

An Incidental Finding of Coronary-cameral Fistulas in a Critically Ill Patient with a Metastatic Cardiac Tumor

Braghadheeswar Thyagarajan¹, Casey Bryant², Ashish K Khanna³

ABSTRACT

It is extremely rare for a cardiac tumor to present with coronary-cameral fistulas. A 66-year-old Caucasian male presented with worsening dyspnea and subsequently had a cardiac catheterization for an ST-elevation myocardial infarction, revealing no evidence of coronary artery disease but multiple coronary-cameral fistulas. Venoarterial (VA) extracorporeal membrane oxygenation was initiated for a severe cardiogenic shock. Workups including a transesophageal echocardiography and a right heart catheterization led to a diagnosis of metastatic melanoma involving both ventricles of his heart. Angiogenesis is well described in melanomas and our novel case reports the extremely rare association of coronary-cameral fistulas with a metastatic cardiac melanoma.

Keywords: Cardiac tumor, Coronary-cameral fistulas, Metastatic melanoma.

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INTRODUCTION

The overall incidence of cardiac tumors is around 0.02%. Metastatic cardiac tumors are more common than primary cardiac malignancies. Presenting symptoms can be classified as obstructive, embolic, constitutional, and arrhythmias.¹ Coronary-cameral fistulas are rare and usually congenital in origin.² There are reports of coronary-cameral fistulas associated with other cardiac tumors.^{3,4} Our case is a rare presentation of a metastatic melanoma of the heart with coronary-cameral fistulas, where we discuss the diagnostic pathways and the critical care management of one such patient in our cardiovascular surgery intensive care unit.

CASE DESCRIPTION

A 66-year-old Caucasian male with no medical history presented with worsening dyspnea and cough for 2 weeks. An electrocardiogram (EKG) obtained showed diffuse ST-segment elevation in leads II, III, aVF, and V3-V5. Cardiac catheterization showed no evidence of coronary artery disease but multiple coronary-cameral fistulas (Fig. 1), connecting the right coronary artery (RCA) to the right ventricle (RV). The systolic pulmonary artery pressure was 90 mm Hg and the wedge obtained was 60 mm Hg. The patient was intubated due to worsening respiratory distress. The rapidly worsening severe cardiogenic shock led to a ventricular tachycardia (VT), requiring cardioversion and amiodarone 150 mg intravenously (IV). Mechanical circulatory support was then initiated with an intra-aortic balloon pump (IABP) through the right femoral artery with 1:1 frequency. He was placed on norepinephrine at 20 µg/minute and epinephrine at 10 µg/minute and subsequently transferred to our center for consideration of venoarterial (VA) extracorporeal membrane oxygenation (ECMO) support.

In the intensive care unit, he had another episode of VT, which required cardioversion, amiodarone 150 mg IV, a lidocaine bolus 1 mg/kg, and a lidocaine infusion at 2 mg/kg/hour. He was also started on milrinone 0.5 µg/kg/hour and inhaled epoprostenol 0.05 µg/kg/minute. Given the unstable hemodynamics, the patient was emergently cannulated onto VA ECMO, using femoral arterial (15 French) and venous cannulation (23 French) for a severe

^{1,2}Department of Anesthesiology, Section on Critical Care Medicine, Wake Forest School of Medicine, Winston-Salem, North Carolina, United States

³Department of Anesthesiology, Section on Critical Care Medicine, Wake Forest School of Medicine, Winston-Salem, North Carolina, United States and Outcomes Research Consortium, Cleveland, OH, United States

Corresponding Author: Braghadheeswar Thyagarajan, Department of Anesthesiology, Section on Critical Care Medicine, Wake Forest School of Medicine, Winston-Salem, North Carolina, United States, Phone: +1 732 804 5949, e-mail: bragmd@gmail.com

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cardiogenic shock. Initial transthoracic echocardiogram (TTE) showed hyperechoic structures in both his ventricles, presumed to be a thrombus formation due to severe left and right ventricular dysfunction. Additional findings included reduced left ventricular (LV) ejection fraction, severe LV global hypokinesis, severe RV dilation, and a small pericardial effusion (Fig. 2). Once stabilized, follow-up transesophageal echocardiogram (TEE) images showed a large triangular mass in the LV apex and a moderate-sized shelf-like mass at the RV apex (Fig. 3). Color Doppler demonstrated flow over the RV mass, confirming the coronary-cameral fistulas (Supplemental Video 1). It was now suspected that these findings represented a cardiac tumor, so a computed tomography (CT) with contrast of the chest, abdomen, and pelvis was obtained that showed metastatic nodules in the lungs, adrenals, kidneys, and liver.

