mesenchymal and lymphoproliferative types.(table 1)

Table 1: WHO histological classification of tumours of the pleura, according to the ICD-O

MESOTHELIAL TUMOURS	LYMPHOPROLIFERATIVE DISORDERS	MESENCHYMAL TUMOURS
1- Diffuse malignant mesothelioma (MM)	5- Primary effusion lymphoma	7- Epithelioid haemangioendothelioma
• Epithelioid mesothelioma	6- Pyothorax-associated lymphoma	• Angiosarcoma
• Sarcomatoid mesothelioma		8- Synovial sarcoma
• Desmoplastic mesothelioma		• Monophasic
• Biphasic mesothelioma		• Biphasic
2- Localised malignant mesothelioma		9- Solitary fibrous tumour of the pleura
3- Well-differentiated papillary mesothelioma		10- Calcifying tumour of the pleura
4- Adenomatoid tumour		11- Desmoplastic round cell tumour

As most of these pleural tumors present with similar clinical symptomatology, the radiological, histological features, precise diagnosis, preoperative histopathological and immunohistochemistry are very essential for better management.³

Diffuse malignant mesothelioma: The most common malignant tumor of the pleura, arising from the mesothelial cells of pleura. Patients present with progressive shortness of breath due to large pleural effusion. The CT scan findings include large pleural effusion with invasion of intercostal muscles, pleural thickening extending along the fissures and mediastinal lymph node deposits. The invasion of chest wall is better appreciated by MRI rather than CT.

Solitary fibrous tumor: Appears as well defined hypodense mass with minimal enhancement with contrast. Areas of hemorrhage, cysts, and necrosis are sometimes seen within the mass

Primary effusion lymphoma: Is usually associated with human herpes virus 8/Kaposi sarcoma herpes virus infection. On CT large pleural effusion with mediastinal lymphadenopathy seen.

Synovial sarcoma: Seen in young patients. It is a localized solid tumor of visceral pleura. The typical histological pattern seen is biphasic pattern with epithelial and spindle cells often associated with pseudocapsule. On CT synovial sarcomas show a large homogenous well defined mass with heterogeneous enhancement with contrast with associated hemorrhage or cystic changes. In primary pleural synovial sarcoma the adjacent ribs may show sclerosis without invasion of chest wall, whereas in extrapleural synovial sarcoma infiltration into adjacent chest wall muscles and erosion of ribs is noted. On MR imaging synovial sarcoma appears as a multilobulated soft tissue mass with signal intensity similar to muscle on T1 weighted images and hyperintensity on T2 weighted images due to the presence of cysts and hemorrhage. Enhancement with contrast is heterogeneous. Tumors that are proved to be synovial sarcomas by histopathological and immunohistochemical analysis are infrequently reported involving the mediastinum^{2,4,5,6,7,8}. Paul H Hartel⁹ reviewed the clinico – pathological findings in 60 cases of primary pulmonary and mediastinal synovial sarcomas. Besides the usual findings of synovial sarcoma described previously, they found certain uncommon features like Verocay body- like formations, vague rosettes, well formed papillary structures. They also concluded that unlike soft tissue synovial sarcoma, pulmonary and mediastinal sarcomas have less calcification, less obvious mast cell influx and decreased vascularity on imaging. Samer Se and Abu Salem¹⁰ reviewed the

different studies involving synovial sarcomas and found that mediastinal sarcoma have a poor prognosis compared to extremity synovial sarcomas as they are large tumors at the time of presentation.

Irappa et al ¹ reported a case of primary mediastinal synovial sarcoma presenting as superior vena cava syndrome which was completely treated with surgery and neoadjuvant chemotherapy. The present cases can be easily differentiated from more common pleural lesions like malignant mesothelioma by the absence of large pleural effusion, nodular pleural thickening or pleural calcification. Solitary fibrous tumors can be benign or malignant and are usually small localized pleural based tumors with occasional calcification , hemorrhage or cystic changes¹¹ . Primitive neuroectodermal tumor of thoraco pulmonary region (Askin tumor) show chest wall and lung invasion on CT and MR imaging. Angiosarcoma and epithelioidhemangioendothelioma of the pleura present with large hemorrhagic pleural effusion, loss of lung volume and diffuse pleural thickening. The treatment options include surgery and postoperative radiation in majority of cases. Combination of adriamycin and ifosfamid as adjuvant chemotherapy is advised for recurrent and unresectable tumors . Conclusions: synovial sarcoma is to be considered in the differential diagnosis of large unilateral intrathoracic tumors with hemorrhage or cystic changes and have to be confirmed by histopathological and immunohistochemical studies for initiating immediate treatment.



