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A Rare Cause of Lung Mass in Young Adult: a Case Report

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Abstract: Synovial sarcoma is usually seen as palpable masses in the large joints of the extremities. Pulmonary involvement is an extremely rare accounting for less than 0.5% of all lung tumors. Commonly seen in young people. Chest pain, dyspnea, cough, and hemoptysis are the common symptoms. Histopathology and cytogenetic analysis are useful in differentiating from other spindle cell neoplasms. Treatment for primary pulmonary synovial sarcoma includes surgical resection, chemotherapy, and radiotherapy with high recurrence rate.
Keywords: Primary pulmonary synovial sarcoma, Histopathology, Lung mass.

INTRODUCTION

Synovial sarcomas (SS) account for nearly 5–10% of all soft tissue sarcomas[1,2].SS usually seen as palpable masses in the large joints of the extremities, but also could occur in neck, tongue, larynx, mediastinum, esophagus, heart, lung, abdomen wall, small intestine, mesentery, vessels, and retroperitoneum.Primary pulmonary synovial sarcoma (PPSS)is aggressive tumor and was first described by Zeren *et al*. in 1995[3]. PPSS is an extremely rare tumor;accounting for less than 0.5% of all lung tumors[4].Here we report a rare case of primary pulmonary synovial sarcoma in a young male.

CASE REPORT

A 30 years old male patient, nonsmoker and non-alcoholic came with left side chest pain, breathlessness, cough with occasional white mucoid expectoration for the past three months.There was no history of fever, wheeze, hemoptysis, anorexia, and significant weight loss. He had no significant comorbidities. On general examination there was no pallor, clubbing, cyanosis, icterus and palpable lymph nodes.His temperature was normal,pulse rate 83 beats/min,Blood pressure was 110/70 mmHg, Respiratory rate 24 breaths/min.Respiratory system examination revealed reduced movement of left hemithorax, shifting of trachea to right, dull percussion note on the left side, diminished breath sounds on left side. Examination of other systems revealed no abnormality.

Complete blood picture, renal function tests, liver function tests were normal.ECG,2-D Echo showed no abnormality and viral screening was normal. Chest radiograph (PA view) showed well defined large opacity in the left upper and mid zone (Fig 1).Sputum culture was negative and no acid fast bacilli detected. USG abdomen was normal. CT scan of the chest revealed a well-defined heterogeneously enhancing mass lesion involving left upper lobe (Fig 2).

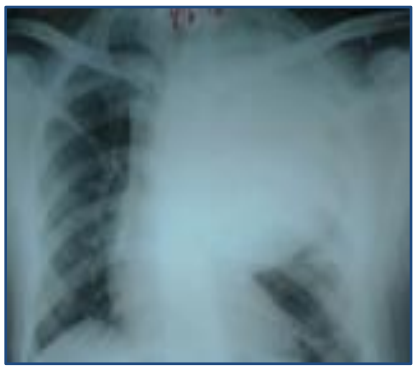

Fig-1: Chest radiograph showing homogenous opacity involving left mid and lower zone

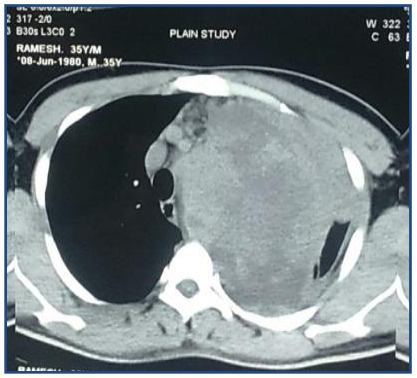

Fig-2: Computed tomography showing large heterogeneous mass lesion involving left upper lobe

