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Resection of Primary Mediastinal Liposarcoma Invading Right Ventricular Outflow Tract

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Liposarcomas, the most common soft tissue tumors, occur predominantly in the lower limbs and retroperitoneum. Primary mediastinal liposarcomas are rare and account for <1% of mediastinal masses. We report a case of a large primary mediastinal liposarcoma invading the right ventricular outflow tract, pulmonary artery, and aortic adventitia that was successfully resected. This report highlights the complex nature of liposarcomas, given their tendency to invade critical structures.

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Liposarcomas are the most common soft tissue sarcoma and predominantly occur in the lower limbs and retroperitoneum.[1] However, primary mediastinal liposarcomas are rare and account for <1% of mediastinal masses.[2] They are typically asymptomatic in their early stages, allowing them to grow and reach significant sizes. As a result, they often invade and compress intrathoracic organs, including the lungs and heart. Presenting symptoms may include cough, chest pain, shortness of breath, and dysphagia. Surgical resection is the primary treatment option, and the degree of intervention depends on tumor invasiveness. We report a case of a large primary mediastinal liposarcoma in a 68-year-old man invading the right ventricular outflow tract (RVOT), pulmonary artery (PA), and aortic adventitia that was successfully resected.

A 68-year-old man with a remote history of stage IIb well-differentiated liposarcoma of the left thigh that underwent radical resection and adjuvant radiotherapy presented with nonspecific chest pain and exercise intolerance for 1 year. Chest radiography showed a lobulated and widened mediastinum (Figure 1). Contrast-enhanced computed tomography of the chest demonstrated a large lobulated heterogeneous anterior mediastinal mass compressing the superior vena cava, right atrium, and right ventricle. It also invaded the RVOT, ascending aorta, and right coronary artery (Figures 2A-2D). Results of laboratory testing, including complete blood count, comprehensive metabolic panel, troponin, β-human chorionic gonadotropin, α-fetoprotein, and lactate dehydrogenase, were within normal limits. Core needle biopsy of the mass revealed an MDM2-positive low grade-appearing spindle cell neoplasm with focal atypical cells and rare mitotic figures. Immunohistochemical stains were negative for S100, β-catenin, smooth muscle actin, and desmin. The sample showed patchy positivity for CD34. These pathologic findings were consistent with well-differentiated liposarcoma. Echocardiography demonstrated mild to moderately dilated right ventricle and mildly decreased right ventricular function with ejection fraction of 53%. The tumor did not respond to chemoradiation, so an operative resection was performed.

A saphenous vein graft was endoscopically harvested prophylactically, given the potential involvement of the right coronary artery. Surgical resection through a primary median sternotomy was performed with a redo oscillating saw because of the underlying mass. The mass was found to be adherent to the RVOT and right ventricular free wall. To decompress the heart and to facilitate further dissection, aortic and bicaval venous cannulation was performed and cardiopulmonary bypass was initiated. The mass was dissected off the right ventricular free wall until it was found invading the right ventricular cavity and RVOT, which was resected en bloc, including the pulmonic valve. The mass was then carefully resected away from the course of the right coronary artery. There was unresectable residual disease near the right coronary artery. The pulmonic valve was

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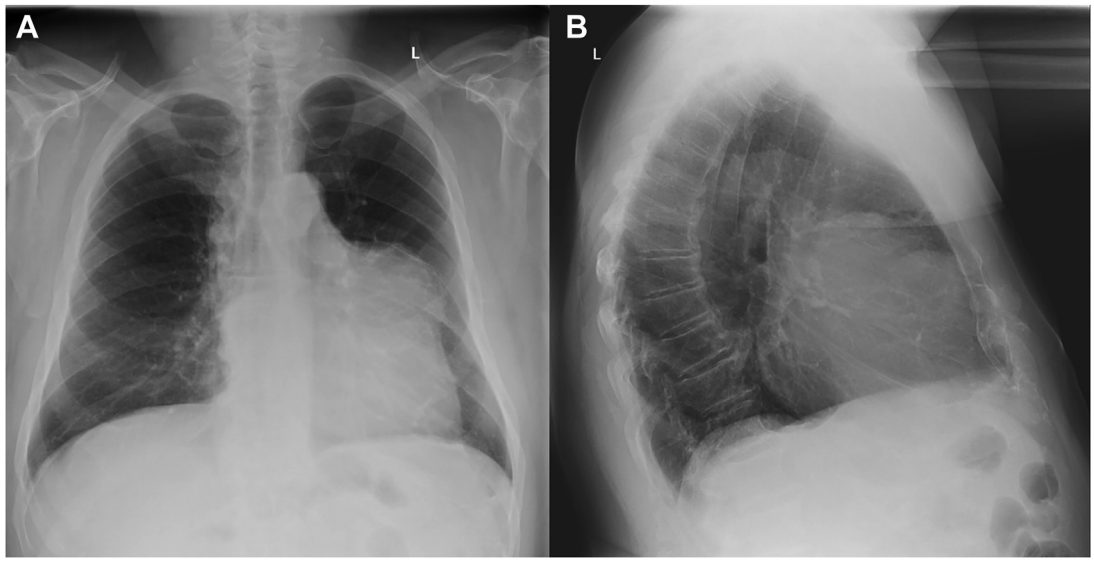

