Surgical Excision for Mediastinal Synovial Sarcoma With Limited Response to Chemoradiotherapy
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Primary synovial sarcoma of the mediastinum is an exceedingly rare neoplasm. We describe a 31-year-old woman who had an incidental diagnosis of mediastinal mass. Histopathology and immunohistochemistry analysis confirmed the diagnosis of primary mediastinal synovial sarcoma. The patient underwent concurrent chemotherapy and radiotherapy, with minimal response radiologically. Resection was subsequently performed, with negative margins. The histopathologic examination revealed the diagnosis with a limited pathologic response. Because of the rarity of primary mediastinal synovial sarcoma, the optimal therapy is still unclear. We report this case of induction therapy followed by en bloc surgical resection.

Primary mediastinal synovial sarcoma is extremely rare [5]. The first cases were reported in 1989 by Witkin and colleagues [6]. The name is a misnomer, inasmuch as “synovial” sarcomas do not originate from synovial cells

The patient then underwent median sternotomy and resection of the mass, which filled the anterior mediastinum and extended posterior to the trachea, abutting the spine. The resection was en bloc, with no visible remaining tumor. The tracheal stent was left in place throughout the perioperative period, and the patient had no respiratory difficulties after the operation. Her postoperative course was uneventful. She was discharged from the hospital on the eighth day.

The tumor measured $14 \times 10 \times 5.4$ cm (Fig 2). Histopathologic examination revealed a high-grade synovial sarcoma with a treatment effect of approximately 20%. The mitotic rate was measured at 3 per 10 high power fields. The positive immunoreactivity for cytokeratins AE1/AE3 and epithelial membrane antigen with negative staining observed for S-100 and CD34 further supported the diagnosis of synovial sarcoma. The margins were tumor free. The patient is receiving routine follow-up care, with no signs of disease or recurrence in the fourth postoperative month.

Comment
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A 31-year-old woman presented with the incidental finding of a mediastinal mass. Computed tomography (CT) demonstrated a $4.5 \times 8.8$ cm heterogeneous mass in the anterior mediastinum, with tracheal displacement. Core-needle biopsy revealed a diagnosis of spindle cell neoplasm positive for B cell lymphoma gene-2, vimentin, and pankeratin, with chromosome 18q11.2 rearrangement, supportive of the diagnosis of high-grade synovial sarcoma. Soon after the biopsy, the patient had a sudden onset of progressive dyspnea and was treated with tracheal stent placement; her symptoms resolved.

She received chemotherapy consisting of one cycle of cisplatin, doxorubicin, cyclophosphamide, and prednisone followed by three cycles of doxorubicin, ifosfamide, and mesna. She was also given concurrent radiotherapy (5600 cGY as a total dose). She was then referred to our thoracic surgery clinic for further evaluation. Her forced expiratory volume in 1 second was 92% of predicted volume, and her diffusion capacity was 80% of predicted values. A posttreatment CT scan demonstrated minimal decrease in size of the mediastinal mass, and the tracheal stent remained (Fig 1). There was no evidence of metastatic disease.
Synovial sarcomas usually originate in the deep soft tissue and may present as asymptomatic growing mass lesions, predominantly in adolescents and young adults between 15 and 40 years of age [5]. Fewer than 40 cases have been described in the literature [1].

Computed tomographic scans demonstrate a heterogeneous soft tissue mass invading the mediastinal structures. The differential diagnosis includes lymphoma, thymoma, germ cell tumors, mesothelioma, metastatic carcinomas, and other sarcomas. Core-needle biopsy may be performed to ascertain the diagnosis. Positron emission tomography can be used in evaluation of the disease, both in diagnosis and during the follow-up period to detect recurrence [7].

With the use of limited existing medical data, synovial sarcoma patients are now classified into two prognostic groups: low risk and high risk. Good prognostic factors for survival include small tumor size (<5 cm), clear resection margins, low mean mitotic activity (<15 mitosis per 10 high power fields), peripheral location of the tumor, absence of necrosis in the tumor, and young patient age (<25 years) [8].

Complete surgical resection remains the mainstay of the treatment for primary synovial sarcoma of the mediastinum [7]. Although concurrent chemotherapy and radiotherapy can be given at induction, the response of the tumor may vary and may be limited, as seen in our case. Experience with this tumor is limited, and the optimal treatment for these patients is therefore uncertain and thus needs more patient results.

References