Clinical Case Report

Medicine®

OPEN

Anterior mediastinal leiomyosarcoma mimicking thymoma

A case report

Xing Xue, MD[a], Wenjie Liang, MD[a], Wenjuan Zhang, MD[b,*]

Abstract

Rationale: Mediastinal leiomyosarcoma is very rare. In this paper, we report one case of anterior mediastinal leiomyosarcoma and retrospectively review the imaging findings of previously reported cases.

Patient Concerns: A 61-year-old male patient was admitted to our hospital due to the presence of a mediastinal space-occupying lesion for two years. Two years previously, chest computed tomography (CT) showed a small nodule in the anterior mediastinum. Twenty days previously, the CT examination was repeated at the local hospital due to respiratory symptoms, which showed a mass in the anterior mediastinum and interstitial inflammation of both lungs. After admission to our hospital, chest contrast-enhanced CT scanning showed a mass in the right anterior mediastinum that was approximately 3.3 × 5.2 cm2 in size that had a clear boundary, slightly heterogeneous internal density and heterogeneous enhancement.

Interventions: The patient underwent a mediastinal lump resection.

Diagnoses: Finally, the pathological diagnosis was anterior mediastinal leiomyosarcoma.

Outcomes: The patient recovered well after the operation.

Lessons: Accidental discovery of anterior mediastinal nodules should be followed up. Mediastinal leiomyosarcoma is common in the posterior mediastinum. Imaging shows a heterogeneous mass with a space-occupying effect that may easily involve adjacent mediastinal vessels or infiltrate surrounding organs.

Abbreviations: CT = computed tomography, DWI = diffusion-weighted imaging, hydroxydaunorubicin oncovin prednisolone, FDG = [^{18}F]-2-fluoro-2-deoxy-D-glucose, MRI = magnetic resonance imaging, PET = positron emission tomography.

Keywords: CT, imaging, leiomyosarcoma

1. Introduction

Mediastinal sarcomas are rare, and mediastinal leiomyosarcomas are even rarer, accounting for approximately 10% of mediastinal sarcoma cases.[1] A leiomyosarcoma is a malignant tumor originating in the mesenchymal tissue. Mediastinal leiomyosarcomas often arise from mediastinal organs, such as the heart and its inflow and outflow tracts, great vessels, and the esophagus. Mediastinal leiomyosarcomas arising from mediastinal soft tissue are very rare.[2]

No more than 40 cases of mediastinal leiomyosarcomas have been reported to date.[3] Most of the reported cases are individual cases, and the clinical and pathological features of mediastinal leiomyoma are reported in few of these cases, whereas imaging findings are available for even fewer cases.[4–16] In this study, we report 1 case of anterior mediastinal leiomyoma confirmed by surgical resection and pathology, with the corresponding clinical and imaging findings. We also summarize the imaging features of mediastinal leiomyosarcoma by reviewing the previous literature.

1.1. Consent

The patient signed the relevant documents agreeing to allow the use of his clinical and imaging data for scientific research or publication.`

2. Case report

The patient is a 61-year-old man who was admitted to our hospital due to a mediastinal mass, interstitial lung disease, and hypertension. Two years ago, chest computed tomography (CT) scanning in the local hospital indicated diffuse interstitial changes in both lungs, pulmonary bullae in the 2 upper lungs, and a small nodule in the anterior mediastinum, which was considered a slightly enlarged lymph node. The patient was not re-examined at specific temporal intervals. More than 20 days ago, the patient coughed up white phlegm and presented with fever and pain in the chest. A chest CT at the local hospital showed that the right anterior mediastinal space was occupied, along with bronchitis

Editor: N/A.

The authors report no conflicts of interest.

[a] Department of Radiology, the First Affiliated Hospital, College of Medicine, Zhejiang University, Hangzhou, Zhejiang, [b] Department of Anesthesiology, the First People's Hospital of Taicang City, Taicang Affiliated Hospital of Soochow University, Taicang, Suzhou, China.

[*] Correspondence: Wenjuan Zhang, Department of Anesthesiology, the First People's Hospital of Taicang City, Taicang Affiliated Hospital of Soochow University, Department and Institution: No. 58, Taicang, Suzhou 215400, China (e-mail: cztchos@163.com).

