Annals of Clinical Case Reports

Case Report
Published: 01 Apr, 2019

Metastatic Pericardial Liposarcoma - A Successful Multimodality Treatment Approach

Krishnasamy S[1], Nair AK[1], Hashim SA[1], Ramesh S[2] and Raja Mokhtar RA[1]*

[1]*Cardiothoracic Unit, University of Malaya Medical Centre, Malaysia*

[2]*Department of Mechanical Engineering, University of Malaya, Malaysia*

Abstract

Liposarcoma is the second most common type of soft tissue malignancy in adults. It usually develops in the retroperitoneum and lower limbs. Although this tumour usually metastasizes to various sites such as the lungs, chest wall and retroperitoneal space, cardiac metastasis especially to the pericardium is extremely rare. This may prove to be a diagnostic and treatment challenge with regards to other types of lesions in the pericardium. An accurate diagnosis, identifying the location, tissue composition and nature of the mass is vital in deciding on early surgical treatment. We report a case of metastatic pericardial liposarcoma that presented with cardiac tamponade after 25 years of the initial liposarcoma in the popliteal fossa.

Case Presentation

A 55-year-old gentleman with underlying hypertension and dyslipidaemia presented to us in December 2015 with increasing shortness of breath for the past 2 weeks. He has a history of left knee liposarcoma at the popliteal fossa and underwent complete surgical excision in 1990, with no evidence of recurrence since then. Chest radiography (Figure 1) revealed an enlarged cardiac shadow with some widening of the mediastinum and atelectasis of the right lung in the mid zone. Electrocardiogram showed normal sinus rhythm and the cardiac enzyme markers were within normal limits. An echocardiogram was also performed and showed global pericardial effusion with features of pericardial tamponade. There was also a mass noted lateral to the left ventricle. Urgent pericardiocentesis was done and 1.3 litres of haemorrhagic fluid was drained.

A CT thorax with contrast (Figure 2) done a week later revealed left posterior pericardial mass measuring 4.6 cm × 4.7 cm × 5.0 cm with minimal right pericardial effusion and scattered non-specific lung nodules in both the lung fields (<5 mm). An initial diagnosis of pericardial cyst was made and patient was kept under surveillance. Hence a repeat surveillance CT thorax (Figure 3) was performed 6 months later and showed the mass had enlarged in size to 6.9 cm × 5.0 cm × 5.7 cm with resolved pericardial effusion and stable non-specific lung nodules. He subsequently underwent a left Video-assisted Thoracoscopic Surgery (VATS) biopsy of the mass which turned out to be a myxoid liposarcoma of intermediate grade. The mass was found to be densely adhered to the pericardium.

He then underwent a cardiac MRI (Figure 4) which showed a soft tissue mass arising from

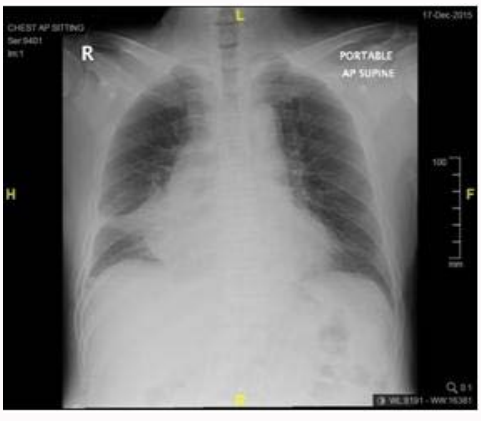

Figure 1: Chest Radiograph on initial presentation.

OPEN ACCESS

Correspondence:
Krishnasamy S, Cardiothoracic Unit, University of Malaya Medical Centre, Kuala Lumpur, Malaysia,
E-mail: sivaprotoss77@yahoo.com

Received Date: *22 Feb 2019*
Accepted Date: *26 Mar 2019*
Published Date: *01 Apr 2019*

Citation:
Krishnasamy S, Nair AK, Hashim SA, Ramesh S, Raja Mokhtar RA. Metastatic Pericardial Liposarcoma - A Successful Multimodality Treatment Approach. Ann Clin Case Rep. 2019; 4: 1632.