Eventually, a right heart catheterization (RHC) was performed with a biopsy of the RV mass and the final pathology demonstrated a heterogeneous population of both epithelioid and spindle cells. Areas of the tumor showed a melanocytic pigment as well as robust reactivity of those areas to antibodies directed against S100, SOX10 (SRY-related HMG box), and HMB45 (human melanoma

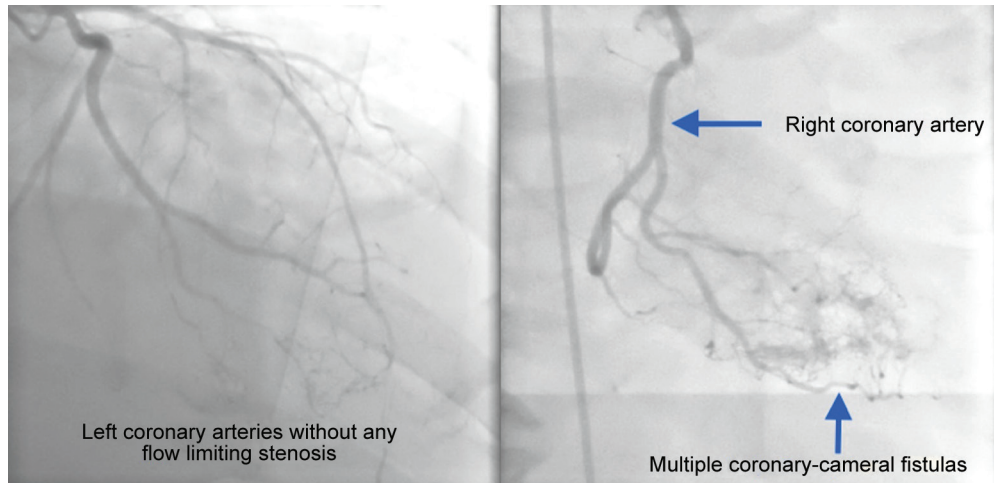


Fig. 1: Cardiac catheterization showed no evidence of coronary artery disease but multiple coronary-cameral fistulas

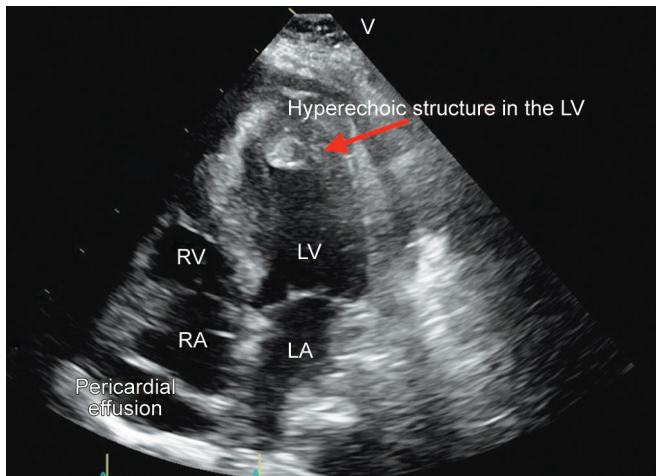


Fig. 2: Transesophageal echocardiogram (TEE) showed hyperechoic structures in both his ventricles and a small pericardial effusion

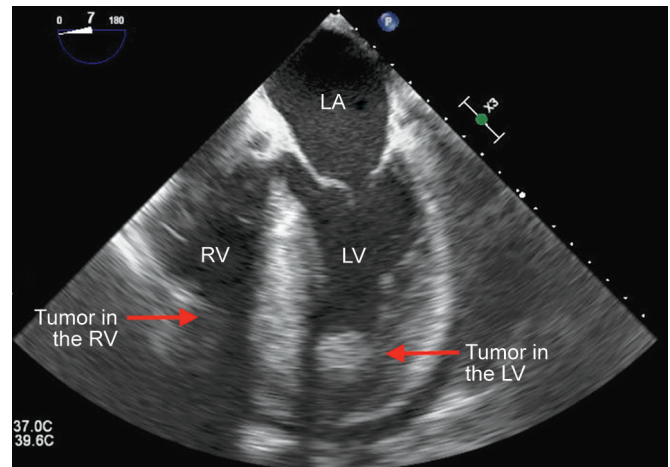


Fig. 3: Transesophageal echocardiogram (TEE) images showed a large triangular mass in the left ventricular (LV) apex and a moderate-sized shelf-like mass at the right ventricular (RV) apex

black). These features are most in keeping with that of a metastatic malignant melanoma. The patient was subsequently weaned off and decannulated from VA ECMO but continued to be on milrinone 0.375 $\mu\text{g}/\text{kg}/\text{minute}$, epinephrine at 4 $\mu\text{g}/\text{minute}$, inhaled epoprostenol 0.05 $\mu\text{g}/\text{kg}/\text{minute}$, vasopressin at 0.04 $\mu\text{g}/\text{minute}$, and IABP with 1:2 frequency. Due to the inoperable nature of the tumor and the lack of chemotherapeutic treatment options, the family decided to pursue comfort care measures.

DISCUSSION

Metastatic cardiac tumors are more common than primary cardiac tumors. The incidence of secondary metastatic cardiac tumors is between 0.7 and 3.5% at autopsy.⁵ The most common metastases are from lung cancer followed by breast cancer. Around 28–56% with metastatic melanoma have cardiac involvement.⁶ The tumor cells often reach the heart by direct extension, hematogenous spread, lymphatic extension, and transvenous extension. The

patient in our case had no past diagnosis of melanoma, so it is possible that he had it in an uncommon site. Cardiac metastases are often silent without any symptoms until at a late stage and most are diagnosed during postmortem.⁷ When present, the clinical presentations of cardiac tumors can be manifested as obstructive, embolic, constitutional, and arrhythmias.¹

Our patient also demonstrated coronary-cameral fistulas on coronary angiography. These fistulas are abnormal vascular connections between the coronary arteries and the cardiac chambers. The incidence of coronary-cameral fistulas is 0.002%. Commonly, they are congenital in origin. Acquired fistulas are most commonly from trauma or iatrogenic causes.² On review of the published literature, there are only a few reports on the association of coronary-cameral fistulas with left atrial myxomas and intracardiac tumors.^{3,4} Based on the size of the coronary-cameral fistulas, these may also present with a steal syndrome, myocardial infarction, dyspnea, angina, and congestive heart failure, either alone or in combination.⁸ There is currently no published literature that reports

coronary-cameral fistulas in conjunction with cardiac melanomas. Angiogenesis has been well described in the development of and metastasis of melanomas.⁹ In our patient, coronary-cameral fistulas arising from the RCA supplying the tumor in the right ventricle are likely due to the process of angiogenesis. The EKG changes initially seen in our patient with ST elevations in leads II, III, and aVF could be related to the presence of coronary-cameral fistulas and a steal phenomenon, affecting blood flow through the RCA and causing an inferior wall myocardial infarction.

TTE has a high sensitivity (90%) and specificity (95%) for the detection of cardiac tumors and can be performed at the bedside.¹⁰ Contrast echocardiography helps in differentiating a tumor from a thrombus.¹⁰ TEE improves image quality and better visualization of small tumors (<5 mm) and tumors in the posterior cardiac segments.¹¹ Multislice CT scans have improved imaging capability of cardiac masses, but a cardiac magnetic resonance imaging has a superior soft tissue characterization.¹² Positron emission tomography-CT remains the gold standard to diagnose metastasis and staging of known malignant melanomas.¹³ Historically, coronary-cameral fistulas have been diagnosed with cardiac catheterization, but with more advancement in imaging modalities, they are also diagnosed with echocardiography and CT scans.¹⁴ In our patient, apart from cardiac catheterization images, we were also able to view the coronary-cameral fistulas on the TEE. Surgical resection and heart transplantation have been described for benign and nonmetastatic cardiac tumors.¹ However, the 5-year survival rate is 15–20% in stage IV melanomas.¹⁵ There are reports describing successful palliative surgical resection of cardiac melanomas for the relief of obstructive symptoms.¹⁵ Given the significant hemodynamic instability and extensive nature of the tumor, our patient was deemed to not be an operative candidate and the family pursued comfort care measures.

CONCLUSION

Clinical presentations of metastatic melanomas to the heart depend on the location and extent of tumor burden. The tumor can masquerade as a myriad of acute cardiac emergencies, making the diagnostic pathway a challenge. Coronary-cameral fistulas rarely can develop in a metastatic melanoma to the heart due to its predilection for angiogenesis. Palliative surgical resection can be considered, but the overall prognosis is poor, and critical care management is primarily supportive.

CONTRIBUTIONS

Braghadheeswar Thyagarajan—Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work.

Casey Bryant—Drafting the work or revising it critically for important intellectual content.

Ashish K. Khanna—Final approval of the version to be published.

ORCID

Braghadheeswar Thyagarajan  <https://orcid.org/0000-0003-4290-6436>

Casey Bryant  <https://orcid.org/0000-0001-5702-5899>

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