A Case of Primary Myxoid Liposarcoma of the Heart Masquerading as Massive Pericardial Effusion- A Case Report

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Abstract

Tumors of the heart are very rare and are mostly benign. Sarcomas are extremely rare in the heart. Myxoid liposarcomas can arise primarily from the heart and the pericardium or can be metastatic disease from elsewhere. The patients present with pericardial effusion of pressure effects. Echocardiography, cardiac MRI and biopsy usually clinches the diagnosis. Surgery is the only curative option for this tumor and is seldom possible due to the location and advanced nature at presentation. We are reporting this case due to its rarity and to emphasize the need for multi-institutional studies to improve the outcomes.

Keywords: Myxoid liposarcoma; Surgery; Cardiac fluid; Massive Pericardial Effusion

Introduction

Primary tumours of the heart are very rare. Nearly 75\% of them are benign. The most common cardiac tumours are myxomas, papillary fibroelastomas and lipomas. Sarcomas being extremely rare in the heart almost all types have been reported involving the heart. Liposarcomas are the second most common sarcomas involving soft tissues and heart is an unusual location for these tumours\cite{1}. With this background, we are presenting a middle-aged woman who presented with cardiac failure. She was evaluated and was found to have a massive pericardial effusion. On further evaluation she was found to have a myxoid liposarcoma of the heart.

Case presentation

A 52-year-old lady presented with cardiac failure. On evaluation, she was found to have a massive Pericardial effusion, for which she underwent pericardiocentesis and about 500 ml of fluid was drained. Further evaluation for
the cause of the pericardial effusion led to a CT scan of the chest, which showed an intra pericardial mass. A guided biopsy revealed a myxoid liposarcoma. Figure 1 shows PET CT was done on 20/02/20 to rule out metastasis from elsewhere. It showed a mild FDG avid hypodense lesion of size 58 x 50 mm along the lateral wall of the right atrium with complicated lobulated pericardial effusion. It also revealed mediastinal (pre vascular, pre tracheal and aorto caval nodes) and retroperitoneal (retro crural, bilateral common iliac and external iliac lymph nodes) lymphadenopathy. Biopsy of the right external iliac node was suggestive of granulomatous inflammation. She was evaluated for excision of the pericardial mass. Echocardiogram revealed an intra pericardial mass attached to the right atrium with no intra-cardiac extension and a minimal pericardial collection.

Figure 1: CT axial images showing the tumor with pericardial infusion and CT guided biopsy

She underwent excision of the tumor on 24/03/20. Intraoperatively, the pericardium was found adherent to the epicardium by gelatinous material. A pearly white mass of size 6 x 6 cm was found attached to the right atrium compressing it. A thin plane was found between the mass and the atrium. It was removed in toto with a rim of right atrial attachment. Pre tracheal node biopsy was taken. Histopathological examination revealed a tumor with atypical spindle cells and numerous lipoblasts in a myxoid stroma consistent with Myxoid liposarcoma. No malignant round cells were noted. The right atrial margin was involved by tumor. The tracheal nodes showed lymphoid hyperplasia as shown in Figure 2.
Figure 2: H&E: A-100X & B-200X- Numerous multivacuolated lipoblasts and spindle cells in a myxoid stroma. C-100X & D-200X- Spindle cell predominant areas

The case was discussed in the multi-disciplinary team, and she was taken up for adjuvant chemotherapy in view of the size of the tumor. She received 6 cycles of Ifosfamide and Epirubicin from April 2020 to August 2020. She presented in July 2021 with disseminated disease with a disease-free interval of 10 months and died soon after.

Discussion

Primary cardiac tumors constitute less than 1% of all tumors(2), of which a vast majority are benign. Among the malignant tumors, primary cardiac sarcomas constitute 10-20% of the cardiac tumours. They resemble their soft tissue counterparts. They are thought to arise from pluripotent mesenchymal cells. Undifferentiated pleomorphic sarcomas and angiosarcomas are the most common sarcomas that have been reported in the heart [2].

Myxoid liposarcoma (LPS) accounts for 30% of all the Liposarcoma cases. They usually present in the limbs with a median age of presentation around 35 - 55 years of age [3]. Only a handful of cases of Liposarcoma involving the heart have been reported. They can be either primary or secondary and the latter is more common. Secondary involvement can be either a metastatic disease from elsewhere or primary mediastinal tumors with pericardial infiltration [4]. Primary cardiac myxoid liposarcomas are extremely rare and have been reported involving cardiac chambers, great vessels and pericardium. They often reach a considerable size before causing symptoms. Pericardial LPS present with cardiac tamponade or with cardiac dysfunction due to the pressure caused on the cardiac chambers.
Multimodal imaging is mandatory for preoperative assessment of cardiac tumors, thereby facilitating appropriate management. Echocardiography is the most vital investigation for assessment. It is often able to give the site, size and cardiac mobility details. Trans oesophageal echocardiography (TOE) can further throw light on the attachment of the tumor. It thereby can differentiate an invading sarcoma and distinguish multiple masses. Contrast echocardiography can distinguish tumors based on their vascularity. In addition, CT scan and cardiac MRI have been shown to be useful. Cardiac MRI (CMR) has been found to be superior to trans thoracic and trans oesophageal echocardiography in finding out the location, attachment to adjacent structures and differentiate masses of various aetiology. Additionally, any associated hemodynamic effect is better known with a CMR.

On microscopic examination they resemble their soft tissue counterparts. They show atypical spindle cells, stellate shaped cells and lipoblasts in a myxoid background. Curvilinear blood vessels are typically present. Round cell component may or may not be present. However the percentage of round cells determines the biology of the disease.

The genetics of primary cardiac sarcomas are poorly understood. They are a result of some specific gene alterations with simple karyotypes (rhabdomyosarcomas and Ewing, synovial, and gastrointestinal stromal tumours) or nonspecific gene alterations with complex karyotypes. Pleomorphic sarcomas and leiomyosarcomas are probably the result of nonspecific translocation.

Very little data is available regarding the management of cardiac sarcomas. Surgery has been the only curative option for cardiac liposarcomas. However, complete surgery has often been difficult due to the critical location of the tumors and often due to local infiltration. Role of adjuvant therapy has been controversial. Reports of post-operative Radiotherapy and chemotherapy giving benefit in the form of better local control are available, albeit not significant. This is probably partly due to the rarity of the disease and the absence of effective methods to address the local area without causing major morbidity.

Prognosis of this tumor depends on various factors such as size, location, histomorphology with the presence of round cells and the presence of metastases. Most of the reported cases show that the clinical outcome is poor especially in tumors that could not be resected completely.

**Conclusion**

Myxoid liposarcomas are very rare neoplasms of the heart. Surgery is the mainstay of treatment. However, due to the location, complete resection is seldom possible. Post-operative radiotherapy has not been shown to improve survival. Chemotherapy has also been tried. The patients tend to recur locally and in distant areas due to these factors. They have a very short disease-free survival just like our patient. Further larger studies would help us in addressing the therapeutic strategies better, thereby improving the prognosis of these tumors.
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References