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Long Term Survival with Primary Angiosarcoma: A Case Study of Survival after Simple Resection

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Case Study

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ABSTRACT

Here we report a case of primary angiosarcoma found in the typical location of the right atrium. Based upon a literature review the average documented survival is 6-14 months with maximal therapy. This patient had a simple resection with no cardiac transplant, chemotherapy, or radiation and has survived with no symptoms for over 7 years. We were unable to find any cases of a patient surviving this long with a primary angiosarcoma treated with simple resection. This indicates the possibility of survival for a prolonged period assuming the tumor does not infiltrate or alter cardiac function by obstruction.

LEARNING OBJECTIVE

Several treatment modalities for primary cardiac angiosarcoma have been proposed and attempted. However, apart from complete cardiac transplant, no long-term survival benefit from any treatment method has been reported. We present a case of a primary cardiac angiosarcoma treated with simple resection that has resulted in a symptom free survival benefit of over 7 years.

INTRODUCTION

Primary cardiac tumors are a rare occurrence in the general population with an incidence ranging from 0.0017-0.003% of all cancer diagnoses [1]. While the majority of these cardiac tumors are benign, approximately 25% of them are found to be malignant. Within that 25% of malignant tumors, angiosarcoma represents approximately 31% of the neoplasms [2]. Sarcomas arise from mesenchymal tissue (cancellous bone, cartilage, fat, muscle, vascular, or hematopoietic tissue) and will resemble these specific cells of origin. Cardiac angiosarcomas are most often located in the right atrium of middle aged males and may produce the signs and symptoms as right sided heart failure. Complete resection of the primary mass is usually difficult due to its location and the fact that most patients already have distant metastases.

CASE DESCRIPTION

This 75 year old Hispanic male was being followed annually in the cardiology clinic after a two-vessel bypass was performed with a left internal mammary artery to left anterior descending artery graft and a saphenous vein to right coronary artery graft. At follow-up he began to complain of intermittent left sided chest pain. This pain would improve with coughing and was mild enough that he could still engage in regular physical activity. He was taken for coronary angiogram and found to have 2+ aortic regurgitation with a normal ejection fraction and a totally occluded right coronary vein graft. Due to chest pain an echocardiogram was ordered to evaluate ventricular function and imaging revealed an unidentified mass in the roof of the right atrium **(Figure 1)**.

