CASE REPORT

Cystic Pleural Synovial Sarcoma
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ABSTRACT
Fewer than 40 cases of primary pleural synovial sarcoma have been reported so far with only 3 cases of cystic synovial sarcoma including cases originating from sites other than the pleura. Here, we present an exceedingly rare case of cystic synovial sarcoma originating from the mediastinal side of the visceral pleura in a 25-year man presenting with hemoptysis. On contrast-enhanced computed tomography (CT), cystic synovial sarcoma and cystic thymoma were difficult to be distinguished due to mediastinal location. Histopathological examination showed spindled morphology of tumor cells with hypercellularity and nuclear atypia. As these features are associated with both monophasic fibrous synovial sarcoma and type A thymoma, immunohistochemistry was performed. A diagnosis of synovial sarcoma was confirmed by detection of CD99 and EMA and negativity of other markers. Fluorescence in situ hybridization (FISH) was not done. Surgical excision was done and followed by oncology referral.

Key Words: Thymoma. Cystic synovial sarcoma. Pleura. Mediastinum.

INTRODUCTION
Synovial sarcoma is a rare histological and clinical entity and very rarely it arises in thorax.¹ Primary synovial sarcoma of the pleura was first reported by Gaertner et al. in 1996.² Less than 40 cases have been reported to date.³,⁴ It is a malignant soft-tissue tumor of uncertain differentiation. Although focal cystic change is often present in synovial sarcoma, cystic synovial sarcoma is extremely rare.¹ To the best of authors' knowledge and research, there have been only three documented cases in the English literature, including cases originating from other than the pleural origin.¹

Herein, we present an extremely rare case of cystic synovial sarcoma in the anterior mediastinum.

CASE REPORT
A 25-year man presented at another facility with cough and hemoptysis. He was started on anti-tuberculous therophy, which he took for 15 days. His chest radiograph showed an opacity. His computed tomography (CT) scan showed 12 x 13 x 13 cm homogenous oval opacity along the left mediastinal border with no lymphadenopathy, lung, tracheal or bronchial involvement. Although focal cystic change is often present in synovial sarcoma, cystic synovial sarcoma is extremely rare.¹

Patient was put on Adriamycin + Ifosfamide + Mesna (AIM) chemotherapy for malignant fibrous tumor, got 6 cycles of chemotherapy and was on follow-up. Two years later, he presented with weakness and shortness of breath for 2 months.

His chest radiograph showed huge opacity. CT scan chest (Figure 1a) showed large low attenuation, sharply demarcated, homogeneous, soft tissue density, minimally enhancing mass in mediastinum bulging into left hemithorax causing collapse of the surrounding lung parenchyma, measuring 9.9 x 14.7 x 17.3 cm. Medially, the fat planes with pulmonary artery and left atrium were preserved with splaying of pulmonary veins. Rest of thorax was normal.

Core biopsy was done which showed spindle cell tumor; morphology and IHC, however, favoured synovial sarcoma (CD 99, EMA positive and CL, LCA, CD34, S100, SMA negative). Ultrasound abdomen and pelvis and bone scan were normal.

Median sternotomy (Figure 1b and c) was done. There was 20 x 22 x 25 cm huge cystic tumor compressing left lung laterally and encroaching upon left bronchus and main blood vessels, causing mediastinal widening.

Debulking of the tumor was done. The wall of the cystic lesion was ruptured during the operation and jelly-like material discharged. The tumor was found to be originating from the mediastinal aspect of the visceral pleura and was firmly adherant to the left lung, middle bronchus, superior vena cava and pulmonary vessels. Left pneumonectomy along with tumor removal was thus performed, separating it from the mediastinal pleura. Tumor was shaved at the region of SVC and pulmonary vessels.

Macroscopically, many mucoid tan brown tissue fragments collectively measuring 35 x 38 x 14 cm were received in formalin along with left lung with ruptured...
pleura. Cut surface was variegated tan brown to black. On opening along the bronchus, lung parenchyma was studded with tumor deposits, spongy in nature with hemorrhagic foci. Similar deposits were seen along the wall of bronchus. Histopathologically (Figure 1d), it was grade 3 synovial sarcoma. Microscopically, tumor cells were spindle shaped forming interlacing fascicles. They had mildly enlarged hyperchromatic nuclei with scant eosinophilic cytoplasm. Although nuclear atypia was moderate, cellularity was very high. Mitotic figures were approximately 40 per 10 high-power fields. Necrosis was apparent and its extent was 50%. There was no lymphovascular invasion. Lymph nodes were negative for tumor pT2b and pN0. Diagnosis of monophasic fibrous synovial sarcoma was considered, based upon morphological and immunohistochemical characteristics. CD99 and EMA were positive, CKAE1/AE3, Cam 5.2, LCA, CD34, CD5, ASMA, CD117, S100 and SMA were negative. The diagnosis of synovial sarcoma was thus confirmed.