Fig 1. A)



Fig 1.B)

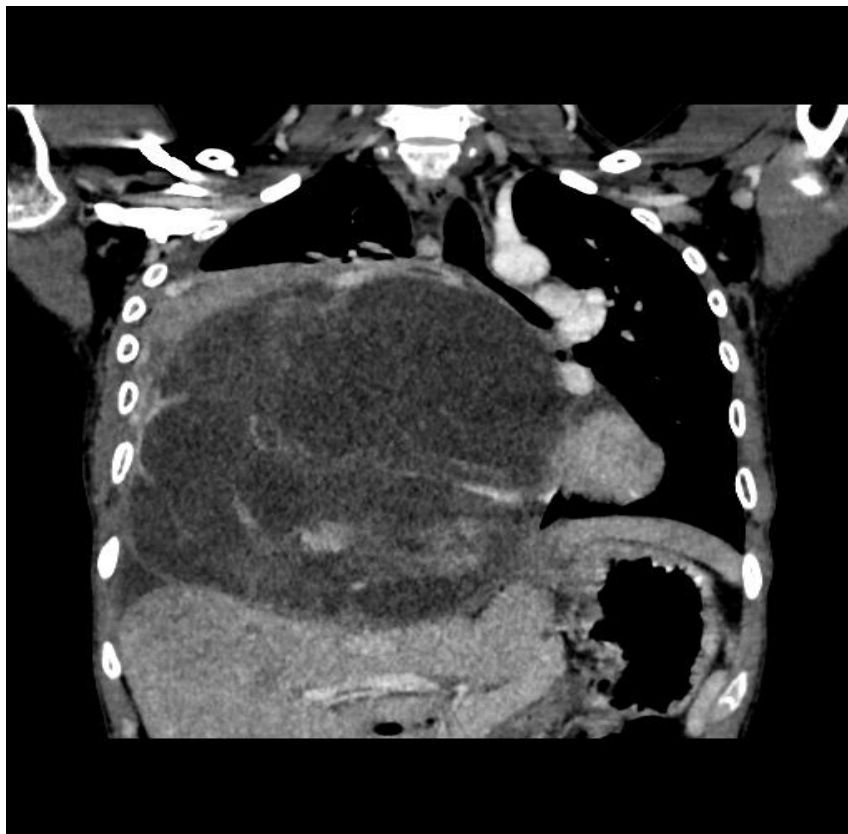


Fig 1.C)

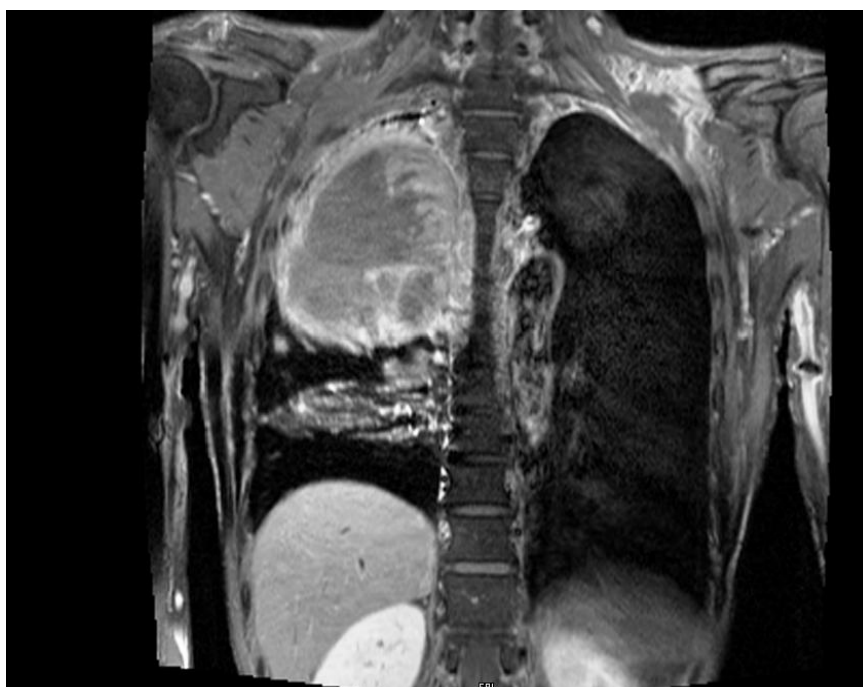


Fig 2 .A)

