Rare Cardiac Papillary Fibroelastoma: Right Atrial, Non-Valvular, Large, Symptomatic With Pulmonary Embolism

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Case Report

A 59-year-old female with a history of hypertension, hyperlipidemia, chronic obstructive pulmonary disease, and anxiety developed sudden-onset shortness of breath. Upon arrival of emergency medical service, the patient was unresponsive and in asystole. Cardiopulmonary resuscitation was immediately started. After 4 rounds of compression, the patient achieved return of spontaneous circulation. In the emergency room, she was intubated for airway protection. Otherwise, a physical exam revealed stable vitals and bilateral wheezes with diminished breath sounds. Initial laboratory investigations showed uncompensated respiratory acidosis, hyperkalemia of 6.2 mEq/L, and an elevated high sensitivity troponin of 263 pg/mL (Table 1). A head computed tomography (CT) was obtained and was negative for any acute abnormality, including hemorrhage. Chest radiograph demonstrated cardiomegaly, central vascular prominence, and absence of lung nodules. A CT angiogram of the chest noted an acute right pulmonary embolism (PE) and a right atrial (RA) filling defect. Furthermore, an echocardiogram illustrated a left ventricular ejection fraction of 55% to 60% and a large (32 × 26 mm), pedunculated, mobile, and non-valvular echodensity that was attached to the RA endocardium (Figure 1).

In the emergency room, the patient was started on heparin infusion and then catheter-directed thrombolysis, but the mass was still seen on repeat echocardiogram. Consequently, she underwent a surgical RA excision and an atrial septum repair with cardiopulmonary bypass. A 40 mm mass was removed, and pathology demonstrated acellular tissue, papillary configuration, and elastic stain positive (Figure 2). The patient was safely extubated to a nasal cannula on day 2 of her hospital stay and discharged home on room air on day 7. She was instructed to take rivaroxaban for hemorrhage.

Case Narrative

ABSTRACT

Introduction: Primary cardiac tumors are rarely seen in the general population and only a subset are classified as cardiac papillary fibroelastoma.

Case Presentation: A 59-year-old female that presented for unresponsiveness and cardiac arrest required 4 rounds of cardiopulmonary resuscitation and intubation. Laboratory investigations showed uncompensated respiratory acidosis, hyperkalemia, and elevated troponins. A chest computed tomography angiogram illustrated an acute right pulmonary embolism and a right atrial filling defect. Furthermore, an echocardiogram demonstrated a normal ejection fraction and a large, pedunculated, mobile, and non-valvular echodensity that was attached to the right atrium endocardium. Therefore, the patient was started on a heparin infusion and catheter-directed thrombolysis; however, the mass persisted. A surgical excision was performed, and a 40 mm was removed. The patient was diagnosed with a papillary fibroelastoma based on the clinical symptoms, imaging, and histological findings.

Conclusion: This patient’s papillary fibroelastoma had multiple rare features including right atrial origin, large size, non-valvular location, and developed symptoms. Although this disease can be initially fatal, the patients typically have a favorable prognosis after a successful excision.

INTRODUCTION

Autopsy reports have shown that cardiac tumors are present in 0.001% to 0.28% of cases. However, these tumors are diverse in etiology, consisting of metastatic, primary, and those that originate from infradiaphragmatic organs. Primary cardiac tumors are rare and are further classified as either myxoma, papillary fibroelastoma (PFE), or lipoma.2–3 PFEs are small, benign, and have a predilection for involving aortic and mitral valves.2–5 Echocardiogram findings that are consistent with PFEs are a speckled interior, peripheral stippling, pedunculated, echo-luent areas, and mobility or vibration in the blood–tumor interface. PFEs are grossly characterized as multiple papillary fronds and similar to a sea anemone after being submerged in water. Histologically, they originate from endothelial cells and are avascular due to elastin and immature collagen fibers.2–6–8 This case report was prepared following the CARE guidelines.9

Keywords: papillary fibroelastoma, right atrial, non-valvular, symptomatic, pulmonary embolism

Abbreviations: CT = computed tomography; PE = pulmonary embolism; PFE = papillary fibroelastoma; RA = right atrial

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6 months and colchicine for 1 month. At her routine 1 month cardiology follow-up, the patient denied any acute symptoms or complaints. Additionally, a repeat echocardiogram did not show recurrence of the RA mass.

