Case Report

A rare case of primary synovial sarcoma of lung

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ABSTRACT

Synovial sarcoma of lung is a very rare tumor accounting for 0.5% of all primary lung malignancy. It presents clinically with cough, chest pain, shortness of breath, or hemoptysis, with a mass lesion on X-ray and computerized tomography scan. Diagnosis is made by histopathology and immunohistochemistry. Here, we report a case of 48-year-old male, who presented with right-sided chest pain, cough with blood-tinged sputum, and found to have primary pulmonary synovial sarcoma of lung.

KEY WORDS: Lung sarcoma, synovial sarcoma lung, sarcoma, primary pulmonary sarcoma

INTRODUCTION

Most common site of synovial sarcoma is periarticular tissue, but it has also been reported from variety of other sites.[1] The lung is a rare site of primary synovial sarcoma. Primary pulmonary sarcoma accounts for 0.5% of all lung malignancies.[2] Metastasis to lung, from other organ, is far more common than primary pulmonary synovial sarcoma. Pulmonary synovial sarcoma requires multimodality treatment with surgery, radiation therapy, and chemotherapy.

CASE REPORT

A 48-year-old male presented with the right-sided chest pain, cough associated with blood-tinged sputum for the last 2 months. Pain was insidious in onset, dull aching in nature. He had one episode of hemoptysis, of around 70 ml, for which he was admitted in intensive care unit and supportive care was given.

Patients’ history was unremarkable with no prior history of pulmonary tuberculosis. There is no history of smoking or alcohol intake. On general physical examination, he was pale. There was no cervical lymphadenopathy or clubbing. On systemic examination, there was decreased respiratory movement on the right supraclavicular and infraclavicular area. The corresponding area was dull on percussion with decrease breath sound.

Chest X-ray revealed large homogenous opacity in right upper and middle zone. Contrast-enhanced computed tomography (CT) scan showed a 5.4 cm × 4.4 cm × 6.1 cm soft tissue density in apical segment of right upper lobe. The lesion was abutting superior vena cava. CT-guided biopsy from lung mass showed infiltrating spindle cell neoplasm into the lung parenchyma. The tumor cells were monomorphic in fascicle having high nuclear cytoplasmic ratio. Lymphovascular tumor emboli and perineural invasion were seen. On immunohistochemistry, tumor cells were positive for EMA and Bcl2 while negative for CK, S100, CD34, desmin, SMA, CK7, CK20, HMB45, and mesothelin. Ki67 was 20%. Based on above characteristics, the final diagnosis of monophasic synovial sarcoma of lung was made [Figure 1].

The patient underwent right upper lobectomy of lung with negative margins. After surgery, six cycles of adjuvant therapy were given.

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How to cite this article: Rajeev LK, Patidar R, Babu G, Suresh Babu MC, Lokesh KN, Patil Okaly GV. A rare case of primary synovial sarcoma of lung. Lung India 2017;34:545-7.
chemotherapy with ifosfamide and doxorubicin were given. Postadjuvant chemotherapy patient is kept on regular follow-up.

**DISCUSSION**

Synovial sarcoma occurs most commonly in adolescent and young adults. It is a rare mesenchymal tumor, accounting for 10% of all soft tissue sarcoma.[1] 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.

Synovial sarcoma arising from the lung has rarely been reported.[2] 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 wall pain, cough, shortness of breath, or hemoptysis. The average age at presentation is 25 years.[3] The diagnosis of primary pulmonary synovial sarcoma requires clinical, radiological, pathological, and immunohistochemical investigations to exclude alternative primary tumors and metastatic sarcoma.[3]

Primary pulmonary synovial sarcomas are of four subtypes – monophasic fibrous (spindle), monophasic epithelial, biphasic, and poorly differentiated, monophasic subtype being most common.[4,5] Biphasic subtype has both epithelial and spindle cell components. Differential diagnoses of monophasic subtype are fibrosarcoma, hemangiopericytoma, leiomyosarcoma, and spindle cell variant of squamous cell carcinoma as all are spindle cell neoplasms. Hence, to differentiate monophasic subtype of synovial cell sarcoma from others, immunohistochemistry is essential. 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.[6]

Cytogenetic study by reverse transcriptase-polymerase chain reaction helps differentiate monophasic and biphasic form. Synovial sarcoma is characterized by a reciprocal chromosomal translocation (X;18) (p11.2; 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.[7] SYT–SSX 1 gene is associated with biphasic subtype and prognosis is bad, whereas monophasic subtype may have either one of two fusion transcripts, SYT–SSX 1 or SYT–SSX 2. All tumors with SYT–SSX 2 gene show monophasic morphology. Despite its high sensitivity, molecular testing is not required if the diagnosis of synovial sarcoma is certain or probable on the basis of clinical, histological, and immunohistochemical evaluations.[7]

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.[7] SYT-SSX translocation is a characteristic of synovial sarcoma, found in more than two-third patients. SYT-SSX1 variant is associated with more aggressive phenotype and more tumor cell proliferation.

The present treatment includes surgical resection, followed by adjunctive chemotherapy or radiotherapy.

**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 surgical excision with negative margins, whenever feasible. Adjuvant chemotherapy and radiotherapy have limited role. Due to high rates of recurrence, a very close follow-up is needed.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Acknowledgment**
I would like to thank Dr. Vikas Asati for technical support.

**Financial support and sponsorship**
Nil.

**Conflicts of interest**
There are no conflicts of interest.
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