



## Giant primary liposarcoma of the anterior mediastinum resected by median sternotomy: A rare case report

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### Abstract

**Background:** Primary liposarcoma of the mediastinum accounts for less than 1% of all mediastinal tumors and 2% of all liposarcomas, making them exceedingly rare. When they do occur, they typically originate in the posterior mediastinum. These tumors are slow growing, and thus are found incidentally or present later in the course with compressive symptoms.

**Case presentation:** We present the case of a previously undoctored 77-year-old man who was found to have a giant liposarcoma of the anterior mediastinum. The patient initially presented to the emergency department complaining of two weeks of shortness of breath, most prominent while sleeping or lying flat, and an occasional non-productive cough. CT chest showed a large right anterior mediastinal mass abutting the anterior pericardium, and pre-operative biopsy determined the mass to be an atypical lipomatous tumor versus a well-differentiated liposarcoma. The patient underwent complete resection of the mass and total mediastinal exenteration via median sternotomy 3 months after his initial presentation. Entire resection of the mass was accomplished with the final specimen measuring 34 x 28.7 x 10.5 cm. Surgical pathology confirmed a well-differentiated liposarcoma. He is currently followed with serial imaging and has had no evidence of disease at one year.

**Conclusion:** Liposarcoma of the mediastinum is rare and typically discovered late in its' disease progression, potentially complicating surgical resection. Surgical resection is the mainstay of treatment for these tumors, but the role of adjuvant therapy in these cases is poorly understood. Due to its' high risk of recurrence, long-term follow up is critical.

### Background

Liposarcomas are malignant tumors of mesenchymal origin that typically occur in the lower limbs and retroperitoneum [1]. They account for 20% of all soft tissue sarcomas, making them the most common subtype [2]. Primary liposarcoma of the mediastinum, however, is rare, constituting only 0.1-0.75% of all mediastinal tumors and 2% of all liposarcomas [1,3]. These tu-

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**Received:** Apr 08, 2024

**Accepted:** May 20, 2024

**Published Online:** May 27, 2024

**Journal:** Annals of Surgical Case Reports & Images

**Online edition:** <https://annscri.org>

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**Cite this article:** Heutlinger O, Painter R, Keshava H, Mahtabifard A. Giant primary liposarcoma of the anterior mediastinum resected by median sternotomy: A rare case report. Ann Surg Case Rep Images. 2024; 1(3): 1030.

**Keywords:** Liposarcoma; Anterior mediastinum; Median sternotomy; Liposarcoma presentation.

mors are slow growing and are typically found incidentally on imaging or after they have grown large enough to cause compressive symptoms, such as dyspnea, chest pain, hoarseness, tachypnea, wheezing, and/or superior vena cava syndrome [3,4]. Surgical resection is the mainstay of treatment for all liposarcomas, though tumor grade correlates with prognosis and the aggressiveness of the disease. The role of adjuvant chemo-

immuno- and radiotherapy is not well established. In this article, we present the case of a giant primary mediastinal liposarcoma that was successfully removed via surgical resection.