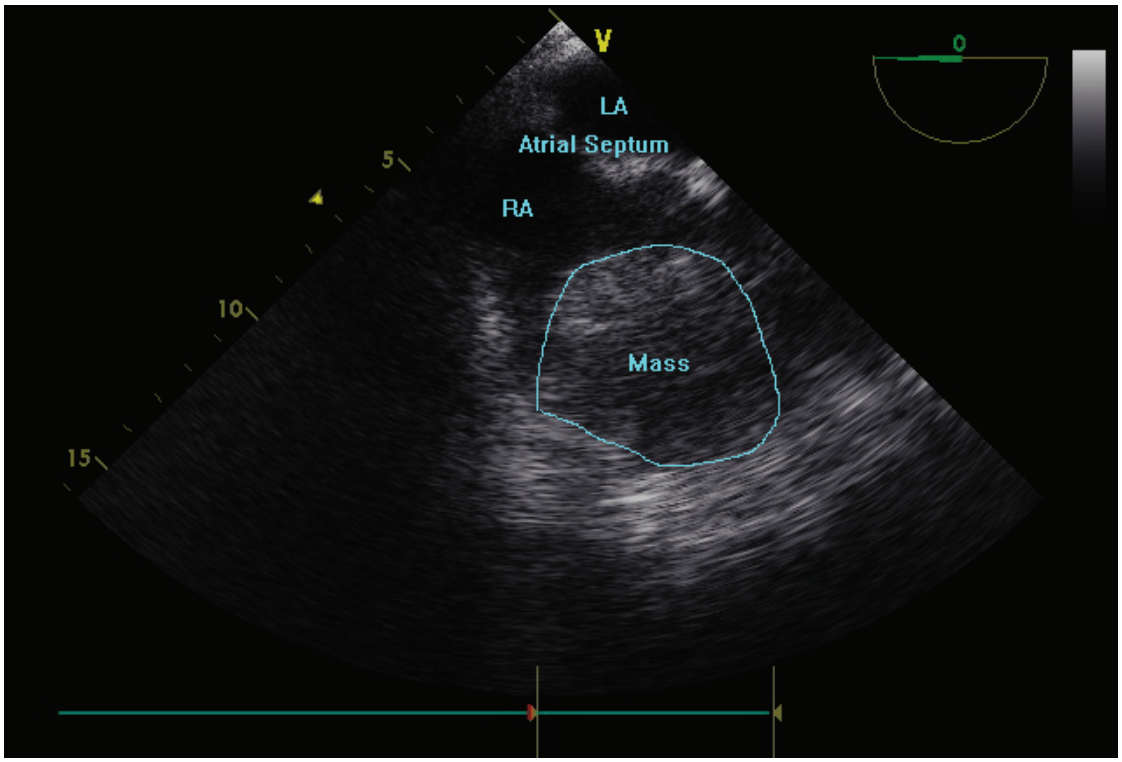

Figure 1. Echocardiogram demonstrating a large mass within the right atrium.

Subsequent cardiac MRI showed a large right atrial mass with bi-atrial enlargement, 2+ aortic regurgitation and findings consistent with a previous infarct **(Figure 2)**.

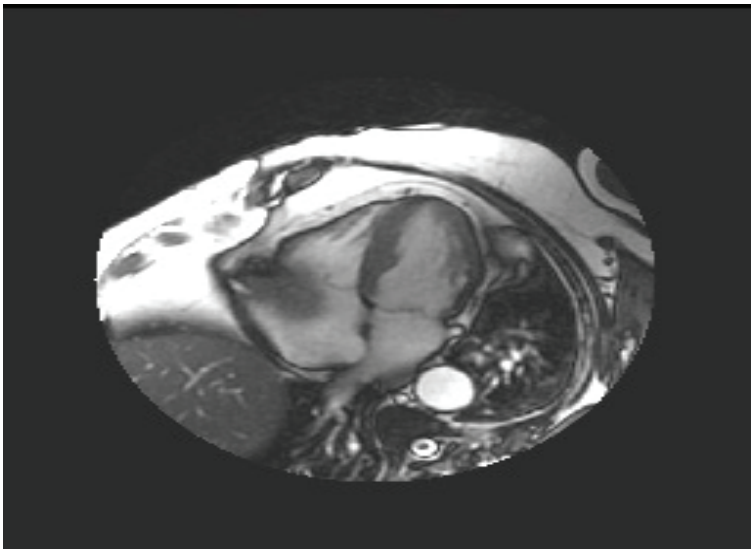

Figure 2. Cardiac MRI demonstrating right atrial mass prior to surgery.

After verifying vascular anatomy with the previous angiogram he was taken to the operating room for exploration and possible resection. Sternal entry was achieved without incident and revealed significant cardiac adhesions. Upon direct visualization, the right heart appeared distended and contained suspicious tissue in the atrioventricular groove. The origin of this tissue was difficult to identify, though it did not appear to be invading nearby structures. No mass was appreciated after entering the right atrium however when the tissue overlying the atrioventricular groove was dissected a large amount of thrombus was found along with abnormal tissue lining a distinct cavity that lacked communication with any cardiac chamber. The tumor had a clear cleavage plane which permitted the mass to be scraped from the atria, ventricle, and aortic sinus. Lymph nodes sent for frozen section were negative for malignancy. Echo immediately following resection demonstrated a complete resolution of the mass. Biopsy sections sent to on-site pathology confirmed a diagnosis of high grade cardiac angiosarcoma. Additional samples were sent to Mayo clinic in Rochester and found to be CD31 and OSCAR cytokeratin positive which is consistent with angiosarcoma. No additional therapy has been performed since the resection. Serial CT scans taken after surgery has shown slight recurrence of a mass in the same anatomic location; however, it appears to be extremely slow growing and free of any distant metastasis. The patient has deferred

any possible chemotherapy or radiation treatments at this time and currently reports no negative impacts or restrictions in his daily life. No additional surgical therapy has been recommended.

