International Journal of Research in Medical Sciences
Bhagya Lakshmi A et al. Int J Res Med Sci. 2015 May;3(5):1269-1271
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: 10.5455/2320-6012.ijrms20150543

Case Report

Primary monophasic synovial sarcoma, a rare pulmonary mesenchymal neoplasm: report of a case with review of literature

A. Bhagya Lakshmi[1]*, K. Satya Sri[1], B. V. S. Kartheek[1], Nagarjun Rao[2]

[1]Department of Pathology, Andhra Medical College, Visakhapatnam, Andhra Pradesh, India
[2]Director of Surgical Pathology and Cytopathology, Department of Pathology, Medical College of Wisconsin, Milwaukee, WI 53226, USA

Received: 18 April 2015
Accepted: 21 April 2015

***Correspondence:**
Dr. A. Bhagya Lakshmi,
E-mail: dr.a.bhagyalaxmi@gmail.com

ABSTRACT

Synovial sarcoma is an uncommon malignant soft tissue tumour which occurs primarily in young adults. It most commonly occurs in the extremities near large joints, particularly the knee joint. Primary pulmonary synovial sarcoma has only occasionally been reported in the literature. We report a case of primary pulmonary monophasic synovial sarcoma in a 23 year old male patient presenting with recent onset of hemoptysis, cough and expectoration. Although an uncommon primary site, the characteristic histological and immunohistochemical features, and fluorescence in situ hybridization analysis for the characteristic (X;18) translocation, with thorough clinical and imaging correlation allowed a definitive diagnosis of primary pulmonary monophasic sarcoma. The mainstay of treatment for these unusual tumours remains complete surgical excision followed by adjuvant radiation and/or chemotherapy.

Keywords: Monophasic synovial sarcoma, Lung, Immunohistochemistry

INTRODUCTION

Synovial Sarcoma (SS) accounts for 10% of all soft tissue sarcomas.[1] Originally described by Simon in 1865 and named by Sabrazes in 1934.[2] Synovial sarcoma typically occurs around joints, mainly the knee.[2] Other sites include the head and neck, mediastinum, retroperitoneum, prostate and peripheral nerves.[3] Primary pulmonary SS, although rare with only a handful of published reports, represents one of the common subtypes of primary mesenchymal tumours of the lung. Males are affected more commonly than females, and most cases occur in adolescents and young adults (15 to 40 years).[4] Because of its rarity, the diagnosis of SS in unusual sites such as the lung is challenging. Pre-operative diagnosis on limited cytology and needle biopsy samples is particularly difficult, with potential for misdiagnosis. A definitive diagnosis requires detailed histologic examination on resection specimens supplemented by immunohistochemical staining and molecular analysis, with clinical and radiological correlation to exclude alternative primary sources.

CASE REPORT

Clinical presentation - A 23 year old male presented with hemoptysis and cough with expectoration of 6 months' duration. Physical examination revealed right infrascapular and infra-axillary basal crepitations on chest auscultation. Routine laboratory investigations were unremarkable. A chest radiograph revealed left pleural effusion. Contrast Enhanced Computed Tomography (CECT) of the chest revealed a large, well demarcated, smooth surfaced, mixed cystic/necrotic and solid,15 cm lesion occupying majority of the left thoracic cavity, confined by the visceral pleura, with associated pleural effusion. The underlying lung was completely collapsed. The radiologic differential diagnosis included several

different neoplastic processes including teratoma, Ewing's sarcoma/peripheral neuroectodermal tumor (PNET), neuroblastoma and pleural fibroma. The patient underwent a left pneumonectomy.

