A dedifferentiated large posterior mediastinal liposarcoma – An incidental finding successfully resected

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SUMMARY

This is a case of a posterior mediastinal mass in an asymptomatic gentleman, which was resected successfully and he has been disease free for more than a year of follow up. The histopathology findings happen to be a rare occurrence.

KEY WORDS:
Posterior mediastinal mass, liposarcoma, dedifferentitated

INTRODUCTION

Primary mediastinal liposarcoma is an extremely rare tumour, with a <1% incidence of all liposarcomas. Most liposarcomas present in the limbs and retroperitoneum.[1,2] We report a 48-year-old lorry driver with an incidental finding of large left sided intrathoracic mass during a routine medical check-up.

CASE REPORT

Mr S is a 48-year-old lorry driver, who is asymptomatic, presented with an incidental finding of a large left hemithorax mass on a chest X-ray (CXR) done during a routine medical check-up (Figure 1A). Computed tomography (CT) scan was performed and showed a 14 x 10 x 15 cm heterogenous, well encapsulated and lobulated mass residing in the left posterior mediastinum abutting the pericardium (Figure 1B). No other mass or lymph nodes were seen. His lung function test showed severe restriction, probably due to the compression by the mass. Echocardiogram was normal.

He underwent left posterolateral thoracotomy and excision of the mass. Intraoperatively, the mass was seen arising from the attachment of the left inferior pulmonary ligament; and adhered to the left lower lobe and diaphragm without infiltration. The blood supply to the mass originated from the inferior pulmonary ligament itself, and was resected completely. No pleural effusion or pleural nodules noted. Postoperative recovery was uneventful and he was discharged on Day 5 of surgery.

Histopathological report showed mixed type liposarcoma with evidence of dedifferentiation into leiomyosarcoma. The spindle cell present in the dedifferentiated area were positive to smooth muscle actin and desmin (Figure 2). The immunohistochemistry markers, S100 and CD34 were positive in the differentiated part of the tumour, but negative in the dedifferentiated area. The few primitive mesenchymal cells were positive to p53. Margins of resection were all clear of tumour.

No adjuvant treatment was advocated for him. He was seen in the clinic at two weeks and three months after the surgery and had no pending issues. Repeated lung function test was normal. He was followed up at six months and CT thorax at one year post-surgery showed no evidence of recurrence.

DISCUSSION

The common types of sarcomas are leiomyosarcoma (18%), fibrosarcoma (14%) and liposarcoma (13%).[3] Liposarcoma usually occurs in the limbs or the retroperitoneal region. Mediastinal liposarcoma is rare, with less than 1% of incidence.[1,3]

Mediastinal liposarcoma is more commonly seen in the anterior mediastinum, followed by posterior mediastinum, pleura and pulmonary. The incidence of posterior mediastinal liposarcoma is around 9%.[4] There are very few cases reported worldwide. There is no association between the site of the tumour and the prognosis of the patient; however, the surgical difficulty in accessing an anterior mediastinum tumour may increase the morbidity.[3,6,7]

In a series of 24 cases of mediastinal liporsarcoma by Hahn et al, the average age of the patients was 51 years.[7] Another series of 25 patients over a span of 42 years by Liang *et al* showed the average age of their patients to be 45 years.[6] Our patient was diagnosed at the age of 48 years and he fits into the age group found by these two considerably large series.

The presenting symptoms will be associated with the size of the mass and its compression of adjacent structures. The symptoms can be shortness of breath, arrhythmia, gastroesophageal reflux disease, dysphagia, chest discomfort or persistent cough.[1] Some of the patients may be asymptomatic, similar to Mr S.