DISCUSSION

Unfortunately, with the exception of complete heart transplant for non-metastatic tumors [3,4] no single therapy has been found to be universally efficacious for the treatment of primary cardiac angiosarcoma. Radiation therapy is contraindicated due to the risks of constrictive pericarditis and other thoracic side effects. Chemotherapy shows poor patient outcome with an average survival of 6-11 months in treatment groups [5]. Conservative surgery is not known to be curative and is utilized mostly for local control and symptomatic relief. Previous attempts at more aggressive surgical resection of a mass have also been discouraging with recent data showing an average survival time of between 6 and 14 months [5,6].

Our case illustrates that long-term survival, while unlikely, is possible in rare circumstances of primary angiosarcoma after complete tumor resection. "Complete or partial surgical resection is still the best option for palliation, with little hope for cure" [7,8]. We choose to proceed with resection based upon the best data available after a careful conversation about the risks and benefits with the patient. Surgically wide resection with negative margins is of course preferred but challenging due to the infiltrate nature of the disease itself. Local recurrence is a frequent occurrence hence the recommendation of additional therapies including chemotherapy and radiation. Surgical approaches have changed little in the past few decades but case reports from different patients seems to suggest that without metastasis survival is significantly improved [9]. Overall standard therapy with resection and adjunct chemotherapy and/or radiation is the ideal strategy with the consideration of heart transplantation in young patients [3].

CONCLUSION

Reviewing his case there are multiple possibilities for his continued survival even though he declined radiation therapy or chemotherapy. First is that the tumor itself was localized to an anatomic region without distant or any metastasis. Next there was a clear cleavage plain allowing gross removal of the entire mass. Finally, while the mass was filled with thrombus, likely bleeding from neovascularization of the tumor, it was relatively small sized in that there were no signs or symptoms of heart failure associated with it prior to excision. The actual nature of adhesion with this specific type of malignant cell may also play a factor and while this is outside of the purview of a case report it may be a direction of future research to consider. Also, worth noting is that the malignancy has returned and continued to grow, however it's extremely slow growth rate has prevented additional problems. Yet, it must be noted that it was not present during the patients' initial coronary artery bypass surgery so it appears to have had a variable rate of growth which also may be worth comparing in a larger study with long term survival in cardiac angiosarcoma.

REFERENCES

1. Reynen K. Frequency of primary tumors of the heart. Am J Cardiol. 1996;77:107
2. Centella T, et al. A patient with cardiac angiosarcoma who survived for four years. Case report and literature review. Rev Esp Cardiol. 2005;58:310-312
3. Baay P, et al. Successful treatment of a cardiac angiosarcoma with combined modality therapy. J Heart Lung Transplant. 1994;13:923-925.
4. Überfuhr P, et al. Heart transplantation: An approach to treating primary cardiac sarcoma? J Heart Lung Transplant. 2002;21:1135-1139
5. Antonuzzo L, et al. Primary cardiac angiosarcoma: A fatal disease. Case Rep Med. 2009;10:1155
6. Herrmann MA, et al. Primary cardiac angiosarcoma: A clinicopathologic study of six cases. J Thorac Cardiovasc Surg. 1992;103:655-664.
7. Patel SD, et al. Primary cardiac angiosarcoma - a review. Med Sci Monit. 2014:103-109.
8. Centofanti PR, et al. Primary cardiac tumors: Early and late results of surgical treatment in 91 patients. Ann Thorac Surg. 1999;68:1236-1241.
9. Truong PT, et al. Treatment and outcomes in adult patients with primary cardiac sarcoma: The British Columbia cancer agency experience. Ann Surg Oncol. 2009;6:3358-3365.