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A rare cause of mediastinal mass: Primary liposarcoma

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Summary

Liposarcoma is the most commonly diagnosed soft tissue sarcoma in adults and occurs predominantly in the lower limbs and retroperitoneum. Primary mediastinal liposarcomas are rare.

Herein we report a case of a 68-year-old man with primary mediastinal liposarcoma involving the diaphragm and pericardium, which was successfully managed by complete surgical excision. The patient's postoperative course was uneventful with no evidence of recurrence 9 months after the operation.

Surgical removal is the optimal treatment for a mediastinal liposarcoma, as it is in other sites. If the entire tumor can not be resected, surgical debulking often results in symptomatic relief. International literature suggests that recurrent disease occurs and therefore a long-term careful follow up is required.

Key words: liposarcoma, mediastinum, surgery

Introduction

Primary mediastinal liposarcoma is an uncommon neoplasm of intrathoracic origin. It is usually diagnosed in later stages when the tumor has become large and symptomatic due to mass effect. Liposarcoma originating in the mediastinum represents less than 1% of mediastinal tumors [1].

A case of primary mediastinal liposarcoma involving the diaphragm and pericardium, which was successfully managed by complete surgical excision, is presented here.

Case presentation

A 68-year-old man who was otherwise well, presented with mild shortness of breath of 6-month duration and a recent onset of chest pain. Physical examination showed dullness on percussion and decreased breath sounds in the lower zone of the left lung. Laboratory data, respiratory function tests and arterial blood gas analyses were within normal limits. Chest x-ray showed a large, well defined soft tissue mass in the anterior mediastinum (Figure 1). On computed tomography (CT), an inhomogeneous fatty mass in the left hemithorax showing invasive features to the heart and left hemidiaphragm was detected (Figure 2). Search for distant metastases including bone scan, cranial and abdominal CTs showed no abnormal findings. Esophagoscopy and bronchoscopy revealed extrinsic compression effects, but no evidence of intraluminal tumor. Therefore, surgical intervention was proposed. The patient underwent a left anterolateral thoracotomy but, due to diaphragmatic invasion, an abdominal extension of the incision was needed. A large, well-demarkated and slightly lobulated mass located in the left pleural cavity, showing invasive features to the heart and left hemidiaphragm, was explored. The tumor was attached to the inferior pericardial wall causing significant compression to the heart. A pericardial incision was necessary to check possible myocardial infiltration.

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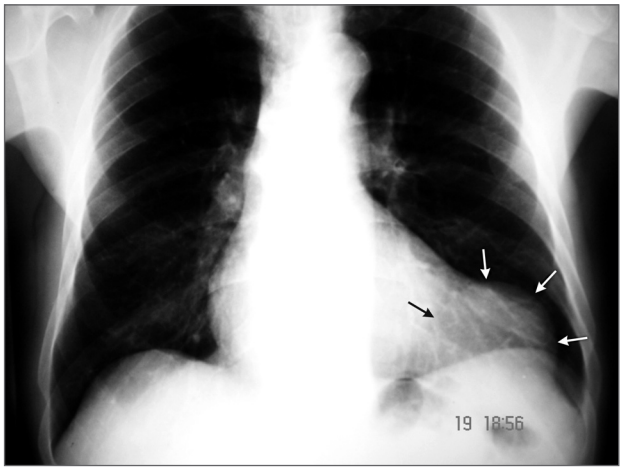

Figure 1. Chest x-ray revealed a soft tissue density mass on the left (arrows).

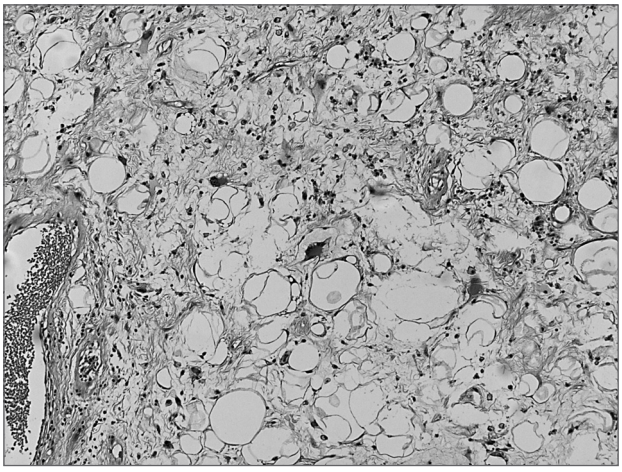

Figure 3. Photomicrograph shows fibrous bands containing atypical cells intermixed with fatty areas (H&E × 200).

