Successful Resection of a Giant Primary Liposarcoma of the Posterior Mediastinum

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Primary liposarcomas of the mediastinum are extremely rare neoplasms, comprising less than 1% of all mediastinal tumors. These tumors occur most commonly in the lower extremities (75%) and less frequently in the retroperitoneum. We present a case involving a successful radical resection of a large myxoid liposarcoma and its recurrence, both located in the posterior mediastinum.

Key words: Liposarcoma, Mediastinum, Surgery, Recurrent liposarcoma.

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In September 2002, a 29-year-old woman was referred to our surgical department because of massive opacification of the left hemithorax on chest radiograph. The patient, who was a non-smoker, presented with a persistent cough that was not responsive to sedative drugs. Mild dyspnea at rest and tachycardia had developed over a 1-month period. She had no significant medical history, and, on physical examination, absent breath sounds and dullness to percussion were noted on the left side of the chest.

A total body computed tomography (CT) scan (Figure 1) showed a mass involving the entire left hemithorax and the posterior mediastinum, with a marked mediastinal shift to the right. The tumor was composed of large lobulated masses with various soft tissue densities, and the adjacent structures appeared compressed but not clearly infiltrated. No other localization of the tumor was seen. A magnetic resonance imaging (MRI) scan did not show signs of invasion of the mediastinal structures or diaphragm. A core needle biopsy performed under CT guidance was not conclusive for a preoperative pathological diagnosis and only showed normal fat tissue composed of mature adult fat cells. However, on the basis of the CT scan and the large size of the mass, surgical exploration was performed. A complete resection of the tumor from the left pleural cavity and posterior mediastinum was made through a posterior–anterior left thoracotomy; during the operation, the substantial size of the mass created problems with its manipulation, causing compression of mediastinal structures and temporary arrhythmias. The tumor was pseudo-encapsulated and had an expansive, non-infiltrative growth, thus making the radical resection possible. Major adhesions were noted in the paraaortic–paraesophageal space, where the base implant seemed to be located. Macroscopically, the resected specimen weighed 4910 g, and its dimensions were $28 	imes 22 	imes 16$ cm (Figure 2). The cut surface was slimy with a yellowish tinge and was an admixture of fleshy sarcoma-like areas and areas resembling ordinary adipose tissue. Microscopically, the tumor was composed of small uniform cells with scant cytoplasm set in a myxoid matrix containing numerous capillaries arranged in a plexiform pattern (Figure 3A). Scattered among these cells was a variable amount of cells with a clear vacuole or vacuoles that distorted and pushed aside the thin nucleus (signet ring lipoblasts; Figure 3B). The final diagnosis was consistent with a paucicellular myxoid liposarcoma (MLS) originating from the posterior mediastinum. The postoperative course was uneventful; chest radiograph documented the re-expansion of the left lung and realignment of the mediastinum, and the patient was discharged on the 11th day post-surgery. At 24-month follow-up, evaluation with a chest CT scan showed a relapse (dimensions, $5 \times 3 \times 2$ cm) in the right paraesophageal space that was radically resected through a right thoracoscopy. The tumor showed the same morphologic features as the original mass, with clearly evident fat-laden cells and signet ring lipoblasts merged into a mixoid matrix containing thin-walled capillaries. At 24 months after the second intervention, the patient was well and free of disease.