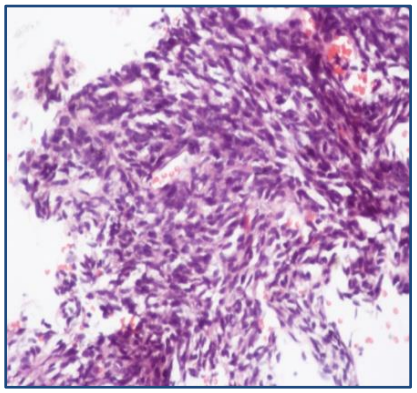

Fig-3: H&E stain showing prominent hemangiopericytomatous pattern. The lesional cells are spindle shaped with oval to elongate hyper chromatic nuclei and scant cytoplasm. These cells are interrupted by vascular channels lined by flat endothelial cells. Scattered mitotic figures are also seen

Histopathological examination of CT guided biopsy from mass lesion suggested mesenchymal neoplasm possibility of synovial sarcom(Fig3).On immunohistochemistry; spindle-shaped tumor cells were strongly positive for bcl-2 and vimentin, nonspecific positivity for synaptophysin.CD_{34}positive in endothelial cells and negative in lesional cells. TTF-1 and Pan CK were negative(Fig 4a-f).Further studies with FISH revealed positive for SSX1 and 18 (SYT) rearrangements which is gold standard for diagnosis of synovial sarcoma.Bone scan showed no uptake anywhere in the body.

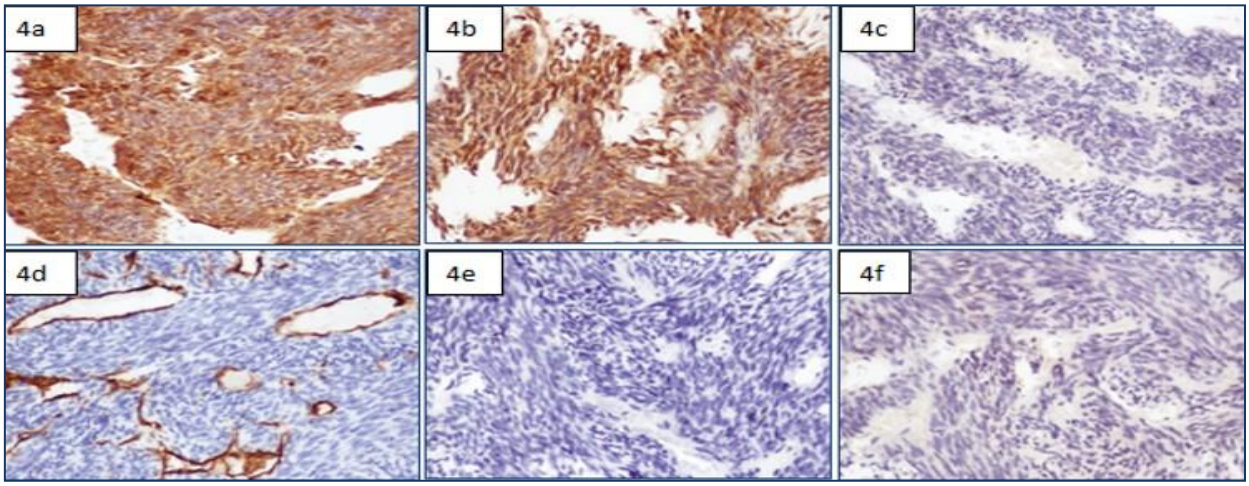

Fig-4a-f: Immunohistochemistry of tumor:Tumor cells were strongly positive for bcl-2(4a) and vimentin (4b), nonspecific positivity for synaptophysin (4c).CD_{34} positive in endothelial cells and negative in lesional cells(4d). TTF-1(4e) and Pan CK (4f) were negative.

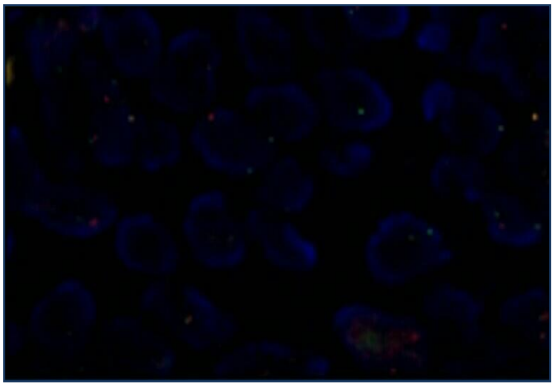

Fig-5: FISH revealed positive for SSX1 and 18 (SYT) rearrangements.

