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An Enormous Angiosarcoma in the Right Atrium of the Heart with an Impressive Outcome

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Abstract

Angiosarcomas of the heart are extremely rare tumors and tend to be located in the right atrium. Echocardiogram, computed tomography, and magnetic resonance imaging are imaging techniques to detect and consider precisely. Surgery is the cornerstone of the therapy modality, but it can be supplied by chemotherapy and radiotherapy. Survival and outcomes of heart sarcomas are unfortunately poor. Our study was about angiosarcoma of the heart. We have managed both surgery and oncologic procedures well. Thus, the patient is still alive 21 months after the surgery. Our aim is to share our successful experiments and contribute to the literature.

Keywords: Coronary artery disease, doxorubicin, heart failure, heart neoplasms, hemangiosarcoma, neoplasm, residual

Introduction

Primary sarcomas of the heart are defined as malignant neoplasms that originate from mesenchymal cells[1]. Primary sarcomas of the heart are extremely rare cases with a rate of incidence is less than 0.1[1]. However, metastatic cardiac tumors are more common than primer ones[1]. The most frequent of them are cutaneous melanoma and breast and lung sarcomas[1]. Echocardiography is the most useful tool in order to investigate cardiac tumors, especially small ones and tumors that have relation with valve structures[2]. It is also the most common imaging technique[1]. Nevertheless, magnetic resonance imaging (MRI) is the most delicate technique to notice the extension of a tumor[2].

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Computed tomography (CT) may detect metastases as easily as MRI does[1]. Coronary angiography can provide information about coronary involvement of mass[1]. It can be more appropriate for elderly patients with a high risk of coronary artery disease[1,2]. The combination of these tools provides detailed information to physicians[1]. Symptoms depend on the location of the tumor is placed[1]. Treatment options for sarcomas are surgery, chemotherapy, and radiotherapy[1]. Our case was a huge angiosarcoma, which is the most frequent primary cardiac sarcoma[1,2]. This case report contributes to the literature via a compelling case that ends with a huge angiosarcoma.

Case Presentation

A 57-year-old female patient was admitted to the cardiology department of our institute with chest pain. Laboratory tests were normal, and in terms of liver and renal function tests, troponin levels were normal. There was an abnormality in the right atrial shadow on chest X-ray; it appeared to be enlarging. Echocardiography and angiography were performed. Angiography was clear, but there was a mass in the right atrium on echocardiogram. CT was used for detailed assessment of the mass. There was metastasis on the liver. The cardiac team was accompanied and decided to perform surgery.

The surgery was performed by wide thoracotomy from the fourth intercostal space to have a better operational view (Figure 1). The cardiopulmonary by-pass was used via bi-caval cannulation. Atriotomy was performed, and the tumor was excised widely with normal tissue border (Figures 2, 3). Although the tumor was not small, there was no need to use a patch. The right atrium was repaired easily via the continuous suture technique without any narrowing. Afterwards, standard closure was accomplished (Figure 4). After an uneventful intensive care unit follow-up, the patient was discharged on the 5th postoperative day. A few days later, the histopathological investigation revealed a microscopic tumor margin in the material we sent. According to detailed pathologic evaluation, the mass we sent was depicted as high-grade angiosarcoma with wide hemorrhagic areas without necrosis. Immunohistochemical investments revealed positive CD 31 and CD 34. The Ki-67 proliferation index was between 50% and 60%.

