

Subtotal Resection for the Symptom Relief of a Fatal and Aggressive Primary Cardiac Liposarcoma: A Case Report

Demiao Kong, MD,^{1,2} Chengming Fan, MD¹

¹Department of Cardiovascular Surgery, the Second Xiangya Hospital, Central South University, Middle Renmin Road 139, 410011 Changsha, China; ²Department of Thoracic Surgery, Guizhou Provincial People's Hospital, Guiyang, Guizhou 550002, China

ABSTRACT

Primary cardiac liposarcoma is a rare malignant soft tissue lesion, but there are still no diagnosis and treatment guidelines for this disease. This is the case report of a 59-year-old male with cardiac liposarcoma infiltrating the mitral valve and the left atrium. He achieved satisfactory symptom relief with subtotal resection.

INTRODUCTION

Primary malignant cardiac tumors are extremely rare neoplasms that originate from cardiac structures and display biologically aggressive behavior. It only has a prevalence of 0.001 to 0.28% [Hudzik 2015], which is found in only 1% of primary malignant cardiac tumors [Butany 2005; Neragi-Miandoab 2007]. Cardiac liposarcoma usually manifests as a large, infiltrating mass and can involve in any part of the heart, including the chambers, great vessels, and pericardium [Sandoval 2012]. Here, we present a case of primary cardiac liposarcoma with the feature of diffuse growth in the left atrium and mitral valve involvement.

CASE REPORT

A 59-year-old man presented with a four-month history of chest pain, swelling in the legs, and a recent weight loss of 20 kg. Laboratory tests and physical examination were unremarkable. Electrocardiography revealed sinus rhythm and incomplete right bundle branch block. Transthoracic echocardiography demonstrated normal ventricular function, left atrial dilation, and a heterogeneous mass in the left atrium, involving the atrial septum and posterior wall of the left atrium with the size of 4.5×2.8cm and 3.5×1.2cm, respectively. The margins of the two masses were connected with each other. And, the two abnormal structures were immobile for the whole period of the cardiac cycle (Figure 1A). Cardiac computed tomography further confirmed these detections, in addition to

posterior leaflet of the mitral valve involved (Figure 1B). No other metastatic mass was detected. The preoperative coronary angiogram was normal. The patient was evaluated by the cardiovascular surgery service for tumor excision.

A standard median sternotomy incision was performed. The aorta was cannulated. Separate cannulas were placed in both the superior vena cava and inferior vena cava. After full heparinization, cardiopulmonary bypass (CPB) was routinely applied, the aorta was cross-clamped, and cold cardioplegic solution was instilled via the aortic root to arrest the heart. After the opening of the left atrium by a bi-atrial and transeptal incision, the whole atrium was found invaded and infiltrated by the tumor. The entire posterior and part of the anterior mitral leaflet as well as the orifices of bilateral pulmonary veins also were infiltrated. Based on the intra-operation findings and further discussion with the patient's family, a cardiac sarcoma was considered and complete resection could not be achieved. After complete resection of the leaflets and chordae tendineae of the mitral valve, partial resection of the tumor was performed (Figure 2). Part of the infiltrated left atrium had been removed for the subtotal resection of the tumor. Mechanical heart valve (size 27) replacement was executed in the end. The patient tolerated the procedure well, and the heart was then beating in sinus rhythm. After careful hemostasis and closing the wound in layers, the patient carefully was transferred to the intensive care unit (ICU) in stable condition.

Pathology confirmed the diagnosis of liposarcoma and showed that the cells were positive expression for SMA,

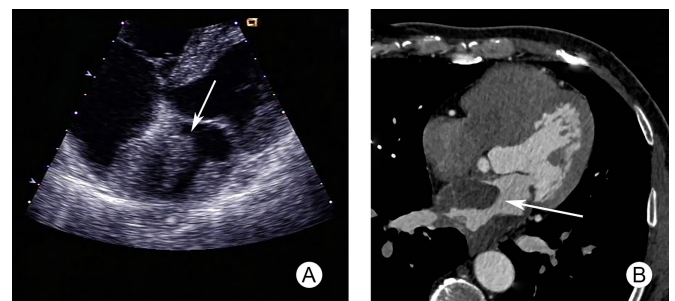


Figure 1. Preoperative echocardiogram. A, showed a heterogeneous mass located in the left atrium (arrow), atrial septum, and the posterior wall of left atrium with the size of 4.5×2.8cm and 3.5×1.2cm, respectively. Computed tomography. B, in four chamber view showed a tumor-like mass located in the left atrium (arrow) with posterior leaflet.

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Correspondence: Chengming Fan; +86-731-85296107; fax: +86-731-85296606 (e-mail: fanchengming@csu.edu.cn).