Due to the sudden onset of shortness of breath and CT findings, the initial list of differentials included PE, organizing thrombus, acute anxiety attack, and acute exacerbation of chronic obstructive pulmonary disease. Due to the RA filling defect in adjunct with the aforementioned symptoms and images, the following were also considered: myxoma, PFE, lipoma, rhabdomyoma, fibroma, and amorphous tumors. The patient was eventually diagnosed with a PFE based on the cardiac arrest presentation, acute PE, mass on echocardiogram, histological stains of papillary configuration, and the absence of lung nodules.

**DISCUSSION**

This case report depicts several features that are rarely seen in patients with PFEs. Firstly, the PFE was located in the right atrium and did not involve the valves. Data, however, suggest that PFEs are commonly in the left heart and involve a valve with a prevalence of 95% and 60% to 93%, respectively.2,6 A large PFE is defined as larger than 10 mm, but typical sizes range from 2 mm to 40 mm.1,3 This patient’s echocardiogram showed a 32 mm mass, but was found to be 40 mm grossly. PFEs are benign endocardial papillomas and are usually asymptomatic, especially when located in the right side of the heart.6 However, 30.8% of patients develop symptoms, which can be stroke, PE, recurrent pulmonary edema, pulmonary hypertension, cardiac obstruction, and heart failure.5,6 Right-sided PFEs are typically asymptomatic until they progress to form a large enough thrombus or embolus that obstructs blood flow.10 There is a 34% incidence rate of thromboembolism in patients with PFEs, compared to only 24% with myxomas.6 Moreover, right-sided PFEs present as PEs in only 0.4% of cases.7

This patient also confirms that the pathophysiology behind PFEs is unknown, as she did not have any significant
history, but still developed a severe, symptomatic mass. PFEs have been identified on the endocardial surfaces, ventricular septum, papillary muscles, and chordae tendineae.\(^2,6,11\) The embolic nature is thought to be a consequence of the mobility, tumor disruption, and embolization of fibrin fragments. Their mobility is a result of their pedunculated nature. PFEs may be described as organizing thrombi: the fronds contain fibrin, hyaluronic acid, and laminated elastic fibers.\(^10\)

Commonly, PFEs are diagnosed by a transthoracic echocardiogram, as it has a high sensitivity, specificity, and overall accuracy (88.9%, 87.8%, and 88.4%, respectively).\(^6\) Alternative forms of detection are CT angiography and magnetic resonance imaging.\(^7\) Compared to PFEs, myxomas can be histologically distinguished by the presence of blood vessels and muscle-specific actin. Rhabdomyoma and fibroma are differentiated from other tumors by their gross appearance.\(^6\) The majority of the cases are treated with surgical resection during a cardiopulmonary bypass, but clear indications for surgery have not been defined in asymptomatic patients.\(^6,7\) The rate of recurrence of a PFE mass after successful excision is 1.6%.\(^3\) For non-surgical candidates, clinicians can consider a long-term course of antiplatelets.\(^3\)

Initially, colchicine was widely used in the 19th century to treat gout and in the 20th century for familial Mediterranean fever, acute pericarditis, and Behcet disease.\(^12,13\) Due to its anti-inflammatory effects, it is now known that 1 month of colchicine significantly prevents the incidence of constrictive physiology and atrial fibrillation in patients who recently underwent a cardiac surgery, including cardiopulmonary bypass.\(^14,15\) Therefore, our patient was prophylactically given a month’s supply of colchicine on discharge.

**CONCLUSION**

This case demonstrates the rare presentation of a cardiac PFE, as it was RA, non-valvular, large, and symptomatic. Therefore, it is important to know that cardiac PFE can contribute to life-threatening PE and subsequent cardiac arrest. With suspicious imaging findings, therapeutic surgical excision should be considered as patients can have a favorable prognosis.

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**Authors’ Contributions**

Rohan M Prasad wrote the case narrative, conducted the literature review, and edited and revised the final draft. Abdul-Fatawu Osman wrote the introduction and abstract, conducted the literature review, and edited and helped with revisions of final draft. Christopher C Garces wrote the Discussion, edited and helped with revisions of the final draft, and obtained consent. Robert Gumbita wrote the Conclusion, conducted the literature review, and edited the final draft. Ahmed Elshafee assisted in writing the
Introduction, Abstract, and Conclusion; completed the work cited, and edited final draft. Pranay Pandrangi assisted in writing the Case Narrative and Discussion, obtained the figures, and edited the final draft. Michael Kehdi assisted in all phases of writing and edited the final draft.

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