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A Successful Resection of Cardiac Metastasis of Round Cell Liposarcoma

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ABSTRACT

Myxoid is a common type of liposarcoma. Cardiac metastasis of sarcoma is a rare diagnosis. Transthoracic echocardiography is a useful tool for beginning the diagnosis. Curative resection of metastasis and then combination systemic chemotherapy for advanced disease is recommended. We report a 50 year old woman who presented with a cardiac metastasis of high grade round cell liposarcoma to right atrium and right ventricle. The tumor was diagnosed with computed tomography and transthoracic echocardiography. The tumor size was 10*10 cm. This is the second report of successful resection of a large intracardiac metastasis of liposarcoma.

Introduction

Liposarcoma is a tumor of adults. The most common sites are thigh and retroperitoneum (1). Round cell type is a poorly differentiated subtype of liposarcoma (2). cardiac metastasis of sarcoma is a rare diagnosis, showing 0.6% in autopsy series of patients with malignant tumors (3).

There is no standard treatment for cardiac metastasis. In selected patients , whose tumor appears to be stable, resection of cardiac metastasis, if technically feasible, offers the best chance for prolonged survival , although the perioperatively mortality rate remains high (40%) (4, 5).

Case presentation:

A 50 year old woman with a history of primary liposarcoma of left knee was referred to our institution because of severe dyspnea. Detailed study revealed that the primary tumor had been a high grade round cell liposarcoma originating from left knee diagnosed 4 years ago. The therapy had consisted tumor resection, chemotherapy and radiotherapy. Two years later, patient was readmitted with chest wall metastasis and underwent partial sternumectomy and insertion of intravenous catheter in right subclavian vein for chemotherapy.

At her recent presentation, symptoms which were accelerated in the past 2 months were: resting dyspnea, tachypnea and dry coughs. Positive findings in physical exam consisted: Jugular vein distention and a diastolic rumble on the left lower sternal border exaggerating with respiration. She had no fever.

Electrocardiogram showed sinus tachycardia rhythm and right axis deviation. In Chest x-ray, cardiothoracic ratio was normal with a prominent right pulmonary trunk. Laboratory tests revealed mild leukocytosis with 92% neutrophilia. Primary differential diagnosis include: infected venous catheter, pulmonary thromboembolie, cardiac or pulmonary metastasis.

Patient underwent further paraclinical evaluations: chest computed tomography revealed a high density mass located on the central venous catheter with low density extension to right atrium (RA) and right ventricle (RV) (Fig 1).

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Figure 1. Thoracic chest computed tomography revealed a low density mass extended to right atrium (RA) and right ventricle (RV) with bilateral pleural effusion.

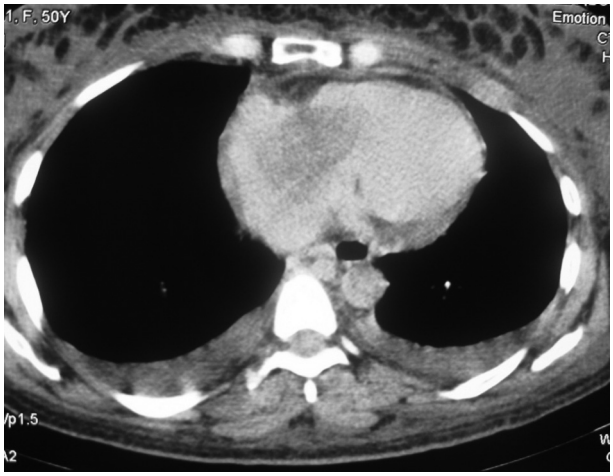
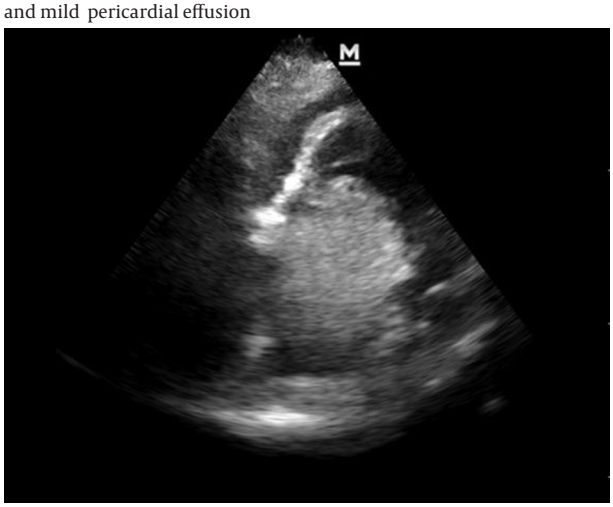


Figure 2. Transthoracic echocardiography in modified four – chamber view shows a large mass in right atrium with functional tricuspid stenosis and mild pericardial effusion



Two dimensional transthoracic echocardiography (TTE) revealed a large lobulated hyperecho mass with diameters of (4.2 * 4.5 cm) in RA. The mass was hypermobile and protruded to RV with every diastole, leading to functional tricuspid stenosis (TS). (Fig 2, 3)

At this point the most probable diagnosis was metastasis of relapsed liposarcoma of knee with superimposed thrombosis and/or vegetation. The patient was undergone a cardiac surgery as a diagnosis and therapeutic option.

