INTRODUCTION

Primary cardiac tumors are rare neoplasms. Their incidence on the basis of autopsy findings is 2:10,000.1 Cardiac papillary fibroelastomas (CPF) account for <10% of primary cardiac tumors, but CPF is the most common primary tumor of the cardiac valves.2,3 All valves can be affected, including the chordae tendineae, and the most common location is the aortic valve.4 CPFs are usually diagnosed randomly during echocardiography or in relation to workup of a cardioembolic source of stroke.

Here, we present two atypical and rare manifestations of CPF leading to cardiac surgery.

CASE PRESENTATION

Informed consent was obtained from both patients in the presented cases.

Case 1

A 66-year-old woman with a medical history of hypertension and hypercholesterolemia presented to the cardiac clinic with dyspnea on exertion. Results of physical examination, laboratory testing, and electrocardiography were unremarkable, and initial transthoracic echocardiography (TTE) revealed a mobile 1.1 × 1.3 cm mass in relation to the pulmonary valve (Figure 1, Video 1). Further evaluation revealed mild regurgitation and no signs of forward flow obstruction, no tricuspid regurgitation, normal vena cava inferior dimension with full collapse during inspiration, and normal right ventricular systolic function. A ventilation/perfusion lung scan was subsequently performed, demonstrating subsegmental perfusion defects in both lungs, indicating pulmonary embolisms (Figure 2). Anticoagulation was initiated, and the patient was referred for surgery to prevent further embolization. During the procedure, the mass was removed without damaging the valve and without progression of the existing mild pulmonary valve regurgitation. The postoperative course was uneventful. Histopathology of the mass revealed papillary fronds composed of central avascular collagen and variable elastic tissue lined by endothelial cells, all in accordance with CPF. At 6-month follow-up, symptoms were absent, a control ventilation/perfusion lung scan showed resolution of previous perfusion defects, and anticoagulation was discontinued.

Case 2

A 77-year-old woman with a medical history of hypertension was referred to her local hospital because of dyspnea in relation to physical activity. Results of physical examination, laboratory testing, and electrocardiography were all normal, and initial TTE revealed normal biventricular systolic function and no significant valvular pathology. Overall, there was no evidence of myocardial ischemia. Because of clinical symptoms, cardiac computed tomography was performed without signs of coronary artery disease. However, a mass was found on the noncoronary cusp of the aortic valve (Figure 3). At our institution, transesophageal echocardiography (TEE) demonstrated a 1.0 × 1.7 cm mobile mass on the aortic side of the aortic valve on the noncoronary cusp, causing obstruction of the ascending aorta and coverage of the right coronary ostium during diastole. Cross-sectional area measurement by three-dimensional image analysis of the ascending aorta at the level of the mass was estimated to be 3.4 cm², and the mass itself measured 1.9 cm² (Figures 4 and 5, Videos 2–4). Mild aortic valve sclerosis was noted, without aortic regurgitation. However, because of symptoms and the risk for embolization, the patient was referred for surgical removal of the mass. During cardiac surgery, the mass was removed, but because of the extent of the tumor’s attachment to the cusp, aortic valve replacement was performed. Histopathology of the mass revealed classical findings in accordance with papillary fibroelastoma. The postoperative course was uneventful, and at 3-month follow-up, the patient was asymptomatic.

DISCUSSION

Case 1 constitutes a rare representation of CPF; only 8% of these tumors are located in relation to the pulmonary valve.1 TTE or TEE is an obvious tool for initial evaluation, and recently three-dimensional TTE and TEE, as well as cardiac computed tomography, have become available for more detailed assessment. CPF is generally a benign condition, and patients may be asymptomatic.6 However, systemic arterial embolization may cause serious complications such as stroke and myocardial infarction, but as in case 1, pulmonary embolism might also occur. In general, surgical treatment should be considered in patients with tumor-related relevant symptoms but also in asymptomatic patients when large mobile tumors (>1 cm) are detected.7 Structural damage to the valves or broad-based adhesion of the tumor to leaflets may require valve

From the Department of Cardiology, Aarhus University Hospital, Aarhus, Denmark.

Keywords: Echocardiography, Fibroelastoma, Pulmonary valve

Conflict of interest: The authors have no potential conflicts of interest relative to this document.

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2468-6441
https://doi.org/10.1016/j.case.2018.02.007
repair or in some cases replacement. However, if possible, excision without repair or replacement should be pursued. If surgery is not indicated or the patient is not a surgical candidate, anticoagulation should be considered in relation to the specific case, and involvement of the pulmonary and tricuspid valves should trigger investigation for pulmonary embolism. No randomized controlled data are available on anticoagulation efficacy in relation to CPF. However, if the patient is symptomatic and a pulmonary embolism is present, anticoagulation should be initiated. Tumor recurrence is very low for CPF, and follow-up should be guided only by clinical indication.

CONCLUSIONS

Common to both cases was presentation with dyspnea, not a typical symptom of CPF. Thus, thorough evaluation of the right-sided heart valves should always be considered in patients with unexplained dyspnea. Also, the clinician should always attempt to demonstrate...
the pathophysiologic explanation for the symptoms during echocardiography. This may easily be forgotten when a rare condition is present.

**SUPPLEMENTARY DATA**

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2018.02.007.

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**Figure 3** (Top) Long-axis computed tomographic images showing the location of the mass in the ascending aorta (AA). (Bottom) Corresponding short-axis computed tomographic images. AV, Aortic valve; CPF, cardiac papillary fibroelastoma; RCA, right coronary artery.

**Figure 4** (Top) Two-dimensional transesophageal echocardiographic still frame of a midesophageal short-axis view during diastole illustrating a large mass in the ascending aorta (AA) with close relation to the right coronary artery (RCA). (Middle) Biplane transesophageal echocardiographic still frame illustrating the mass to originate from the noncoronary cusp of the aortic valve. (Bottom) Combined biplane and three-dimensional transesophageal echocardiographic view of the mass in the ascending aorta obstructing a large area of the lumen. CPF, Cardiac papillary fibroelastoma; LA, Left atrium; LV, left ventricle; RA, right atrium; RVOT, right ventricular outflow tract. For additional details and live images, see Videos 2 to 4.
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Figure 5 (Top) Color flow Doppler images of the left ventricular outflow tract (LVOT) and ascending aorta. (Bottom) Continuous-wave Doppler flow velocity profile from the LVOT and ascending aorta. CPF, Cardiac papillary fibroelastoma.