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Research Article

UNVEILING THE RARE AND GIANT: A CASE OF PRIMARY SYNOVIAL SARCOMA OF PLUERA

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Abstract

Background: Synovial sarcoma is a mesenchymal malignant spindle cell neoplasm. It most often occurs in the soft tissue of the extremities (lower around ankle, knee). Most pleural synovial sarcomas are metastatic. Primary synovial sarcoma of the pleura and lungs is extremely rare. Most frequently misdiagnosed as malignant mesothelioma. Case presentation: 55year-old female with a large synovial sarcoma of the right pleura. Patient presented with shortness of breath since 3 months duration. Chest x ray lateral view shows homogenous radio opacity with lobulated anterior margin extending from anterior mediastinum to posterior. Chest computed tomography (CT) revealed a 11 x 10.5 x 12.1cm large, well-defined, lobulated, pleuralbased solid lesion noted in the right hemithorax. Image guided biopsydone. Diagnosed as undifferentiated spindle cell sarcoma. She underwent right anterolateral thoracotomy and resection of tumour. The resected specimen contained a 13x 11x 8cm tumour diagnosed as biphasic synovial sarcoma based on its morphologic and immunohistochemical features. Advised Patient adjuvant radiation and chemotherapy. **Conclusion:** We presenting our experience of a large synovial sarcoma of the pleura in a patient with severe dyspnoea. She underwent right anterolateral thoracotomy and resection of tumour. Although the best treatment for this rare condition has not been defined, tumour resection and adjuvant therapy were able to decrease recurrence and increase in overall survival.

Keywords: Biphasic synovial sarcoma, Pleura, Anterolateral thoracotomy, Excision of tumour, Radiotherapy

INTRODUCTION

Synovial sarcoma is defined as mesenchymal malignant spindle cell neoplasm, featuring variable epithelial differentiation, including gland formation, and harbours a specific chromosomal translocation, t(X;18) (p11;q11), producing a SYT-SSX fusion gene. Patient present at any age, most commonly present in adolescents and young adults between the 10-40 years [1,2]. Male >female (2:1). Synovial sarcoma present around 10% of all soft tissue sarcomas[3],80 to 90 percent lesion seen at periarticular regions most commonly at lower extremities followed by trunk, head, neck (including larynx and hypo pharynx)and mediastinum[4].Synovial sarcoma is also present at visceral sites such as the lungs, pluera, gastrointestinal tract and kidney. Synovial sarcoma is a slow growing tumour, Up to 50% of cases of metastasise to the lungs, locoregional lymph nodes. Bone is the most common sites of systemic spread. Systemic spread to the lungs is the most common cause of death in metastatic synovial sarcoma. Surgical management with adjuvant radiotherapy and/or ifosfamide-based chemotherapy is the mainstay of treatment.

CASE PRESENTATION

A55-year-old female came with a cheif complaint of difficulty in breathing since 3 moths. Patient given a history of NO exposure to asbestos. Physical examination revealed oxygen saturation of 93% (room air) and decreased right vesicular sounds. Chest radiography showed a homogenous radio opacity with lobulated anterior margin extending from anterior mediastinum to posterior (Fig. 1).

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On chest computed tomography (CT), shows large, well defined, lobulated pleural based solid mass lesion measuring approximately 11.0 x 10.5 x 12.1 cm (AP X TR X CC) noted in the right apical region and upper posterior aspect of the right hemithorax with mass effect on the right main bronchus and bronchus intermedius with few non enhancing areas, extension-superiorly till lower border of the first rib; posteriorly till anterior surface of 1-6th posterior ribs without any obvious erosions; posteromedially up to right para vertebral space with fat planes maintained; medially abutting trachea, right main bronchus and right pulmonary artery and its branches without any infiltration. The lesion anteriorly causing mass effect on posterior segment of right upper lobe and displacing the right upper lobe bronchus without infiltration, lesion displacing horizontal fissure inferiorly and causing mass effect on superior segment of the right lower lobe (Fig 2A,B,& C). Image (CT)-guided trucut biopsy was done. The initial histopathology reported as undifferentiated spindel cell sarcoma, grade 2.