### Case presentation

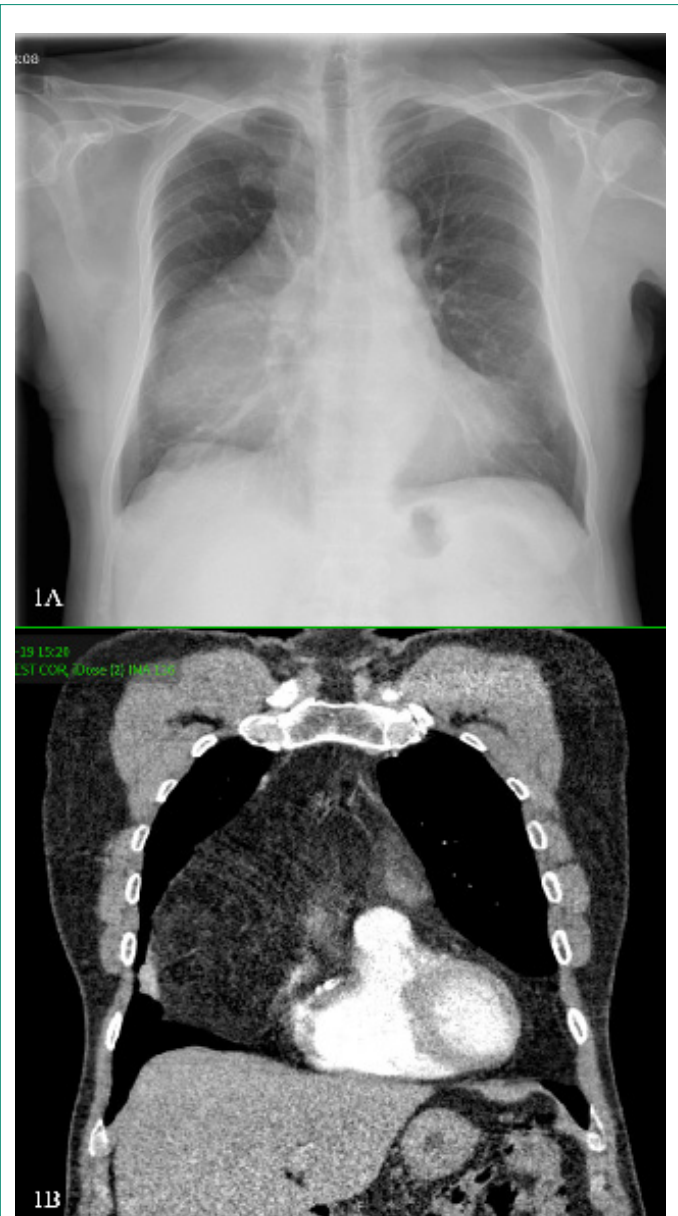
A 77-year-old undoctored male began experiencing shortness of breath at rest while on vacation in the Philippines. He saw a physician there who obtained a Chest X-Ray (CXR) and Computed Tomography (CT) scan which revealed a large mass in the anterior mediastinum. The patient was advised to undergo biopsy of the mass, so the patient returned to the United States for continued workup and management. He arrived at our emergency department shortly after his return. At this time, the patient reported 2 weeks of shortness of breath, most notable when sleeping or lying flat, that was relieved by turning onto his right side. He endorsed an occasional cough with no sputum production or hemoptysis. The patient had no history of fever, chills, chest pain, palpitations, difficulty swallowing, nausea, vomiting, abdominal pain, numbness, tingling, or weakness in the bilateral upper extremities. The patient did not have any medical history at the time of presentation, but later established care within our institution and was diagnosed with hypertension and hyperlipidemia. His family history was non-contributory. Vital signs were notable for a blood pressure elevated to 167/82, but otherwise were within normal limits. Physical examination was unremarkable with regular heart sounds and clear lung fields bilaterally. Laboratory results were also unremarkable. CXR revealed a large soft tissue mass projected over the right middle and lower hemithorax (Figure 1A). CT chest with contrast showed a large right anterior mediastinal mass measuring 15.6 x 8.9 x 16.6 cm consisting of mostly fat density. The mass appeared mostly well-defined and abutted the anterior pericardium, ascending aorta, aortic arch vessels, superior vena cava (SVC), and anterior trachea. The right middle and lower lobes had mild compressive atelectasis. The differential diagnosis based on this image included liposarcoma, thymolipoma, and teratoma (Figure 1B). The X-ray and CT obtained in the Philippines were the patient's first images of the chest, so no earlier comparison images were available.

A PET/CT was obtained which re-demonstrated the anterior mediastinal mass consisting of mostly fat. The mass was mildly FDG avid (1.8 SUV). It also showed other incidental findings unrelated to the liposarcoma (Figure 2). Based on the PET/CT findings an image-guided biopsy of the mediastinal mass was performed with interventional radiology which revealed an atypical lipomatous tumor versus a well differentiated liposarcoma. Immunohistochemical stains for CD34, PAX-8, AE1/AE3, and SOX-13 were all negative.

