

Scientific letter

Liposarcoma of the Anterior Mediastinum Leading to Hemodynamic Compromise



Liposarcoma de mediastino anterior que provoca deterioro hemodinámico

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Dear Editor:

Liposarcomas are malignant mesenchymal tumors. They are the most common type of soft tissue sarcoma.¹ Liposarcomas commonly take place in the retroperitoneum or thigh, while primary mediastinal liposarcomas are rare and they are usually detected late when the tumor has attained large size.² Liposarcomas are classified into 4 subtypes: myxoid, pleomorphic, well-differentiated and de-differentiated liposarcoma. CT images vary depending on the degree of differentiation. Well-differentiated cases typically have septated fatty mass. Imaging without fat attenuation is seen in 20% of cases. Patients usually present late due to the compressive effect of the tumor on adjacent structures. Diagnosis is confirmed by thick needle puncture. Complete surgical resection is still the mainstay of therapy. Liposarcomas could have bad prognosis because of incomplete surgical excision due to its inaccessible location, but debulking will relieve symptoms.³ As the recurrent rate is high, a close long term follow-up is recommended. We present a rare case of liposarcoma in the anterior mediastinum.

A 58 year old, non-smoker women with no history of interest attended our emergency department because of shortness of breath and chest tightness. Thorax CT scan demonstrated a mass of 12 cm × 9 cm × 9 cm in the anterior mediastinum (Fig. 1A). MRI showed a well limited tumor in contact with surrounding structures without invading them (Fig. 1B and C). Tumor core needle biopsy confirmed liposarcoma's diagnosis (Fig. 1D). There was no evidence of distant disease. Clinical course was unfavorable and the patient developed hemodynamic compromise with concomitant pericardial effusion due to the large tumor size that pushed adjacent structures (heart and large vessels). Pericardial window and emergent decompressive surgery were performed. Complete tumor removal was made with evidence of negative surgical

margins and negative pericardial fluid analysis (negative cytology exam of pericardial fluid). So that we assume that pericardial fluid was a consequence of hemodynamic instability. To date, the patient remains stable without any evidence of recurrence.

Liposarcomas are malignant tumors with a mesenchymal origin characterized by amplification of MDM2 and CDK4 genes on chromosome 12.⁴ Mediastinum location accounts for <1% of liposarcomas.⁵ Symptoms are due to compression of adjacent structures. Diagnosis may be confirmed by thick needle puncture. CT scan is usually performed first and varies from a predominantly fat-containing mass to a solid mass, but MRI is the best diagnostic imaging test. Clinical behavior depends on the histopathological patterns, where well-differentiated forms are of low-grade in nature with rare metastatic potential when compared to that of poorly differentiated ones. Definitive diagnosis is made by identifying the stellate shaped lipoblasts on histology. Complete decompressive surgical removal is the treatment of choice.⁶ Repeat surgical resection or radiotherapy should be considered in cases of local recurrence. Chemotherapy is generally ineffective for liposarcoma. Eribulin (microtubule growth inhibitor) has recently been approved for advanced liposarcomas. Prognosis is related with subtype (worst prognosis in pleomorphic cases) and there is a high risk of recurrence. Most commonly recurrences will occur during the first 6 months or it may be delayed for 5–10 years following surgery. Because of this, a close long term follow-up is recommended.

We report a clinical case of mediastinal liposarcoma and present an important clinical issue of this tumor. Because of its size, liposarcomas could compress adjacent organs and cause hemodynamic compromise. We suggest that prompt surgical procedure with complete resection should be initially considered in cases of primary mediastinum liposarcoma in order to reduce complications.

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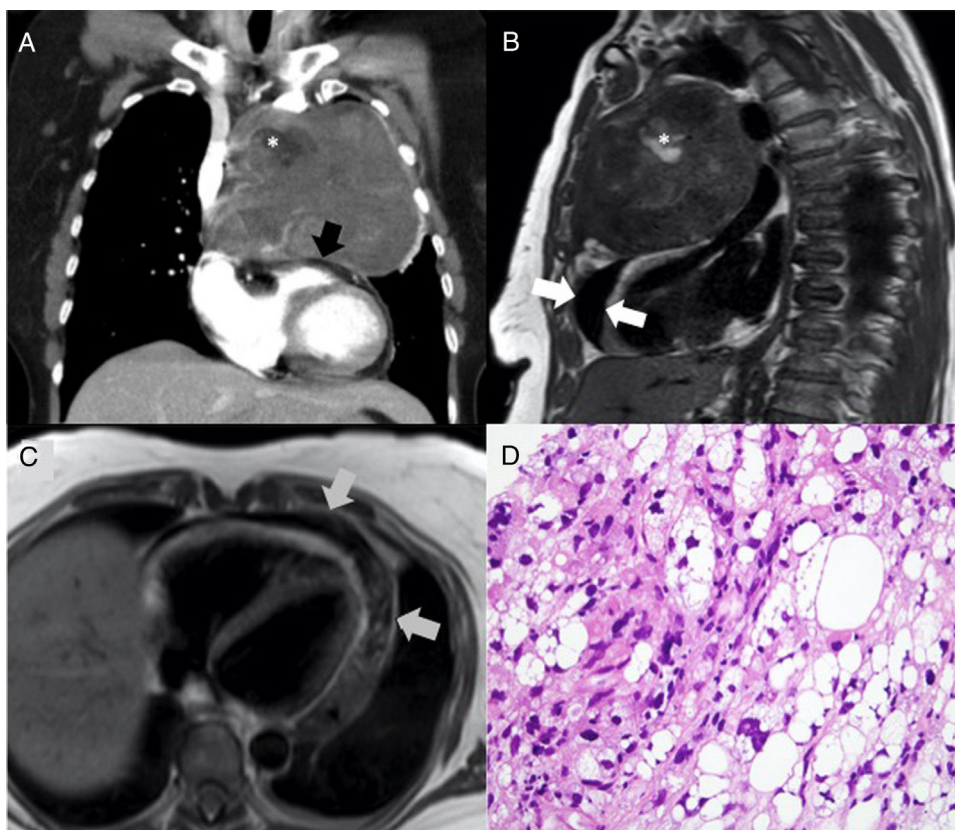


Fig. 1. (A) Thorax CT (coronal) with intravenous contrast that shows a well limited tumor in anterior mediastinum (12 cm × 9 cm × 9 cm) with compression of the heart (black arrows) that has a hypodense central area suggestive of fatty tissue (asterisk). (B) MRI (sagittal, T1 sequence) shows a hiperintense signal in the central zone described, confirming its fatty condition (asterisk). Onset of pericardial effusion (white arrows). (C) MRI (axial, T1 sequence) shows apical and left side pericardial effusion (gray arrows). (D) Histology of the tumor with final diagnosis of liposarcoma: tumor formed by adipocytes with atypical nuclei, high proliferative index and stellate shaped lipoblasts with positive S100 protein immuno-labeling.

Author's contributions

AC, MT and AM were responsible for the conception and design of the study, and wrote and edited the manuscript. AC, MT, AM and LV contributed to the drafting and revision of the manuscript. All authors read and approved the final manuscript.

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