Case Report

A Rare Case of Primary Pulmonary Synovial Sarcoma

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Abstract

Primary pulmonary synovial sarcoma is a very rare tumour. The recommended treatment includes surgery and chemotherapy with or without radiotherapy. We report the case of a 29-year-old female who presented with left-sided chest pain and heaviness, pleural effusion and a large mass filling left hemithorax. She was treated by surgical excision, histopathological and immunohistochemical findings confirmed the diagnosis of synovial sarcoma (monophasic type). **[Indian J Chest Dis Allied Sci 2016;58:251-252]**

Key words: Immunohistochemistry, Lung, Synovial sarcoma.

Introduction

Synovial sarcoma is a highly malignant mesenchymal tumour that occurs mostly in young adults and adolescents, the usual location being in extremities. Primary pulmonary sarcoma is a very rare malignancy and comprises less than 0.5% of all primary lung carcinomas.[1] It originates from multipotent mesenchymal cells. We report here the clinical profile and management of primary pulmonary synovial sarcoma in a young female.

Case Report

A 29-year-old female presented to surgical clinic with left-sided chest pain, breathlessness and occasional cough for the duration of four months. The onset was insidious and complaints were gradually progressive in nature, other history was unremarkable. The patient was a non-smoker and non-alcoholic.

On examination there was a dull percussion note with decreased breath sounds in left hemithorax. Rest of the physical examination did not reveal any abnormality. Chest radiograph revealed a dense shadow in left-side of chest with blunting of cardiophrenic angle. High-resolution computed tomography (HRCT) of chest revealed a mass lesion (20cm×14cm×8cm) involving both left upper and lower lobes with mediastinum shift to the right side with pleural effusion (Figure 1). Computed tomography-guided biopsy done from the left lung mass revealed a spindle cell tumour. Pleural fluid cytology was negative for malignancy.

Patient underwent left-sided pneumonectomy with mediastinal lymph node dissection. The excised pneumonectomy specimen was 22cm×15cm×8cm in size. Gross pathological examination revealed a 20cm×10cm, well-demarcated fleshy, brownish and necrotic mass in continuity with bronchial tree. Histopathological examination showed poorly differentiated monophasic spindle cell sarcoma (Figure 2). On immunohistochemistry (IHC) staining tumour cells were strongly positive for vimentin and

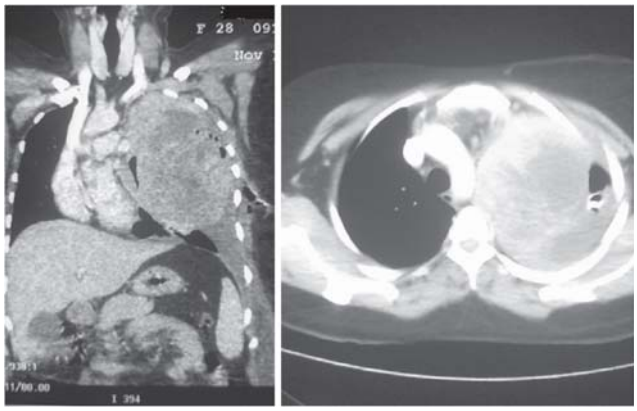

Figure 1. Computed tomography of chest showing a large heterogeneous mass involving left lung with few air loculi within and shifting of mediastinal structures to the right side.

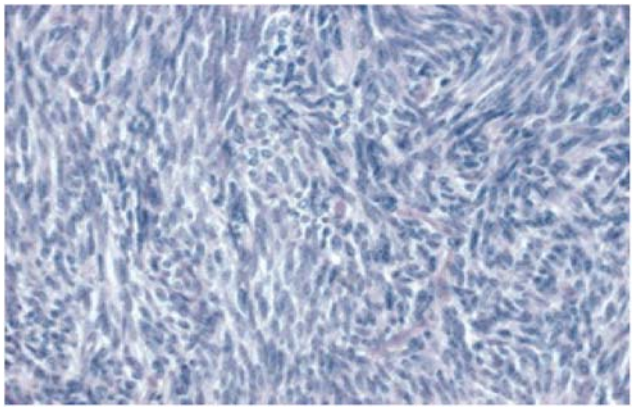

Figure 2. Photomicrograph showing a monophasic synovial sarcoma with apparent fascicular arrangement of spindle cells (Haematoxylin and Eosin × 400).

