Two Rare Manifestations of Cardiac Papillary Fibroelastoma Presenting with Dyspnea

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INTRODUCTION

Primary cardiac tumors are rare neoplasms. Their incidence on the basis of autopsy findings is 2:10,000.[1] Cardiac papillary fibroelastomas (CPFs) 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 cm^2, and the mass itself measured 1.9 cm^2 (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.[5] 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

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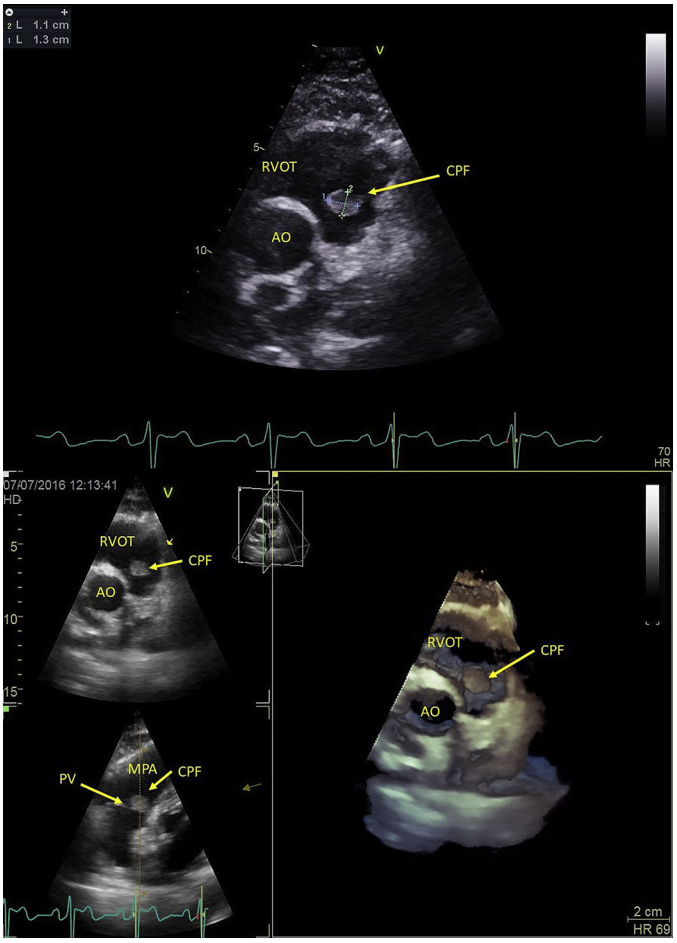

Figure 1 (*Top*) Two-dimensional transthoracic echocardiographic still frame of a modified parasternal short-axis view illustrating a 1.1 × 1.3 cm solitary mass in relation to the pulmonic valve. Histopathology revealed a cardiac papillary fibroelastoma (CPF). (*Bottom left*) Biplane transthoracic echocardiographic still frames illustrating the location of the mass on the pulmonary side of the pulmonary valve (PV). (*Bottom right*) Three-dimensional transthoracic echocardiographic still frame of the mass in the main pulmonary artery (MPA). *AO*, Aorta; *RVOT*, right ventricular outflow tract. For additional details and live images, see Video 1.

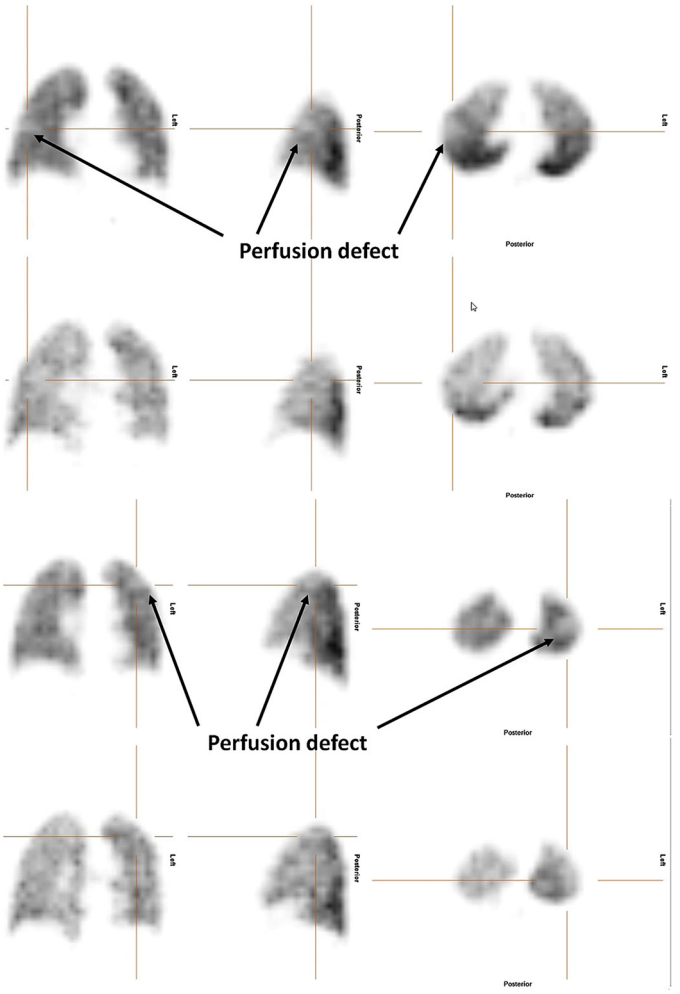

Figure 2 (*Top*) Pulmonary ventilation/perfusion scan showing perfusion defect in the middle lobe of the right lung and concomitant normal ventilation. (*Bottom*) Pulmonary ventilation/perfusion scan showing perfusion defect in the upper lobe of the left lung and concomitant normal ventilation.

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.[8]

Case 2 also presents a very rare manifestation of CPF. The association of CPF with coronary embolism is well documented.[9] However, physical obstruction of the right coronary ostium is reported in only a few previous cases.[10] During two-dimensional TTE, the mass was missed because of suboptimal imaging. However, TEE was suggestive of obstruction of the right coronary artery ostium during diastole. In addition, three-dimensional TEE proved valuable in demonstrating the tumor mimicking a supravalvular aortic stenosis as another possible underlying mechanism of the symptoms. Planimetric analysis of the images allowed quantification of a 56% stenosis.

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