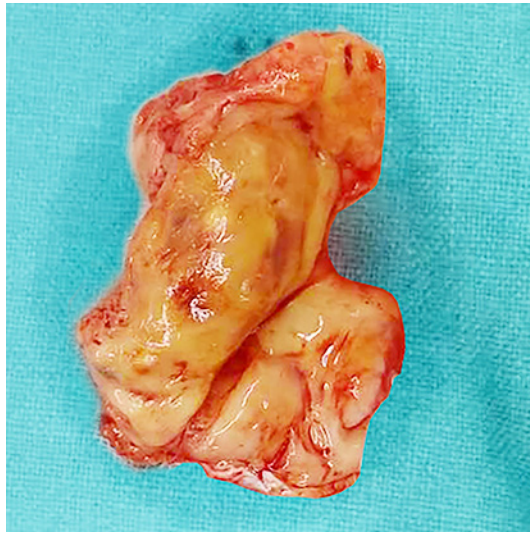


Figure 2. Intraoperative view of the tumor after the subtotal removal.

Vim, and CD117 and partial positive for Ki-67, CD34, S100, Dog-1, Des and D240, but negative for CK and EMA (Figure 3).

The patient recovered without complications and discharged on postoperative day 7 with recommendation for follow up. He died at one year after surgery because of recurrence.

DISCUSSION

Liposarcoma is the second most common soft tissue sarcoma in adults, occurring predominantly in the lower limbs and retroperitoneum, while its occurrence in the heart primarily or metastatically is extremely rare [Neragi-Miandoab 2007; Lococo 2011; Wang 2010; O'Neill 2020]. Cardiac malignant tumor may lead to severe symptoms, including fatal arrhythmia, heart failure, or sudden death [Gazit 2007; Araoz 1999]. Liposarcomas often are asymptomatic and may reach a considerable size before causing any symptoms, due to direct invasion or compression of other thoracic organs [Lococo 2011; Wang 2010]. Therefore, clinical diagnosis is difficult.

Noninvasive imaging plays a critical role in the diagnosis and surgical planning of cardiac masses [Lestuzzi 2015]. Echocardiography remains the first choice for cardiac mass evaluation; however, it is difficult to assess precisely, especially for the margin of an invasive lesion because of the relatively narrow window of the echocardiographic examination. Cardiac magnetic resonance is an alternative imaging modality for cardiac masses, but the main disadvantage is susceptibility to motion artifact [Fan 2015]. With the development of multidetector computed tomography (MDCT) scanners and retrospective ECG gating, cardiac CT is a practical approach for cardiac masses assessment [Samuel 2018].

Despite the significant advances in diagnostic techniques, the imaging characteristics of primary cardiac

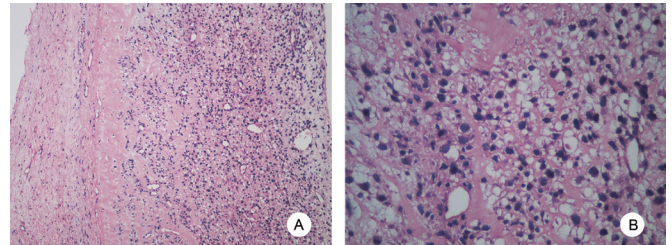


Figure 3. Primary cardiac liposarcoma diagnosed using immunohistochemistry.

liposarcomas were inconclusive. This malignancy usually manifests as a large, infiltrating mass and tends to arise in one of the atria; however, it also has been discovered in both ventricles and in the pericardium [Lococo 2011]. Pericardial involvement may take the form of effusion, tumor nodules, or irregular masses. There have been several reports of pericardium involvement as effusion, tumor nodules or an irregular mass primarily arising in the pericardium [Wang 2010; O'Neill 2020]. In this case, the cardiac liposarcoma is characterized by an invasive and infiltrating growth rather than exophytic growth, which forms a huge mass. Furthermore, the entire posterior and part of the anterior mitral leaflet as well as the orifices of bilateral pulmonary veins were rarely invaded.

Surgical treatment is the mainstay of therapy for primary cardiac tumors [Samuel 2018], yet due to the rarity of these malignancies, their management remains a challenge [Shen 2020]. Very few data are available for the most appropriate treatment of cardiac sarcomas. Evidence shows that the outcome is generally poor, especially in aggressive tumors that are characterized by infiltrative growth and in-situ recurrence tendency [Ramlawi 2016]. Total surgical resection rarely is of benefit for malignant cardiac tumors [Wu 2012]. Furthermore, complete surgical resection usually was considered to be impossible because of the extent of invasion of the tumor into surrounding tissues. Subtotal resection of the tumor often was performed for the diagnosis and palliation of symptoms under these circumstances. The role of chemotherapy or radiation therapy is controversial, and radical resection of the tumor seems to be the best treatment option [Xiao 2020]. Thus, alternative strategies, such as radical tumor resection along with the following techniques (heart transplant, total artificial heart, and auto-transplant), can be considered even though tumor recurrence cannot totally be avoided [Xiao 2020; Kremer 2016; Reardon 2006]. In the present case, the whole left atrium and some crucial adjacent tissue were infiltrated by the liposarcoma. Subtotal resection was performed. To avoid transplanted valve infiltration, a mechanical valve was chosen instead of a tissue valve. The patient's symptom significantly was relieved after surgery, but he had a local recurrence and died at one year after the surgery. In conclusion, we present a very rare case of primary cardiac liposarcoma located in the left atrium and mitral valve. Subtotal resection of tumor is a satisfactory method for symptom relief.

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