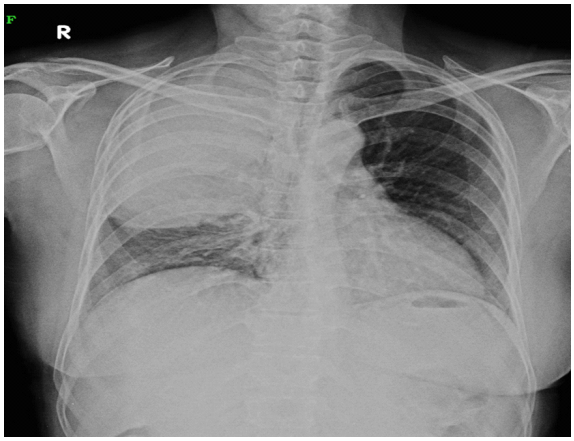
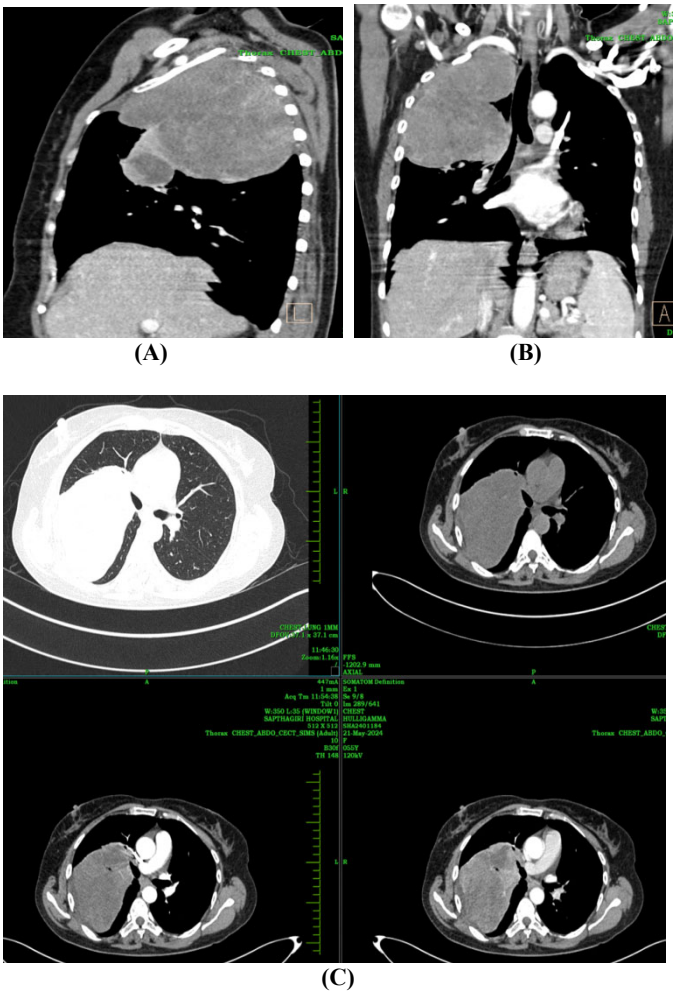