FIGURE 1 Chest radiograph showing a lobulated prominence of the mediastinum in (A) posterior-anterior view; (B) lateral view.

replaced with a stented bovine pericardial valve; the RVOT and PA were reconstructed with a bovine pericardial patch.

During mass resection, the aortic root became denuded by tumor invasion of the aortic adventitia, which subsequently led to significant bleeding. Therefore, the heart was arrested; the aorta was opened and reconstructed with a bovine pericardial patch. The patient was separated from cardiopulmonary bypass; however, because of ongoing hemorrhage and persistent right ventricular dysfunction, the decision was made for the patient to leave the operating room on central venoarterial extracorporeal membrane oxygenation. The chest was left open and packed. An Esmarch dressing was affixed over the wound. The patient was decannulated from venoarterial extracorporeal membrane oxygenation on postoperative day (POD) 2, and his chest wound was closed. The postoperative course was prolonged because of multiple complications, including left cerebellar ischemic infarction, altered mental status, metabolic encephalopathy, acute hypoxemic respiratory failure secondary to ventilator-associated pneumonia, bilateral deep venous thrombus, and small bowel ischemia that required a small bowel resection. Because of his prolonged hospital course, he also required a tracheostomy and percutaneous endoscopic gastrostomy tube placement. However, by POD 26, the patient's neurologic examination findings significantly improved; he was alert, able to follow commands, and moving all extremities to command. By POD 34, the patient's condition had improved, and he was communicating. The patient was discharged on POD 50 to a skilled nursing facility without focal neurologic deficits.

On pathologic examination, the tumor was well encapsulated. It weighed 684.8 g and measured 17.5 × 15.3 × 5.5 cm (Figure 3). On review of the specimen, findings were consistent with the original biopsy, and a final diagnosis of well-differentiated liposarcoma was established. Although the patient had a remote prior history of liposarcoma in his left thigh, the mediastinal tumor was believed to represent primary mediastinal liposarcoma as well-differentiated liposarcoma has no propensity to metastasize.

COMMENT

Primary mediastinal liposarcomas are rare and account for <1% of all mediastinal tumors.[2] In general, mediastinal liposarcomas are identified after they have grown to significant sizes and begin to compress or to invade adjacent structures in the thorax, such as the lungs or heart. Symptoms range from dysphagia to dyspnea, tachypnea, wheezing, and chest pain. In this case, the primary symptom was nonspecific chest pain.[2-6]

Surgical resection is the optimal and necessary treatment of mediastinal liposarcomas because of their malignant nature and their ability to compress and invade vital thoracic organs.[3-6] If complete surgical resection is not possible, tumor debulking is recommended for symptomatic relief. Owing to the aggressive invasion of the RVOT, PA, and aorta, nearly complete resection and reconstruction were feasible only by putting the patient

