

Received: 2020.01.25

Accepted: 2020.04.24

Available online: 2020.05.18

Published: 2020.06.16

Papillary Fibroelastoma of the Right Ventricle in the Setting of Multifocal Pulmonary Hemorrhages and Pulmonary Embolization in a 39-Year-Old Man: A Case Report

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

EF **Shahnaz Duymun**
E **Asad Nasir**

Department of Medicine, Arnot Ogden Medical Center, Elmira, NY, U.S.A.

Corresponding Author: Shahnaz Duymun, e-mail: sduymun23@gmail.com
Conflict of interest: None declared

Patient: Male, 39-year-old
Final Diagnosis: Papillary fibroelastoma
Symptoms: Chest pain • fever • hemoptysis • weight loss
Medication: —
Clinical Procedure: —
Specialty: Cardiac Surgery • Cardiology • Critical Care Medicine • Pulmonology

Objective: Rare disease

Background: Papillary fibroelastoma accounts for approximately 24% of all primary cardiac tumors and is the third most common primary cardiac neoplasm. It mostly involves the cardiac valves; however, there have been cases of involvement of the endocardium. Papillary fibroelastoma, although typically benign, can cause a plethora of complications, including systemic embolization, myocardial infarction, arrhythmia, and sudden cardiac death. In this article we present a unique case of a man diagnosed with a papillary fibroelastoma of the right ventricle in the setting of multifocal pulmonary hemorrhages and pulmonary embolization.

Case Report: A 39-year-old man presented with hemoptysis and dyspnea due to bilateral lower-lobe pneumonia and a left pleural effusion. Further imaging throughout his hospitalization revealed worsening multifocal consolidations, pulmonary hemorrhages, and a pulmonary embolus. A mobile cardiac mass measuring 30.2×20 mm, detected by echocardiography, was found in the right ventricle, partially fixed to the intraventricular septum via a 14.4-mm stalk, which was surgically excised and was found to be consistent with papillary fibroelastoma. He underwent an IVC filter and was discharged on warfarin, which he tolerated well.

Conclusions: Papillary fibroelastomas are benign infrequent findings but carry a high risk of systemic complications, as demonstrated in our patient. He had no known cardiac disease or risk factor for tumor growth, yet developed a papillary fibroelastoma of the right ventricle, which is rare. Given the potential of fatal outcomes, it is imperative that patients with unexplained embolic phenomena undergo early diagnosis by echocardiography and early surgical treatment.

MeSH Keywords: Cardiac Surgical Procedures • Heart Neoplasms • Pulmonary Embolism

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/923074>



1693



—



3



9



Background

Primary cardiac tumors are uncommon findings, with an incidence of only 0.02%, 10% of which are papillary fibroelastomas. Papillary fibroelastoma, known for its resemblance to a sea anemone, is the third most common primary cardiac tumor after myxomas and lipomas [1]. It is also the most common tumor of cardiac valves; primarily the aortic, followed by the mitral and then the pulmonic valves. They can also involve nonvalvular endocardium, mostly left-sided but also right-sided chambers [2–4]. Though benign, severe complications can arise due to the tumor's mobility, usually thrombi formation, which in turn can lead to myocardial infarction, heart failure, systemic embolization, or sudden cardiac death [3,4]. In light of these complications, treatment consists of complete tumor resection with close follow-up and serial echocardiography for several years after resection to evaluate for recurrence, although it is uncommon [2,3].

Case Report

A 39-year-old man with a history of tobacco and alcohol use, chronic obstructive pulmonary disease, hypertension, and gastric esophageal reflux disease presented to our facility with hemoptysis and dyspnea. He had been experiencing multiple upper-respiratory infections for the past 8 weeks prior to presentation, which had been progressively worsening, resulting in hemoptysis, with up to 30–40 cc production per day. The hemoptysis began 4 days prior to hospitalization, at which time he sought medical attention at his primary care physician's office, where he underwent routine lab work and imaging. He was noted to have an elevated white blood cell count, with imaging showing an evolving right lower-lobe opacity. He was prescribed levofloxacin; however, despite treatment, his dyspnea and hemoptysis worsened. Thus, he presented to our Emergency Department a few days later. He described fever, fatigue, and generalized weakness, as well as nausea, headache, and flushing. He also reported dyspnea, an unintentional weight loss of 9 pounds, and a band-like chest pain that worsened with coughing.

