Giant Meditational Biphasic Synovial Sarcoma – A Rare Clinical Presentation

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Abstract

Mediastinal synovial sarcomas are very rare tumors. These sarcomas are challenging to treat because of its anatomic proximity to vital structures in the mediastinum, difficulty in diagnosis and high likelihood of recurrence and metastasis. We report a patient with large mediastinal biphasic synovial sarcoma and discuss the clinical presentation, pathology, clinical relevance of various histologic variants, immunohistochemistry and multi-modality treatment.

Keywords: Mediastinum; Sarcoma; Cancer

Introduction

Synovial sarcoma as the name implies commonly arise from the synovium in the extremities. But these sarcomas can also occur rarely in the mediastinum. These are tumors with anatomic and histological diversity. They have a poor prognosis. Prognosis of these tumors also depends on the location, size, stage, histology, completeness of surgical resection and metastasis. Multi-modality treatment improves survival. We publish this report on synovial sarcoma due to its rarity and challenges in its management. Soft tissue sarcomas account for less than 0.01 % of all thoracic malignant neoplasms [1].

Case Report

A 60yrs old male presented with history of chest pain and difficulty in breathing since two months. Chest X-ray showed widening of the mediastinal border. Computed Tomography (CT) of the thorax (Figure 1) revealed a large (9.7 × 8.2 cm) well circumscribed heterogeneous mass with peripheral rim calcifications in the left anterior mediastinum. There was no evidence of pleural effusion and the lung parenchyma appeared normal. There was no mediastinal lymphadenopathy. The tumor was huge and compressing on the left lung leading to breathing difficulty. On systemic examination there was no evidence of metastasis to other organs which was also confirmed by whole body positron emission tomography (PET-CT) scan. CT guided biopsy from the lesion suggested it to be a benign mesenchymal neoplasm. Bronchoscopic examination revealed extrinsic compression of left main stem bronchus. Bronchial washings were not done.

In view of the well circumscribed mass not invading neighboring structures surgical excision was planned. Median sternotomy was done and the mass was located anterior to pericardium and hilum of the left lung causing collapse of the lung. There were only fibrous adhesions and no invasion of surrounding structures. Complete surgical excision of the well encapsulated mass was performed (Figure 2). Pathological examination revealed a tumor consisting of pattern-less sheets and lobules of short spindle to epithelioid...
cells with short oval nuclei, opened up to granular chromatin, scanty cytoplasm and very rare mitosis (Figure 3A, 3B). Many spaces lined by palisading tumor cells were present, containing a homogenous eosinophilic to hemorrhagic material. Microcystic pattern was focally seen. Areas of necrosis and calcification are also noted. The capsule was not infiltrated. It was a spindle and epithelioid cell tumor with lymphovascular and cystic spaces. Immunohistochemistry was positive for Vimentin, CD-99, BCL-2, epithelial membrane keratin (EMA) and Ki-67 which is suggestive of biphasic synovial sarcoma (Figure 4A, 4B). The post-operative period was uneventful. The patient was discharged home on the seventh post-operative day.

Our patient received adjuvant chemotherapy and radiation therapy. He received intensity modulated radiation therapy (IMRT) comprising of 60Gy in 30 fractions to the tumor bed. Adjuvant chemotherapy with MAID schedule comprising of mesna, adriamycin, ifosfamide and dacarbazine were administered for six cycles. The patient has been followed up for six months with no evidence of recurrence and has returned to normal life style.

Discussion

Synovial sarcomas are rare tumors arising from deep soft tissues [2]. They account for < 1% of malignant neoplasms [3]. In retrospective study by Burt et al, only 1.4% of total 3149 soft tissue sarcomas examined were primary mediastinal sarcomas [4]. Among these, synovial sarcomas accounted for only 2% [5]. Classically synovial sarcoma is a mesenchymal spindle cell tumor. Three subtypes are recognized they are biphasic, monophasic & poorly differentiated varieties. Biphasic variety is characterized by proliferation of bland spindle cells laid in a collagenous background and hemangiopericytomatous vascular growth pattern and epithelial differentiation ranging from well-formed gland like structures to aggregates of cuboidal cells. Monophasic variety contains purely the spindle cells and occasionally only epithelial cells which are very rare. The spindle cells are uniform, small, plump and elongated. They have dark stippled chromatin, scant cytoplasm and indistinct cell margins. They are usually seen in sheets or fascicles. The poorly differentiated variety shows increased cellularity, pleomorphism, mitosis and necrosis. It is characterized by the presence of either two of the following in two low power fields like cellular areas with nuclear crowding, nuclear irregularity, prominent nucleoli and irregular clumped chromatin.

Immunohistology can aid in the diagnosis [6]. The glandular component of synovial sarcoma expresses carcinoembryonic antigen (CEA) and spindle cells are frequently CD 99 and BCL-2 positive. The spindle cells of monophasic and poorly differentiated synovial sarcoma express cytokeratin and EMA. Calretinin and CK 5/6 which are useful in differentiating mesothelioma from adenocarcinoma are positive in synovial sarcoma but not in mesothelioma. WT1 is expressed in mesothelioma but not in synovial sarcoma. In our patient, the histopathology showed spindle and epithelioid cells. The capsule was not infiltrated and the resected margin was free. Size (> 5 cm) is the primary predictor of metastasis and survival. There is no difference in survival rates between the biphasic and monophasic variety. Whereas poorly differentiated areas indicate poor survival with increase in recurrence, metastasis and death.
Surgical resection is the cornerstone of the therapy for such tumors [7]. The complete resection of the tumor is the main factor which determines the survival. Radiotherapy is recommended for positive margins. Chemotherapy is reserved for metastasis. The surgical excision in our patient was complete with no evidence of residual tumor or any recurrence at six months which was confirmed by PET scan. Since soft tissue sarcomas are high grade tumors with chances of local recurrence and distant metastasis and considering the size of the tumor (> 5 cm) our patient received adjuvant chemo radiation therapy as per our tumor board policy.

Conclusion

Giant mediastinal synovial sarcoma is a rare presentation. The mainstay of treatment is prompt diagnosis and complete surgical resection, which will result in long term survival. Radiation and chemotherapy is to be added as a part of multi-modality treatment as indicated. Our patient had complete surgical excision with negative margins, but since it was a giant tumor the prognosis is guarded.

References