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Case Report

Primary Pulmonary Synovial Sarcoma-Rare Case Presentation

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Abstract

Pulmonary Synovial Sarcoma is a rare primary lung malignancy with 0.1 - 0.5% incidence rate. Surgical excision followed by adjuvant chemotherapy with or without radiotherapy is the treatment of choice in these malignancies due to poor prognosis. In this case report we discuss a 39 years old male, who presented with right sided chest pain and occasional hemoptysis, diagnosed to have pulmonary mass lesion confined to right upper lobe infiltrating middle lobe. Thoracotomy followed by bilobectomy and mediastinal lymph node dissection of right side was performed. Histopathology was suggestive of biphasic synovial sarcoma. Immunohistochemical staining was positive for TLE-1, EMA, cytokeratin, CD56 and negative for CD34, TTF-1 suggestive of intermediate grade spindle cell sarcoma consistent with biphasic synovial sarcoma with no evidence of lymphovascular invasion. Postoperatively, patient was subjected to six cycles of adjuvant chemotherapy.

Keywords: *Pulmonary Synovial Sarcoma; Bilobectomy; Primary Malignancy; Chemotherapy*

Introduction

Pulmonary Synovial Sarcoma is one of the rare primary lung malignancy. It is considered to be distinct from other lung tumours based on clinical presentation, pathological features and prognosis. Surgical excision followed by adjuvant chemotherapy with or without radiotherapy is the treatment of choice.

Case Report

Thirty-nine years old male presented with right sided chest pain associated with hemoptysis occasionally for 9 months. There were no other symptoms present and patient had no history of smoking. Patients vitals were stable. Routine blood investigations were within normal limits.

Chest x-ray was suggestive of mass lesion in the right upper and middle zone (Figure 1A). Contrast enhanced computed topography (CECT) thorax was suggestive of heterogeneously enhancing mass lesion in right upper lobe abutting right middle lobe and costal pleura with no evidence of invasion to mediastinal lymph nodes and chest wall (Figure 1B). CT guided biopsy attempted twice was inconclusive. CECT Brain, CT abdomen and bone scan were normal.

Bronchoscopy did not show endobronchial growth and broncheoalvelolar lavage was negative for malignant cells. Endobronchial ultrasound was negative for mediastinal lymph node metastasis.

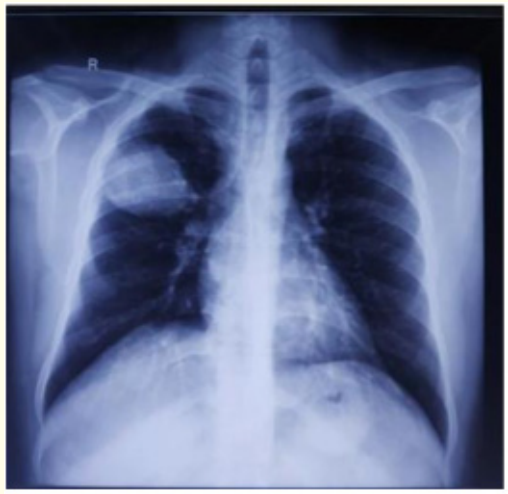

Figure 1A: *Chest x-ray showing mass lesion.*

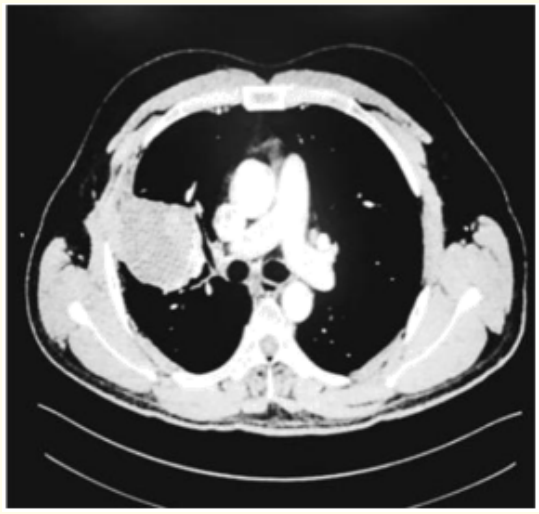

Figure 1B: *CECT thorax showing mass lesion.*

He was planned for posterolateral right upper lobectomy with mediastinal lymph node dissection under general anaesthesia. Intraoperatively patient had large mass lesion in right upper lobe infiltrating middle lobe with fused fissure. Hence he underwent bilobectomy with mediastinal lymph node dissection of stations 2, 4, 7, 9, 10. Postoperative condition was uneventful. Drains were removed on third postoperative day.

Gross specimen (Figure 2A) pathologically consisted of large cavitatory lesion in the right upper and middle lobe with fused fissure. Histopathology (Figure 2B) was suggestive of intermediate grade spindle cell sarcoma consistent with biphasic synovial sarcoma with no evidence of lymphovascular invasion. Immunostaining showed diffuse strong positivity for TLE-1, patchy positivity for EMA, cytokeratin, CD56 and negativity for CD34 and TTF1. Postoperatively patient has received adjuvant chemotherapy.

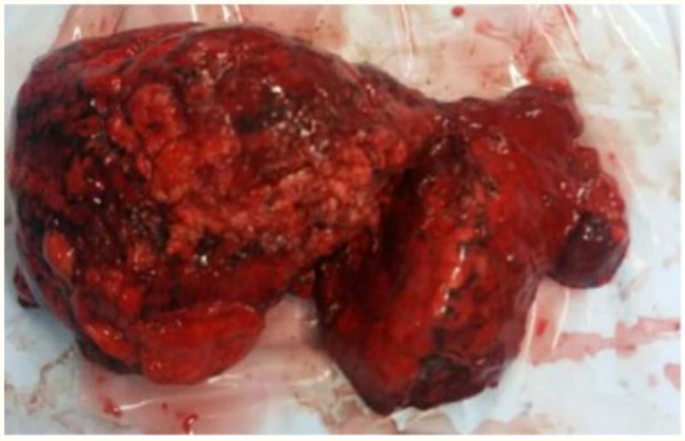

Figure 2A: *Gross specimen.*

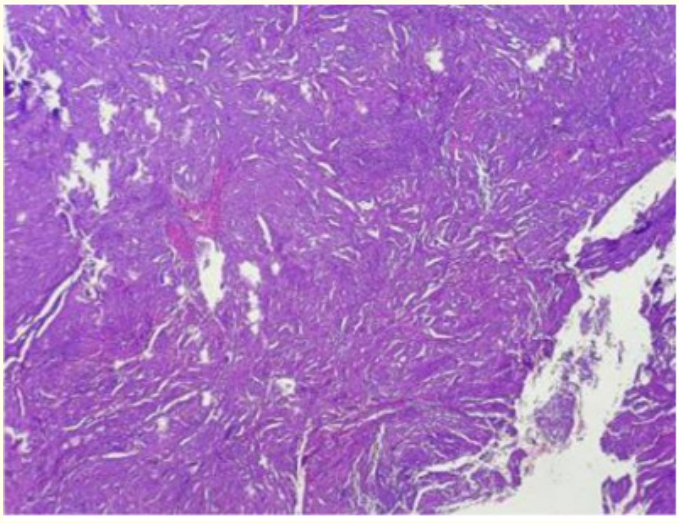

Figure 2B: *Histopathology.*

Discussion

Synovial sarcoma is a rare mesenchymal tumour accounts for 8 - 10% of soft tissue sarcoma. It was described in 1865 by Simon. More than 90% synovial sarcomas are located in the extremities and 0.1 - 0.5% constitute primary lung malignancy [1]. It can arise from the parenchyma, tracheobronchial tree or pulmonary artery and it is classified by World Health Organization as a mesenchymal tumour [2].

Clinical manifestation depends on the histological type, region, size and degree of differentiation of the tumour. Symptoms vary from being asymptomatic to symptomatic with chest pain, hemoptysis, dyspnea. Peak incidence is between 4^{th} and 7^{th} decade of life. It usually as male preponderance.

Histological subtype includes biphasic, monophasic (spindle), monophasic epithelial and poorly differentiated tumours [3]. Most commonly observed subtype is monophasic. Biphasic variety is easily diagnosed on the basis of presence of epithelial and spindle cells.

Immunohistochemistry is essential when making a differential diagnosis as monophasic variety can be misdiagnosed as other type of sarcoma. Cytogenetic studies have reported a translocation t(X;18) (p11.2; q11.2) for definitive diagnosis [4].

There is no current standardized therapy for patients with primary pulmonary synovial sarcoma. Complete surgical resection is the treatment of choice. Prognosis is generally poor with overall five-year survival rate of 50%. Prior to surgery extra pulmonary metastasis or primary focus of synovial sarcoma has to be excluded by imaging studies.

In advanced or unresectable tumours, chemotherapy is advisable. Role of radiotherapy is usually indicated in patients with positive resectable margins. Tumour size, higher histological grade, male gender, mitotic rate, extensive tumour necrosis and neurovascular invasion are negative prognostic factors. The expression of SYT-SSX1 variant has been associated with worse prognosis.

Conclusion

Primary pulmonary synovial sarcoma is a rare and aggressive tumour. Definitive diagnosis depends on clinical imaging and immunohistochemical staining. Surgical resection along with adjuvant chemotherapy with or without radiotherapy is the treatment of choice. Prognosis is poor.

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