Upon admission he was noted to have decreased breath sounds of bilateral posterior lower lobes with increased dullness to percussion of the left lower lobe and crackles to the right middle lobe. CT chest angiography revealed a pulmonary embolus of the right lower lobe, as well as bilateral consolidation, left pleural effusion, and right middle-lobe atelectasis (Figure 1). He was started on a vancomycin, piperacillin-tazobactam, and a heparin infusion, and was admitted to the general telemetry floor. Due to inability to obtain therapeutic levels on heparin, he was switched to enoxaparin for anticoagulation. Within 24 h of admission, he developed worsening shortness

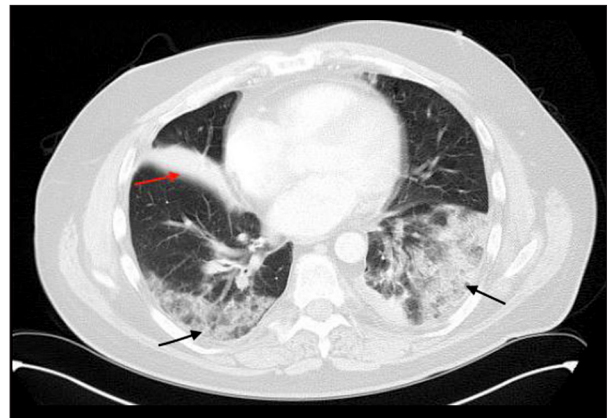


Figure 1. CTA chest with and without IV contrast, axial lung view, demonstrating bilateral lower-lobe consolidation (black arrows) and right middle-lobe atelectasis (red arrow).

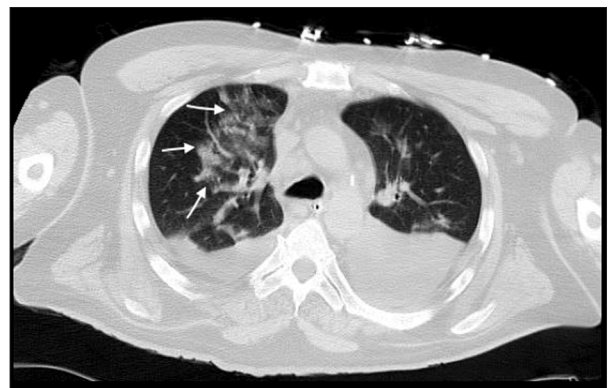


Figure 2. CT chest without IV contrast, axial lung view, suggesting pulmonary hemorrhages (white arrows).

of breath, increasing oxygen requirements, and tachycardia, and was subsequently transferred to the Intensive Care Unit.

He underwent a bronchoscopy with biopsy, which was negative for any acute pathology. Sputum cultures, respiratory panel, urine streptococcal antigen, and blood cultures were negative, thus lowering suspicion of an infectious etiology as the cause of his presentation. Hypercoagulable and vasculitis workups were negative. Repeat CT chest imaging revealed worsening multifocal consolidations and effusions, with concern for pulmonary hemorrhages given his ongoing hemoptysis (Figure 2). His respiratory status continued to worsen, prompting emergent intubation. He began to develop worsening anemia, so anticoagulation was held and an inferior vena cava filter was placed.

Transthoracic echocardiography was performed and revealed a highly mobile mass measuring 30.2×20 mm in the right ventricle, partially fixed to the intraventricular septum via a 14.4-mm stalk (Figure 3). Also noted on echocardiography were moderately elevated pulmonary artery pressure, a right-to-left atrial



Figure 3. Apical 4-chamber view of a transthoracic echocardiogram of our patient revealing papillary fibroelastoma in the right ventricle measuring 30.2×20 mm, delineated by green arrows.

