Pulmonic Papillary Fibroelastoma: Cause of Recurrent Pulmonary Emboli and Treatment

Alexander E Urban¹, Masoor Kamalesh², Daniel Gutteridge³

1. University of Tennessee Health Science Center, Internal Medicine, Memphis, TN
2. Indiana University School of Medicine, Cardiology, Indianapolis, IN
3. Indiana University School of Medicine, Pulmonary Critical Care, Indianapolis, IN

Cardiac papillary fibroelastomas are rare, typically benign primary cardiac tumors. The pulmonary valve is the least common site for valvular papillary fibroelastomas. They commonly present with dyspnea on exertion and can show right ventricular outflow tract obstruction on echocardiography. Embolic phenomena are one of the most serious consequences. Treatment usually consists of surgical excision. We report the first case of pulmonary embolism from pulmonary valve papillary fibroelastoma treated with anticoagulation. This occurred in only the second case of pulmonary embolism from pulmonary papillary fibroelastoma reported in literature to date.

Key words: papillary fibroelastoma; pulmonary valve; pulmonary embolus; anticoagulation

Citation: Urban, A.E., Kamalesh M. and Gutteridge, D. Pulmonic Papillary Fibroelastoma: Cause of Recurrent Pulmonary Emboli and Treatment. International Cardiovascular Forum Journal. 2015;4:92-93. DOI: 10.17987/icfj.v4i0.161

A 74 year old white male presented with increasing shortness of breath of four days. He denied chest pain, orthopnea, paroxysmal nocturnal dyspnea, palpitations, fever, chills, change in weight, and edema. He quit smoking 20 years ago. He was diagnosed with severe acute respiratory distress syndrome, acute on chronic kidney injury, acute liver injury, and non-ST elevation myocardial infarction (NSTEMI).

Physical exam revealed an afibrile patient with heart rate 112, blood pressure 144/82 mmHg, respiratory rate 22, oxygen saturation 60% on room air, improving to 70% on 100% FiO₂ via CPAP mask. Arterial blood gas revealed pH 7.35, pCO₂ 34 mmHg, pO₂ 21 mmHg, and a bicarbonate level of 18.8 mmol/L. Lactic acid was 9.5 mmol/L. He continued to deteriorate and was intubated. Cardiac exam had a regular rhythm with a gallop noted but no murmurs or jugular venous distention. Peripheral pulses were 1+ bilaterally and extremities were warm. Scattered wheezes were heard throughout lung fields.

Leukocytosis of 16,420 cells/μL with elevated neutrophil count with 20% bandemia was seen on laboratory testing. Blood and sputum cultures were negative. BUN and creatinine were 19 and 2.1, respectively. Liver enzymes were also elevated with AST 463 and ALT 346. Troponin at time of admission was 0.98 ng/mL but trended up to 5.68 ng/mL. BNP was 405 pg/mL. D-dimer was elevated at 574 ng/mL. EKG showed lateral lead ST segment depression. Chest x-ray findings included multilobar interstitial opacities concerning for pneumonia.

Two days after admission, the patient was...
stable enough to travel to radiology for a computed tomography scan of the chest with contrast, which revealed diffuse ground glass opacities and bilateral subsegmental pulmonary emboli involving the right and left upper lobes. The right ventricular outlet at the area of the pulmonic valve showed a filling defect. Lower extremity venous ultrasound did not reveal evidence of deep vein or superficial thrombus. Transesophageal echocardiography found a single, mobile, filamentous, echogenic mass in the right ventricular outflow tract crossing the pulmonic valve, measuring about 5 cm with a stalk of approximately 10 mm (Figure 1).

The patient was started on heparin to prevent further thrombus formation on the surface of the tumor and embolization. However, he was diagnosed with heparin-induced thrombocytopenia when his platelet count decreased, so he was switched to argatroban. He remained asymptomatic during the remainder of his admission. He was discharged home on lifetime anticoagulation with warfarin and follow up was arranged to re-evaluate potential surgical needs when his functional status improved. At the two month follow up appointment, he remained asymptomatic without need for further intervention.

The most common presentation from papillary fibroelastoma of all locations are stroke or transient ischemic attack (19.6%), angina (8.0%), myocardial infarction (4.6%), heart failure (4.0%), sudden death (3.4%), presyncpe or syncpe (2.0%), blindness (1.1%), pulmonary embolism (0.5%), peripheral emboli (0.5%), mesenteric ischemia (0.3%), and renal infarction (0.2%)1. Of those involving the pulmonary valve, patients commonly present with dyspnea on exertion (34.6%), atypical chest pain (11.5%), and intermittent palpitations (7.6%). Other symptoms include cough, fever, transient blindness, syncpe, weakness, anorexia, and tingling and numbness of the hands3.

Characteristic findings on echocardiography for papillary fibroelastomas include: a) the tumor is round, oval, or irregular in appearance, with well-demarcated borders and a homogenous texture; b) most cardiac papillary fibroelastomas are small (99% were <20 mm in the largest dimension); c) nearly half had small stalks, and those with stalks were mobile; and d) they are usually single (91%) but may be multiple and are most often associated with cardiac valvular disease5. Hakim et al. found that transesophageal echocardiography detected 100% of cases involving the pulmonary valve while transthoracic echocardiography only detected 90%. The average size of pulmonary valve papillary fibroelastoma was 1.36 x 1.38 cm (range 0.1-2.8 cm)2. Most have a stalk 1-3 mm in length4.

This tumor is larger with longer stalk than average. Bacterial and Libman Sack’s endocarditis are less likely given negative blood cultures and anti-nuclear antibody. The presence of a round, mobile, pedunculated lesion attached to the leaflet of the pulmonary valve confirms the presence of tumor-like lesion instead of infection or debris6. Libman Sack’s endocarditis rarely presents on the right heart and has no stalk. Lamb’s excrescences are multiple in greater than 90% of cases5. Echocardiography findings and lack of support for other diagnoses make papillary fibroelastoma the most likely diagnosis. Consensus from literature review recommends surgical excision1-3,6,8, but anticoagulation was used given the patient’s poor functional status. Since papillary fibroelastomas have the potential to cause thrombus formation4, anticoagulation will reduce clot formation and embolization. To our knowledge, this is the first case in which a patient with pulmonary valve papillary fibroelastoma has been managed with anticoagulation alone. Two other case reports note successful management with anticoagulation over a four month interval for a left atrial appendage papillary fibroelastoma8 and a four year period for a left ventricular papillary fibroelastoma10.

Statement of ethical publishing
The authors agree to abide by the requirements of the “Statement of publishing ethics of the International Cardiovascular Forum Journal.11”

Conflict of interest:
The authors declare there is no conflict of interest.

Address for correspondence:
Alexander E. Urban, MD
University of Tennessee Health Science Center
E-mail: aurban@uthsc.edu

References