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A RARE CASE OF PRIMARY PULMONARY SYNOVIAL SARCOMA

General Medicine

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ABSTRACT

Background: A 22 years old young married female presented with complaints of dry cough, breathlessness, decreased appetite and low-grade fever for 2 months. No history of chest pain, hemoptysis and loss of weight. The patient was on treatment for infertility for the last 3 months. Examination revealed trachea shifted to the left side, stony dull note on percussion, reduced intensity of breath on the right side of the chest. A diagnosis of right-sided pleural effusion -? tubercular etiology was made. Pleural fluid analysis revealed exudative effusion with lymphocytic predominance. Therapeutic thoracocentesis was done along with CAT-1 ATT. Repeat imaging presence of a mass lesion in the right pleural cavity. The patient was further evaluated with Mediastinal cyst fluid analysis, biopsy, PETCT. The patient underwent right pleural cyst excision. The immunoprofile of mediastinal solid mass is consistent with "SYNOVIAL SARCOMA". As PETCT was normal, the patient was not started on chemo or radiotherapy and was advised to review on a regular basis.

Conclusion: Primary synovial sarcoma is a very rare malignancy of lung, 0.5% of overall lung cancers. The most appropriate management is surgical excision. Due to high rates of recurrence a very close follow up is needed. Highly aggressive and with poor prognosis.

KEYWORDS

Primary pulmonary sarcoma, Lung sarcoma, synovial sarcoma lung, sarcoma.

INTRODUCTION:

Synovial sarcoma is a soft tissue tumor which arises from pleuripotent mesenchymal tissue. It most commonly occurs in the extremities and also reported to occur in the lung, mediastinum, abdomen, head and neck. Metastatic synovial sarcoma from extremities is the most common in pulmonary parenchyma and pleura. Only 0.5% of all primary pulmonary malignancies is due to pulmonary sarcomas. Herein, we present a case report of young female with primary synovial sarcoma of lung.

CASE REPORT:

A 22 years old young married female presented with 2 months' history of dry cough, breathlessness, decreased appetite and low grade fever. No history of chest pain, haemoptysis and loss of weight. She was on treatment for infertility for last 3 months. No prior history of tuberculosis.

On examination, the patient was vitally stable without signs of respiratory distress or nail clubbing. Cardiorespiratory examination was notable for left-sided tracheal deviation, displaced apical heartbeat. Reduced chest movements, stony dull note on percussion and absent breath sounds on right hemi thorax.

Complete blood count, Renal and Liver function tests were normal, ESR: 25mm/1st hour. HIV, HBAsAg, HCV were non-reactive. Pleural fluid analysis showed exudate with lymphocytic predominance, normal ADA levels, no atypical or malignant cells. Ultrasound abdomen showed 2 cysts in left ovary measuring 2.1x1.6cm and 2.9x1.9cm with septations and haemorrhage.

Chest X-ray showed complete opacity of the entire right hemi thorax with shifting of trachea and heart to the left side [Fig1]. Contrast-enhanced computed tomography (CT) scan of Thorax showed a cystic lesion in the right hemi thorax measuring 13x11x8 cm with compression atelectasis of right upper lobe of lung and mediastinal shift to left [Fig2].

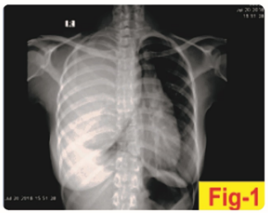


Fig-1

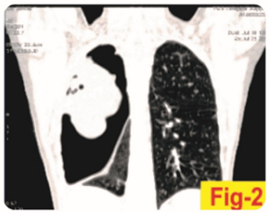
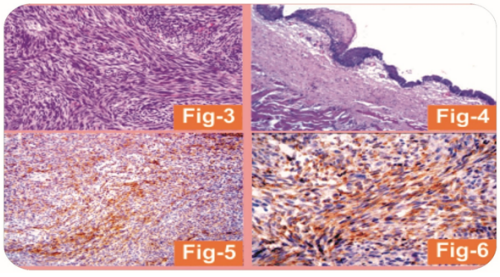


Fig-2

CT guided biopsy showed spindle shaped cells arranged in whorls and solid nests [Fig3]. The portions of cyst wall lined by epithelium showing ulcerations [fig4]. The neoplastic cells showed membrane positively for CD99, cytoplasmic positivity for Bcl2, CK, EMA and a proliferative index of 70-80% [fig5&6]. PETCT showed no evidence of residual disease, any other visceral or skeletal lesions.