shunt, tricuspid regurgitation, and severe hypokinesia of the right ventricle. He was evaluated by Cardiothoracic Surgery, who postulated that this right ventricular mass was the primary cause of his pulmonary infarcts. Therefore, he was transferred to a tertiary care facility for further evaluation and treatment. There, he underwent resection of the right ventricular mass with cryoablation, repair of his tricuspid valve, and pulmonary embolectomy. His post-operative course was complicated by acute kidney failure requiring continuous renal replacement therapy, and intermittent hemodialysis, as well as heparin-induced thrombocytopenia. He was initially treated with bivalirudin for heparin-induced thrombocytopenia and was eventually transitioned to warfarin for anticoagulation. More pertinently, the pathology of the excised right ventricular lesion was consistent with fibroelastoma. It is believed that the fibroelastoma was indeed the cause of his pulmonary embolus and hemorrhages.

Repeat echocardiography performed postoperatively did not show any evidence of a right heart mass, but there was noted to be mild right-heart dysfunction and mild evidence of a ventricular septal defect at the resection site, which was repaired. The patient remained hemodynamically stable postoperatively and was discharged to a short-term rehab facility and went home thereafter. He has since seen his cardiologist, cardiothoracic surgeon, pulmonologist, and primary care physician, and has been doing well. His IVC filter was removed and he completed a 6-month course of warfarin for his pulmonary embolus.

Discussion

Primary cardiac tumors are rare findings, with an incidence rate of only 0.02%. Papillary fibroelastomas are the third most

common primary cardiac tumor, with an incidence rate of 10%. These tumors consist of gelatinous material and a stalk with multiple papillary projections resembling a sea anemone when once immersed in water, which is its pathognomonic feature [1].

Papillary fibroelastomas characteristically grow on cardiac valves and are the most common cardiac valvular neoplasm. In an extensive study by Tamin et al., 511 cases of papillary fibroelastomas were reviewed, 78% of which involved cardiac valves, with the remaining 22% involving nonvalvular endocardium. Of that 22%, only 6% involved right-sided chambers [2]. A study by Ngaage et al. reviewed 88 cases of fibroelastomas, which also demonstrated predominant involvement of cardiac valves, followed by cardiac chambers; 52% of the fibroelastomas were attached to the aortic valve, followed by 16% on the mitral valve, and 28% in the cardiac chambers. Of that 28%, the majority of fibroelastomas occurring in the cardiac chambers occurred in the left ventricle, with only 3 cases found to involve the right ventricle [3]. Similarly, Gowda et al. reported valvular surfaces were the predominant location for these tumors [4]. Additionally, these tumors tend to gravitate towards areas of endocardial damage such as valves that have undergone repair, are damaged from degenerative and rheumatic processes, or are prolapsed [5]. Our patient had a fibroelastoma of his right ventricle involving endocardium that was previously healthy, which has only been seen in a few cases.

Papillary fibroelastomas are mostly asymptomatic and are usually diagnosed incidentally. Symptoms that do occur tend to be nonspecific, including dyspnea, syncope, angina, or signs of heart failure. Complications, however, arise with the embolization of the tumor or secondary thrombi formation, which occur in about 35% of reported cases. These systemic embolizations are known to cause myocardial infarctions, stroke, retinal artery embolization, and sudden death [3,4]. Papillary fibroelastomas of left-sided chambers have a high risk of embolic complications, such as neurological phenomena. Right-sided papillary fibroelastomas may remain asymptomatic; however, there have been a few reports of pulmonary emboli due to these right-sided tumors, such as in the present case [6].

The mainstay for diagnosing papillary fibroelastomas continues to be echocardiography. These tumors are visualized as round, oval, or irregular on echocardiography, with well-demarcated borders and occasional areas of lucency within the tumor [5,7]. Most are small, with the largest documented diameter being 5 cm [3]. Nearly half of papillary fibroelastomas are attached to the endocardium via a stalk, which makes these tumors mobile [7]. It is the mobility, rather than the size and location of the tumor, that leads to systemic complications such as pulmonary embolizations and stroke [3]. Limitations of traditional transthoracic echocardiography include masking of the tumor by another lesion, preexisting valvular damage, small

size of the tumor, and the technique of echocardiographic examination [7]. In these cases, transesophageal echocardiography or even 3D echocardiography may be needed for further evaluation.

The best treatment for these patients is clean and complete surgical excision of the tumor. Ngaage et al. demonstrated that simple shave excisions were adequate for most nonvalvular and valvular lesions [3]. Valve repair or replacement may be warranted if the lesion is extensive, if there is preexisting valvular disease, or if there is damage during the excision [8]. Prophylactic anticoagulation should be advised once the diagnosis of papillary fibroelastoma has been made, due to their tendency to embolize [9]. If left untreated, there is an increased risk of cerebral vascular events and death. Tamin et al. compared patients who underwent surgical excision of their tumor with those who did not. Those who did not undergo surgical resection had a 6% risk of stroke 1 year after diagnosis, with a mortality risk of 13%, compared with 2% stroke risk and mortality 1 year after surgical removal [2]. Due to these risks, close follow-up care and serial echocardiography after initial diagnosis are recommended and can help improve outcomes and monitoring for recurrence or further growth. Recurrence was seen in 1.6% of patients after surgical removal, arising at or near the site of resection, due to endocardial injury or due to prior incomplete excision [2]. Our patient underwent complete surgical excision of the lesion with valvular repair and was treated with anticoagulation, with excellent results. Follow-up echocardiography has not shown recurrence of any lesions, and his valvulopathy has improved.

References:

1. Darvishian F, Farmer P: Papillary fibroelastoma of the heart: Report of two cases and review of the literature. *Ann Clin Lab Sci*, 2001; 31: 291–96
2. Tamin SS, Maleszewski JJ, Scott CG et al: Prognostic and bioepidemiologic implications of papillary fibroelastomas. *J Am Coll Cardiol*, 2015; 65: 2420–29
3. Ngaage D, Mullany CJ, Daly RC et al: Surgical treatment of cardiac papillary fibroelastoma: A single center experience with eighty-eight patients. *Ann Thorac Surg*, 2005; 80: 1712–18
4. Gowda RM, Khan IA, Nair CK et al: Cardiac papillary fibroelastoma: A comprehensive analysis of 725 cases. *Am Heart J*, 2003;146: 404–10
5. Klarich K, Enriquez-Sarano E, Gura G et al: Papillary fibroelastoma: Echocardiographic characteristics for diagnosis and pathologic correlation. *J Am Coll Cardiol*, 1997; 30: 784–90
6. Waltenberger J, Thelin S: Papillary Fibroelastoma as an unusual source of reported pulmonary embolism. *Circulation*, 1994; 89: 2433
7. Sun JP, Asher CR, Yang XS et al: Clinical and echocardiographic characteristics of papillary fibroelastomas. A retrospective and prospective study in 162 patients. *Circulation*, 2001; 103: 2687–93
8. Shaihan D, Labib SB, Chang G: Cardiac papillary fibroelastoma. *Ann Thorac Surg*, 1995; 59: 538–41
9. Georghiou GP, Eraz E, Vidne BA, Aravot D: Tricuspid valve papillary fibroelastoma: An unusual cause of intermittent dyspnea. *Eur J Cardiothorac Surg*, 2003; 23: 429–31

Conclusions

Papillary fibroelastomas are known to be benign infrequent findings but they carry a high risk of systemic complications, such as embolization, as demonstrated in our patient. These tumors primarily occur on cardiac valves, especially those that have been damaged from prior disease. After cardiac valves, fibroelastomas preferentially grow in the left ventricle followed by the right ventricle, with estimated occurrences of 3.4% of all documented cases. Our patient, with no known cardiac disease or risk factor for tumor growth, was found to have a papillary fibroelastoma of the right ventricle, making this case unusual.

Given the potential of fatal outcomes, it is imperative that patients with unexplained embolic phenomena undergo transthoracic echocardiography and, if necessary, transesophageal echocardiography, to exclude cardiac sources of embolization, especially tumors such as papillary fibroelastoma. Surgical resection should be offered to symptomatic patients, as well as asymptomatic patients who are at high risk of embolization. These tumors, though uncommon and mostly benign, pose great serious to patients; therefore, early diagnosis and treatment are vital in anyone with unexplained embolic phenomena.

Conflicts of interest

None.