Common radiological presentation will be evidence of widened mediastinum on CXR. CT scan of thorax will reveal characteristic features that enables the radiologist to have a high suspicion of liposarcoma. Those characteristics are

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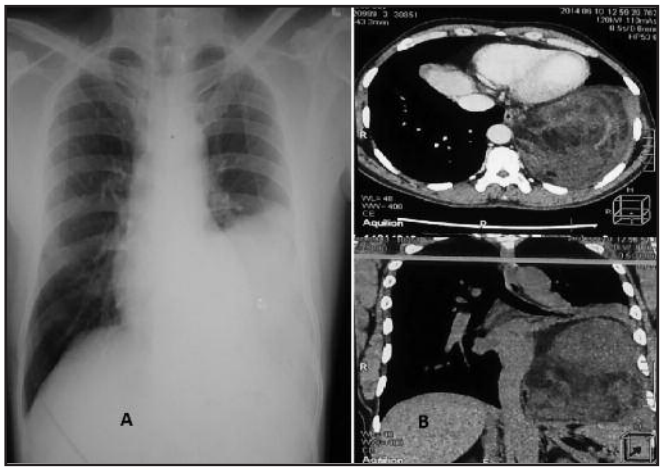

Fig. 1: (A) Chest X-ray of the patient, posterior-anterior view. (B) Computed tomography scan of the patient, both axial and coronal view.

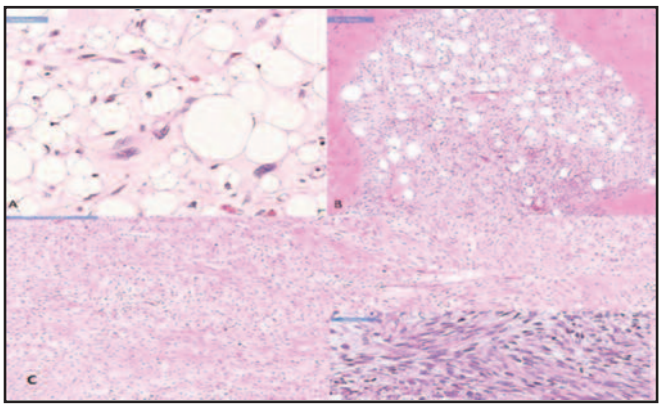

Fig. 1: (A) Section from the tumour composed of variable sized adipocytes, lipoblasts and atypical stromal cells with hyperchromatic nuclei.
(B) In another section of the tumour, the neoplastic adipocytes are disposed in a myxoid background, associated with distinctive plexiform capillary network.
(C) Area of dedifferentiation within the tumour made up of spindle cells forming interlacing smooth muscle fascicles, containing moderately pleomorphic hyperchromatic nuclei, positive to SMA and desmin.

large intrathoracic mass (>70mm); well defined, smooth or lobulated margins; presence of intramural vessels and absence of lymphadenopathy.[5] In the CT images of Mr S, all these characteristics were present, which could be a learning point to refer similar patients early to a thoracic centre once identified.

There are six different subtypes of liposarcoma; well differentiated, myxoid, round cell, pleomorphic, mixed and dedifferentiated.[2] Well differentiated liposarcoma have the best prognosis compared to the rest. The overall survival amongst patients with pleomorphic or dedifferentiated liposarcoma was significantly shorter compared to the well differentiated type.[3] This patient's histopathology revealed a mixed type liposarcoma with dedifferentiation and a scattered positivity to p53, hence his prognosis is not good.

Liang *et al* described a significant difference in the survival between patients who underwent complete resection and those with positive margin. Radiotherapy did not show any significant improvement in survival.[6] Complete surgical resection is the most important criteria in determining the survival of the patient, as demonstrated in this case.[4] Chemoradiotherapy has shown to be of minimal benefit and owing to the rarity of the condition, a randomised study of any manner will be hard to come by.

CONCLUSION

Posterior mediastinal liposarcoma with the histology of mixed type with dedifferentiation is very rare and to our knowledge, have never been reported before locally. Complete surgical resection is the cornerstone of the treatment at the moment, provided there is no evidence of distant metastasis. There is minimal role of chemoradiotherapy as neoadjuvant or adjuvant treatment. By identifying these cases with clinical presentation and imaging, early referral should be made for resection.

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