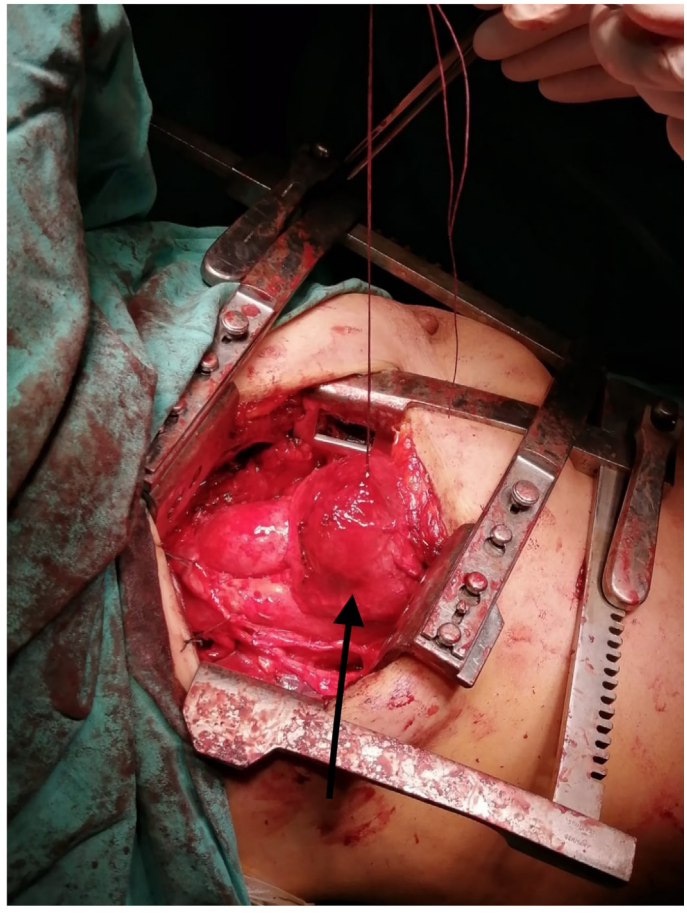

Figure 1. The mass is seen at the tip of the arrow as part of the right atrium from the extended thoracotomy space

Follow-up was maintained by medical oncologic interventions. The patient received chemotherapy six times via doxorubicin and docetaxel. There was liver metastasis on positron emission tomography-CT 18 months after the operation, despite the cardiac functions being hassle-free. Metastasis in the liver was intervened with radiofrequency ablation after detection. Thus, she is still alive approximately 2 years after the operation.

A written informed consent was obtained from the patient.

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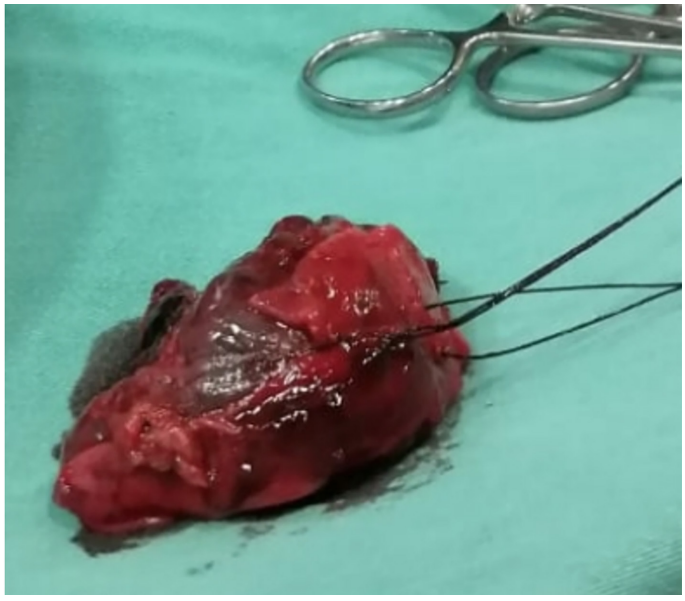

Figure 2. The excised mass is seen macroscopically

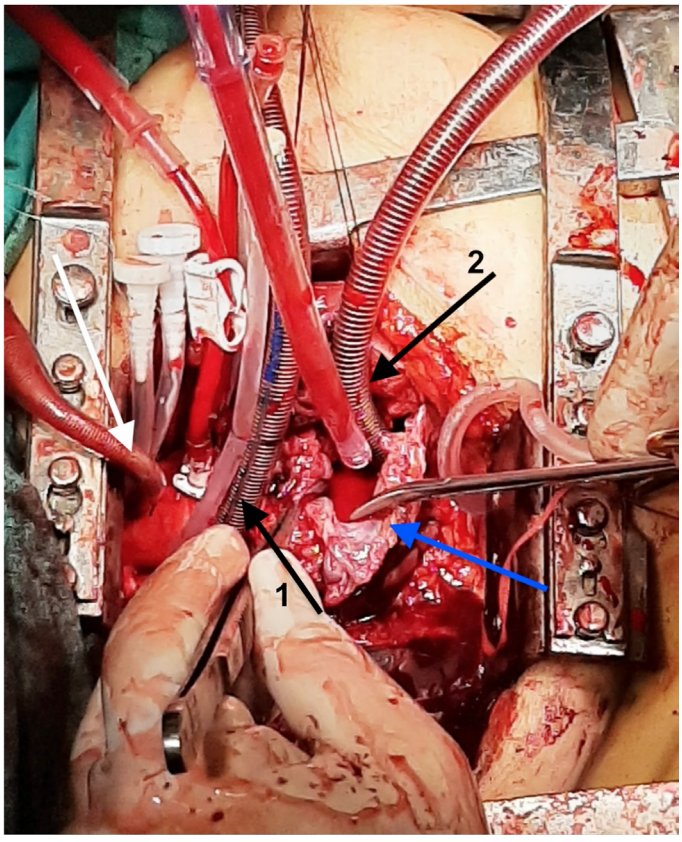

Figure 3. The blue arrow shows the auricle of the right atrium after excising the tumor. The white arrow indicates the aortic cannula. The first and second arrows indicate the cannulas of the superior and inferior vena cava

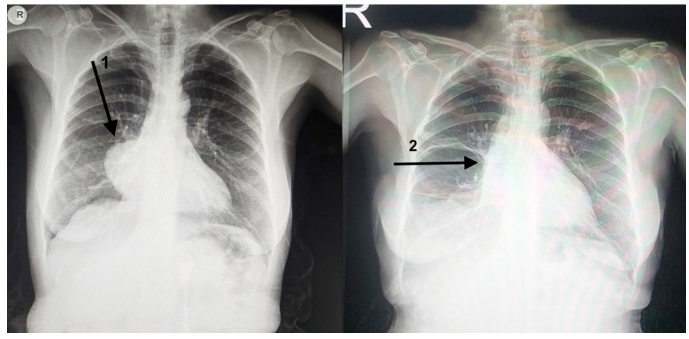

Figure 4. First and second arrows show the right atrial space preoperatively and postoperatively

Discussion

Angiosarcomas are the most frequent primary sarcomas of the heart[1,2]. It constitutes almost 40% of all sarcomas[3]. Even if an extraordinary placement of angiosarcomas has been shown in the literature[4], angiosarcomas are typically placed in the right atrium[1-3]. They tend to invade the atrial wall and pericardium[3]. The most frequent symptom of angiosarcoma is chest pain[2]. Symptoms related to right heart failure, hemopericardium, and supraventricular arrhythmias may be seen as well[2]. Sunray appearance may be observed on CT after receiving IV contrast[2]. Given the immunohistochemical panels, CD34, Factor 8, CD31, and vimentin are positive for angiosarcomas[1]. The macroscopic character of angiosarcoma is generally a gray-brown mass with bleeding sites[1]. Most angiosarcomas have vascular differentiation with significantly atypic endothelial cells in a microscopic view[1,3]. Survival of angiosarcoma is less than 1 year unless the tumor is not resected completely[3]. Metastasis at the beginning of the diagnosis is blamed for poor prognosis[2]. Metastasis of angiosarcomas of the heart is frequently observed in the lung and liver[2].

Surgery is the mainstream therapy for cardiac sarcomas[5]. Furthermore, it has been demonstrated that surgical resection without any macroscopic and microscopic residual tissue is associated with increased survival rates[5]. Even though chemotherapy and radiotherapy might have beneficial effects, it may make surgery to postpone[5]. However, surgery is the

only treatment modality that has evidence of a positive relationship with survival[5]. Look Hong et al.[5] mentioned that their median follow-up is 12 months and the time frame of the median survival is 13 months. Despite the poor outcomes of the tumor, our patient is alive 21 months after the surgery. In the study by Bakaeen et al.[6], 11 of 27 patients had angiosarcomas. Three of them resulted in R1 and the other eight patients were R0[6], which is a clear, successful outcome. However, the resection board in our study was considered R1 even if we attempted R0 resection. Despite the existence of a microscopically residual tumor, no local recurrences were encountered in our patients during the 21-month follow-up period. Thereupon, in the study of Bakaeen et al.[6], local recurrence developed in only 3 of 26 patients undergoing R0 and R1 resection. However, it should be recognized that it is among all sarcomas included in the study. Alassal et al.[7] shared their experiments in this respect. They excised a vast mass from the right atrium and repaired it with the bovine pericardium successfully[7]. However, despite their patient receiving chemotherapy and radiotherapy, he died 7 months after the surgery[7]. On the other hand, the study of Bakaeen et al.[6] has significantly superior outcomes of survival.

In conclusion, angiosarcomas of the heart require further investigation. Literature on this valid topic should be expanded. Angiosarcomas of the heart have pathetic outcomes. Surgery is the cornerstone therapy, but not the only therapy choice. Furthermore, our study claims that multiple treatments for angiosarcoma of the heart are more beneficial than surgery alone. Expanding the life span of these patients is possible by supplying surgery with radiotherapy and chemotherapy modalities. Thus, our study is a comprehensive case to contribute to the literature.

Ethics

Informed Consent: Informed consent was obtained.

Authorship Contributions

Surgical and Medical Practices: - Concept: - Design: - Data Collection and/or Processing: - Analysis and/or Interpretation: - Literature Search: - Writing: All authors contributed equally to the article.

Conflict of Interest: The authors declare no conflicts of interest concerning the authorship or publication of this article.

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