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Right Atrium Primary Cardiac Lymphoma Causing Heart Failure

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Abstract

Background Patients with primary cardiac tumors may present with symptoms based on the size and location of the tumor. Symptoms may include congestive heart failure secondary to intracardiac obstruction, systemic embolization, arrhythmias, and constitutional symptoms.

Case Description A patient presented with new onset atrial fibrillation and heart failure. Workup including open surgery revealed a primary cardiac lymphoma.

Conclusion Cardiac tumors present with a variety of symptoms and are best evaluated by echocardiogram, computed tomography angiography, and magnetic resonance imaging. Tissue diagnosis is necessary. Although primary cardiac lymphoma is rare, long-term survival after chemotherapy and rituximab is superior to other cardiac malignancies.

Keywords
- cardiac
- heart failure
- imaging (all modalities)
- off-pump surgery
- positron emission tomography
- tumor

Introduction

Patients with primary cardiac tumors may present with symptoms based on the size and location of the tumor. Symptoms may include congestive heart failure (CHF) secondary to intracardiac obstruction, systemic embolization, arrhythmias, and constitutional symptoms. The identification of a cardiac tumor relies heavily on echocardiogram, computed tomography angiography (CTA), and magnetic resonance imaging (MRI). Some cardiac masses may not require tissue diagnosis such as pseudotumors, thrombus, lipomas, and papillary fibroelastoma.[1] All other tumors require a tissue diagnosis that will dictate further treatment.

Case Presentation

A 74-year-old male presented to his primary care physician with a 1-week history of paroxysmal nocturnal dyspnea and chest discomfort. He had never experienced these symptoms. He was an active smoker with a 20-pack year history but had no history of cardiac disease. The rest of his medical and surgical history was unremarkable. He denied syncope, chest pain, fever, chills, cough, abdominal pain, nausea or vomiting, use of illegal drugs, or family history of cardiac disorders.

In the emergency department, his heart rate was 150 beats per minute, with other vital signs within normal limits. Heart examination showed irregular tachycardia without murmurs. Electrocardiogram identified new onset atrial fibrillation and no signs of ischemia. Chest X-ray demonstrated cardiomegaly and pulmonary edema. Pro-B-type natriuretic peptide was elevated at 1,406 pg/mL (0–150) with normal troponin levels. (►**Fig. 1A**)

Echocardiogram revealed a large pericardial effusion without tamponade, a right atrial mass, and a left ventricular ejection fraction of 25%. He was admitted to the hospital with intravenous heparin and diltiazem therapy. A pericardial window relieved his symptoms, but no diagnosis was

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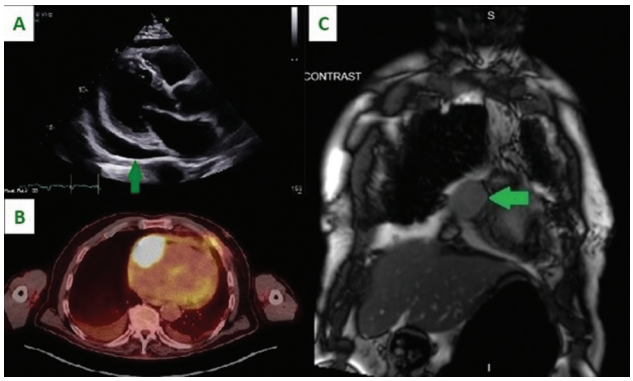

Fig. 1 Preoperative imaging. (**A**) Transthoracic echocardiography demonstrated pericardial effusion. (**B**) positron emission tomography scan. (**C**) Magnetic resonance imaging.

obtained from examination of the fluid and pericardial tissue. Given concerns for a cardiac malignancy, better characterization of this mass was pursued with transesophageal echocardiogram, a positron emission tomography, and MRI.

Transesophageal echocardiography identified a right-sided cardiac mass in the atrioventricular groove measuring 4.1 × 4.2 × 5.3 cm. This mass was positive on positron emission tomography scan (►**Fig. 1B**), with no other positive area. MRI (►**Fig. 1C**) demonstrated right coronary involvement.

A coronary CTA (►**Fig. 2**) was obtained, and this confirmed that a nondominant right coronary was encased by the mass. Cardiac catheterization revealed no obstructive coronary disease. After evaluation by the tumor board and review of previous imaging, no mass was identified on a previous CT scan of the chest 4 years prior.

