

# Cardiac Papillary Fibroelastoma: Pulmonic Valve Involvement with Pulmonary Embolism and Pulmonary Hypertension

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Case Report

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# Abstract

Pulmonic valve papillary fibroelastomas are rarely seen tumors of the cardiac valves. In this case, a pulmonic valve mass was seen on echocardiogram, which was diagnosed as papillary fibroelastoma due to characteristic imaging findings. Complications of pulmonary embolism and pulmonary hypertension were present. Due to contraindications, the patient was managed with anticoagulation instead of surgery.

## Background

Primary cardiac tumors are rare. Their incidence has been estimated to be between 0.00017 and 0.033% at autopsy and 0.019% on echocardiography [1]. Tumors typically seen include myxomas, papillary fibroelastomas (PFE), rhabdomyomas, and fibromas. Papillary fibroelastomas are benign tumors derived from endothelial cells. They are small, pedunculated avascular tumors. 95% of these are found in the left-sided chambers [2]. They account for three-fourths of all cardiac valve tumors [3]. It most commonly affects the aortic valve (35%), followed by the mitral valve (29%) and then tricuspid and pulmonic valve (10% each) [4]. These tumors were previously found postmortem, however, with better imaging modalities their premorbid detection is increasing.

## Case

We present a case of an 83-year-old female who was found to have an incidental pulmonic valve mass with characteristic findings of papillary fibroelastoma seen on echocardiogram. The patient had a history of ischemic cardiomyopathy with an ejection fraction of 15%, atrial fibrillation, sick sinus syndrome with a biventricular implantable cardioverter defibrillator, pulmonary embolism, pulmonary hypertension, hypertension, and obstructive sleep apnea. Medications included aspirin, amiodarone, warfarin, furosemide, carvedilol and losartan.

Echocardiogram showed a well-circumscribed, pedunculated mass attached to the ventricular aspect of the pulmonic valve leaflet which measured 1.4x1.5 cm and prolapsed back-and-forth

in the right ventricular outflow tract without evidence of obstruction (Fig. 1). There was minimal pulmonic regurgitation and moderate pulmonary hypertension. CT scan with contrast also showed the mass in the right ventricle (Fig. 2). The patient's pulmonary embolism is suspected to have been a complication of the mass. Given the patient's multiple comorbidities, advanced age, and low ejection fraction, it was decided not to surgically intervene and she was maintained on anticoagulation. She was followed with echocardiographic imaging for over 5 years due to multiple hospitalizations secondary to congestive heart failure exacerbation. During this time, the pulmonic valve mass remained stable in size. She ultimately passed away after a complicated hospital course with septic shock.

## Discussion

Echocardiographic imaging is the initial imaging of choice and has a high sensitivity, specificity, and overall accuracy (88.9%, 87.8%, and 88.4%, respectively) [5]. Other diagnostic modalities include cardiac CT and MRI. MRI can be beneficial in determining vascularity [6]. Live 3D

echocardiograms can also be used which help provide additional information regarding static and dynamic spatial location of the tumor and also in discerning neighboring structures [7]. Transesophageal echocardiogram should always be performed before surgical intervention for guiding surgical approach [5]. Some of the characteristic echocardiographic findings of PFEs include small size (usually 1.5cm), mobile with a short pedicle or stalk attachment. They are well-circumscribed, homogenous, and sometimes have a speckled interior with stippling near the edges, typically originating from the valvular surface [1, 4, 5]. Compared to PFEs, myxomas are primarily located in the atrium (75% left atrium and 15 – 10% right atrium), are larger, have a heterogeneous appearance, and are attached by a longer stalk to the interatrial septum [8].

Grossly upon resection, the tumor has a sea anemone-like appearance with multiple papillary fronds. Histopathology shows that they are covered by endothelium with a mucopolysaccharide, and vascular core of connective fiber, which may contain variable content of collagen, smooth muscle, and elastic fiber [9].

If left untreated, the complications commonly include embolization. If left-sided, they can cause strokes, and if right-sided, as in our case, they can result in pulmonary embolism and pulmonary hypertension. Coronary ostial occlusion may be seen in larger-sized or highly mobile masses.

Acute valvular dysfunction may also be seen however, valvular insufficiencies are not commonly seen [5].

Definite management is not agreed upon; surgery is recommended for all patients with prior embolic complications and may be offered to asymptomatic patients

given the risk of embolization. Recurrence after surgery has not been reported. If a patient has a high surgical risk, especially elderly patients, anticoagulation should be considered.

## **Conclusion**

This case demonstrated a patient with characteristic imaging findings of pulmonic valve PFE with complications including pulmonary embolism and pulmonary hypertension. Though surgery is recommended in these patients, our patient had multiple comorbidities and was maintained on anticoagulation.

## **Declarations**

## Statements and Declarations

The Authors declare that there is no conflict of interest. The authors declare that no funds, grants, or other support were received during the preparation of this manuscript.

