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Primary Cardiac Sarcoma Remission after Surgery: 5-Years Follow-up: Case Report

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ABSTRACT

Background: Primary cardiac sarcoma is a rare and atypical clinical entity. We present a patient with long-term remission after primary cardiac sarcoma resection.

Case Report: A 42-year-old previously healthy female presented to the emergency department after an effort-induced 30-minute episode of chest pain and extreme shortness of breath. Physical examination upon admission was remarkable for a pulse of 99/minute; blood pressure was 101/73 mmHg. Transthoracic echocardiography showed a mass measuring 5.5 × 5.6 cm extending from the left septum to the mitral valve anterior leaflet. A multilobulated broad-based 5.5 × 5.6 × 4 cm^3 mass invading a large portion of the left septum to the mitral valve anterior leaflet was completely excised in the open heart surgery. Chemotherapy regimen (paclitaxel 175 mg/m^2/day on day 1, every 21 days) was started after operation. Full remission was provided. Metastasis and recurrence have not been observed for 5 years of follow-up by PET. We observed during 5 years and used a PET. And the finally we did not see metastasis at the 5 years of follow-up.

Conclusions: We strongly recommend that a patient-specific multidisciplinary approach involving radical resection, chemotherapy, and radiation therapy in these cases results in patient survival and a significant improvement in quality of life. We also think that it is necessary to perform MRI to exclude other illnesses that are considered to be a myxoma.

INTRODUCTION

Primary cardiac sarcoma is a rare and atypical clinical manifestation. It is usually detected in an advanced stage [Ramlawi 2014]. Treatment for cardiac sarcoma is radiotherapy and/or chemotherapy after surgical resection [Randhawa 2016]. Cardiac sarcomas are rapidly fatal. Because sarcomas are characterized by a brief clinical course, aggressive behavior, and a delays that delay diagnosis [Zhang 2008]. The median survival of patients treated conservatively ranges from 9 to 12 months [Ramlawi 2014]. We present a patient with long-term remission after primary cardiac sarcoma resection.

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CASE PRESENTATION

A 42-year-old previously healthy female presented to the emergency department after an effort-induced 30-minute episode of chest pain and extreme shortness of breath. Physical examination upon admission was remarkable for a pulse of 99 beats per minute, blood pressure of 101/73 mmHg, temperature at 36.6ºC, room-air oxygen saturation at 100%, and intense pallor. Laboratory workup was normal. The patient did not have hypertension and arrhythmia. There was no murmur or additional sound on the cardiac osculation. Transthoracic echocardiography showed a mass measuring 5.5 × 5.6 cm extending from the left septum to the mitral valve anterior leaflet prolapsing through the mitral valve during diastole, resulting in severe mitral stenosis. The patient was diagnosed as with left atrial myxoma on the basis of echocardiographic findings, and no further tests were needed. Because the patient was thought to have a myxoma, neither CT nor MRI was performed before the operation.

Following median sternotomy, 100-300 U/kg heparin was administered, and cardiopulmonary bypass was performed after aorto-bicaval cannulation. Left atriotomy was performed, and we completely excised a multilobulated broad-based 5.5 × 5.6 × 4 cm3 mass invading a large portion of left septum to the mitral valve anterior leaflet (Figure 1). After mass excision, the left atrium was closed. The final pathology result was a high-grade unspecified pleomorphic sarcoma. The postoperative course was unremarkable, and the patient was discharged

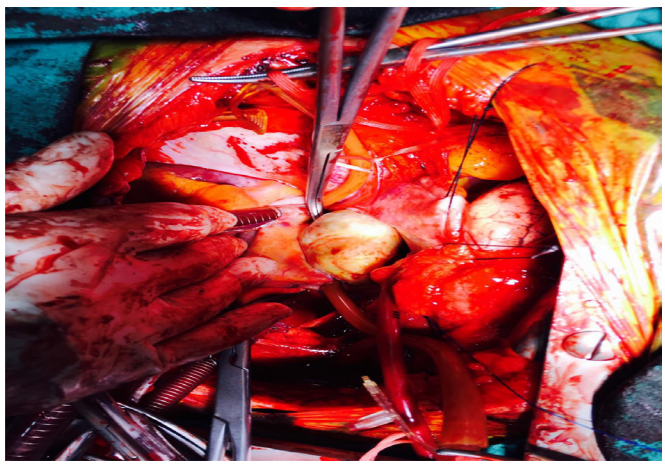

Figure 1. Primary cardiac sarcoma during operation.

