Case Report

Primary synovial sarcoma of lung

Devleena, Vikram Bansal, Tamohan Chaudhuri, Shravasti Roy

Department of Radiation Oncology and Pathology, Saroj Gupta Cancer Centre and Research Institute, Mahatma Gandhi Road, Thakurpukur, Kolkata, West Bengal, India

ABSTRACT

A synovial sarcoma (SS) is a rare form of cancer which usually occurs near the joints of the arm, neck, or leg, but has been documented in most human tissues and organs, including the brain, prostate, and heart. Primary pulmonary SS is an extremely rare tumor. We report a case of primary SS of lung who presented with severe chest pain and a large right lung mass with right-sided pleural effusion in computed tomography (CT) scan of thorax. The diagnosis was made on the basis of CT-guided core biopsy and immunohistochemistry. On immunohistochemistry, tumor cell expressed epithelial membrane antigen, bcl 2, Vimentin and smooth muscle actin and were immunonegative for S100 and cytokeratin. So, the final diagnosis was primary SS.

KEY WORDS: Immunohistochemistry, lung, synovial sarcoma

Address for correspondence: Dr. Devleena, Flat A 6/13, 1, Raja Ram Mohan Roy Road, Kolkata - 700 041, West Bengal, India. E-mail: dr_devleena@yahoo.com

INTRODUCTION

Primary synovial sarcoma (SS) arising in lung is very rare and comprise <0.5% of all primary lung carcinoma.[1] There are very few case reports in literature. It is believed to be originated from multipotent mesenchymal cells. There are four basic histological variants that includes biphasic, monophasic spindle cell or fibrous, monophasic epithelial, and poorly differentiated subtypes. In lung, monophasic subtype is most common. The overall prognosis in primary SS is very poor.

CASE REPORT

A case of 53-year-old male patients presented to us on 1st December 2011 with right-sided chest pain, cough, and associated occasional fever. His history was unremarkable. He had no previous surgeries. He was a nonsmoker and nonalcoholic. On examination, patient was healthy male with no abnormality. Chest examination revealed a dull percussion note and decreased breath sounds at right upper lung. Thorough clinical examination of patient did not reveal any abnormality elsewhere in the body. X ray chest showed a large right lung mass with shift of mediastinum to the right and blurring of right costophrenic angle. High-resolution computed tomography (CT) scan of thorax, dated September 2011, revealed a huge right lung space occupying lesion (SOL), right pleural effusion and erosion of 4th rib [Figure 1]. CT guided fine needle aspiration cytology (FNAC) done from right lung SOL revealed spindle cell neoplasm [Figure 2]. Pleural fluid analysis was negative for malignant cells. Further work up for metastasis was negative. He was sent to cardiothoracic surgery for consultation. But in view of huge mass and pleural effusion, it was considered inoperable. To further confirm the diagnosis, he was advised CT-guided trucut biopsy and immunohistochemistry. Histology showed a spindle cell neoplasm with oval to spindle cells, hyperchromatic nuclei and occasional mitosis, suggesting a sarcoma [Figure 3]. Immunohistochemical staining was diffusely positive for Vimentin, strongly positive for bcl 2, patchy positive for EMA, and immunonegative for S-100 and cytokeratin. So, final diagnosis was made as synovial cell sarcoma. He was treated with six cycles of ifosfamideand doxorubicin-based chemotherapy, completed in July 2012. He was then reassessed with contrast-enhanced computed tomography (CECT) thorax and brain. CT chest revealed a huge soft tissue mass (24 × 4 × 15 cm) in right hemithorax with erosion in 4th and 5th rib. CT brain revealed lucent areas in bones of right wall of orbit and occipital bone suggestive of

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metastasis. After that as per our multidisciplinary board advice, he was advised palliative radiotherapy to right lung mass. Radiotherapy was started with palliative intent with 30 Gy in conventional fractionation using three-dimensional conformal radiotherapy technique in 6 MV Linear accelerator. As patient improved symptomatically, radiotherapy was continued till 50 Gy in conventional fractionation, keeping dose to organs at risk within tolerance limit. After completion of radiotherapy, he was kept on supportive care and follow-up. In January 2013, he came for review. Chest examination showed no breath sounds in right side. CECT thorax revealed neoplastic lesion in right lung (18 × 10 × 11 cm) and moderate pleural effusion. He was advised to do a positron emission tomography (PET) scan. PET scan report dated 06/05/2013 showed a non-fluorodeoxyglucose (FDG) avid large mass lesion with central necrosis in right lung measuring 20 × 11.3 × 9.2 cm. Right lower hemithorax showed a small non-FDG avid pleural effusion. Adjacent pleura showed FDG avid (Standardized uptake value max 3.8) pleural thickening. A centimetric calcified precarinal node with mild FDG avidity (Standardized uptake value max 2.18) and a small non-FDG avid aortopulmonary node are also seen [Figure 4]. Patient is stable till date and is kept on supportive care.

