Rare disease

Giant liposarcoma of the posterior mediastinum and retroperitoneum

Katsunobu Taki, Masayuki Watanabe, Shiro Iwagami, Yohei Nagai, Masaaki Iwatsuki, Takatsugu Ishimoto, Yoshifumi Baba, Yuji Miyamoto, Hideo Baba

Department of Gastroenterological Surgery, Kumamoto University, Kumamoto, Japan

Correspondence to Dr Masayuki Watanabe, masanabe@fc.kuh.kumamoto-u.ac.jp

Summary

A 39-year-old Japanese man has been complaining of chest pain for 3 months and a CT revealed a huge tumour in the posterior mediastinum and retroperitoneum. The mediastinal tumour was continuous with the retroperitoneal one through the oesophageal hiatus and the lower oesophagus was completely surrounded by the tumour. MRI suggested lipogenic tumour. Resection of the tumour with oesophagectomy was performed through bilateral thoracotomy and laparotomy. Pathological examination revealed well-differentiated liposarcoma which was completely resected. The patient has been alive without recurrence for 14 months after surgery.

BACKGROUND

Liposarcoma is a common soft tissue sarcoma which usually occurs in adults. The common sites of occurrence are the extremities, retroperitoneum and inguinal region. We have experienced a rare case of a giant liposarcoma located in the posterior mediastinum and retroperitoneum, surrounding the oesophagus. We successfully resected the tumour with an oesophagectomy through bilateral thoracotomy with a laparotomy. Here, we describe the feature of imaging studies of this rare tumour and demonstrate the operative procedures we undertook.

CASE PRESENTATION

A 39-year-old Japanese man complained of chest pain for 3 months before admission. He had a good general condition and physical examination and laboratory data demonstrated no abnormality.

INVESTIGATIONS

CT of the thorax and abdomen revealed a huge tumour, 40×30×15 cm in size, in the posterior mediastinum and the retroperitoneum (figure 1). The mediastinal tumour was continuous with the retroperitoneal tumour through the oesophageal hiatus, and the lower oesophagus was completely surrounded by the tumour. The tumour contained areas of both fat and soft tissue density, and was scarcely enhanced with intravenous contrast medium. An ¹⁸F-fluorodeoxyglucose positron emission tomography (FDG-PET) revealed that small amount of FDG accumulated in the soft tissue density area. MRI demonstrated high intensity activity in the tumour by both T1-weighted and T2-weighted imaging, and there were many fibrous septa observed in the tumour (figure 2). These imaging studies indicated a lipogenic tumour and liposarcoma was highly suspected.

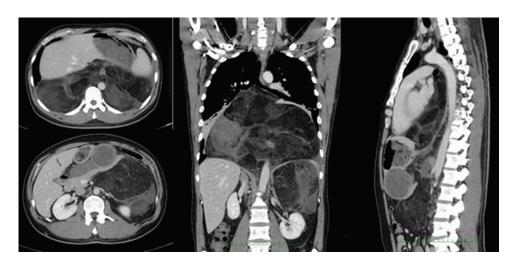


Figure 1 A CT revealed a huge tumour located in the posterior mediastinum continuous to the retroperitoneum.

BM Case Reports

TREATMENT

As a surgical resection was considered to be the only treatment to cure, surgery was performed. At first, under a general anaesthesia with the patient in a left lateral position, a right thoracotomy was undertaken through the fifth intercostal space. The tumour was located in the posterior mediastinum and adherent to the pleura, pericardium and oesophagus. The tumour was dissected sharply

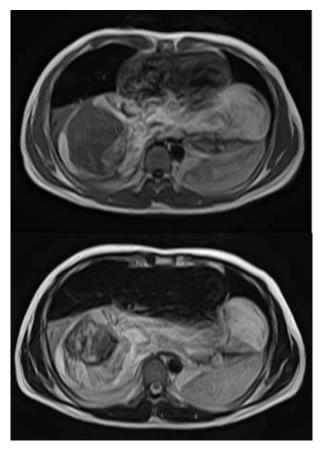


Figure 2 A MRI demonstrated low intensity tumour both in T1-weighted and T2-weighted images. A thick fibrous septa is observed in the tumour.

and bluntly from the surrounding organs. After the dissection was completed, the patient position was changed to a supine one and an upper midline skin incision was made for laparotomy. The tumour was located mainly in the retroperitoneum and adherent to the pancreas and mesocolon. The spleen, pancreas tail and descending colon were mobilised from the retroperitoneum, and the tumour was dissected from these organs. Finally, the patient was changed to a right hemi-lateral position, a left thoracotomy was made through the sixth intercostal space continuous to the laparotomy. The tumour was dissected from the left pleura and became free from the surrounding organs except the oesophagus, which ran through the tumour (figure 3A). At this point, it was decided to perform a combined resection of the oesophagus. A subtotal oesophagectomy was performed and the tumour was completely resected (figure 3B). As there was risk of local recurrence, a gastric conduit reconstruction was performed through the retrosternal route. Histopathologic examination revealed a well-differentiated liposarcoma (figure 4).