[Received: March 16, 2015; accepted after revision: September 18, 2015]

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B–cell lymphoma 2 (BCL2) and focally positive for epithelial membrane antigen (EMA) and S-100 (Figure 3). Tumour cells were negative for cytokeratin (CK), smooth muscle actin (SMA), cluster of differentiation 99 (CD99) and cluster of differentiation 34 (CD34) with mitotic rate of 12/10 high power field.

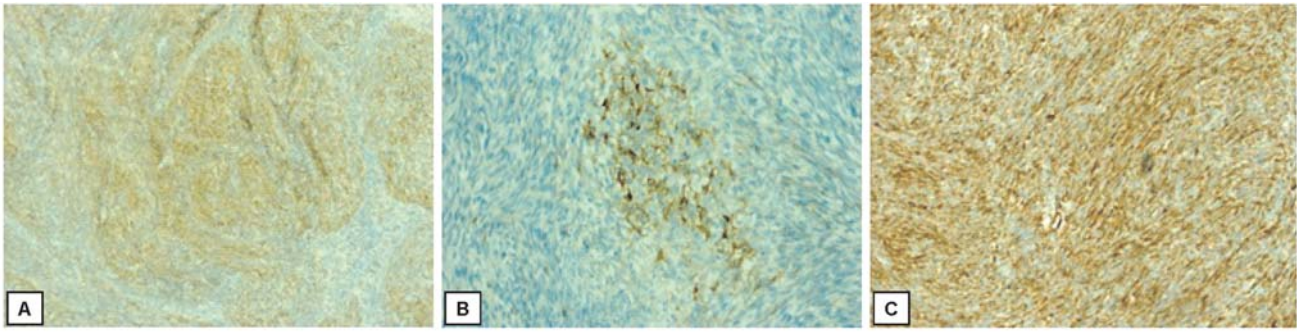

Figure 3. Photomicrographs showing immunohistochemistry staining patterns (× 200) of synovial sarcoma with (a) B–cell lymphoma, (b) epithelial membrane antigen and (c) vimentin.

On the basis of morphological and immunohistochemical findings, a diagnosis for spindle cell synovial sarcoma (monophasic type) was made. Post-operative period was uneventful. Following surgery ifosfamide and adriamycin based chemotherapy was administered. Patient is on regular follow-up and is asymptomatic at present.

Discussion

Synovial sarcoma is a rare mesenchymal tumour and accounts for 10% of all soft tissue sarcomas, being more common in men.[1] Synovial sarcoma typically presents in young adults between 15-30 years of age and commonly occurs in extremities, but neck, heart, lung, mediastinum and abdominal wall sites have also been reported.

Primary pulmonary sarcoma is a very rare lung malignancy in clinical practice.[2] There are four histological subtypes which include biphasic, monophasic epithelial, monophasic spindle and poorly differentiated.[3] The monophasic subtype is most common subtype in lung and it is difficult to diagnose because of uniform spindle cell pattern.[2] Therefore, immunohistochemistry is essential to differentiate monophasic type from other types of sarcoma. Synovial sarcoma is usually positive for vimentin, BCL2 and EMA but negative for S-100 and desmin.[3] In our patient, the excised specimen revealed poorly-differentiated monophasic spindle cells which were positive for vimentin, BCL-2 and EMA.

Both monophasic and biphasic types are characterised by a reciprocal translocation (X; 18) (p11.2; q11.2) in most of the cases. This translocation results from the fusion of SYT gene from chromosome 18 to either of the two highly homologous genes at Xp11, SSX1 or SSX2. SYT-SSX1 and SYT-SSX2 are thought to function in aberrant transcriptional regulation.[4,5] This is usually the only abnormality that occurs in all variants of synovial sarcoma.

Primary pulmonary synovial sarcoma are rare tumours with 60% of cases being centrally located. These patients usually present with obstructive pneumonia, haemoptysis, dyspnoea, cough and fever as in this case.[6] The overall prognosis of primary pulmonary synovial sarcoma is poor and 5-year survival is approximately 50%. The bad prognostic factors includes male gender, old age (>20 years), tumour size (>5cm), extensive tumour necrosis, high grade and large number of mitotic figures (>10/high power fields).[7]

The primary treatment of pulmonary synovial sarcoma is surgical resection with negative margins with or without adjuvant chemotherapy and or radiotherapy.[6] In the present case, patient underwent left pneumonectomy with lymph node dissection and was started on chemotherapy post-operatively.

References

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