Abstract

Primary cardiac tumors are rare, present in roughly 0.05% of the population. Cardiac papillary fibroelastoma (CPF) is the second most common, accounting for 10% of primary cardiac tumors. Most cases of CPFs are discovered incidentally on autopsy; however, they may present clinically with systemic embolization or heart failure symptoms. The recommended treatment for symptomatic CPF patients is surgical resection. Treatment in asymptomatic patients remains somewhat controversial with incidentally discovered tumors presenting a clinical dilemma. We present a case of an atypically located CPF that was discovered incidentally on intraoperative transesophageal echocardiography (TEE) during a routine coronary artery bypass graft operation. This case highlights several important points for cardiac anesthesiologists. The first is the importance of performing a comprehensive intraoperative TEE. Next, this case reinforces the broad utility of TEE for evaluation of intracardiac tumors. Finally, this case demonstrates the importance of precise localization of intracardiac tumors.

Keywords: Cardiac papillary fibroelastoma, cardiac tumor, incidental finding, intraoperative transesophageal echocardiography

Introduction

Primary cardiac tumors are rare, present in roughly 0.05% of the population on autopsy studies. Cardiac papillary fibroelastoma (CPF) is the second most common, accounting for 10% of primary cardiac tumors. Typically, CPFs are valvular in their attachment with a predominance for the left side of the heart. Most cases of CPFs are discovered incidentally on autopsy; however, clinical presentation includes systemic embolization such as transient ischemic attack or stroke, angina or myocardial infarction, sudden cardiac death, pulmonary embolism, or heart failure symptoms if affecting left atrial filling or function. The recommended treatment for symptomatic CPF patients is surgical resection. The treatment for CPF in asymptomatic patients is controversial. Because tumor mobility is an independent predictor of tumor-related embolization and morbidity, some authors suggest using this as a metric to guide management.

We present a case of an atypically located papillary fibroelastoma that was discovered incidentally on intraoperative transesophageal echocardiography (TEE) during a routine coronary artery bypass graft (CABG) operation.

Case Report

A 77-year-old man with a medical history of coronary artery disease, mild aortic stenosis, diabetes mellitus type 2, hypertension, and hyperlipidemia presented with symptoms of unstable angina. Cardiac catheterization revealed three-vessel coronary artery disease, and he was scheduled for a CABG. A preoperative transthoracic echocardiogram demonstrated a left ventricular ejection fraction of 47% with global hypokinesis and mild aortic stenosis with a mean gradient of 20 mmHg. The left atrium was mildly dilated but without echogenicity or irregularity. No obvious mass was identified in the left atrium.

The patient was brought to the operating room for CABG. Before the start of the procedure, TEE confirmed mildly reduced left ventricular systolic function and mild aortic stenosis. Imaging also revealed a 12 mm × 6 mm mass in the left atrium, attached to the ridge of epicardium dividing the left ventricle and the right ventricle.

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upper pulmonary vein and the left atrial appendage [Figure 1]. The mass was small, highly mobile with a homogenous central core and filamentous projections. Differential diagnosis included thrombus, benign cardiac tumor, or other malignancy such as a sarcoma. Suspicion was particularly high for an atrial myxoma due to the location, mobile nature of the mass, and somewhat globular appearance. However, the mass was much smaller in size than a typical myxoma and demonstrated peripheral filamentous elements more consistent with either thrombus or papillary fibroelastoma. Due to the potential for malignancy or embolization, the decision was made to resect the tumor. Given the precise echocardiographic localization of the mass to the left atrial appendage, the mass was approached directly through the left atrial appendage. The mass identified on TEE was easily visualized at the junction of the left atrial appendage and left atrium proper, and easily excised in its entirety, revealing an irregular tan colored, soft, and gelatinous mass with focal hemorrhage [Figure 2]. Attention was then turned to the coronary artery bypass component of the operation, which proceeded as planned. His postbypass TEE demonstrated complete excision of the mass, and postoperative course was unremarkable. Final pathology of the left atrial mass revealed fibroelastoma [Figure 3].

**DISCUSSION**

CPF are rare, benign, primary cardiac tumors. CPF arise from endocardium and are affixed to cardiac valves in approximately 80%–90% of cases. CPF demonstrates a predilection for the left side of the heart, with the most common location being the aortic valve (35%) followed by the mitral valve (30%). The incidence of CPF in the left atrium but not attached to valvular tissue is not well defined.

CPF is generally smaller than myxomas. They tend to be singular in nature and are often associated with other cardiac lesions, such as valvular disease, HOCM, or congenital heart disease. Echocardiographically, CPF may be round or irregular with a characteristic shimmering along the edges. CPF often have a short pedicle, and the distal portions may be highly mobile. While the diagnosis of CPF has increased substantially with improved echocardiographic modalities, the small size of these tumors may evade echocardiographic detection, particularly when utilizing TTE. A 16-year analysis of pathologically confirmed CPF encountered at the Cleveland Clinic reported an overall sensitivity of TTE at 62%, with TEE demonstrating a 77% sensitivity. TEE may also be superior in delineating the attachment site and the extent of tumor involvement.

While CPF is considered benign, they may result in severe morbidity and mortality related to tumor embolization. Clinical presentation for left-sided lesions includes stroke, transient ischemic attacks, myocardial infarction, retinal artery occlusion, or renal infarction. Right-sided lesions may result in pulmonary complications. Management of CPFs may vary based on the presence or absence of symptoms, size and location of tumor, and various patient factors, most notably, surgical risk. It is generally agreed that symptomatic patients should undergo surgical resection. Systemic antiplatelet agents or anticoagulation may be a viable option for patients in whom surgery is contraindicated. Asymptomatic patients with large (>10 mm) or mobile tumors should also be considered for surgery, as mobility is an...
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independent risk factor for embolization.[1] Right-sided tumors rarely require surgical intervention.

This case highlights several points for cardiac anesthesiologists. The first is the importance of performing a comprehensive intraoperative TEE exam. A 15-year retrospective study of intraoperative TEE revealed that new findings on pre-CPB exam influenced surgical decision in 7% of cases.[9] Next, this case reinforces the broad utility of TEE for evaluation of intracardiac tumors. Direct comparison of transthoracic and TEE has shown TEE to be superior when evaluating left-sided intracardiac lesions. Finally, this case demonstrates the importance of the precise description of intracardiac tumors, as the atypical location of the CPF in our case altered the surgical approach.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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