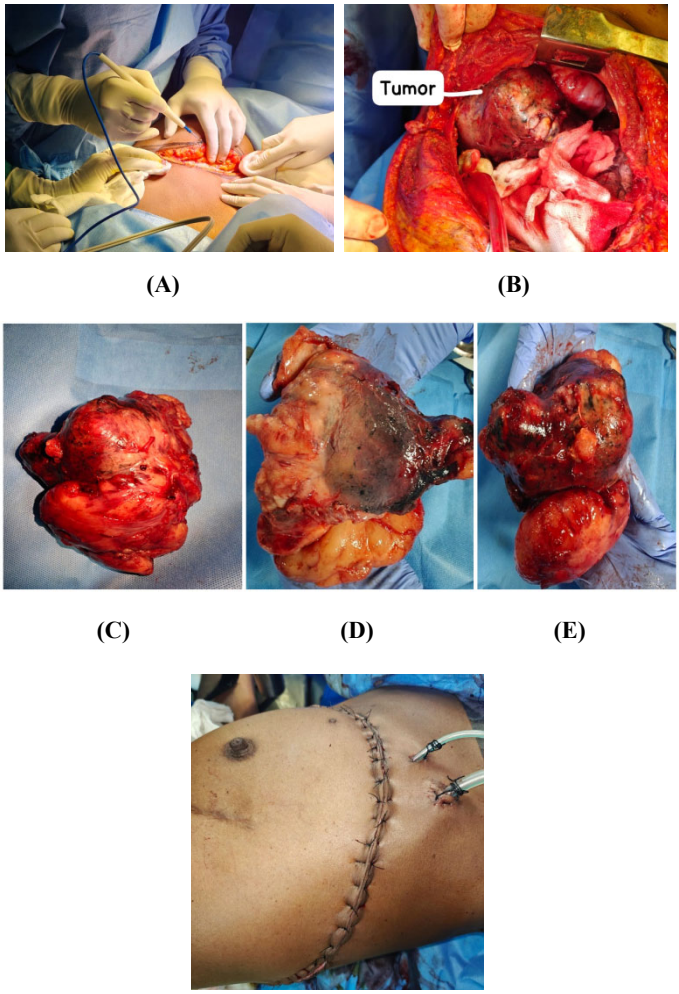
Fig. 1. Chest X ray



(A) (B)

(C)

Fig. 2. CT thorax



(A) (B)

(C) (D) (E)

(F)

Figure 3.

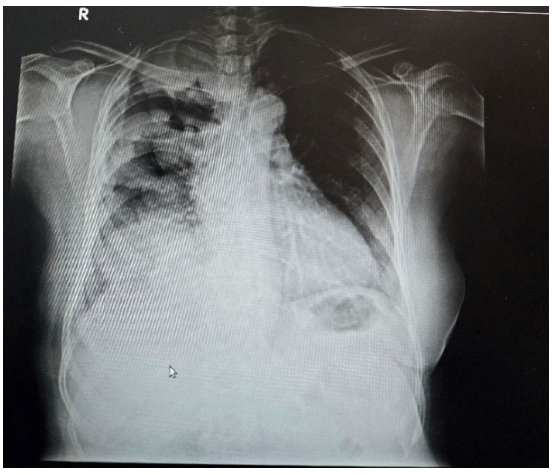


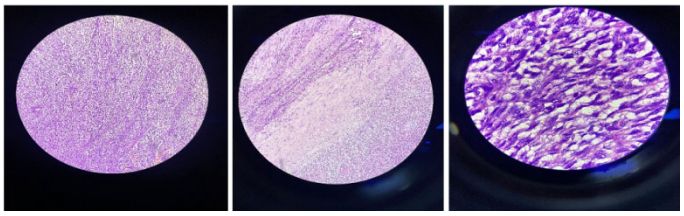
Fig 4. Post OP X Ray Chest

After preoperative evaluation and optimisation planned for surgical intervention. Under general anaesthesia with double lumen endotracheal tube. Position of the patient left Iateral decubitus with large pressure bag positioned beneath contralateral flank & ipsilateral hip was elevated. Right shoulder was in parallel plane. Followed by reverse trendelenburg. Parts painted and drapped. Right 5th intercostal space anterolateral thoracotomy incision taken (Fig 3 A,B,C,D,E, &F). Right lung collapsed. Firstly pulmonary artery identified in the oblique fissure and posterior oblique fissure was divided. Space was developed between superior and inferior veins. Then the anterior oblique fissure was divided. Solid lesion identified extra luminally in right middle lobe long with lung Parenchyma.Lesion was mobilised in its inferior endfor visualisation of the hilar region. The inferior vein is dissected free from surrounding tissue and divided. Tumour was identified along with the middle lobe, superiorly Retracted towards mediastinum. Tumour was separated from the surrounding structures by blunt dissection,tumour was adherent to the pleura membrane superiorly which was divided using electrocautery and was resected in toto. Haemostasis isachieved. Normal saline irrigationgiven, icdplaced and connected to water seal.Thoracotomy incision was closed in Layers. After shifting to icu bed side chest x ray done (Fig 4)

The surgical operating time was 210 min, and total blood loss was 1100 cc. Her postoperative course was uneventful and patient was discharged on postoperative day 7.Before discharging icd removed according to drain protocol.