Pathologic findings - On gross examination, the left pneumonectomy specimen measured 18x15x8 cm, with a 12x8x8 circumscribed, yellow-white, soft, fleshy tumour involving the peripheral lower lobe, with compression of adjacent lung parenchyma. The cut surface showed areas of haemorrhage, necrosis and cystic change (Figure 1). Hematoxylin and Eosin (H&E) stained sections showed a neoplasm with alternating densely cellular and relatively hypocellular areas. The cellular areas comprised of fascicles of spindle cells with oval to elongated nuclei with moderate nuclear pleomorphism, inconspicuous nucleoli, and scanty eosinophilic cytoplasm. Variable mitotic activity was present. The hypocellular areas had a slightly myxoid background. Other features included a focally prominent hemangiopericytic pattern, cystic change and necrosis (Figure 2, 3). Differential diagnostic considerations were malignant solitary fibrous tumor and monophasic synovial sarcoma. Immunohistochemistry revealed tumor cell positivity for Vimentin**,**CD99 (MIC-2) and CD56, with focal staining for bcl-2; whereas pancytokeratin, Epithelial Membrane Antigen (EMA), Smooth Muscle Actin (SMA), chromogranin A and CD34 were negative. The immune-profile was consistent with SS. Fluorescence *In Situ* Hybridization (FISH) using "breakapart" DNA probes for SYT locus on chromosome 18, had the characteristic split signals indicative of the t(X;18) translocation (Figure 4). A final diagnosis of monophasic SS, fibrous type, was made. The patient is now undergoing adjuvant chemoradiation treatment.

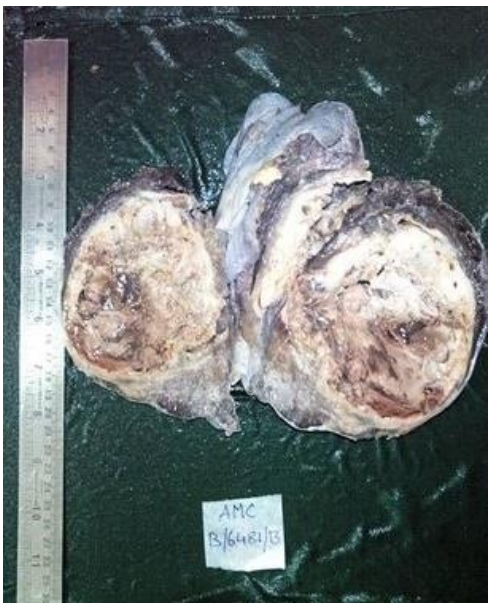

Figure 1: Gross photograph of the left pneumonectomy specimen on cut section showing a 12x8x8 cm circumscribed, yellow-white, soft, fleshy tumour with areas of haemorrhage, necrosis and cystic change.

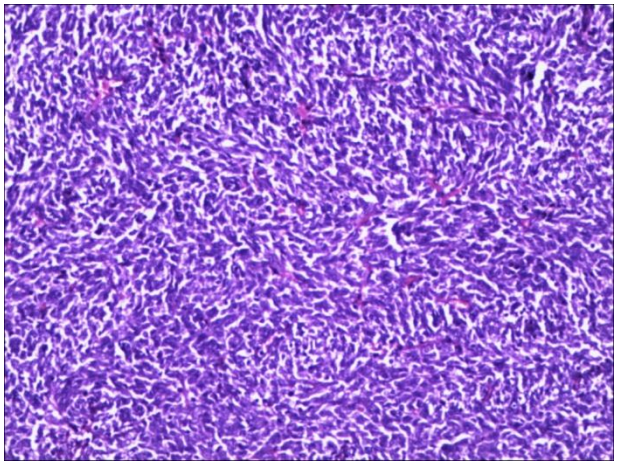

Figure 2: Photomicrograph showing fascicles of spindle shaped tumour cells. (H&E; 100x).

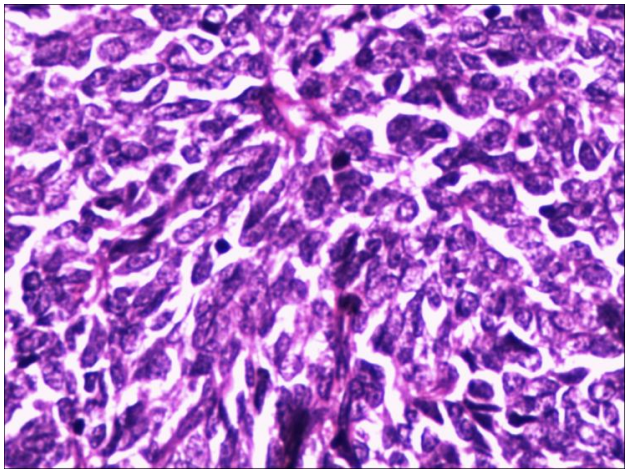

Figure 3: Photomicrograph showing spindle cells with oval to elongated nuclei with moderate nuclear pleomorphism, inconspicuous nucleoli, and scanty eosinophilic cytoplasm. (H&E; 400x).

