INTRODUCTION

Synovial Sarcoma is an uncommon soft tissue tumor typically occurring in young and middle aged adults. It arises in the vicinity of extremities in close proximities of large joints and has a very aggressive behavior. It accounts for approximately 5% of all the adult soft tissue sarcomas. It does not originate from the synovial tissue but arises from pleuripotent mesenchymal tissues; hence the term “synovial sarcoma” is a misnomer. Other reported sites for synovial sarcoma are head and neck, lung, heart, mediastinum, and abdominal wall. Synovial sarcoma is associated with a local recurrence and distant metastases with the most common site affected being lungs. In patients with extremity sarcoma around 20% will develop pulmonary metastases at some point in the disease course. Metastatic pulmonary synovial sarcoma from extremities is much more common as compared to pulmonary sarcomas which carries a poor prognosis having an overall 5-year survival rate of 50% and comprise only 0.5% of all the primary lung malignancies. On histology these tumors are classified as biphasic, monophasic (purely epithelioid or purely fibroblastic) or poorly differentiated. Molecular testing is pathognomonic for t(x;18) chromosomal translocation which has enabled diagnostic confirmation in approximately 90% of cases. In t(x;18)-negative cases, diagnosis must rely on histological and immunophenotypic features. The mainstay of treatment remains surgical resection.

Case Report

A 68 years old lady presented in chest outpatient department with complaints of dyspnea on exertion, cough with minimal expectoration and right sided chest pain for 2 months. There was no history of fever, wheezing, hemoptysis, significant weight loss and decreased appetite. The severity of chest pain and dyspnea were gradually increasing.

Her past medical history and family history was unremarkable. She was a non-smoker. On general examination there was no evidence of pallor, clubbing, enlarged neck veins, cyanosis and palpable lymph nodes. Her temperature was 37.8 °C, respiratory rate was 30 breaths/min, pulse rate 90 beats/min, blood pressure 130/80 mm Hg and SPO2- 90%. Chest examination showed bilateral VBS (vesicular breath sound) decreased air entry on right side and no added sounds. Examination of other systems revealed no abnormality.

Her complete hemogram and blood biochemistry were within normal limits. The sputum was negative for acid fast bacilli, Gram-stain and culture. The Chest X-ray showed right sided homogenous opacity. On contrast enhanced computed tomography (CECT) chest there was a soft tissue mass in right hilar region which was causing obliteration of right upper lobe.
bronchus and subsegmental collapse. Few subcentimetric mediastinal lymph nodes were also identified. Fiberoptic Bronchoscopy (FOB) revealed an endobronchial mass. A biopsy was taken from the mass and sent for histopathology.

The pathological specimen consisted of multiple grey brown soft tissue pieces measuring 0.8x0.6x0.3 cm. Microscopic examination revealed spindled and epithelioid malignant cells arranged in sheets, clusters and cords containing large irregular hyperchromatic nuclei and scanty cytoplasm. On immunohistochemistry (IHC) these cells were positive for cytokeratin, p63, vimentin, CD99 and negative for S-100, NSE, chromogranin, LCA, WT-1, CK20, TTF, Bcl2, CD56, ER, PR. The final diagnosis rendered was pulmonary Synovial Sarcoma.
DISCUSSION

The term Synovial Sarcoma (SS) is derived from the morphological similarity to the embryonic synovial glands and is often misinterpreted to mean that the tumor originates from synovial tissue, which does not hold true. SS has been proposed to originate from myogenic cell lines and occurs in soft tissues almost anywhere in the body, most frequently in the lower (62%) and upper (21%) extremities. Synovial sarcoma usually occurs in young and middle-aged adults and has a predilection for the extremities and accounts for approximately 5% of all adult soft tissue sarcomas. Primary pulmonary synovial sarcoma is an extremely rare tumor and has a very aggressive course. In previous reported cases males were commonly affected with an average age of 25 years and presenting symptoms of chest pain, cough, dyspnea or hemoptysis. In most of the cases large pleural based heterogenous intraparenchymal masses were seen. Histopathological types are: Biphasic, monophasic fibrous, monophasic epithelial type, and poorly differentiated type. Biphasic synovial sarcoma being the most common and classical type. It comprises of epithelial cells surrounded by spindle shaped cells forming solid, compact cords or sheets. Pulmonary synovial sarcoma should be differentiated from other primary pulmonary malignancies such as malignant fibrous histiocytoma, fibrosarcoma, intrapulmonary solitary fibrous tumor, malignant mesothelioma, adenocarcinoma, and carcinomas. Histology is often supplemented with IHC studies for a better diagnosis. On IHC synovial sarcomas are uniformly positive for cytokeratin, EMA, bcl2 and vimentin and negative for S-100, desmin, smooth muscle actin. CD99 is positive in 50-100% cases. FNAC of the lung mass may be inconclusive, which is very helpful in the diagnosis of lung carcinoma. Hence image-guided needle biopsy, open lung biopsy, thoracoscopic biopsy is advised to make a proper diagnosis.

The clinical course of synovial sarcoma is frequently protracted and may mimic other primary pulmonary tumors but in large proportion of cases the tumor eventuates in the death of the patient. Thus, synovial sarcoma has generally been regarded as a high grade sarcoma and treated with adjuvant radiation therapy and chemotherapy. Primary synovial sarcoma arising from the lung is closely associated with smoking and carries a very poor prognosis as it is a very aggressive tumor with a 5-year survival rate of 50%. Factors accounting for poor prognosis are: size> 5cm, male gender, older age (>20 years), extensive tumor necrosis, high count of mitotic figure (>10/10 HPF), neurovascular invasion and local eradication of the growth. Few recently done cytogenetic studies have states that synovial sarcoma has a consistent chromosomal translocation t(x;18)(p11;q11) and that this translocation fuses the SYT gene to either of the two homologous genes SSX1 or SSX2. These fusion genes are thought to be associated with tumor initiation, but the target gene is still remains unidentified.

There is no standardized therapy but due to the relative resistance of sarcoma to either chemotherapy or radiotherapy compared to other solid tumors, surgical management has been a pivotal therapy in the disease. However some studies report that synovial sarcomas are chemosensitive to ifosfamide and doxorubicin, with a response rate of around 24%. Adjuvant chemotherapy may improve the time to local recurrence and recurrence free survival rate and may result in a better overall survival rate.

CONCLUSION

Not all the pulmonary malignancies are bronchogenic carcinoma, pulmonary synovial sarcoma although rare, should be considered while making a diagnosis. Clinically it may mimic other lung tumors. Thus, histopathology is a must for establishing a diagnosis. Immunohistochemistry is preferably done for subtyping of this rare tumor. Cytogenetic studies for t(X;18) should be advised for the confirmation.

References