A Giant Primary Liposarcoma of the Anterior Mediastinum

Ön Mediasten Yerleşimli Dev Primer Mediastinal Liposarkom

Dev Mediastinal Liposarkom / Giant Mediastinal Liposarcoma

Erdal Yekeler[1], Hakkı Ulutas[2], Sevilay Akalp Ozmen[3]
[1]Ataturk Chest Diseases and Thoracic Surgery Center, Department of Thoracic Surgery, Ankara,
[2]University of Inonu, School of Medicine, Department of Thoracic Surgery, Malatya,
[3]Region Teaching and Research Hospital, Department of Pathology, Erzurum, Turkey.

Bu olgu Türkiye Solunum Araştırmaları Derneği (TUSAD) 33. Ulusal kongresinde poster olarak sunulmuştur.

Özet

Liposarkomlar; primitive mezenşimal hücrelerden kaynaklanan nadir tümörlerdir. Mediastinal yerleşim çok nadirdir ve tüm mediastinal tümörlerin %1'den azını oluşturur. Bu tümörler akciğerin elastikiyeti ve plevral boşluk nedeniyle büyük boyutlara ulaşıncaya kadar asemptomatik kalabilirler. Mediastinal hayati organları invaze etmiş kötü diferansiye tiplerde tam rezeksiyon her zaman mümkün olmayabilir. Biz bu makalemizde; son 3 aya kadar hiç bir şikâyeti olmayan, ön mediasten yerleşimli, dev boyutlara ulaşmış, 34 yaşında erkek hastamızı ve onun başarılı cerrahi tedavisini sunduk.

Anahtar Kelimeler

Miksoid Liposarkom; Mediasten; Cerrahi

Abstract

Liposarcomas are uncommon tumors originated from the primitive mesenchymal cells. Mediastinal liposarcoma is a very rare localization, accounting less than 1% of all the mediastinal tumors. These tumors may remain asymptomatic until they reach a giant size due to pleural space and elasticity of the lungs. Complete resection is not always possible in the poorly differential types invading the mediastinal vital organs. In this article, we presented a 34 years-old man patient with an anterior mediastinum localized liposarcoma that was asymptomatic until last 3 months and reached to giant sizes and complete successful surgical intervention.

Keywords

Myxoid Liposarcoma; Mediastinum; Surgery

DOI: 10.4328/JCAM.939 Received: 27.02.2012 Accepted: 09.03.2012 Printed: 01.03.2015 J Clin Anal Med 2015;6(2): 257-259
Corresponding Author: Erdal Yekeler, Osmangazi Mah. 230 Sok. Taner Apt. B-Blok No: 4 Yildizkent, Palandoken 25070 Erzurum, Turkey.
GSM: +905326761759 F.: +90 3123552135 E-Mail: drerdalyekeler@hotmail.com

Introduction

Liposarcomas are uncommon neoplasms originated from the primitive mesenchymal cells, accounting for 15-20% of all the sarcomas. Liposarcomas have been often reported in the lower extremities, retroperitoneum, vulva and as rarely retropharyngeum [1-5]. Primary liposarcoma of the mediastinum is one from very rare localizations, and they are seen as less than 1% among all the mediastinal tumors. Mediastinal liposarcomas are often anterior mediastinal localized and mostly reported to originate from the thymus- related fatty tissue [1,3,4,8]. In this study, we present a case with anterior mediastinal localized liposarcoma, which was originated from paracardiac fatty tissue and remained asymptomatic until reached to a giant size and successful surgical resection.