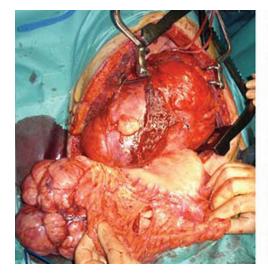
OUTCOME AND FOLLOW-UP

The postoperative course was uneventful and he was discharged on 18 postoperative day. At present, he has been alive without recurrence for 14 months.

DISCUSSION

The incidence of sarcoma is relatively low, composing approximately 0.8% of all newly diagnosed malignancies. Of these sarcomas, liposarcoma accounts for approximately 20%, and 13% of them originate from the retroperitoneum. A mediastinal liposarcoma is extremely rare ¹² and there are few report of the mediastinal extension from the retroperitoneal liposarcoma. This is the first case report of liposarcoma which extended from the retroperitoneum to the mediastinum.

In most patients with liposarcoma, definite symptoms are lacking and the diagnosis is usually made by CT or MRI. Well-differentiated liposarcomas resemble lipomas, with attenuation (CT) and signal intensity (MRI) equal to those of fat. Fibrous septa may be thicker, more irregular, or more nodular than those seen in lipoma.³ In this case,



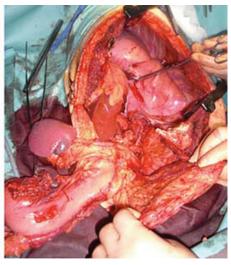


Figure 3 (A,B) A complete removal of the tumour was achieved with combined resection of the oesophagus.



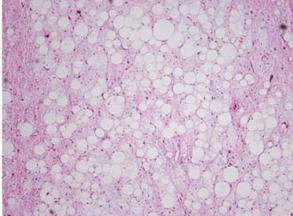


Figure 4 Macroscopic and microscopic appearance of the resected tumour. Pathologic examination revealed well-differentiated liposarcoma (X100).

both CT and MRI suggested a lipogenic tumour and thick fibrous septa was clearly observed in MRI.

Current recommendation in treatment for liposarcoma is en bloc removal if possible, as this is the only definitive treatment option. As well as the other soft tissue sarcomas, local control is critical and largely depends on the quality of surgery. In this case, we chose bilateral thoracotomy and laparotomy to approach the tumour, as it was the only method for en bloc removal of the tumour. Moreover, we performed oesophagectomy with the tumour resection, although it was a highly invasive surgery for the patient. As a result, a complete resection of the tumour was achieved. The patient is alive without disease at 14 months after surgery without any signs of recurrence.

Learning points

- Mediastinal liposarcoma or mediastinal extension of the abdominal liposarcoma is extremely rare.
- ► Although a CT and a MRI are useful to diagnose lipogenic tumour, differential diagnosis of liposarcoma from lipoma is usually difficult.
- A complete resection of the tumour is the only treatment for cure.

Competing interests None.

Patient consent Obtained.

REFERENCES

- Hsu JS, Kang WY, Liu GC, et al. Giant fibrolipoma in the mediastinum: an unusual case. Ann Thorac Surg 2005;80:e10–12.
- Lin YY, Su WF. Liposarcoma of the anterior mediastinum and neck. N Engl J Med 2010;363:864
- Pereira JM, Sirlin CB, Pinto PS, et al. CT and MR imaging of extrahepatic fatty masses of the abdomen and pelvis: techniques, diagnosis, differential diagnosis, and pitfalls. *Radiographics* 2005;25:69–85.
- Wiedemann D, Schistek R, Gassner E, et al. Mediastinal liposarcoma. J Card Surg 2011;26:162–4.
- Stojadinovic A, Leung DH, Hoos A, et al. Analysis of the prognostic significance of microscopic margins in 2,084 localized primary adult soft tissue sarcomas. Ann Surg 2002;235:424

 –34.

This pdf has been created automatically from the final edited text and images.

Copyright 2011 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit http://group.bmj.com/group/rights-licensing/permissions.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Taki K, Watanabe M, Iwagami S, Nagai Y, Iwatsuki M, Ishimoto T, Baba Y, Miyamoto Y, Baba H. Giant liposarcoma of the posterior mediastinum and retroperitoneum. *BMJ Case Reports* 2011;10.1136/bcr.06.2011.4341, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- ► Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow