Atrial Thrombus Mimicking Ewing’s Sarcoma

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INTRODUCTION

Catheter-related intracardiac thrombosis is a documented complication of central venous catheters (CVCs), specifically within the right atrium. We report a case of an incidental finding of right atrial mass on routine transthoracic echocardiography for post-chemotherapy evaluation in a patient with Ewing’s sarcoma. The mass was evaluated as a calcified atrial thrombus using minimally invasive investigations of echocardiography, cardiac MRI (magnetic resonance imaging), and positron emission tomography.

CASE PRESENTATION

A 17-year-old male patient presented for echocardiography as part of routine postchemotherapy assessment for localized Ewing’s sarcoma of his left hip. Genetic analysis of the Ewing’s sarcoma was fluorescence in situ hybridization positive for EWSR/FLI1 rearrangement. His oncologic management included surgical excision with clear margins and adjuvant chemotherapy with vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide. The management of Ewing’s sarcoma was otherwise uncomplicated. A CVC was in situ for 10 months for the administration of chemotherapy. On transthoracic echocardiography, there was an incidental finding of an echo density measuring roughly 1.3 × 2.0 cm within the right atrium (Figure 1). The patient was asymptomatic, with optimal response and recovery to chemotherapy. Differential diagnoses at this time included cardiac tumor with potential metastatic invasion of Ewing’s sarcoma, intracardiac thrombus, and a prominent crista terminalis.

Urgent transesophageal echocardiography was performed to further evaluate the right atrial mass. The findings were of an echoropic, amorphous structure in the inferoposterior aspect of the right atrium adjacent to the tricuspid annulus with no valvar involvement (Figure 2). The structure appeared immobile and adherent to the right lateral, posterior, and inferior wall of the right atrium (Figure 3, Videos 1 and 2). Of note, the CVC was seen projecting toward and abutting the atrial mass (Figure 3, Video 3). The initial impression was fibrosis and possible organized thrombus secondary to continuous irritation from the CVC tip on the right atrial wall and less likely cardiac malignancy or changes related to Ewing’s sarcoma.

The patient was immediately commenced on a course of therapeutic anticoagulation. He underwent uneventful removal of the CVC. Repeat transesophageal echocardiography was performed after 6 weeks, showing no significant change of the mass, and the impression was of an calcified, organized thrombus from central line-induced trauma (Figure 4).

For further clarification, cardiac MRI was also performed. It confirmed the mass to be an amorphous, avascular structure with only minimal contrast enhancement on the undersurface of the mass on delayed imaging. The estimated size was 0.5 × 1.2 cm (Figure 5).

The patient remained on therapeutic anticoagulation with routine cardiology and oncology follow-up. As part of his oncology follow up, 18F-fluorodeoxyglucose positron emission tomography was performed, which did not identify 18F-fluorodeoxyglucose-avid recurrence of his Ewing’s sarcoma. Cardiac-focused images were taken, showing no notable suspicious tracer avidity of the atrial mass, compatible with presumed diagnosis of a calcified thrombus (Figure 6).

Finally, repeat transthoracic echocardiography was performed 5 months after initial imaging. Although the patient has completed an extended course of therapeutic coagulation, the atrial mass remained present, with no significant change in size or location. On the basis of the maintained calcific appearance of the mass, it is likely that it will remain indefinitely despite prolonged anticoagulation. On repeated imaging, the calcified thrombus has remained fixed and immobile, with an exceedingly low risk for an embolic change. The patient has remained asymptomatic with an otherwise optimal recovery from his oncologic treatment. He has remained in remission with ongoing oncology follow-up.

DISCUSSION

Cardiac malignancy is rare, with an autopsy incidence of 0.02% to 0.056% for primary tumors and 1.23% for secondary tumors. The prognosis of malignant cardiac tumors is very poor, although distant metastasis is the cause of death in the majority of patients. Although it is uncommon, cases of cardiac metastasis of Ewing’s sarcoma have been documented.

In this case, we reviewed a young patient with an incidental finding of an intracardiac mass at the completion of his oncologic therapy for Ewing’s sarcoma. Although there are reported cases of cardiac metastasis of Ewing’s sarcoma, the incidence and prognosis of cardiac metastasis of Ewing’s sarcoma is not well established. On review of the available literature, the spread of cardiac Ewing’s sarcoma is also variable, with reported in cases of involvement in the right ventricle alone, the right and left ventricles, and within the left atrium. The prognosis remains poor in widespread metastatic Ewing’s sarcoma with or without cardiac involvement, with <20% of cure.

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280
Figure 1 Apical long-axis right atrial–focused view on initial transthoracic echocardiography. As indicated by the red arrow, there is an undifferentiated echo density measuring roughly 1.3 × 2.0 cm within the right atrium.