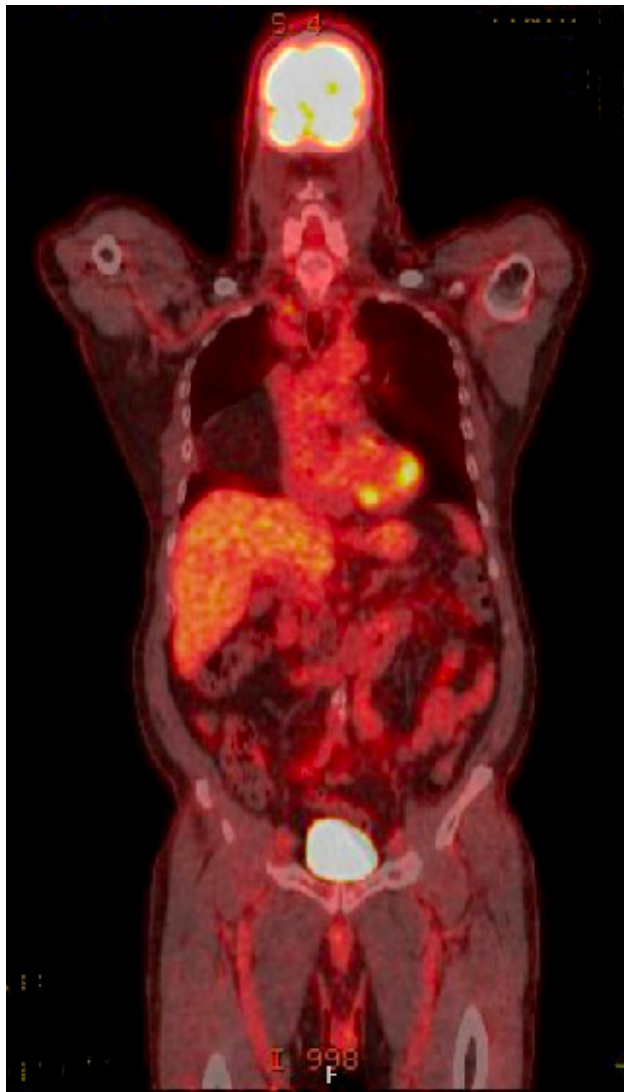
Complete resection of the mass and total mediastinal exenteration was performed via median sternotomy under general anesthesia three months after his initial presentation in the emergency room. The tumor was well-encapsulated and took up the entire anterior mediastinum, expanding into bilateral hemithoraces with more significant expansion on the right side. The pericardium, aorta, and great vessels were easily swept off and not involved with the mass. Given the size of the mass, an extensive resection of all the anterior mediastinal structures including thymectomy was done. The dissection extended up into the neck to the level of the thyroid gland and into both pleural spaces. The operation was performed without complications and about 200 cc of blood loss. Surgical pathology confirmed a

grade 1 well-differentiated liposarcoma measuring 34 x 28.7 x 10.5 cm. One level 5 lymph node was negative for malignancy. The tumor was characterized as American Joint Committee on Cancer (AJCC) stage 1B (pT4 N0). Florescence in situ hybridization (FISH) analysis showed MDM2 gene amplification.

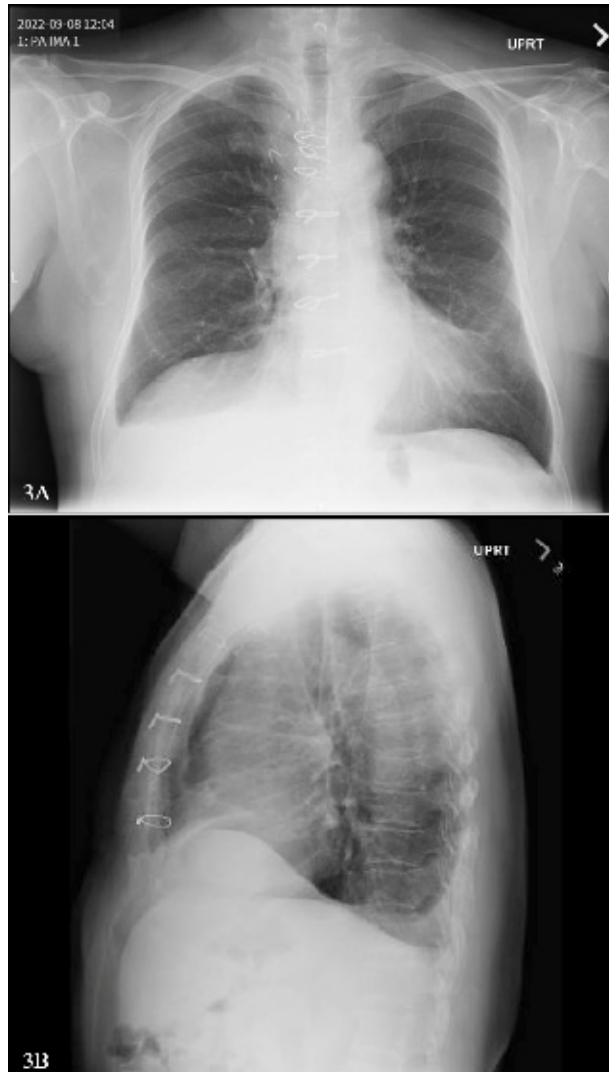
The inpatient postoperative course was unremarkable. CXR taken two weeks post-operatively showed elevation of the right hemithorax and mediastinal postoperative changes consistent with inpatient CXRs (Figure 3). These findings were unchanged at two months after surgery (Figure 4). The patient followed up with medical oncology who did not recommend adjuvant radiation therapy or systemic chemotherapy. Given the low-grade nature of his tumor, adjuvant therapy was unlikely to pose any benefit. The patient continues to be closely followed as an outpatient with imaging every 6 months and has had no evidence of recurrence since his surgery around one year ago.



