

Case Report



Complete resolve of primary cardiac sarcoma by chemotherapy: A case report

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Abstract

Primary cardiac tumors (PCTs) are rare tumors with high mortality rate and low survival. There are several treatment approaches in the management of PCT. Complete surgical resection is the main base of treatment, while adjuvant chemotherapy or radiotherapy after surgical resection are points of disagreement. In this case report, complete remission of the relapsed non-metastatic cardiac synovial sarcoma was observed after salvage chemotherapy and complete surgical resection.

Introduction

Primary cardiac tumors (PCTs) are rare, with an incidence between 0.0017 to 0.019%.¹⁻⁶ Nearly 25% of these tumors are malignant.²⁻⁴ PCTs are more common in the right atrium than left.⁷ PCTs are often diagnosed during evaluations of nonspecific symptoms, such as dyspnea, syncope or tachycardia.⁴ Because of atrium obstruction, the symptoms are the same as seen in heart failure. Right atrium involvement includes increased jugular vein pressure, lower extremities edema, and ascites, while left atrium involvement symptoms are dyspnea or shock state. PCTs are diagnosed based on echocardiography, Computed tomography (CT) scan, magnetic resonance imaging (MRI), and pathology. These tumors show an aggressive manner with high recurrence in their management.⁷ Expected average survival for PCTs is about 9 to 11 months.¹⁻⁴ The most common site for metastases is the brain.⁸ The main cornerstone of primary cardiac sarcoma treatment is still complete surgical resection of diagnosed mass based on echocardiography and CT or MRI.⁹⁻¹² 9 to 12 months of survival has been seen in patients who were treated only by medical therapy.¹³ In this paper, we describe a case of intracardiac sarcoma which was successfully treated by high dose ifosfamide as salvage chemotherapy.

Case Presentation

Our patient is a 53 years old man with no past medical history who referred with occasional dyspnea and right

upper quadrant pain in the last two years. The patient was visited by a cardiologist because of progressive exertional dyspnea and fatigue, and an intracardiac mass in the right atrium was diagnosed by echocardiography. After coronary angiography, he was referred to a cardiac surgeon and mass resection was performed. Pathology reported benign tumor. Two months after mass resection, exertional dyspnea FC III/IV started again. In re-evaluation echocardiography, tumor regrowth was observed that was bigger than before in size. After MRI, because of non-metastatic regrowth, the patient undergoes surgery again, and this time, synovial sarcoma was reported (Figure 1).

Chemotherapy with gemcitabine and docetaxel was started, but because of unresponsiveness and relapse, adjuvant chemotherapy with high-dose ifosfamide (3 g/m² IV, day 1-3, q 3 weeks) was also started. In follow up echocardiography, no tumor was observed, and despite that 14 months have been passed from the second surgery, our patient was completely cured. In this case, this is important that complete remission and perfect cure of intracardiac synovial sarcoma was obtained by salvage high-dose ifosfamide after a course of gemcitabine and docetaxel.

Discussion

PCTs prognosis is generally unpleasant.^{3,12} The most common symptoms are dyspnea, and respectively, chest pain, congestive heart failure, palpitation, fever, and anemia.³ Therapeutic difficulty and high recurrence such

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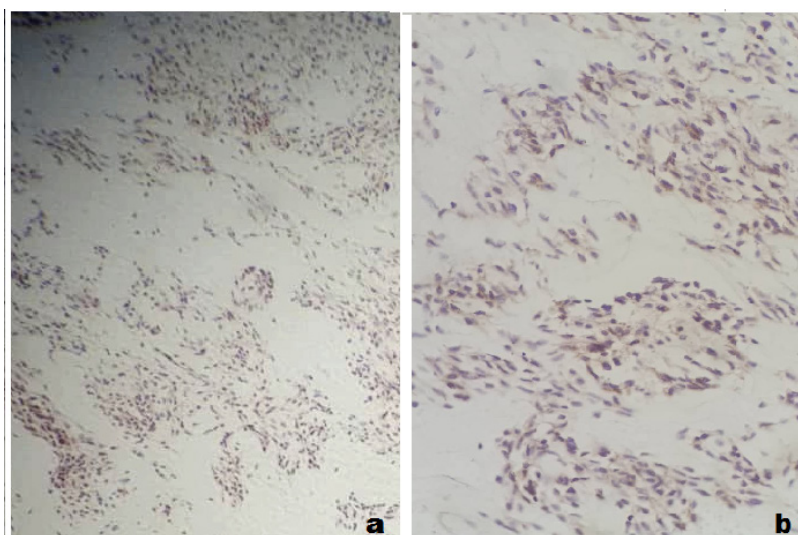


Figure 1. Pathologic image of intracardiac mass in the biopsy, showing synovial sarcoma.

as metastases causes prognosis would poor.¹⁰ Factors in favor of better outcome are left atrial position, low mitotic proliferation count, no necrosis, and no metastases at the time of diagnosis.¹² Due to the lack of comprehensive studies in PCTs management, the beneficial effects of adjuvant chemotherapy are not yet known.^{2,3,14} In some studies, and case reports, adjuvant chemotherapy is recommended after surgery in order to improve survival.^{5,10,14} Vaitiekiene et al reported a case of primary cardiac sarcoma with 26 months of survival after complete surgery and chemotherapy.⁵ Some studies emphasized on individual approach to patients in multidisciplinary teams, even in metastatic tumors, to improve the outcome.^{5,8,15} Successful treatment and improved survival are observed in patients with multimodality approach (surgery, chemotherapy, and radiotherapy) more than single treatment (surgery, chemotherapy, or radiotherapy only).¹⁶ Akishima reported a case of primary cardiac sarcoma with two years of survival after surgery, proton beam radiotherapy and molecularly targeted drugs chemotherapy.¹⁷ In soft tissue sarcomas, adjuvant chemotherapy with or without radiotherapy is recommended to decrease local or metastatic recurrence of the tumor.^{15,18} In our case, relapsed synovial sarcoma completely revealed by salvage chemotherapy by high-dose ifosfamide (3 g/m² IV, day 1-3, q 3 weeks) after unresponsiveness with gemcitabine and docetaxel, and so far, after 14 months, the patient does not have any problems. The limitation of this study was the lack of treatment other than adjuvant chemotherapy in the relapsed case, which successfully responded to high dose ifosfamide.

Conclusion

There is no doubt that surgical resection is the main cornerstone in the management of sarcoma. However, despite disagreements about adjuvant chemotherapy or radiotherapy after surgery in approach to synovial

sarcoma, it seems that the current approach has satisfactory results in sarcoma survival prolongation. It is hoped that in the near future, according to extensive studies and case reports, adjuvant chemotherapy in combination with surgery, enters the main protocol treatment of sarcomas in order to improve survival.

Conflict of Interest

All authors have no conflicts of interest to declare.

Ethical Approval

This study was approved by the Ethics Committee of the Tabriz University of Medical Sciences, Tabriz, Iran. Additionally, informed consent was obtained from the patient.

Authors' Contribution

NG: Investigation, project administration, supervision, validation, visualization. ST: Investigation, writing – original draft, writing – review & editing. All authors read and approved the final manuscript.

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