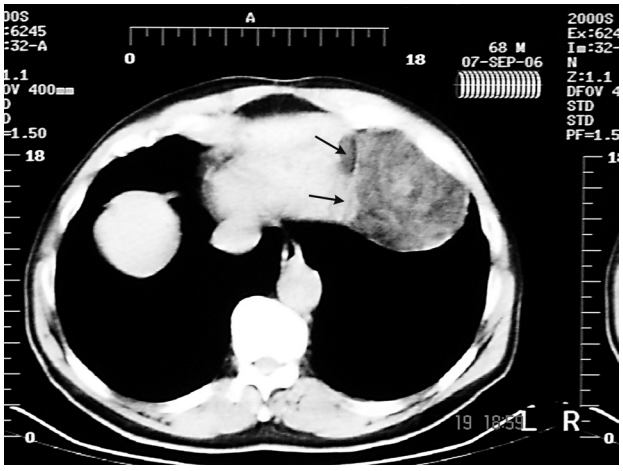

Figure 2. Computed tomography revealed an inhomogeneous fatty mass in the left hemithorax with invasive features to the heart and left hemidiaphragm (arrows).

This manipulation revealed that the tumor was in close association with the left atrium and ventricle anteriorly but showed no invasion to these vital structures.

En bloc resection of the tumor with part of pericardium and the left hemidiaphragm was performed. The diaphragmatic defect was covered with bovine pericardium (peri-guard, synovis). The patient tolerated the operation well and had an uneventful postoperative recovery. Nine months later he is free of disease and in a very good condition.

The entire tumor measured 9×7×5 cm in diameter and weighed 430 g in total. The mass was soft and pale-yellow in color on cut section. The final pathologic diagnosis was well differentiated low grade liposarcoma (atypical lipomatous tumor, Figure 3).

Discussion

Liposarcoma comprises approximately 1% of all malignancies and is the second most common soft tissue sarcoma in adults. The commonest primary site is the lower limb followed by retroperitoneum. Primary liposarcoma of the mediastinum is extremely rare, representing less than 1% of mediastinal tumors, with less than 150 cases reported in the literature. It usually occurs in adults, with most cases developing in patients more than 40 years of age [1,2]. Malignant liposarcoma develops more commonly in the posterior mediastinum. Liposarcoma of the anterior mediastinum is very rare and only a few cases have been reported in the international literature [3].

Mediastinal liposarcomas may extend into the pleural spaces and achieve a large size before detection. The presenting signs and symptoms are related to size and direct invasion of contiguous structures like the pericardium or superior vena cava [4]. Dyspnea, chest pain and tachypnea are the most common symptoms. Asymptomatic cases discovered by radiological imaging have also been reported [5]. In our case chest pain and mild shortness of breath were the main symptoms.

The predominant finding of mediastinal liposarcoma on conventional chest radiography is a widened mediastinum most of the times. On CT, the appearance of mediastinal liposarcomas, as of liposarcomas located in any part of body, varies from a predominantly fat-containing mass to a solid mass. Low attenuation values between –50 and –150 Hounsfield Unit (HU) are consistent with a tissue composed of fat. Greater values are related to necrosis, heterogeneity and soft tissue component in liposarcomas. On magnetic reso-

nance imaging (MRI), T1-weighted images show the fatty tissue with a high signal intensity, whereas the signal intensity diminishes in T2-weighted images. A differential diagnosis should be made between lipoma, thymolipoma, teratoma, lymphoma, germ cell tumor or even herniated peritoneal fat [6].

Pathologically, 4 main types of liposarcomas have been described: myxoid, well differentiated, dedifferentiated and pleomorphic. Evans reported that survival in patients with dedifferentiated or pleomorphic liposarcomas was significantly shorter than in patients with myxoid or well differentiated liposarcomas [7]. Well differentiated low-grade liposarcomas, also known as atypical lipomatous tumors, have histologic features in many areas resembling mature adipose tissue. The cytoplasm of the atypical cells is usually indistinct or amorphous and occasional cytoplasmic vacuoles are noted [8]. Evans also reported that atypical lipomatous tumors may transform to dedifferentiated liposarcomas and usually do not metastasize [7].

Conclusion

Surgical removal is the optimal treatment for a mediastinal liposarcoma, like in other sites of the body. If the entire tumor can not be resected, surgical debulking often results in symptomatic relief. Radiotherapy and chemotherapy may be added as adjuncts to surgical excision but liposarcomas seem to have low sensitivity to those therapeutic modalities [9].

Recurrence is common in deep-seated liposarcomas and it becomes apparent within the first 6 months in most cases, but it may be delayed for 5 or 10 years following the initial excision [10]. Recurrence is related to incomplete excision and tumor tissue left behind at the time of surgery. Therefore a close follow up is strongly recommended.

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