A minimally invasive approach was considered but decided against given concern for potential iatrogenic right

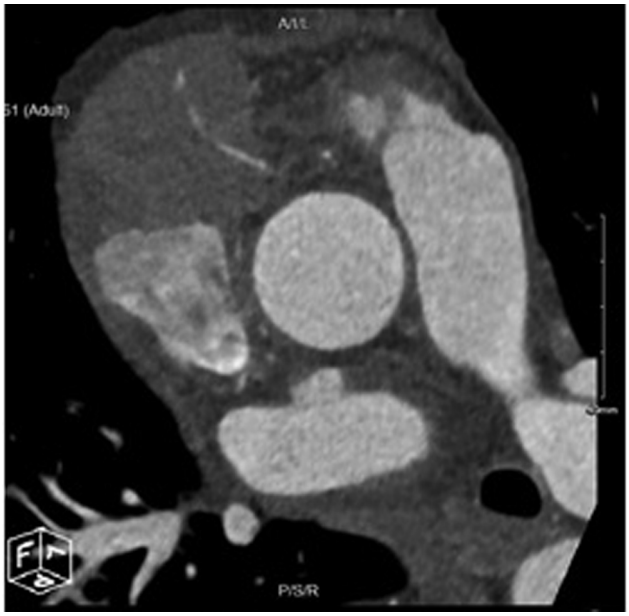

Fig. 2 Computed tomography coronary angiography with right coronary encased by tumor.

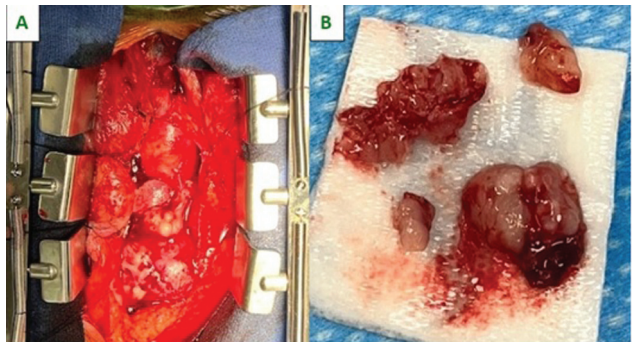

Fig. 3 Intraoperative images: (**A**) Surgical site. (**B**) Specimen.

coronary artery injury by this method. He was returned to the operating room for open biopsy (►**Fig. 3A, B**) through a median sternotomy. Pathology revealed a large B cell lymphoma, and he was treated appropriately.

Discussion

Patients with primary cardiac tumors may present with symptoms based on the size and location of the tumor. Symptoms may include CHF secondary to intracardiac obstruction, systemic embolization, arrhythmias, and constitutional symptoms. The identification of a cardiac tumor relies heavily on echocardiogram, CTA, and MRI to confirm the size and location. Some cardiac masses may not require tissue diagnosis such as pseudotumors, thrombus, lipomas, and papillary fibroelastoma.[1] All other tumors require a tissue diagnosis that will dictate further treatment.

Primary cardiac tumors represent 0.3 to 0.7% of all cardiac tumors. Cardiac metastasis from an extra cardiac primary tumor is 30 times more likely.[1] Cardiac myxomas are a form of benign primary cardiac tumors, and although rare, myxomas are the most common primary tumor of the heart. Myxomas typically develop within specific regions of the heart, 75% of cases originate in the left atrium, while around 20% arise from the right atrium.[2] Because benign right heart tumors are less common than left-sided benign tumors and are often very large, these are frequently seen early before surgical resection has been attempted because of the suspicion of malignancy.[1]

Only 25% of primary cardiac tumors are malignant and of these, 75% are sarcomas. Without surgical resection of a sarcoma, the survival rate is 10% at 9 to 12 months.[1] Primary tumors usually arise in a much younger population than our patient. Right-sided tumors usually grow in an outward pattern. If imaging does not characterize the cardiac mass during the evaluation, a biopsy is warranted for diagnosis and to exclude lymphoma, which are best treated nonsurgically. A retrospective review of 54 patients that underwent surgical resection with bovine pericardial reconstruction of primary cardiac malignancy reported a 9% 30-day mortality. Survival was not improved for patients with positive surgical margins but improved to a 5-year survival of 17% for patients with negative surgical margins.[3,4]

Primary cardiac lymphomas (PCLs) represent 1% of all primary cardiac malignancies. A review of the National Cancer Database identified 305 patients with PCL between 2004 and 2016. Eighty-two percent patients were white and 54.5% male. The average age was 68.7 (± 14.2) years. About 92.7% were B cell lymphoma. Seventy-one percent of patients were treated with chemotherapy, 21.6% with immunotherapy, 16.7% with surgery, and 8.2% with radiation. Overall survival was 46.6% with a median survival of 45.4 months. The most common chemotherapy regimen was CHOP—cyclophosphamide, doxorubicin, vincristine, and prednisone. Increasing age was correlated with poor survival. Improved survival was associated with chemotherapy and further improved survival was statistically significant with the addition of rituximab resulting in a 1-, 3-, and 4-year survival of 71.6, 60.2, and 53.1%.[5]

Cardiac tumors present with a variety of symptoms and are best evaluated by echocardiogram, CTA, and MRI. Tissue diagnosis is necessary. Although PCL is rare, long-term survival after chemotherapy and rituximab is superior to other cardiac malignancies. If left untreated, prognosis is grim, with survival reported at less than a month. However, if treated, survival can be prolonged to 5 years.

Conclusion

Cardiac tumors present with a variety of symptoms and are best evaluated by echocardiogram, CTA, and MRI. Tissue diagnosis is necessary. Although PCL is rare, long-term survival after chemotherapy and rituximab is superior to other cardiac malignancies.

Conflict of Interest
None declared.

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