Case Report

A 34-years-old male patient who had not any complaint previously was admitted to our hospital with complaints of shortness of breath increasing on effort and right chest pain for 3 months. On X-ray radiograph, an opacity was detected, which filled the 2/3 lower part of right hemithorax (Figure-1a). On subsequent contrast-enhanced computed tomography (CT), an anterior mediastinal localized mass lesion of 36x22x17 cm in size, shifting the mediastinal organs particularly the heart leftward, pressing the diaphragm downward was found (Figure-1b). The tumor

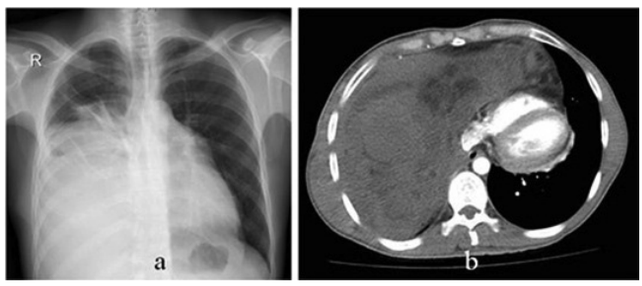


Figure 1. The opacity filling 2/3 lower part of the right hemothorax is seen on the Chest X-ray (A). On thoracic computed tomography, smooth contoured, heterogeneous density giant mass lesion completely filled the right hemothorax, and deviated the heart and mediastinal organs to leftward (B).

was smooth contoured with different density areas. Because of the low attenuation values (-65 HU) of the mass on CT it might be diagnosed as lipoma or liposarcoma. A fine needle aspiration cytology attempted from the mass was non diagnostic. Surgery was planned and then right anterior thoracotomy and inferior partial median sternotomy was performed. The mass was found to have minimal adhesions with the surrounding tissue, the tumor was encapsulated and had an expansive, non-infiltrative growth, thus making the radical resection possible. The mass was seen to be originated from the paracardiac fatty tissue and invaded at this point (Figure-2a). The tumor was en-bloc resected with the right, left paracardiac and mediastinal fatty tissue (Figure-2b). There was no evidence of pericardial invasion and mediastinal or hilar lymphadenopathy. The tumor was weighed 3.200g which was detected to be lobulated, and smooth surfaced. On histopathological evaluation of the tumor revealed the diffused distribution of lipoblastic mesenchymal cells with increased cellularity and atypical nuclei, and a plexiform capillary network, all set in a myxoid matrix, scattered strands of fibroconnective tissue, with positive results of protein S-100, Vimentin and CD34 and negative results of protein smooth muscle actin (SMA) and

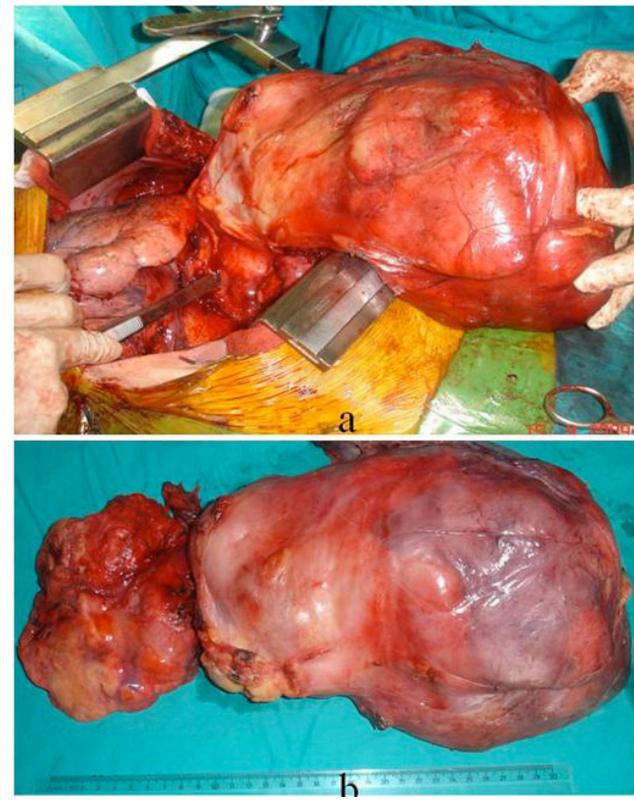


Figure 2. On the intra-operative and post-operative view, invasion in the right paracardiac area is seen after the mass was dissected from the surrounding tissues and removed out of the thorax (A, B).

desmin, examined by immunohistochemical methods, also surgical margin was tumor negative on paracardiac localization. The final diagnosis was made as a primary myxoid liposarcoma of anterior mediastinum (Figure-3). Surgical margin was tumor negative on paracardiac localization The postoperative course was uneventful; chest radiograph documented the re-expansion of the right lung and re-alignment of the mediastinum, and the patient was discharged on the 13th day post-operative. No recurrence was seen in 1-year follow-up of the case.