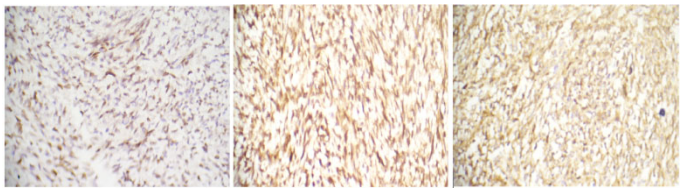
The final histopathology reported as 13 x 11 x 8cm, multifocal, undifferentiated, grade 2, necrosis <50%,0-9 mitosis/10 hpf no lymphovascular invasion, positive margins-medial, lateral, superior, UNDIFFERENTIATED SPINDEL CELL SARCOMA (Fig:5A, B,C). Differential diagnosis:Biphasic synovial sarcoma, fibrosarcoma. Immunohistochemicallythe tumour cells were positive for BCL-2, CD99, TLE1, CD56, vimentin, SMA and calponin (Fig. Negative for desmin, caldesmon, EMA, myogenin, betacatenin,STAT6, WT1, calretinin, CD117, DOG1, CD34, CD31, FL-1, S100 and SOX 10. Because the tumour cells were positive for CD99 and

BCL-2, which are usually negative in a case of malignant mesothelioma, malignant sarcomatoidmesothelioma seemed to be less of a possibility. Immunohistochemistry suggestive of biphasic synovial sarcoma. We diagnosed the tumour as a Biphasic synovial sarcoma based on its morphologic and immunohistochemical features. There was residual microscopic tumour, planned adjuvant radiation.



(A) (B) (C)

Figure 5.



(A) (B) (C)

Figure 6.

DISCUSSION

Overall, survival at 5 years is about 60% but drops to 30% at 10 years with no significant difference between monophasic and biphasic subtypes. Grossly, synovial sarcoma tends to be multinodular and well circumscribed, and relatively often features blood-filled cystic spaces (Fig. 4-172). Size is very variable and ranges from 1 to 10 cm. Poorly differentiated synovial sarcoma typically shows hemorrhagic areas and abundant necrosis. Microscopically, synovial sarcoma is further classified based on morphology into three main subtypes: (1) monophasic spindle cell synovial sarcoma; (2) biphasic synovial sarcoma; and (3) poorly differentiated synovial sarcoma. Monophasic spindle cell synovial sarcoma is composed of a cytologically uniform spindle cell population, organized in cellular sheets and fascicles, and set in a variably collagenous background. Biphasic synovial sarcoma is defined by the presence of variable amounts of epithelial differentiation in context with a spindle cell component that exhibits the same morphologic features of monophasic spindle cell synovial sarcoma. The poorly differentiated variant (which is associated with more aggressive behavior) accounts for approximately 20% of all synovial sarcomas and may exhibit three main cytomorphologies: large cell, spindle cell, and round cell. In retrospect, primary synovial sarcoma misdiagnosed as malignant sarcomatous mesothelioma. Despite having an aggressive clinical history, patients with synovial sarcoma typically live longer than those with mesothelioma. To confirm a diagnosis, ancillary testing such cytogenetic or immunohistochemical analyses are useful. Positive immunohistochemical results for cytokeratin and epithelial membrane antigen help differentiate synovial sarcoma from other pleural sarcomas. Neural (S100) and smooth muscle (desmin, smooth muscle act in) markers do not stain in synovial sarcomas[5] . Its utility is limited since this staining pattern resembles that of sarcomatous mesothelioma. Because it continuously stains biphasic synovial sarcoma but only becomes focally positive in certain mesotheliomas, the epithelial marker BerEp4 may be useful in differentiating between biphasic synovial sarcoma and malignant mesothelioma. It is less helpful, though, in differentiating between monophasic synovial sarcoma and mesothelioma, which also stains differently with BerEp4 . Regardless of histologic subtype, almost 90% of synovial sarcomas have been shown to have the chromosomal translocation t(X;18) (p11.2;q11.2)[6,7,8,9]. The SYT gene on chromosome 18 fuses with either the SSX1 or SSX2 gene on chromosomal X as a result of this translocation. The translocation or fusion transcript can be identified using a variety of methods, such as reverse transcriptase–polymerase chain reaction assay traditional cytogenetic analysis, and molecular cytogenetic analysis (FISH)[10,11,12]. Pleural synovial sarcoma has no known best course of treatment. Radiation, chemotherapy, and surgery have all been employed as multimodal therapies [13] . More resections may be necessary to treat recurrences, which are likely to occur. Synovial sarcoma in the extremities is very sensitive to chemotherapy, particularly doxorubicin and ifosfamide[14] .

Conclusion

We want to share our experience of treating a rare case of large synovial sarcoma of the pleura in a patient with dyspnoea, who underwent resection of tumour. For synovial sarcoma treatment has not been defined. Surgical management followed by adjuvant therapy were able to decrease local recurrence and increase in overall survival.

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