Medicine (2018) 97:25(e11132)

Received: 4 March 2018 / Accepted: 15 May 2018

http://dx.doi.org/10.1097/MD.0000000000011132

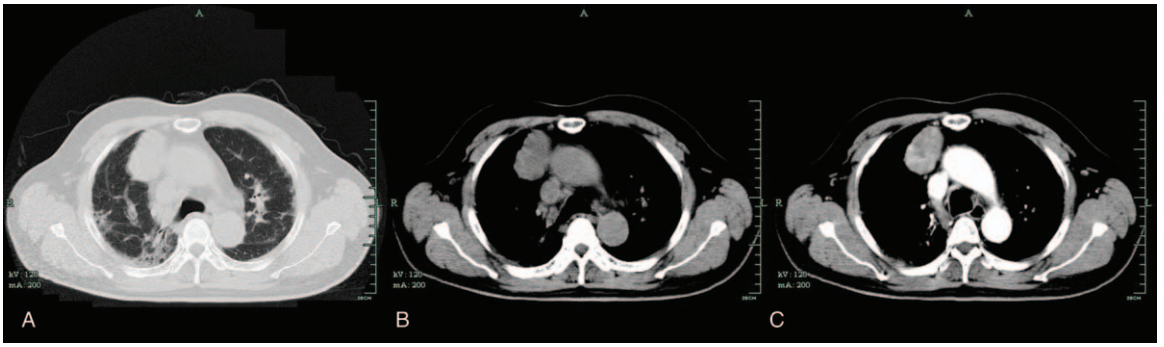

Figure 1. (A) Lung window. Lung CT shows 2 lungs with patchy grid hyperdensity remarkable under the pleura. A mass is located in the anterior mediastinum. (B and C) Mediastinum window. The anterior mediastinal mass shows an heterogeneous texture, a clear boundary, and inhomogeneous enhancement. CT = computed tomography.

and an enlarged heart with a thickened pericardium. The patient was then admitted to our hospital for further diagnosis and treatment. He had recently shown no significant reduction in weight. The patient had exhibited hypertension for more than 5 years, controlled with nifedipine. He had a history of alcoholic liver and rheumatoid arthritis for more than 5 years, for which he had previously taken medication (eg, methotrexate), which had been discontinued for several months. He also had a long history of dust exposure and drinking habits (100 mL every day, continuously for 40 years). The patient had no smoking habits. Upon the discovery of pulmonary auscultation, heavy lung respiratory murmurs were recorded.

At admission, the patient's blood indexes were basically normal, with the exception of slightly higher eosinophil levels (6%). He tested positive for perinuclear antineutrophil cytoplasmic antibody (p-ANCA), and his hypersensitive C-reactive protein level was slightly increased. Tumor markers were not elevated. At our institution, chest-unenchanced and contrast-enhanced CT was performed, which showed a right anterior mediastinal soft tissue mass with a size of approximately 5.2 × 3.3 cm,[2] the boundary was still clear, and the internal density was homogeneous (about 35 HU). Contrast-enhanced CT scanning showed the mass with slightly heterogeneous enhancement, and the CT value was approximately 32 to 81 HU. There was no obvious enlargement of the lymph nodes in the mediastinum (Fig. 1). In this case, thymoma was first considered, based on the clinical and imaging features of middle-aged men, the anterior mediastinum, and inhomogeneous enhancement.

After evaluation of the operation, mediastinal mass resection was performed. In the operation, the tumor was located in the anterior superior mediastinum and was found to be hard in texture and to exhibit a pale flesh color in cross-section. The tumor invaded the right upper lung, and there was no metastatic nodule in the thoracic cavity. The lung exhibited interstitial fibrosis, especially in the right lower lung. Microscopy showed that the tumor cells were braided and palisade in shape, with a large number of strange nuclei. The cells were markedly irregular; nuclear division was easily visible (>5/10 high power field); and hemorrhage and necrosis could be seen in some regions (Fig. 2). The immunohistochemistry results showed the following: CD117 (−), S-100 (−), desmin (+), CD34 (−), smooth muscle actin (−), anoctamin–1 (−), Ki-67 (40%), cytokeratins (−), and myogenic differentiation 1 (−). The final pathological diagnosis was (anterior mediastinal) leiomyosarcoma. The patient recovered well after the operation. Unfortunately, the patient was lost in the follow-up.

3. Discussion

Mediastinal leiomyosarcoma appears in adults (26–79 years of age) and occurs preferentially in males.[2,3,4,7,13] Mediastinal leiomyosarcoma often appears in the posterior mediastinum (10/15 cases),[7] but can also appear in other mediastinal locations, including the anterior mediastinum.[7,13] Leiomyosarcomas of the mediastinum can be asymptomatic and can result in nonspecific symptoms, such as coughing, chest pain, back pain, and shortness of breath.[7,13] The case of leiomyosarcoma reported herein involved an older males, similar to previous reports. However, our case involved an anterior mediastinal location and rheumatoid arthritis (RA); we have not yet determined whether there is any correlation between mediastinal leiomyosarcoma and RA. Furthermore, in our case, the mediastinal lesion was detected during a health examination and was very small at that time, similar to the size of lymph nodes; therefore, not enough attention was initially paid to the lesion. Two years later, the tumor was markedly enlarged. Thus, we suggest that small nodules in the anterior mediastinum should be closely followed up.

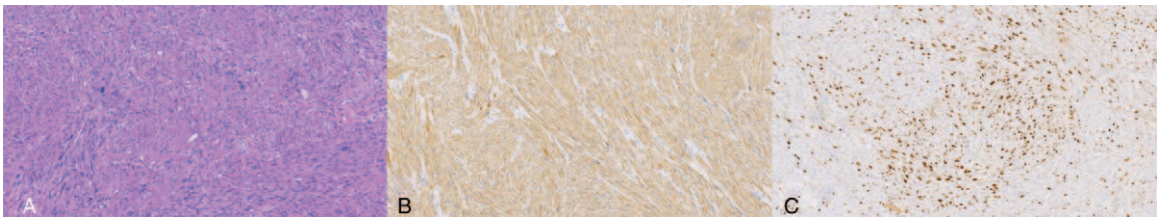

Figure 2. (A) The tumor cells are arranged in a braid-shaped pattern with a distinct nucleus, and bleeding and necrosis can be seen in the region (HE staining; 100× magnification). (B) Desmin staining is positive (100× magnification). (C) Ki-67 shows that tumor cell proliferation is active (100× magnification).

In chest radiography or CT, mediastinal leiomyosarcomas are usually characterized by very large soft tissue masses with a maximum diameter of 4.5 to 18.0 cm.[4–13,15,16] Mediastinal leiomyosarcomas can show an obvious occupying effect. When a leiomyosarcoma is located in the posterior mediastinum, the tumor can push against surrounding organs, such as the esophagus, descending aorta, the spinal canal, and the inferior vena cava, which can occasionally be affected.[4,7,11,15] When a leiomyosarcoma is located in the anterior mediastinum, the boundary of the lesion is clear and occasionally calcified.[4–6,14,16] It may surround the vena cava and form an embolism.[4–6,14,16] When the volume of tumor becomes large, it can protrude into 1 side of the pleural cavity, and effusion can occur in the pleura.[5,6,16] The tumor can show heterogeneous enhancement because of bleeding, necrosis, and a heterogeneous texture.[2,4,6,16] Magnetic resonance imaging (MRI) examination may allow a clear diagnosis of paraneoplastic thrombosis.[14] When a leiomyosarcoma is located in the visceral mediastinum, it is related to the tumor originating from the mediastinal great vessels.[8] A visceral mediastinal leiomyosarcoma can infiltrate the adjacent pulmonary hilum, pulmonary vein, and left atrium.[8–10] Occasionally, a middle mediastinal leiomyosarcoma may be accompanied by fatal intra-aortic thrombosis.[13] A mediastinal leiomyosarcoma is usually not associated with enlargement of the lymph nodes.[10,16] The performance of MRI and positron emission tomography (PET) for the identification of mediastinal leiomyosarcomas is addressed little in the literature. Upon MRI, leiomyosarcomas are seen with enhancement and have the appearance of water molecule diffusion confined in the diffusion-weighted imaging (DWI) sequence. Mediastinal leiomyosarcomas show high [^{18}F]-2-fluoro-2-deoxy-D-glucose (FDG) uptake under PET.[10] In our case, similar to previous reports, contrast-enhanced CT also showed heterogeneous enhancement. Although marked bleeding and necrosis were lacking, the pathology still showed regional hemorrhage and necrosis. Therefore, the possibility of leiomyosarcoma should be considered when large mediastinal tumors are accompanied by bleeding and necrosis in imaging examinations.

In the case of leiomyosarcoma present in the anterior mediastinum, it should be differentiated from thymoma and mediastinal germinoma on CT. When leiomyosarcoma is present in the posterior mediastinum, it should be differentiated from neurogenic tumors. In this case, the following characteristics: the site and regular morphology, homogeneous density, and also clear border, are similar to those of low-risk thymoma. While high-risk thymoma generally presents with irregular morphology, heterogeneous enhancement, and infiltration of the peripheral space.[17,18] The clinical characteristics may be useful to identify leiomyosarcoma from thymoma because approximately one-third of the thymoma patients are accompanied with myasthenia gravis. Different from leiomyosarcoma in this case, the malignant mediastinal germinoma is usually irregularly shaped, with heterogeneous enhancement and invasion of adjacent structures, occasionally accompanied by enlarged lymph nodes. In addition, patients with mediastinal germinoma are relatively young. Abnormal elevations of plasma tumor markers, such as alpha-fetoprotein and human chorionic gonadotropin, also contribute to the identification with anterior mediastinum. The leiomyosarcoma in the posterior mediastinum is generally large in size, irregular in morphology, with obvious occupying effect. In contrast, neurogenic tumors often present with regular shape, homogeneous density (or cystic degeneration), and clear borders. Further more, the close relationship between tumors and nerve roots contributes to the diagnosis of neurogenic tumors as well.

Mediastinal leiomyosarcomas are usually treated with surgical resection.[4,7] However, a certain proportion of mediastinal leiomyosarcomas are prone to recur, and patients die postoperatively after approximately 2 to 7 years.[3,4,7]

4. Conclusions

In conclusion, leiomyosarcomas are common in the posterior mediastinum and may also be seen in the anterior mediastinum. A leiomyosarcoma can surround adjacent mediastinal vessels or infiltrate surrounding organs. Leiomyosarcomas should be included in the differential diagnosis when the mediastinal mass shows inhomogeneous enhancement.

Author contributions

Conceptualization: Xing Xue, Wenjuan Zhang.
Data curation: Xing Xue, Wenjie Liang, Wenjuan Zhang.
Formal analysis: Xing Xue.
Project administration: Wenjie Liang.
Writing – original draft: Xing Xue, Wenjie Liang, Wenjuan Zhang.
Writing – review & editing: Xing Xue, Wenjie Liang, Wenjuan Zhang.

References

[1] Burt M, Ihde JK, Hajdu SI, et al. Primary sarcomas of the mediastinum: results of therapy. J Thorac Cardiovasc Surg 1998;115:671–80.
[2] Gladish GW, Sabloff BM, Munden RF, et al. Primary thoracic sarcomas. Radiographics 2002;22:621–37.
[3] den Bakker MA, Marx A, Mukai K, et al. Mesenchymal tumours of the mediastinum: part II. Virchows Arch 2015;467:501–17.
[4] Moran CA, Suster S, Perino G, et al. Malignant smooth muscle tumors presenting as mediastinal soft tissue masses. A clinicopathologic study of 10 cases. Cancer 1994;74:2251–60.
[5] Van Schil PE, Colpaert CG, Van Look R, et al. Primary mediastinal leiomyosarcoma. Thorac Cardiovasc Surg 1993;41:377–8.
[6] Eroğlu A, Kürkçüoğlu C, Karaoğlanoğlu N, et al. Primary leiomyosarcoma of the anterior mediastinum. Eur J Cardiothorac Surg 2002;21:943–5.
[7] Hirano H, Kizaki T, Sashikata T, et al. Leiomyosarcoma arising from soft tissue tumor of the mediastinum. Med Electron Microsc 2003;36:52–8.
[8] Conner WC, Fink GW, McGinnis KM, et al. Surgical management of leiomyosarcoma of the mediastinum. Ann Thorac Surg 2004;77:334–6.
[9] Cakir O, Topal U, Bayram AS, et al. Sarcomas: rare primary malignant tumors of the thorax. Diagn Interv Radiol 2005;11:23–7.
[10] D'Aiuto M, Veronesi G, Pompilio G, et al. Extended right pneumonectomy with partial left atrial resection for primary leiomyosarcoma of the mediastinum. J Thorac Cardiovasc Surg 2005;129:694–5.
[11] Lee DH, Park CK, Keum DY, et al. Leiomyosarcoma of the posterior mediastinum extending into the adjacent spinal canal. Korean J Thorac Cardiovasc Surg 2012;45:192–5.
[12] Vaziri M. Primary mediastinal leiomyosarcoma. Gen Thorac Cardiovasc Surg 2012;60:522–4.
[13] Yoshida M, Ando S, Naito Y, et al. Mediastinal leiomyosarcoma concurrent with intra-aortic thrombosis. BMJ Case Rep 2013;2013: bcr2012007527.
[14] Labarca E, Zapico A, Ríos B, et al. Superior vena cava syndrome due to a leiomyosarcoma of the anterior mediastinum: a case report and literature overview. Int J Surg Case Rep 2014;5:984–7.
[15] Chen F, Muranishi Y, Minakata K, et al. Complete resection of a giant mediastinal leiomyosarcoma. Ann Thorac Surg 2015;99:e69–71.
[16] Liao CY, Chien ST, Huan SF. Primary leiomyosarcoma of the anterior mediastinum. QJM 2015;108:429–30.
[17] Priola AM, Priola SM, Di Franco M, et al. Computed tomography and thymoma: distinctive findings in invasive and noninvasive thymoma and predictive features of recurrence. Radiol Med 2010;115:1–21.
[18] Hu YC, Wu L, Yan LF, et al. Predicting subtypes of thymic epithelial tumors using CT: new perspective based on a comprehensive analysis of 216 patients. Sci Rep 2014;4:6984.