ISSN: 2474-1655

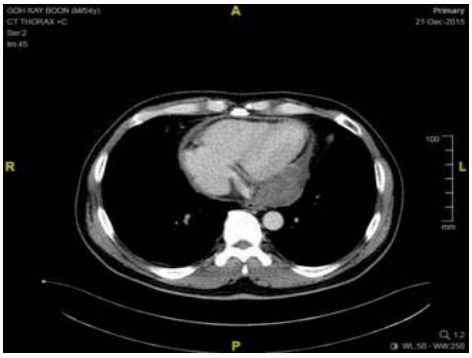

Figure 2: CT Thorax in Dec 2015 showing the left posterior pericardial mass with minimal right pericardial effusion.

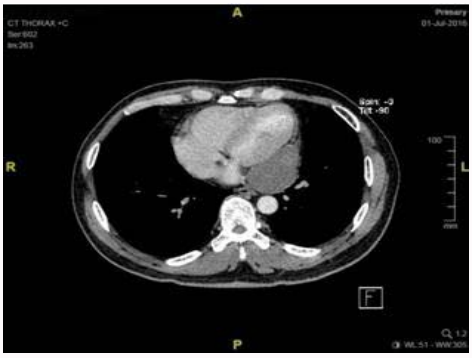

Figure 3: Repeated CT Thorax in Jul 2016 showing an enlarged mass with resolved pericardial effusion.

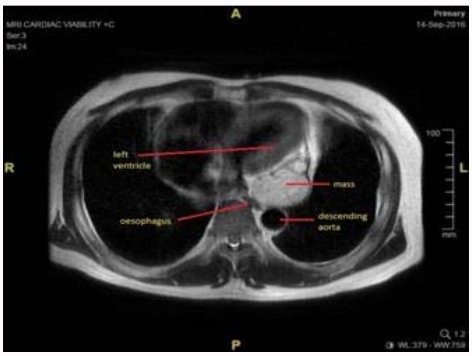

Figure 4: Cardiac MRI demonstrating adherence of the mass to the descending aorta and oesophagus.

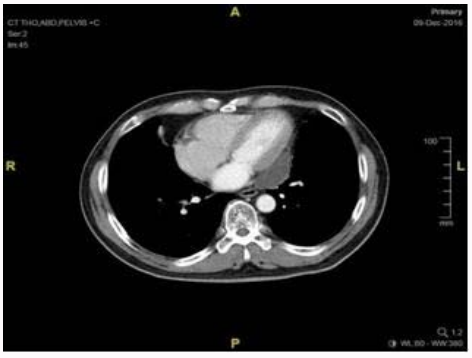

Figure 5: CT Thorax after completion of neoadjuvant radiotherapy showing a reduction in tumour size.

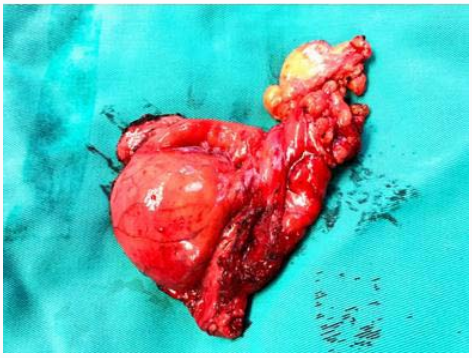

Figure 6: Left pericardial mass which has been resected en bloc.