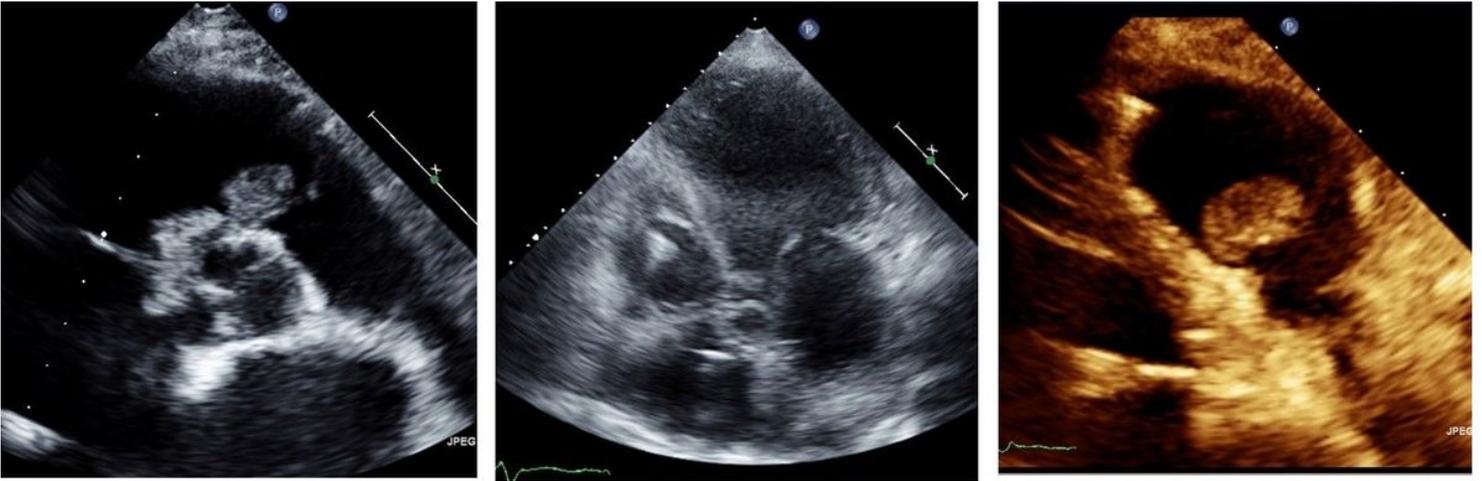
## Informed consent

Informed consent was obtained from the patient for publication of this case report.

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## Figures



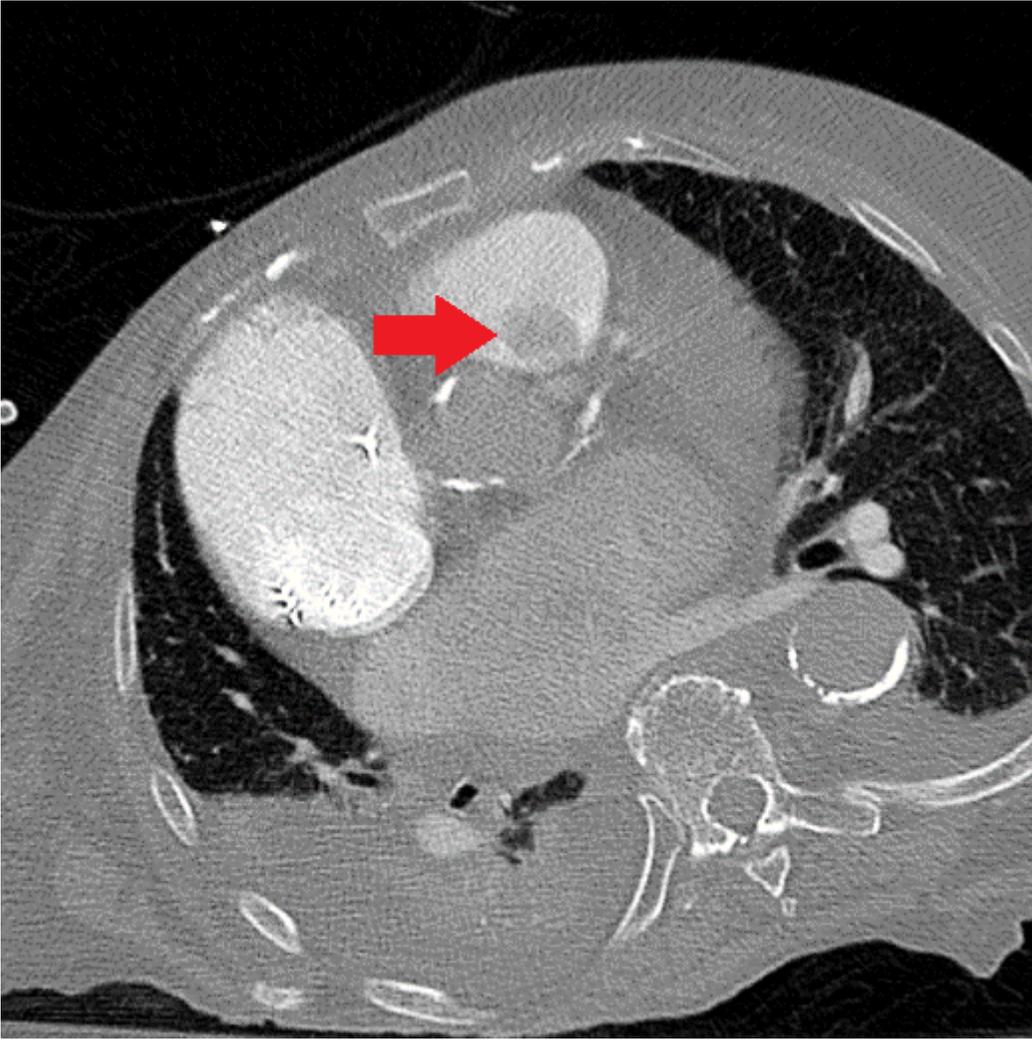
**Figure 1**

Transthoracic echocardiogram showing papillary fibroelastoma

a: A pedunculated mass attached to the pulmonic valve is seen

b: Mass can be seen in the right atrium

c: Well-circumscribed and homogenous mass visible



**Figure 2**

Computed tomography scan showing papillary fibroelastoma in the right ventricle