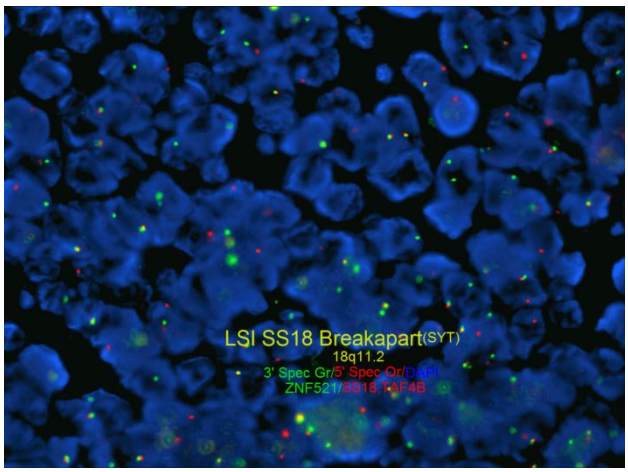

Figure 4: Fluorescence in situ hybridization (FISH) using "breakapart" DNA probes for the SYT locus on chromosome 18, showing the characteristic split signals indicative of the t(X;18) translocation.

DISCUSSION

Metastases from peripheral soft tissue sarcomas account for the vast majority of mesenchymal neoplasms of the lung. Primary pulmonary sarcomas account for less than 0.5% of all lung cancers.[5] SS accounts for 5-10% of all soft tissue sarcomas.[6] In a recent large retrospective review comprising over 6000 soft tissue sarcomas, the incidence of SS was approximately 6%.[7] It is most prevalent in adolescents and young adults between 15 and 40 years of age, with an almost equal male:female ratio (1.2:1).[2]

2% of all mediastinal sarcomas are SS, and pulmonary SS comprise a small proportion of them, with a handful of case reports and small case series.[8] The average age at presentation is 25 years.[9] Histologically, there are two major categories of SS: biphasic and monophasic types. Biphasic SS has distinct epithelial and spindle cell components, in varying proportions. Vast majority of SS (including our case) are of the monophasic fibrous type. Monophasic fibrous SS may histologically resemble a number of other spindle cell neoplasms, of which the important ones include Solitary Fibrous Tumour (SFT), Malignant Peripheral Nerve Sheath Tumor (MPNST), leiomyosarcoma and spindle cell carcinoma. Immunohistochemistry is quite useful in resolving the differential diagnosis. SS show frequent positivity for bcl-2, CD99 (MIC-2) and vimentin; with focal staining for epithelial markers such as cytokeratin and Epithelial Membrane Antigen (EMA), even in monophasic SS. S-100, CD34, desmin and Smooth Muscle Actin (SMA) are typically negative. Although cytokeratin and EMA were negative in our case; the characteristic morphologic findings and positive immune-staining for vimentin, MIC-2 and bcl-2 (focally) allowed a reasonably definitive histologic diagnosis. Cytogenetic studies on synovial sarcomas have shown a consistent, specific translocation, most commonly a balanced reciprocal translocation [t(X;18)(p11;q11)] involving the SYT-SSX loci, in virtually all SS, regardless of subtype.[10] The sensitivity of this test for diagnostic purposes approaches 100%. Therefore a confirmatory diagnosis of primary pulmonary SS, as in our case, should include a thorough clinical and radiological investigation to exclude alternative primary sources, detailed immunohistochemistry, and identification of the specific translocation by conventional cytogenetics or FISH.[5] Great caution should be applied in making the diagnosis of pulmonary SS on limited biopsy and/or cytology material (unless supported by molecular analysis), due to its uncommon occurrence and unusual location.

Overall prognosis of SS is related to disease stage. Information on pulmonary SS is limited due to its rarity. In available case series and reports, the main prognostic factor appears to be the ability to achieve a complete resection. The five-year survival has been reported to range between 36-76%.[5] Current treatment modalities include surgical resection (lobectomy or pneumonectomy) followed by adjuvant radiotherapy or chemotherapy.[5]

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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DOI: 10.5455/2320-6012.ijrms20150543
Cite this article as: Bhagya Lakshmi A, Satya Sri K, Kartheek BVS, Rao N. Primary monophasic synovial sarcoma, a rare pulmonary mesenchymal neoplasm: report of a case with review of literature. Int J Res Med Sci 2015;3:1269-71.