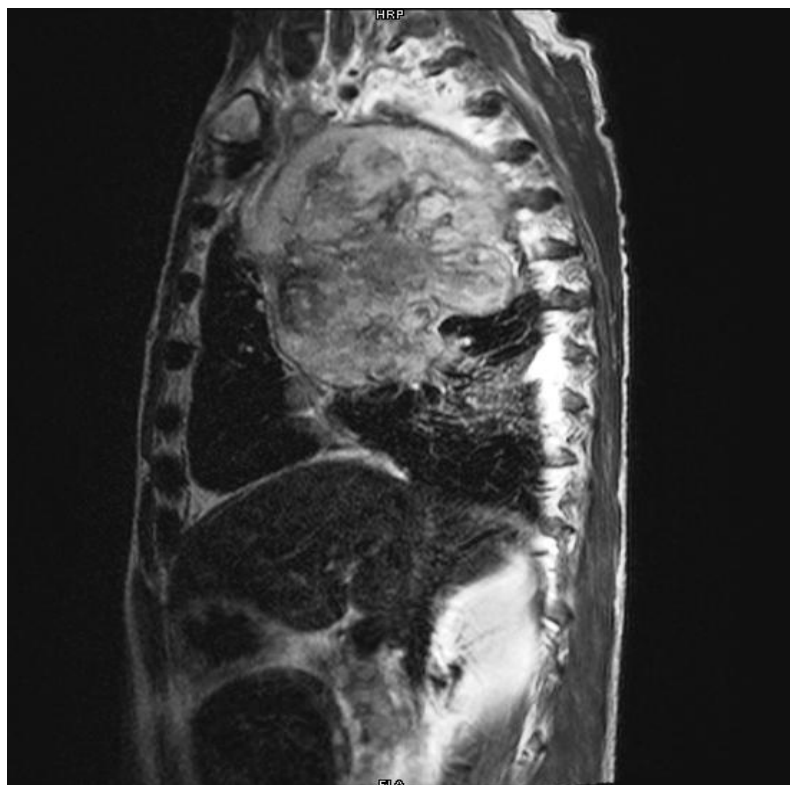


Fig 2.B)

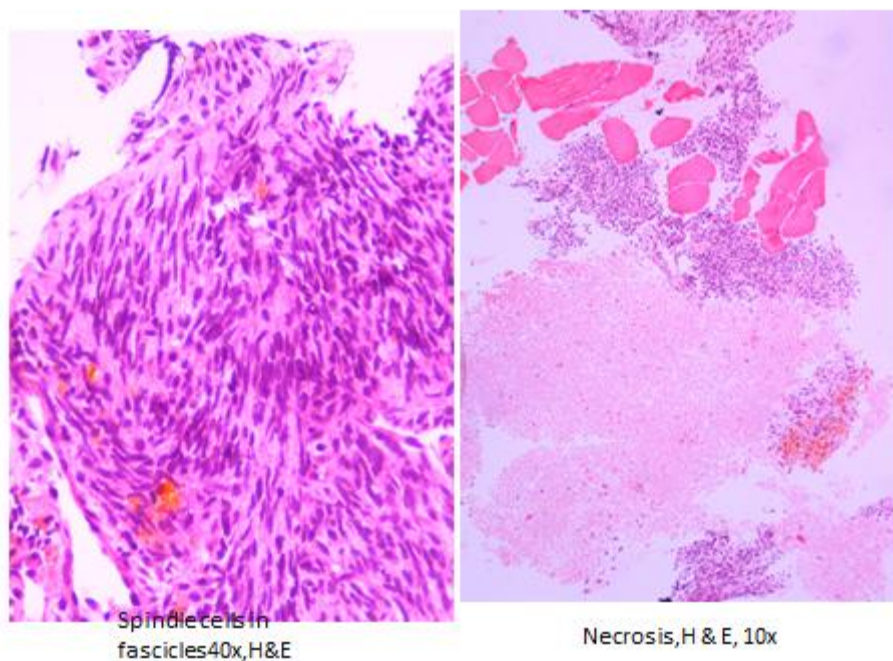


Fig 3) A& B

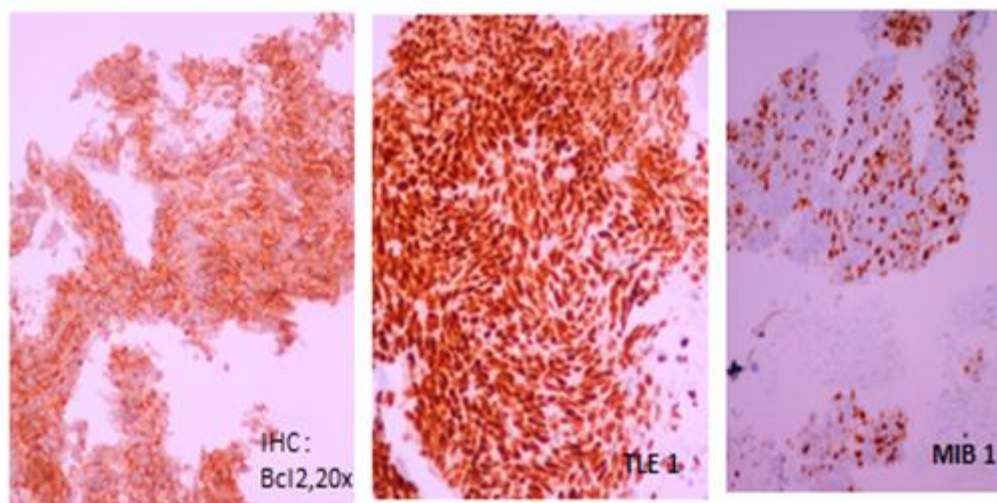


Fig 4)

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LEGENDS:

Fig 1. CE CT images show large heterogeneously enhancing mass in posterior mediastinum insinuating to left side.

Fig 2. Contrast MR Images show enhancing mass right hemithorax with segmental lung collapse

Fig 3. Histopathology showing spindle cells infascicles and foci of necrosis.

Fig 4. IHC show bcl2 stain in tumor cells, TLE-1 positive tumour cell nuclei, MIB-1 high in tumour cells

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