Postoperative course was uneventful for a period of time, except for wound infection. At the time of third month postoperatively he was fine and referred to the oncologist for further management.

DISCUSSION

Synovial sarcoma is a rare mesenchymal tumor, accounting for 8 - 10% of all soft tissue tumors. It commonly occurs in adolescents and young adults, with slight male predilection. It is a highly aggressive malignant neoplasm and is not related to cigarette smoking. The diagnosis of primary pleural synovial sarcoma requires clinical, radiological, pathological, and immunohistochemical investigations to exclude alternative primary tumors and metastatic sarcomas.

Primary pleural synovial sarcomas have four subtypes - monophasic fibrous (spindle), monophasic epithelial, biphasic, and poorly differentiated. Monophasic subtype is most common subtype. Close differential diagnoses of monophasic subtype are fibrosarcoma, hemangio-pericytoma, leiomyo-sarcoma, and spindle cell variant of squamous cell carcinoma – as all are spindle cell neoplasms, consisting of spindle cell variety. Hence, immunohistochemistry is pivotal for differentiation among these and monophasic spindle cell synovial sarcoma. This case was characterized by the presence of spindle cell sarcoma on histopathological examination – the tumor cells being negative for cytokeratin, but positive for epithelial membrane antigen and CD99. Thus, diagnosis was primary synovial sarcoma of pleura invading left lung. A similar case of primary pleural synovial sarcoma was reported by Tajima et al.

About 66% of primary pleural synovial sarcomas are centrally located and usually present with post-obstructive pneumonia, atelectasis, and hemoptysis. Peripheral tumors are less common and usually asymptomatic. In this case, the adjacent lung and bronchus were infiltrated by tumor. Prognosis of primary pulmonary synovial sarcoma is poor.

Cytogenetic study by reverse transcriptase-polymerase chain reaction (RT-PCR) differentiates between monophasic and biphasic form. Synovial sarcoma is characterized by a reciprocal chromosomal translocation (X;18) (p-11.2; q11.2) which results from fusion of SYT gene on chromosome 18 to either of two genes, SSX 1 and SSX 2 on chromosome X. If the diagnosis of synovial sarcoma is certain or probable on the basis of clinical, histological, and immunohistochemical evaluations then molecular testing is not required despite its high sensitivity.

Treatment includes surgical resection, followed by adjuvant chemotherapy or radiotherapy. Surgical resection with clear resection margins is the main treatment modality, chemo and radio are less effective. Prognosis is variable, depending on grade of tumor. 5-year survival rate is estimated to be 50%. Adverse prognostic factors include stage of disease, increasing age (>20 years), male gender, large tumor size (>5 cm), and high mitotic activity (>9 mitoses per 10 HPF). This patient fell in the poor prognosis group.

Synovial sarcoma is an aggressive tumor with a poor prognosis. Recurrences are very common and patients need to undergo consecutive surgical resections. Cadmen and co-workers reported recurrence in 91.5%, MacKenzie in 28%, Vieta in 46.4%, and Cantin in 59%.

The 5-year disease-free survival was 20.9%. Long-term follow-up in literature indicated that the outcomes in terms of recurrence and death was poor with a 2-year disease-free survival rate of 35.7%. The results also showed that the most common site of recurrence was thorax. Due to minimal clinical data, exact follow-up strategy is not known; and each clinician individualises...
the follow-up to 3-monthly CT scan chest and suspicious lesions be biopsied. Spindloid cells with specific immunohistochemistry clinches the diagnosis of recurrence.

This is an exceedingly rare case of cystic synovial sarcoma. Due to its occurrence in the visceral pleura near the anterior mediastinum and its adherence to the mediastinal pleura, distinguishing between cystic synovial sarcoma and cystic thymoma, was difficult on contrast-enhanced CT. Histopathological examination revealed a tumor composed of spindle cells. This feature alone could not differentiate between the two suspected tumor types because monophasic fibrous synovial sarcoma and type A thymoma (i.e. the spindle cell variety), both take the form of spindled cells. Therefore, immunohistochemistry was essential for determining the tumor type. CD99 and EMA positivity and negativity of all the other markers caught the definitive diagnosis of synovial sarcoma. Thus extensive clinical examination, followed by full body CT scan, and histopathology with immunohistochemistry led to the diagnosis of primary pleural synovial sarcoma in this case.

REFERENCES