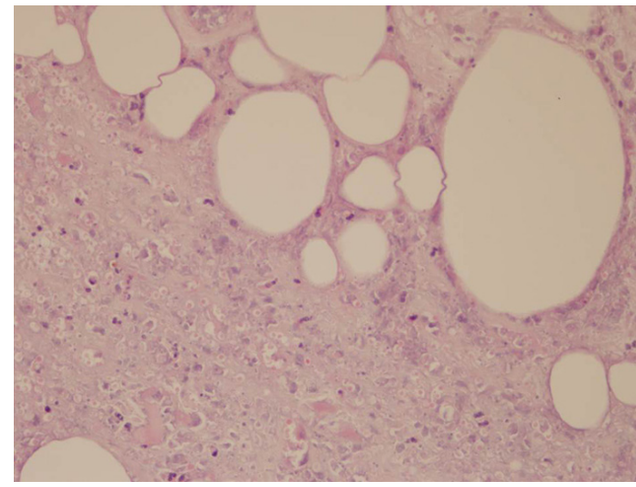


Figure 3. On microscopic examination are showed myxoid differentiation, abondant fibrinoid necrosis, and lipoblasts with vacuolated cytoplasm and atypical nuclei (H&E).

Discussion

Primary liposarcomas of the mediastinum are very rarely seen neoplasms and may remain asymptomatic until reach to a giant size. They generally occur at adults, although they have been reported in children also [1,2,6,7]. The most common symptoms

may emerge with chest pain, dyspnea, coughing, wheezing and compression to adjacent organs developing related to the tumor size. Vena cava superior syndrome, mediastinal shift and atelectasis due to the pulmonary compression and cardiac compression findings may develop [3,4,7].

Mediastinal liposarcomas may not give symptoms until they reach to a large size because of the elasticity of lung tissue and the pleural space. Liposarcoma cases reached to the large size, manifests as a mass appearance on chest X-ray radiography. Many cases are diagnosed during the routine chest graphy evaluation [1-4].

On Thorax CT evaluation, they are seen as the smooth contoured solid mass lesions consist of heterogeneous densities having a mostly fatty content, necrosis areas and soft tissue content. Especially, myxoid type calcifications and ossifications may be seen. Normal fat shows low attenuation values between -50 and -150 Hounsfield Unit (HU), whereas the attenuation of liposarcomas may show higher density because of its different tissue composition [4,6,9]. An important role of especially multislice CT is to guide the pre-operative surgical planning. CT is an effective guide in a correct incision and surgical management. Despite so much guiding informations about the nature of the tumor is obtained from CT, the histopathologic examination is necessary for the definitive pre-operative diagnosis [9,10]. MRI is better than CT-scanning in ruling out invasion of vessels in the mediastinum and thoracic inlet [10].

Primary liposarcomas are histologically divided into 5 subtypes: well-differentiated, myxoid type, round-cell, pleomorphic ve mixed cell type. Of these sub-types, myxoid type is the most common liposarcoma (40-50%)[5], while well-differentiated type has the least common recurrence (53%) and the best 5-year survival rate (70-85%). Whereas pleomorphic and round cell types have been reported to have the most common recurrence rate with 73% and 85% respectively, and the worst 5-year survival rate with 10-21% and 18%, respectively [2,6,7].