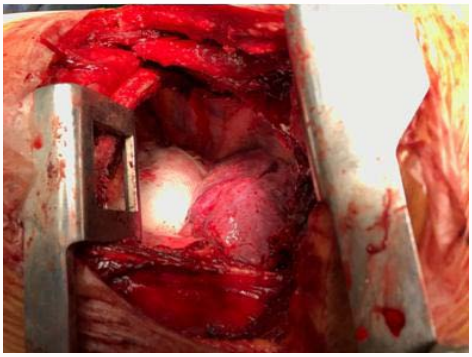

Figure 7: Prolene mesh covering the pericardial defect.

the pericardium adjacent to the left ventricle with involvement of the descending thoracic aorta, thoracic oesophagus and left hemidiaphragm. As the lesion was invading surrounding structures he was then subjected to 25 cycles of neoadjuvant radiotherapy in November 2016 and a repeat CT scan (Figure 5) a month later showed significant reduction in tumour size to 2.1 cm × 5.6 cm × 4.5 cm with a clear fat plane between the mass and adjacent structures plus no evidence of distant metastasis. The non-specific small lung nodules (<5 mm) remain unchanged. In view of this, we planned for a curative resection of his tumour. To further clarify the nature of the lung nodules, we carried out a PET-CT scan which confirmed no FDG-avid lesions elsewhere.

In January 2017 patient underwent a left posterolateral thoracotomy with en bloc resection of the left pericardial mass (Figure 6) and implantation of a prolene mesh over the pericardial defect with plication of the left hemi-diaphragm (Figure 7). The tumour which had a solid and cystic component was densely adhered to the lateral epicardial surface of the heart. A portion of the epicardium of the lateral wall of the heart was taken together as en bloc resection with the mass. An on table frozen section of the epicardial surface adjacent to the tumour showed no evidence of malignancy with just hypertrophied myocytes (Figure 8). Enlarged lymph node samples taken from stations 6, 9, and 10 also showed no evidence of malignancy. The histopathology result of the tumour itself confirmed a myxoid liposarcoma. The patient recovered well from his surgery and was discharged home within a week. He has remained well since. He is currently on a 6-monthly follow up under the Oncologist and his latest repeat CT thorax (Figure 9) in June 2018 which showed no evidence of tumour recurrence.

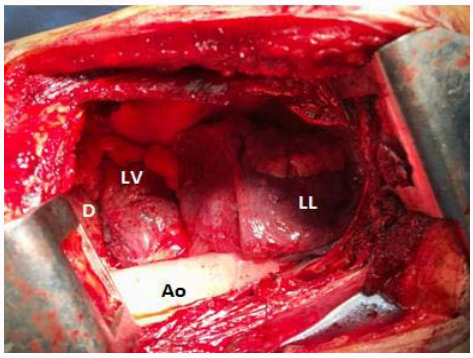

Figure 8: Tumour has been successfully resected away from the lateral surface of the heart.
LL: Left Lung; Ao: Aorta; LV: Left Ventricle Exposed; D: Diaphragm

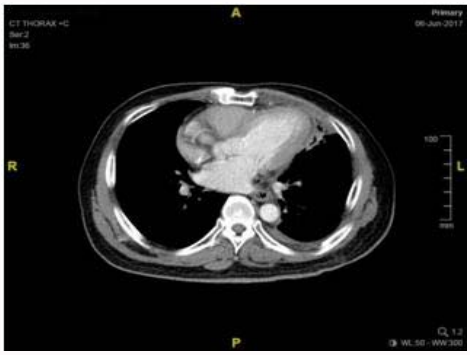

Figure 9: CT Thorax 6 months post tumour resection.

Discussion

Liposarcoma has been classified by WHO into four distinct histological subtypes as follows: well-differentiated, myxoid/round cell, pleomorphic and dedifferentiated [1]. They have a propensity for local recurrence in about 40% of the cases, but very rarely metastasise [1]. Despite metastatic lesions of extracardiac liposarcoma being more common than the development of primary cardiac liposarcoma itself, the incidence of metastasis to the pericardium remains extremely low, with only ten cases that have been reported in the literature up to the year 2015 [2-3]. The time interval between the initial presentation of liposarcoma and the cardiac metastasis can be quite long, ranging from 7 to 25 years [3].