Based on above characteristics, the final diagnosis of monophasic synovial sarcoma of lung was made. Patient underwent right pleural cyst excision. Post-operative period was uneventful. As PETCT did not reveal secondaries, the patient was not started on chemo or radiotherapy, and was advised follow up review on regular basis.

DISCUSSION:

Primary synovial sarcoma of lung is a very rare tumor with few cases reported in literature. The term synovial sarcoma in lung is a misnomer, named due to its resemblance to synovium on light microscopy. Ipsilateral pleural effusion is common, which was in this case.

It is a rare mesenchymal tumor, accounting for 10% of all soft tissue sarcoma. Synovial sarcoma occurs most commonly in adolescent and young adults. It occurs most commonly in the soft tissues of the extremities, especially near large joints, but head and neck, lung, heart, mediastinum, and abdominal wall sites also have been reported. Pulmonary sarcomas are very uncommon and comprise only 0.5% of all primary lung malignancies.

Patients with primary synovial sarcomas of the lung may present as chest pain, cough, shortness of breath, or hemoptysis. On imaging, initial chest radiographs may show well-defined opaque lesions of varying sizes; sometimes associated with tracheal deviation, mediastinal shift, and pleural effusion. CT, magnetic resonance

imaging, and PET/CT scans are largely utilized for staging the neoplasm, assessing its resectability, planning the surgical resection and afterward evaluating degree of response to therapeutic interventions.

Histologically there are 4 subtypes in primary pulmonary synovial sarcomas – monophasic fibrous (spindle), monophasic epithelial, biphasic, and poorly differentiated, monophasic subtype being most common. Differential diagnosis of monophasic subtype are fibrosarcoma, leiomyosarcoma, hemangiopericytoma, and spindle cell variant of squamous cell carcinoma as all are spindle cell neoplasms.

Immunohistochemistry is essential, to differentiate monophasic subtype of synovial cell sarcoma from others. On immunohistochemistry, synovial sarcomas are nearly uniformly positive for cytokeratin, EMA, bcl-2, and vimentin and negative for S-100, desmin, smooth muscle actin, and vascular tumor markers. Although rarely used, the most crucial method in establishing the definitive diagnosis of mediastinal synovial sarcomas is cytogenetic testing. This is because 90% of such neoplasms exhibit a characteristic reciprocal chromosomal translocation (t[x; 18] [p11.2; q11.2]).

The diagnosis of primary pulmonary synovial sarcoma requires clinical, radiological, pathological, and immunohistochemical investigations to exclude alternative primary tumors and metastatic sarcoma. The present treatment includes surgical resection, followed by adjunctive chemotherapy or radiotherapy. Complete surgical resections of mediastinal synovial sarcomas whenever technically possible is the standard of care. External beam radiation therapy (EBRT) is the most frequent therapeutic modality used in patients with primary unresectable, metastatic and incompletely debulked mediastinal synovial sarcomas. Synovial sarcomas are characterized by their moderate sensitivity to chemotherapeutic regimens comprising doxorubicin and ifosfamide.

Mediastinal synovial sarcomas carry poorer prognosis compared to the extremity synovial sarcomas, with an estimated overall 5-year survival rate of 35.7%. Hence, long-term follow-up is advised. Prognosis depends on the size of tumor and the age of patient. Other factors are male gender, extensive tumor necrosis, high grade, large number of mitotic figures (>10/hpf), and SYT-SSX1 variant. SYT-SSX1 variant is associated with more aggressive phenotype and more tumor cell proliferation.

CONCLUSION:

Primary synovial sarcoma of lung is a very rare tumor, with poor prognosis. The diagnosis of primary pulmonary synovial sarcoma requires clinical, radiological, pathological, and immunohistochemical investigations to exclude alternative primary tumors and metastatic sarcoma. The most appropriate management is the surgical excision with negative margins, whenever feasible. Adjuvant chemotherapy and radiotherapy have a limited role. Due to high rates of recurrence, a very close follow-up is needed.

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