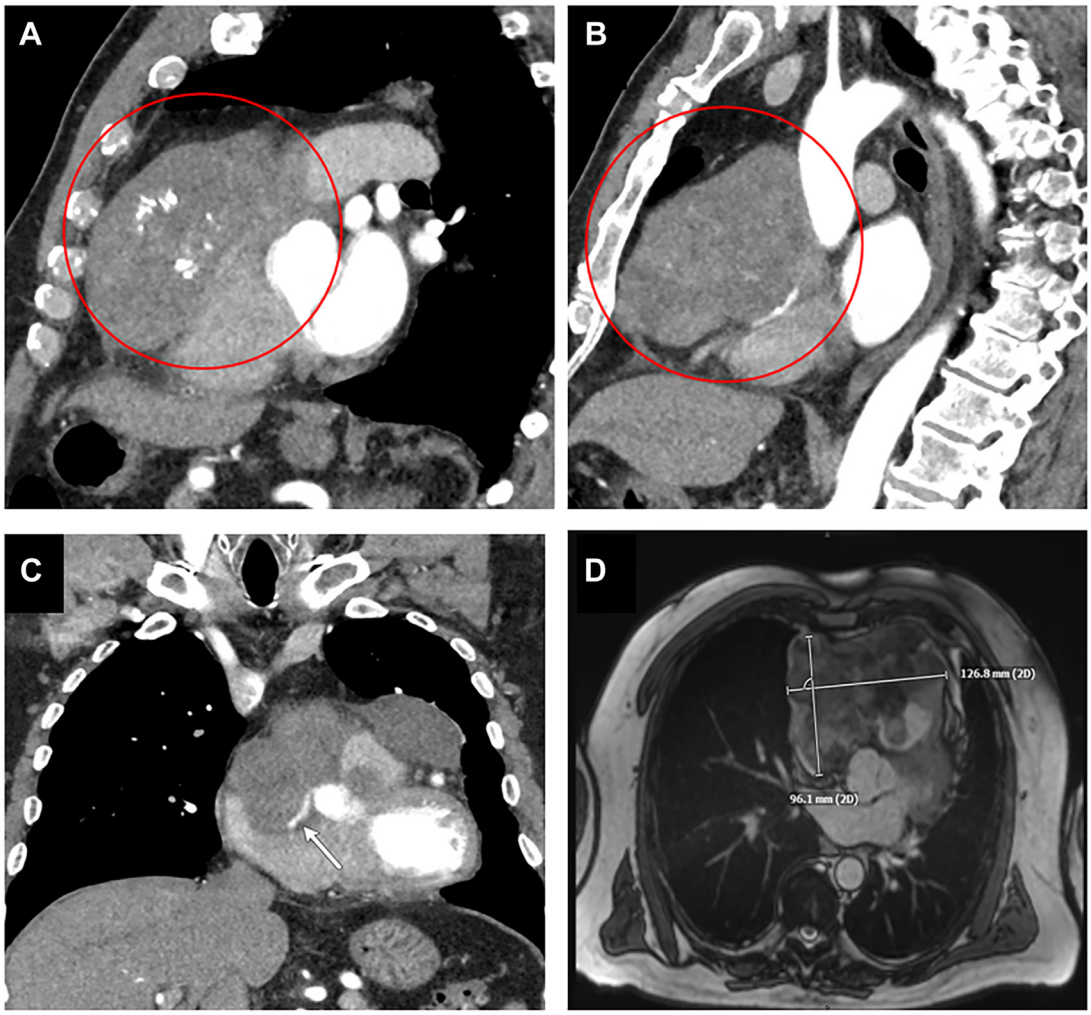

FIGURE 2 Contrast-enhanced computed tomography of the chest showing (A) 12.8 × 9.1 × 11.2-cm anterior mediastinal mass invading right ventricular outflow tract and compressing right ventricle, right atrium, and superior vena cava (circle); (B) mass partially encircling the ascending aorta (circle); and (C) mass partially encircling the right coronary artery (arrow). (D) Magnetic resonance imaging of the chest mass.

on cardiopulmonary bypass and arresting the heart. Use of radiation therapy or chemotherapy has not been demonstrated to be effective in the treatment of liposarcoma.[5,7] Our patient underwent chemoradiation, which did not have a noticeable impact on the size of the tumor.

In conclusion, primary mediastinal liposarcomas are extremely rare malignant neoplasms. Surgical resection is the primary treatment option and can range from simple debulking to aggressive resection with use of cardiopulmonary bypass and cardioplegic arrest. We successfully performed a radical resection of a large anterior mediastinal liposarcoma that had invaded the RVOT, PA, and aortic adventitia. Long-term follow-up is still necessary to determine any recurrence of the tumor.