The initial presentation of cardiac metastasis of liposarcoma depends on the part of the heart involved and the size of the tumour. Mostly they remain undiagnosed until patients develop symptoms mainly due to compressive effects on the heart chambers which could manifest as congestive heart failure, arrhythmia, pericarditis, angina, syncope and sudden cardiac death [3-5]. Pericardial invasion may also lead to effusions of various sizes that can result in tamponade and diastolic cardiac impairment, as what we experienced with our patient [2,6]. This could turn out to be a critical and life-threatening situation if not addressed in a timely manner.

The first-line treatment for primary liposarcoma is surgical excision with complete resection of the tumour whenever possible [1-2]. Local recurrence or distant metastasis can occur many years after treatment of the primary tumour, hence the past history of liposarcoma is important here [2,4]. Although there is no standard treatment for cardiac or pericardial metastases, radical surgical resection of the metastatic tumour provides the highest chance of long-lasting survival [2,4,6]. When combined with neoadjuvant radiotherapy, this may significantly reduce the rate of disease progression and recurrence [2,5,6]. Chemotherapy has been shown to be effective in a fraction of cases of metastatic or unresectable soft tissue sarcoma, however the overall response rate was only 46% and it is still lacking in evidence [1,4,5].

In our patient, transthoracic echocardiography initially identified the mass, which was further delineated by CT thorax. However the Magnetic Resonance Imaging (MRI) demonstrated that the mass had invaded the surrounding structures, which was in keeping with the feature of a malignant tumour. This was significant in presurgical planning as it enabled the patient to have neoadjuvant radiotherapy. MRI is a well established imaging modality in assessing any suspected cardiac lesions, plus it can be useful for tissue characterization and differentiation between benign and malignant tumours [7,8]. The integration of all the information provided by various imaging methods enabled us to provide the most appropriate treatment for this patient.

Conclusion

Metastatic pericardial liposarcoma can prove to be a diagnostic challenge and multimodal imaging is highly recommended for optimal preoperative assessment to evaluate the differential diagnosis and deciding on appropriate treatment. Aggressive surgical resection with the intention of resolving any cardiac impairment is the treatment of choice and combination with perioperative radiotherapy may help to prolong survival and avoid recurrence. However, continued surveillance and long-term follow-up with serial imaging is required for all cases of cardiac liposarcomas.

References

1. Steger CM. Primary liposarcoma of the heart. BMJ Case Rep. 2001;2011:bcr0320114013.
2. Yamashita Y, Kurisu K, Kimura S, Ueno Y. Successful resection of a huge metastatic liposarcoma in the pericardium resulting in improvement if diastolic heart failure: a case report. Surg Case Rep. 2015;1:74.
3. Papavdi A, Agapitos E. Undiagnosed primary cardiac liposarcoma in an adult - a case report and review of the literature. Am J Forensic Med Pathol. 2013;34(4):299-301.
4. Aoyama A, Isowa N, Chihara K, Ito T. Pericardial metastasis of myxoid liposarcoma causing cardiac tamponade. Jpn J Thorac Cardiovasc Surg. 2005;53(4):193-5.
5. Pápai Z, Bodoky G, Szántó J, Poller I, Rahóty P, Eckhardt S, et al. The efficacy of a combination of etoposide, ifosfamide, and cisplatin in the treatment of patients with soft tissue sarcoma. Cancer. 2000;89(1):177-80.
6. Markovic ZZ, Mladenovic A, Banovic M, Ivanovic B. Correlation of different imaging modalities in pre-surgical evaluation of pericardial metastasis of liposarcoma. Chin Med J (Engl). 2012;125(20):3752-4.
7. Kim EY, Park KY, Jeon YB, Ha SY, Chung WJ. A primary pericardial liposarcoma mimicking intracardiac neoplasm on echocardiography: role of computed tomography and magnetic resonance imaging in the differential diagnosis. Int J Cardiol. 2013;167(4):e92-4.
8. Tilling L, Hudsmith L, Goldman J, Becher H. Imaging of pericardial tumours: a case report. Cardiovascular Ultrasound. 2006;4:29.