After median sternotomy, aorta artery (AO) was cannulated, superior vena cava (SVC) was cannulated directly, and inferior vena cava was cannulated classically. Pump was activated and AO clamped. Cardioplegia solution was used. RA was exposed and tumor was extracted. It was a large tumor (10*10cm) which was expanded from SVC to RA, tricuspid valve (T.V), and to RV.

Figure 3. Transthoracic echocardioghraphy shows a large lobulated mass in right atrium

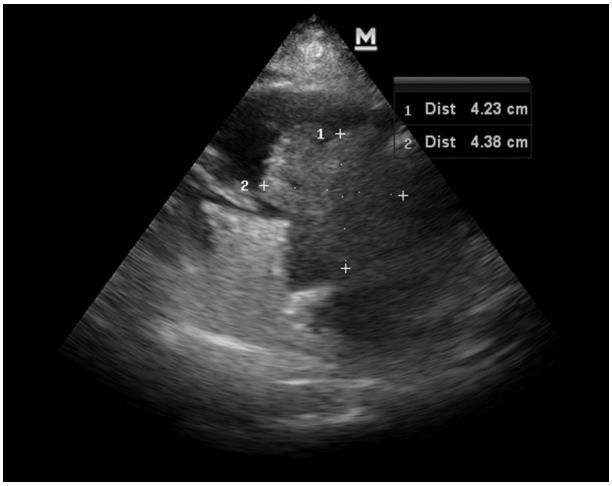
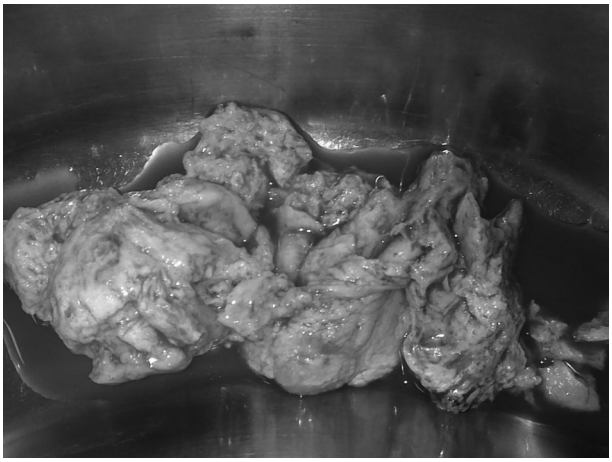
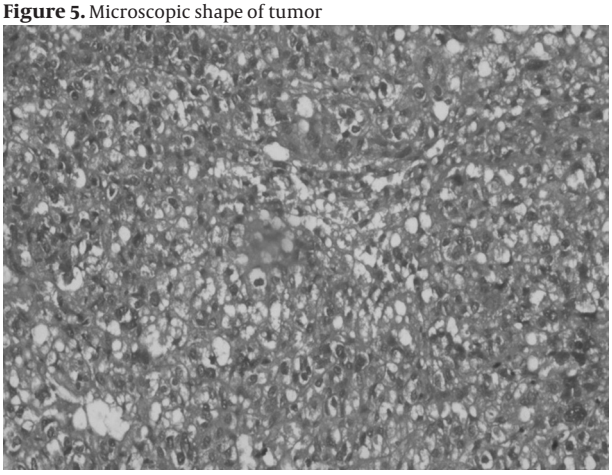


Figure 4. Macroscopic shape of tumor (14*10*7 cm cardiac tumor)



RA & RV tumor were extracted. Then, cannula tape of SVC was opened and tumor residual was extracted from SVC totally. No tumor` adhesion was seen on tricuspid valve. Post operative TTE showed no residual mass, only mild degree of tricuspid regurgitation (TR) was seen.

Figure 5. Microscopic shape of tumor



Macroscopic pathology study reported:an irregular lobulated gray creamy mass measuring 14*10*7 cm (Fig.4). Area of hemorrhage and extensive area of necrosis were seen. Microscopic features included: proliferation of lipoblasts with moderate atypia with mitotic figure, hemorrhage and massive necrosis were seen. Final diagnosis was metastatic liposarcoma (fig 5).

Patient was discharged from hospital after 3 weeks of hospitalization with adjuvant radiotherapy regimen. At the time of submitting the manuscript, patient has experienced seven months relapsed free survival confirmed by follow up visits.

Discussion:

The survival in liposarcoma lesions containing round cell component is 50% within 5 years(1). The clinical presentation is almost due to the development of a mass which is usually large and painless (1). Lungs are the most common site of metastasis in advanced stages of liposarcoma.

Three techniques are considered useful for diagnosing intracardiac tumors. TTE is a useful tool for beginning the diagnosis. CT & MRI are the best ways to show intracardiac invasion (6). The definitive confirmation of the disease is by biopsy (1). Surgery remains the principle therapeutic modality. The optimum combination of radiotherapy and chemotherapy has remained controversial (1).

Our case is one of the rare cases of right heart intracavity metastasis of liposarcoma and the second reported case of successful resection of a more than 10 cm size intracardiac metastasis tumor (7), with promising result of 6 months event free survival.

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