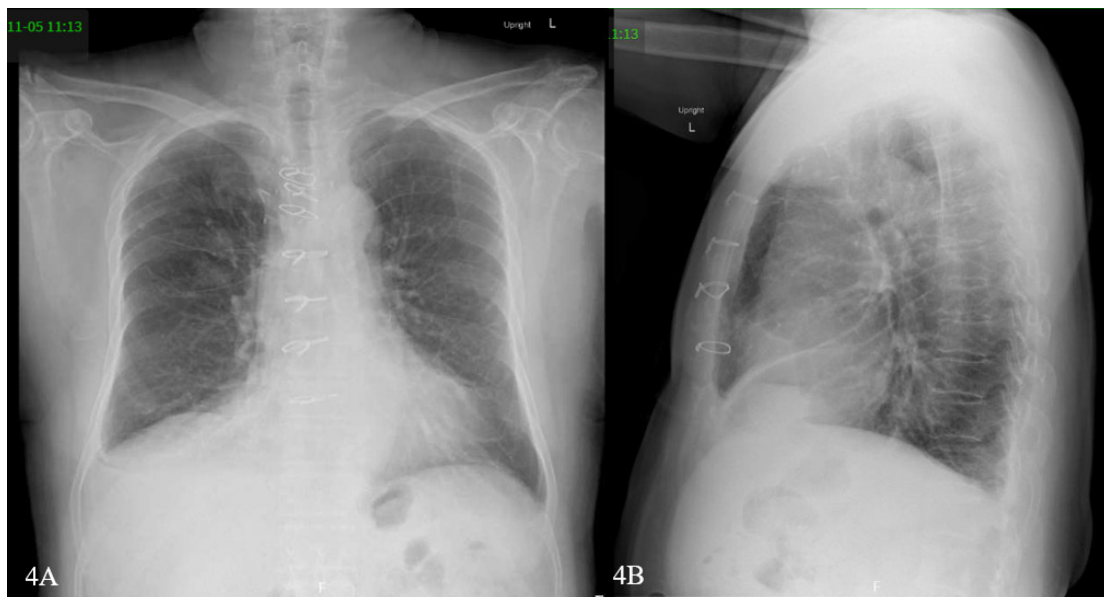
**Figure 1:** Chest X-ray and CT at initial presentation in the emergency department. **(A):** Chest x-ray showing a large soft tissue mass projected over the right middle and lower hemithorax. **(B):** CT chest with contrast showing a large right anterior mediastinal mass measuring 15.6 cm x 8.9 cm x 16.6 cm consisting of mostly fat density with a small left-sided soft tissue component.



**Figure 2: PET/CT at time of diagnosis.** PET/CT showing mildly FDG avid (1.8 SUV) anterior mediastinal mass with mostly fat density and a smaller left-sided soft tissue density.



**Figure 3: Chest X-ray at 2 weeks post-operation.** (A): AP view showing stable mediastinal postoperative changes with elevation of the right hemidiaphragm. No consolidations, obvious effusion, or pneumothorax are seen. (B): Lateral view.



**Figure 4: Chest X-ray at 2 months post-operation.** (A): AP view showing stable mediastinal postoperative changes, bibasilar atelectasis, and mild vascular congestion. No consolidations, obvious effusion, or pneumothorax are seen. (B): Lateral view.