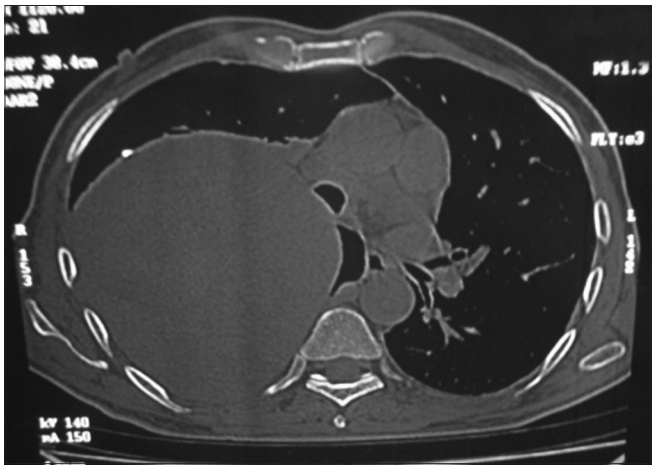

Figure 1: High resolution computed tomography scan of thorax revealed a huge right lung SOL, right pleural effusion and erosion of 4th rib

DISCUSSION

SS is a rare and well-established mesenchymal tumor, accounting for approximately 10% of all soft tissue tumors.[1,2] SS typically presents in adolescents and young adults between 15 and 30 years of age, most commonly in soft tissues of the extremities (especially near large joints), but neck, lung, heart, mediastinum, and abdominal wall sites have been reported.[1] The term "synovial" sarcoma was given because of the synovial differentiation of the tumor that is believed to originate from multipotential mesenchymal cells.

Primary SS arising in the lung is very rarely seen in clinical practice.[3,4] The generally accepted

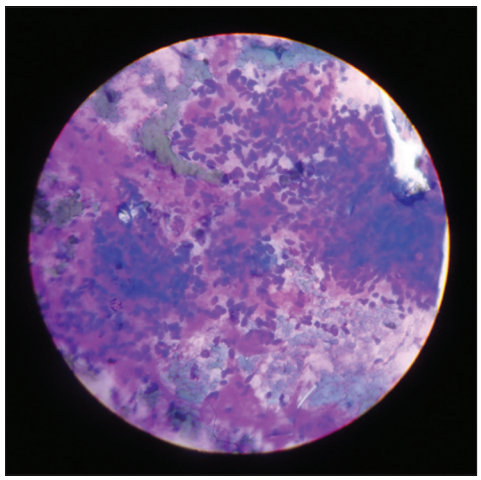

Figure 2: Computed tomography guided fine needle aspiration cytology of the lesion showing spindled cells with relatively bland nuclear features embedded in stromal fragments

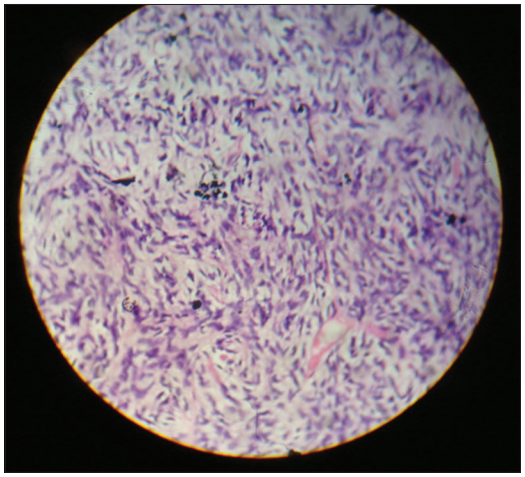

Figure 3: H/E stained section of the computed tomography guided trucut biopsy (×400) showing spindle cells in fascicles

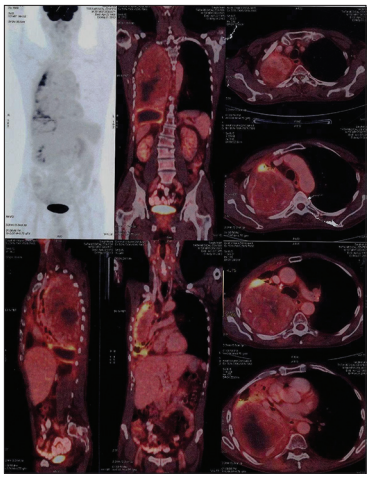

Figure 4: PET scan showed a non FDG avid large mass lesion with central necrosis in right lung, small non FDG avid right pleural effusion

histological subtypes of SS are biphasic, monophasic spindle cell or fibrous, monophasic epithelial, and poorly differentiated subtypes.[5] Among them the monophasic neoplasm subtype occurs most often in lung.[6] The biphasic type is easily diagnosed based on the presence of both epithelial and spindle cell components, whereas monophasic type is difficult to diagnose because of uniform spindle cell pattern. Hence, to differentiate monophasic type of SS from other varieties of sarcoma, immunohistochemistry is essential. Immunohistochemically, SS is usually positive for cytokeratin, EMA, bcl-2, Vimentin and negative for S-100, desmin.[7-9] Immunohistochemistry in this study shows positivity for bcl-2, Vimentin, EMA, and negativity for S-100 and cytokeratin. Cytokeratin negativity was also supported by Dennison *et al*.,[10] Roy *et al*.,[11] and Mermigkis *et al*.[12]

A characteristic balanced translocation exists in SS between chromosomes X and 18, t (X; 18) (p11.2; q11.2) in the majority of cases. The translocation fuses the SYT gene from chromosome 18 to either of two highly homologous genes at Xp11, SSX1, or SSX2. SYT-SSX1 and SYT-SSX2 are thought to function in aberrant transcriptional regulation.[13,14] This translocation is usually the only abnormality and occurs in all variants of SS. The prognosis of patients with SYT-SSX2 abnormality is better than SYT-SSX1 abnormality. Unfortunately, this genetic molecular work up in our case could not be performed due to infrastructural constraint.

The overall prognosis is poor in primary SS with a 5-year survival rate of approximately 50%. Factors responsible for bad prognosis include tumor size (>5 cm), male gender, older age (>20 years), extensive tumor necrosis, high grade, large number of mitotic figures (>10/high powered fields), and recently SYT-SSX1 variant.[15]

There is no standardized therapy; most patients are treated with surgery or surgery with adjuvant radiation therapy. There are limited controlled studies regarding chemotherapy because of rarity of this tumor. SSs are chemosensitive to ifosfamide and doxorubicin, with an overall response rate of approximately 24%.[16]

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