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DISCLOSURES

The authors have no conflicts of interest to disclose.

PATIENT CONSENT

Obtained.

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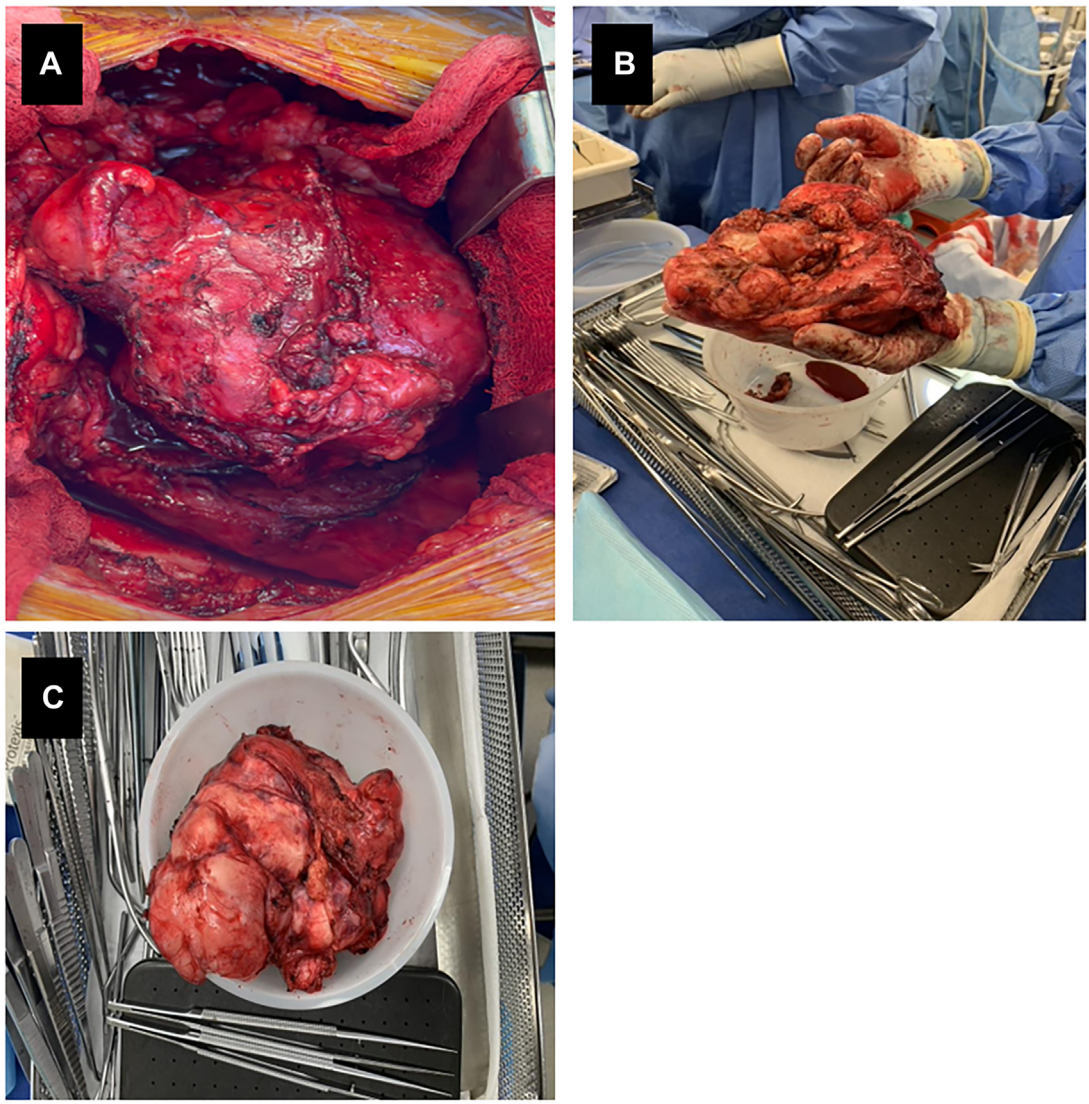

FIGURE 3 (A) Intraoperative view of mass. (B, C) Excised mass.

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