DISCUSSION

Primary synovial sarcomas are most common in the soft tissue near the large joints of the arm and leg but also occur in other organs, including the brain, prostate, and heart. Synovial sarcoma occurs in about 2 per 100,000 people a year [5]. PPSS commonly seen in age between 31 and 50 (range 3–84) years. It affects both sexes equally with similar incidence on right and left lungs [6].

Typical signs and symptoms include chest pain, dyspnea, cough, and hemoptysis. In the largest patient series, 40% of patients were asymptomatic, and PPSS was anincidental finding at chest radiography [3].

On chest radiographs, the mass appears homogeneous, and evidence of cavitation, calcification, or lymphadenopathy is rare. On CT images, the mass usually shows heterogeneous enhancement and comprised nodular soft-tissue components mixed with areas of low attenuation and Ipsilateral pleural effusion is common[7].

Histologically synovial sarcomas are of four subtypes – monophasic fibrous (spindle), monophasic epithelial, biphasic, and poorly differentiated, monophasic subtype being most common [8,9].Epithelial and spindle cell components are present biphasic subtype. Monophasic subtype needs to be differentiated from other spindle cell neoplasms like fibrosarcoma, hemangiopericytomatous, leiomyosarcoma and spindle cell variant of squamous cell carcinoma, for which immunohistochemistry and FISH is essential.

Histology is usually supplemented with immunohistochemistry. Synovial sarcomas are usually uniformly positive for Cytokeratin, vimentin, Bcl-2 and EMA and negative for S-100, desmin, smooth muscle actin and vascular tumor markers [7]. In our case immunohistochemistry positive for bcl-2 and vimentin, nonspecific positivity for synaptophysin,CD_{34}positive in endothelial cells and negative in lesional cells. TTF-1 and Pan CK were negative.Histology and IHC have been recently supplemented by cytogenetic analysis which is gold slandered for diagnosis of synovial sarcoma. Cytogenetic studies of synovial sarcoma have revealed the chromosomal translocation t(x: 19) (p11;q11).This translocation fuses the SYT gene from chromosome 18 to either of homologous genes at

Xp11,SSX1 or SSX2.SYT-SSX1 and SYT-SSX2 are thought to function as aberrant transcription regulator[6]. The sensitivity of this test is 100%.Cytogenetic analysis of our patient revealed presence of t(x: 18).

Although there is no gold standard of treatment for primary pulmonary synovial sarcoma, a multidisciplinary approach, including surgical resection, chemotherapy, and radiotherapy has been suggested. Radical resection is the mainstay of treatment. Adjuvant radiotherapy is usually recommended in cases of incomplete resection or extensive resection of a large tumor. The benefits of chemotherapy are unclear. However, neoadjuvant chemotherapy can be beneficial prior to radical resection since it can cause reduction in tumor volume[10].Synovial sarcoma has a response rate of approximately 50% to ifosfamide and doxorubicin [11].Our patient referred for surgical excision.

The prognosis of the synovial sarcoma is poor with an overall 5 year survival rate of 50%.Factors predicting a worse prognosis include tumor size(>5 cm),male gender,older age(>20 years),extensive tumor necrosis,large number of mitotic figures(>10 per 10 hpf),neurovascular invasion,SYT-SSX1 variant[12].

CONCLUSION

Sinovial sarcoma is rare tumor that commoly effects the large joints of the extremities. Pulmonary involvement is very rare. Commonly seen in young people. Histopathology and cytogenetic analysis are useful in differentiating from other spindle cell neoplasms.Even though a rare entity Primary pulmonary synovial sarcoma should be considered in differential diagnosis of lung masses especially in young patients.

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