However, 5-year survival rates are directly related with the surgical resection of the primary and recurrent tumors [2]. Nevertheless, especially in the retroperitoneal and abdominal localized tumors reached to giant sizes, a tumor-reduction operation, even with an incomplete resection has been reported to improve the life quality. Furthermore, in the mediastinal poor differential type since liposarcomas often invade mediastinal structures and vital organs such as heart, complete resection in tumors localized in these areas is not always possible. Considering the limited benefit of radiotherapy and chemotherapy, we recommend de-bulking operation also in these cases.

Treatment of mediastinal liposarcomas is surgery and complete resection always gives a good outcome. In case of invasion to the mediastinal vital organs, complete resection is not always possible. Tumor-reducing surgery will provide a benefit in order to improve the life quality [1,4,7]. Benefit of the chemotherapy and radiotherapy is limited in such cases. Local irradiation is essential to prevent the recurrence in the incomplete resections. Recurrences often reveal following the invaded and incomplete resections depending on the type of liposarcoma. Treatment is surgery also in the recurrences [2,4,6,8].

Particularly, in preoperative evaluation of the primary mediastinal liposarcomas reached to giant sizes; the tumor is challenging for the surgeon due to its location and compression findings and closeness to the vital organs; however, the resection is mostly possible, because the tumor is encapsulated and its dissection is easy except the local invasions. A giant mediastinal liposarcoma which have a vital organ invasion or not, if complete resection is possible, surgery is very well curative treatment option considering chemotherapy and radiotherapy treatment options provide a highly limited benefit. If complete resection is impossible, we recommend to surgeons de-bulking operation also in these cases.

Competing interests

The authors declare that they have no competing interests.

References

1. Marulli G, Rea F, Feltracco P, Calabrese F, Giacometti C, Rizzardi G, Vincenzo L, Sartori F. Successful resection of a giant primary liposarcoma of the posterior mediastinum. J Thorac Oncol 2007;2(5):453-5.
2. Noji T, Morikawa T, Kaji M, Ohtake S, Katoh H. Successful resection of a recurrent mediastinal liposarcoma invading the pericardium: report of a case. Surg Today 2004;34(5):450-2.
3. Klimstra DS, Moran CA, Perino G, Koss MN, Rosai J. Liposarcoma of the anterior mediastinum and thymus. A clinicopathologic study of 28 cases. Am J Surg Pathol 1995;19(7):782-91.
4. Kara M, Ozkan M, Dizbay Sak S, Kavukçu ST. Successful removal of a giant recurrent mediastinal liposarcoma involving both hemithoraces. Eur J Cardiothorac Surg 2001;20(3):647-9.
5. Cengel S, Hosal AS. Retropharyngeal Liposarcoma Presenting with Dysphagia and Weight Loss: Case Report. Turkiye Klinikleri J Med Sci 2010;30(4):1421-4.
6. Hirai S, Hamanaka Y, Mitsui N, Uegami S, Matsuura Y. Surgical resection of primary liposarcoma of the anterior mediastinum. Ann Thorac Cardiovasc Surg. 2008;14(1):38-41.
7. Chiyo M, Fujisawa T, Yasukawa T, Shiba M, Shibuya K, Sekine Y, Hiroshima K, Ohwada H. Successful resection of a primary liposarcoma in the anterior mediastinum in a child: report of a case. Surg Today 2001;31(3):230-2.
8. Sekine Y, Hamaguchi K, Miyahara Y, Baba M, Yasufuku K, Fujisawa T, Yamaguchi Y. Thymus-related liposarcoma: report of a case and review of the literature. Surg Today 1996;26(3):203-7.
9. Santamaría G, Serres X, Pruna X, Forcada P. Primary mediastinal liposarcoma with moderately high CT attenuation. AJR Am J Roentgenol 1996;167(4):1064-5.
10. Munk PL, Lee MJ, Janzen DL, Connell DG, Logan PM, Poon PY, Bainbridge TC. Lipoma and liposarcoma: evaluation using CT and MR imaging. AJR Am J Roentgenol 1997;169(2):589-94.