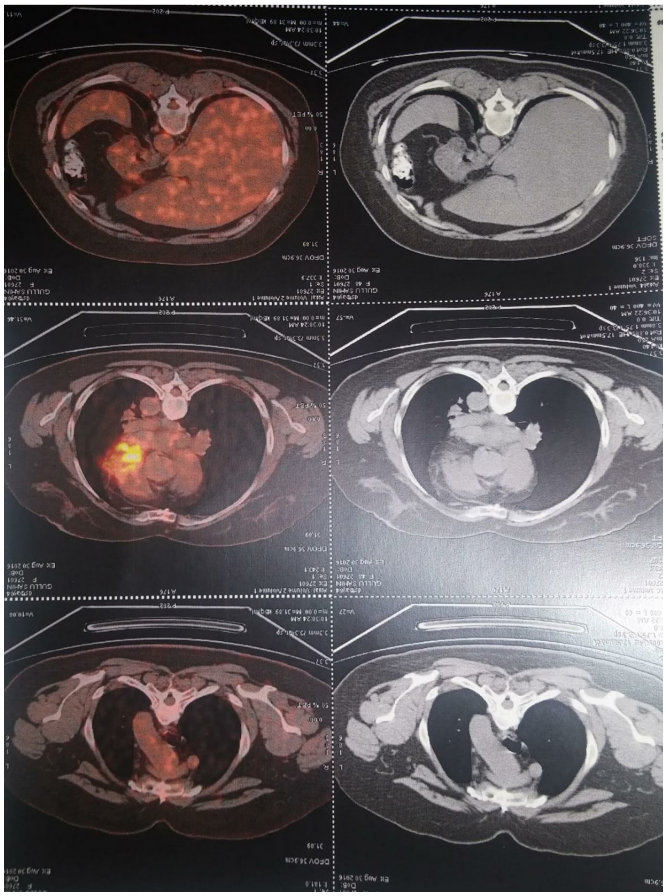

Figure 2. PET image 5 years after operation.

to home on the 10th postoperative day. Chemotherapy regimen (paclitaxel 175 mg/m^2 per day on day 1, every 21 days) was started. After 6 cycles of paclitaxel chemotherapy, clinical findings and imaging findings showed no progression. We observed during 5 years and used a PET (Figure2), and the finally we did not see metastasis at the 5 years of follow-up.

DISCUSSION

Primary cardiac sarcoma is a rare and atypical clinical manifestations [Ramlawi 2014]. Cardiac sarcomas are rapidly fatal. Because sarcomas are characterized by a brief clinical course, aggressive behavior, and a delays that is a delayed diagnosis [Zhang 2008].

Cardiac sarcomas have a poor prognosis with a course of 11 to 17 months. Because the tumor multiplies very rapidly and makes distal metastases [Hamidi 2010]. Hamidi et al showed that non–operated-on patients died within 1 month whereas operated-on patients lived 12 months [Hamidi 2010]. In another similar study Radhawa and colleagues reported operated-on patients lived 14 months; however, non–operated-on patients died in 9 to 12 months [Blackman 2012]. Our patient operated on lived for 5 years.

The management of primary cardiac sarcoma is challenging. The challenges include establishing an accurate diagnosis in a timely fashion, designing a patient-specific treatment plan, achieving complete resection often requiring extensive surgery, and integrating neoadjuvant and adjuvant therapy [Shanmugam 2006].

It remains unclear whether radical surgical resection is an alternative in patients without metastatic disease because heart sarcoma is rare, there are only a limited number of studies on the topic, and no standard treatment is defined. Although aggressive resection of the heart sarcoma is difficult, some studies clearly showed that complete tumor removal is the best method to improve survival [Bakaeen 2009].

Although it appears that most angiosarcomas are resistant to chemotherapy and radiation, aggressive treatment does seem to contribute to reducing tumor size. Chemotherapy and radiation are important in treatment of angiosarcomas because of the high probability of metastases [Oh 2013]. Cisplatin, cyclophosphamide, dacarbazine, doxorubicin, ifosfamide, mitomycin C, paclitaxel, and vincristine are commonly prescribed agents in standard chemotherapy [Simpson 2008]. We used paclitaxel (175 mg/m2/day on day 1, every 21 days) as a chemotherapy regimen. Immunotherapy has also been used, with one report showing a 30-month survival following a combination of surgery, chemotherapy, and recombinant IL-2 therapy [Swetal 2014]. We performed surgery in our case, and then we used chemotherapy.

CONCLUSIONS

Because primary cardiac sarcoma is a rare disease, data regarding the optimal treatment protocol are inconclusive. However, data derived from several single-center studies and isolated case reports, including ours, strongly suggest that a patient-tailored multidisciplinary approach, comprising radical resection, chemotherapy, and radiation therapy provided in specialized centers, results in a significant improvement in patient survival and quality of life.

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