

Case Report

Primary Intrathoracic Biphasec Synovial Sarcoma

Yilmaz Tezcan MD¹, Mehmet Koc MD¹, Husnu Kocak MD², Yusuf Kaya MD²**Abstract**

Synovial sarcomas are most frequently observed in the extremities. Although synovial sarcomas are the third most common histological type of soft-tissue sarcomas of the extremities, primary mediastinal synovial sarcoma is extremely rare. Monophasic synovial sarcoma is the most commonly observed subtype, whereas the biphasic subtype is less common. We present our case which was diagnosed as biphasic synovial sarcoma located in the anterior mediastinum, which is considered to be a rare entity. The patient underwent surgical resection together with multimodal adjuvant radiotherapy and chemotherapy.

Keywords: Chemotherapy, prognosis, radiotherapy, synovial sarcoma

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Case Report

Our case was a 38-year-old married female who presented with complaints of cough, dyspnea, pain, and fatigue for 2 – 3 months. PA chest radiography revealed a mass in the anterior and left side mediastinum (Figure 1). A subsequent CT scan of the thorax showed a soft tissue mass, 13 – 14 cm in diameter, in the anterior and left side mediastinum (Figure 2). The same mass was verified by an MRI of the thorax (Figure 3). Following routine examinations, the patient underwent a left thoracotomy with complete excision of the mediastinal mass.

The surgical procedure was as follows: the patient was placed on her right side with one arm raised. An incision was made on the skin of the rib cage. Muscle layers were cut and a rib removed to gain access to the cavity. Retractors were used to hold the ribs apart, exposing the tumor. The tumor and capsule were removed intact after which the layers of the skin, muscle, and other tissues were closed with stitches and staples. Next, the chest wall was closed. The left thoracotomy procedure was completed without complication.

The pathology specimen consisted of a tumor that resembled encapsulated grey-white soft tissue, which macroscopically measured 13×12×8 cm. There were groups of cells that had oval nuclei, fusiform cytoplasm, and malignant tumor that consisted of solid masses of epithelioid cells with wide eosinophilic cytoplasm and oval, pleomorphic nucleus. Focal necrosis was present in the tumor and mitotic activity was found to be 2/10 BBA. Tumor cells stained positive in solid epithelioid areas with cytokeratin; focal staining was observed with S100, CD99, and cytokeratin 7. Based on histomorphological and immunohistochemical findings, the case was diagnosed as biphasic synovial sarcoma. According to postoperative TNM staging, this case was staged as T2bN0M0.

Adjuvant radiotherapy was planned for the patient due to close surgical margins, young age, and tumor size (13 cm). A total of 66 Gy adjuvant radiotherapy was applied to the primary tumor and consisted of an initial 50 Gy (Phase I) dose with a boost of 16 Gy (Phase II), followed by adjuvant chemotherapy with 4 cycles of ifosfamide and adriablastin. The treatments were well tolerated and the patient was observed each 3 months for follow up visits. A thorax CT performed 6 months after treatment end did not reveal any pathological findings (Figure 4). After approximately 36 months (3 years), the patient was lost due to disease progression.

Discussion

Primary mediastinal synovial sarcomas are extremely rare in the thorax and lungs. Primary pulmonary sarcomas account for < 0.5% of lung cancers.¹ However, an increase has been observed in these tumors in recent years.² Leiomyosarcomas, fibrosarcomas, and hemangiopericytomas are the most common types of primary pulmonary sarcomas.³ Primary pulmonary and mediastinal synovial sarcomas are more aggressive than soft tissue synovial sarcomas. While biphasic synovial sarcomas arise in the pleural cavity,⁴ most are localized within the pulmonary parenchyma⁵ and rarely extend into the bronchial structures.⁶ Mediastinal lymphadenopathy is rare.⁷

Synovial sarcomas are histologically classified into four types: biphasic, monophasic fibrous, monophasic epithelial, and poorly differentiated. Monophasic synovial sarcoma is the most commonly observed subtype, and studies show that epithelial components of biphasic tumors are surrounded by pneumocytes. Direct chest radiography is used primarily for diagnosis where the lesion presents a typically uniform view with well-circumscribed rounded or lobulated borders.⁷⁻⁸ Some patients have mediastinal shift. Bilateral pleural effusion is common. CT scan is more sensitive than chest radiography for detecting calcified tumor matrix and cortical destruction.⁹ These tumors are less vascular and MRI imaging presents three findings (clear, dark, and grey) that reflect tumor, hemorrhage, and necrosis.¹⁰ Clinically, patients may show varied symptoms such as cough, dyspnea, chest pain, and fatigue depending on the size and extent of the mass.

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Figure 1. PA chest radiography prior to treatment.

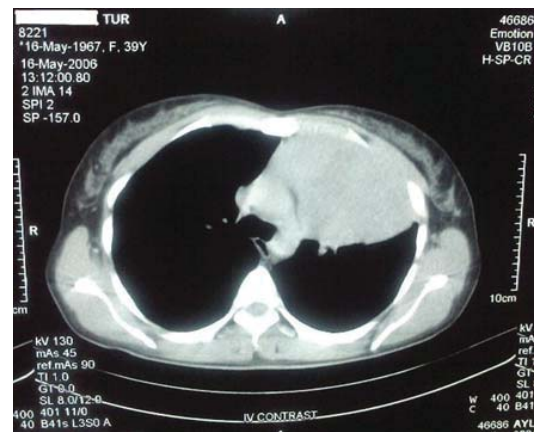


Figure 2. CT scan of the thorax prior to treatment.



Figure 3. Thorax MRI prior to treatment.



Figure 4. Thorax CT scan performed 6 months after the treatments

CT-guided needle biopsy is adequate for diagnosis. Prognosis is related to the phase of the disease, and is generally poor. The five-year survival rate is between 36 – 76%.³

Tumor size (≥ 9 cm), male patients, over the age of 20 years, the presence of extensive tumor necrosis, high grade, large number of mitosis ($> 9 - 10$), neurovascular invasion, and in recent years, the presence of the SYT-SSX1 variant can be listed as poor prognostic factors.¹¹ Complete resection is mentioned as the most significant prognostic factor in a meta-analysis. This meta-analysis has shown that the application of adjuvant radiotherapy and adjuvant chemotherapy following complete surgical resection prolongs the time of local recurrence and survival without recurrence, and accordingly causes an increase in total survival rate.^{12,13}

Synovial sarcomas are tumors which have moderate chemosensitivity, with about 50% response rates to regimens containing ifosfamide and doxorubicin.¹⁴ Radiotherapy is recommended in cases with positive margins.¹⁵

In summary, we have presented a rare case of biphasic synovial sarcoma located in the anterior mediastinum. In these rarely observed cases, survival can be increased through complete resection and aggressive multimodal treatments.

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