Figure 2 Image from initial transesophageal echocardiography from midesophageal position of the probe at 126° transducer angle. The right atrial echo density previously visualized on transthoracic echocardiography is better defined at the inferoposterior aspect of the right atrium, as marked by the red arrow.

Figure 3 Image from initial transesophageal echocardiography from a modified bicaval view. The right atrial echo density, as indicated by the red arrow, is visualized adherent to the lateral/inferior wall of the right atrium. Of note is the sessile nature of the mass with very limited mobility.

Figure 4 Image from repeat surveillance transesophageal echocardiography from a midesophageal four-chamber view. The right atrial echo density, as indicated by the red arrow, is again visualized adherent to the right atrial lateral wall. There is no apparent change in the size or location of the mass compared with previous imaging. The previously visualized CVC had been removed and is no longer appreciated.

Figure 5 Fluorine-18-fluorodeoxyglucose positron emission tomographic images were obtained as part of postchemotherapy surveillance. There was additional focus placed on assessment of the previously identified right atrial mass with dedicated cardiac views. There was no evidence of metabolic activity of the right atrial mass, as indicated by the red arrow, suggestive of a lesion of benign nature.

VIDEO HIGHLIGHTS

Video 1: Initial transesophageal echocardiography on midesophageal four-chamber view, live three-dimensional acquisition. The right atrial mass can be seen adherent to atrial wall. The mass is echo dense, amorphous, and fixed at the inferoposterior aspect of the right atrium. Also seen is the CVC, which appears to be abutting the mass.

Video 2: Initial transesophageal echocardiography, three-dimensional imaging capture. The mass is viewed from above the right atrium. The mass is seen adherent to the lateral/inferior wall of the right atrium. Of note is the sessile nature of the mass with very limited mobility.

Video 3: Initial transesophageal echocardiography from midesophageal modified bicaval view. A biplane capture is shown. The right atrial echo density can be seen fixed to the atrial wall adjacent to the tricuspid valve. A CVC can be prominently seen in the left image extending toward the atrial mass with the tip of the catheter abutting the echo density.

View the video content online at www.cvcasejournal.com.
As such, on the discovery of the undifferentiated intracardiac mass in this patient, extensive workup was conducted. On the basis of the available imaging in conjunction with clinical assessment, there were no immediate indications for further invasive surgical investigation and management. Echocardiography was an essential component in diagnostic evaluation as well as surveillance of progression or lack of progression of the intracardiac mass.

CVCs are important access devices routinely used in health care, with the majority in use for prolonged durations in oncologic therapy and in the critically ill. Catheter-related intracardiac thrombosis is a known complication of CVCs, with most reported cases within the right atrium. In a previous study of CVC-related thrombus, 12.5% of patients with CVC insertion were found to have abnormal right atrial masses consistent with thrombus on echocardiography. In another study of postmortem cases, 29% of patients with CVC insertion were found to have mural thrombi present on autopsy. In general, CVC-associated thrombosis is a well-documented complication with considerable impact of patient morbidity and mortality.

With the continued increasing rate of CVC use, particularly in the care of ill patients in hypercoagulable states, there should be strong consideration of the placement of the CVC. As expected, higher rates of right atrial thrombi have been observed in patients with CVC tips placed within the right atrium.

Echocardiography in conjunction with other minimally invasive imaging modalities, such as cardiac MRI and positron emission tomography/computed tomography, plays an important diagnostic and surveillance role in all cardiac investigations. Specifically for the workup of a right atrial mass, a broad multimodality approach is outlined in Figure 7. Although rare, cardiac metastasis, specifically of Ewing’s sarcoma, has significant associated mortality and should be thoroughly worked up on any index of suspicion. CVC-related intracardiac thrombosis should continue to be evaluated as their role in

Figure 6 Low-dose computed tomographic images obtained for attenuation correction purposes demonstrate evidence of a calcific mass in the right atrial lateral wall, indicated by red arrow.

Figure 7 Flowchart for the approach and evaluation of a right atrial echo density.
health care provision grows, with greater attention to the initial placement of the CVC.

CONCLUSION

Our case demonstrates an incidental finding of an intracardiac mass in a patient on a background of Ewing’s sarcoma. From multiple imaging investigations along with clinical correlation, the concluding impression was of a fixed, calcified atrial thrombus secondary to CVC irritation. Both transthoracic echocardiography and transesophageal echocardiography were essential investigative and surveillance modalities along with cardiac MRI and 18F-fluorodeoxyglucose positron emission tomography as supportive investigations for the evaluation and workup of an undifferentiated intracardiac mass.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.07.006.

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