DISCUSSION

MLS is the most common subtype of liposarcoma, accounting for more than one third of liposarcomas and representing approximately 10% of all adult soft tissue sarcomas. MLS is a disease of young adults; the age at
presentation is, on average, a decade younger than that with other histological subtypes. It has a peak incidence in the fourth and fifth decades of life. These tumors occur preferentially in the deep soft tissue of the lower extremities (thigh, popliteal area, groin, and buttock), followed by the retroperitoneum. The mediastinum is usually involved as a metastatic site, whereas it represents an extremely rare site of primary origin (0.5–1% of all mediastinal tumors); liposarcomas are mainly located in the anterior mediastinum. Microscopically, pure MLS is composed of a hypocellular bland spindle cell proliferation set in a myxoid background. Lipoblasts are often monovacuolated and tend to cluster around vessels. The most distinguishing morphological clue to the diagnosis is the presence of a thin-walled capillary network organized in a plexiform “chicken-wire” pattern. Mitotic figures and necrosis are uncommon. The differential diagnosis includes all the neoplasms with a myxoid stroma. The cells composing myxoid malignant fibrous histiocytoma (myxofibrosarcoma) demonstrate greater nuclear pleomorphism and lack cytoplasmic vacuoles. Diagnostic difficulties involving myxoid liposarcoma sometimes occur, especially in relation to other types of liposarcomas and lipomatous neoplasms. However, atypical lipomatous tumors display a population of cells with enlarged, dense, irregularly shaped nuclei. The histological features of MLS are indistinguishable from those of lipoblastoma and lipoblastomatosis, except for the lobulation found in many lipoblastomas.

The classification of liposarcoma often contains a category formerly known as “round cell” liposarcoma (RC). A subset of MLS shows histological progression to hypercellular or RC morphology, which is associated with a significantly poorer prognosis. The recognition and quantification of hypercellular/RC areas represent critical steps in the evaluation of MLS subtypes because hypercellularity seems to be correlated with the clinical outcome. On the basis of the percentage of hypercellularity/RC formation, MLS/RC (more than 25% hypercellular/RC areas) and RC (more than 75% hypercellular/RC areas) subtypes are recognized. The 5-year survival probability is approximately 90% for pure MLS, 40–50% for MLS/RC, and less than 25% for RC.

The clinical behavior is usually characterized by slow growth that is not infiltrative but that causes a gradual compression of adjacent intrathoracic structures. Symptoms, when present, include chest pain, cough, dyspnea, and manifestations related to the compression of the heart and great vessels (superior vena cava syndrome, cardiac tamponade, tachycardia, arrhythmias). Despite the presence of a huge mass causing complete atelectasis of the left lung and a mediastinal shift of the heart to the right side, our patient did not have significant symptoms. The slow growth of the mass may have allowed the patient to gradually adapt. Some authors have reported the effectiveness of fine-needle or large-core biopsy under CT guidance for preoperative pathological diagnosis; in the present case and in other reports, however, preoperative diagnosis was not possible. Despite the ease with which the biopsy was performed, only mature adipose tissue admixed with myxoid matrix was observed. This may have been because of both the large size of the mass and the peculiar heterogeneous morphological pattern of...
MLS in which highly cellular areas are admixed with foci resembling normal adipose tissue.

The gold standard for the treatment of these tumors is complete surgical excision. In our case, after the successful resection of the tumor, we also obtained the simultaneous re-expansion of the left lung, which had completely collapsed, and realignment of the mediastinum. Our patient was young, and, on pathological examination, we found a low grade of cellularity and necrosis. On the basis of these findings and because of the radical surgical excision of the tumor, we did not proceed with adjuvant treatment. Unfortunately, 2 years after the surgical excision, the patient presented with a local recurrence that probably originated from the base implant of the tumor in the posterior mediastinum; this recurrence was successfully resected. The rarity of this tumor is demonstrated by the few documented cases (approximately 80 patients) in two major reviews and some case reports. At this moment, no clear data are available regarding the efficacy of adjuvant treatments. The proposed chemotherapeutic regimens are anecdotal and have not been proven effective. However, radiotherapy seems to be useful for the local control of disease in cases that are not suitable for surgery or not radically resected, but radiotherapy is of little utility in the dosage that can be used in the mediastinum or chest cavity. Nevertheless, Burt et al. recommended continued exploration of an aggressive adjuvant treatment in consideration of the high percentage of local recurrence (64%) even after radical resection.

REFERENCES