## Discussion

Liposarcomas are derived from primitive mesenchymal cells. There are four histologic subtypes- well-differentiated, dedifferentiated, myxoid, and pleomorphic- with well-differentiated being the most common [4-7]. Well-differentiated and atypical lipomatous tumors carry the best prognosis, whereas pleomorphic and myxoid tend to be more aggressive and carry worse prognoses. Amplification of the MDM2, CDK4, and HMGA2 genes have been found in both well-differentiated/atypical lipomatous tumors and dedifferentiated liposarcomas [8]. Most mediastinal liposarcomas develop in the posterior mediastinum, accounting for almost half of all cases [9]. The next most common location is the anterior-superior mediastinum, followed by the middle mediastinum, then other locations. There is some evidence to suggest that well-differentiated and atypical lipomatous liposarcomas occur more frequently in the anterior mediastinum in contrast to the overall tendency for liposarcomas to originate in the posterior mediastinum [7,9].

The average age at presentation for primary liposarcomas is 50 years old, with a range of 20 to 70 years [10-12]. These tumors grow slowly and generally only produce symptoms once they have reached a large enough size to compress nearby intrathoracic structures [1,4]. This pattern of growth is thought to be related to these tumors tendency to grow circumferentially rather than infiltratively [13]. It is estimated that 85% of cases present with compressive symptoms, whereas the other 15% of cases are incidentally found on imaging [14]. A review done by Schweitzer and Aguam found that 63% of patients with mediastinal liposarcomas present with respiratory symptoms, 50% with chest pain, and 15% with SVC obstruction [15]. The presenting symptoms often mimic common diseases of the heart and lungs, so obtaining imaging is critical in creating a proper differential early in the diagnostic workup.

Surgical resection alone is the gold standard of treatment for mediastinal liposarcomas and is often curative [12,16-18]. Surgical debulking can provide symptomatic relief if complete resection of the tumor is not possible. The role for adjuvant radiation and chemotherapy is not well established for mediastinal liposarcomas, with some evidence suggesting minimal responsiveness to these treatments and other authors recommending adjuvant radiation be reserved for unresectable and poorly differentiated tumors [10-12,17]. One case report showed improvement in long-term survival by five years or more with use of adjuvant radiation therapy after a partial resection [19]. However, further evidence is needed to establish a clear role for adjuvant treatments.

Histologic subtype is the best prognostic factor for liposarcomas [12,17]. Tumor size, gender, and age have not been shown to influence survival [17]. The rates of recurrence for primary mediastinal liposarcomas range from 10% to 50%, and recurrence can occur years after the initial resection [3,12,17,20]. As such, close and long-term follow up is recommended for patients with mediastinal liposarcomas, particularly those with incomplete resections and unfavorable histologic subtypes.

Surgical approach and optimization of risk reduction must be considered on a case-by-case basis. In the case of large liposarcomas, measures should be taken between the surgical and anesthesia team to minimize the risk of mediastinal mass syndrome- a life-threatening, acute cardiorespiratory decompensation that can occur with administration of general anesthesia in patients with large compressive tumors [21,22]. In terms of

surgical approach, median sternotomy is most commonly performed, though posterolateral thoracotomy, classic or hemi-clamshell thoracotomy can be utilized in certain cases [12]. Smaller tumors that are found incidentally could be resected via robot-assisted or video-assisted thoracoscopic surgery, but this is generally not indicated for any tumor exceeding 10 cm [23]. Median sternotomy provides a wide surgical field, allowing for good exposure of the heart, bilateral lungs, and hilar structures. A hemi-clamshell incision can expand on this midline approach to allow better visibility of the lower lobes and posterior hilum. The most common vascular structure resected in these cases is the SVC, followed by the innominate vein [24]. SVC involvement has occurred in multiple cases of primary mediastinal liposarcomas and can be managed with partial SVC resection or complete resection with reconstruction [23].

## Conclusion

Primary liposarcomas of the mediastinum are rare tumors that grow slowly and often present as incidental imaging findings or with compressive symptoms. The primary treatment of these tumors is surgical resection, typically via median sternotomy, with limited evidence on the role of adjuvant chemotherapy and radiotherapy. Liposarcomas have a high tendency to recur and often recur years after initial surgery, so close long-term follow up is imperative for these patients. Our patient continues to be disease free one year after surgery and will continue to be followed closely with serial imaging.

## Declarations

**Acknowledgements:** No acknowledgements

**Ethics guidelines:** Not applicable.

**Conflict of interest:** No conflicts of interest among any participating authors

**Consent for publication:** Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient/parent/guardian/relative of the patient. A copy of the consent form is available for review by the Editor of this journal.

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