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Case Report

Pulmonic valve fibroelastoma—A rare incidental finding ☆☆☆

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ABSTRACT

Primary cardiac neoplasms are a rare, commonly benign, tumor with an approximate incidence rate of 0.02%. Papillary fibroelastoma (PFE), a common form of primary cardiac neoplasms, typically present as a mass on the aortic and mitral valves, while rarely presenting as a pulmonary valve tumor. The majority of PFEs are asymptomatic, however valvular masses can pose a significant health hazard due to their potential to fragment into the bloodstream, facilitate thrombus formation, and restrict blood flow. Due to these risks, careful resection of the mass is recommended for symptomatic patients and asymptomatic patients if the tumor is large (>1 cm), mobile, or on left-sided valves. Here we present a case of an incidental finding of a pulmonic valve papillary fibroelastoma in a 65-year-old man by transesophageal echocardiography during a coronary artery bypass graft procedure.

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Introduction

Primary cardiac neoplasms are a rare, commonly benign, tumor with an approximate incidence rate of 0.02% [1]. Papillary

fibroelastoma (PFE), a common form of primary cardiac neoplasms, typically present as a mass on the aortic and mitral valves, while rarely presenting as a pulmonary valve tumor. The majority of PFEs are asymptomatic; however, valvular masses can pose a significant health hazard due to their po-

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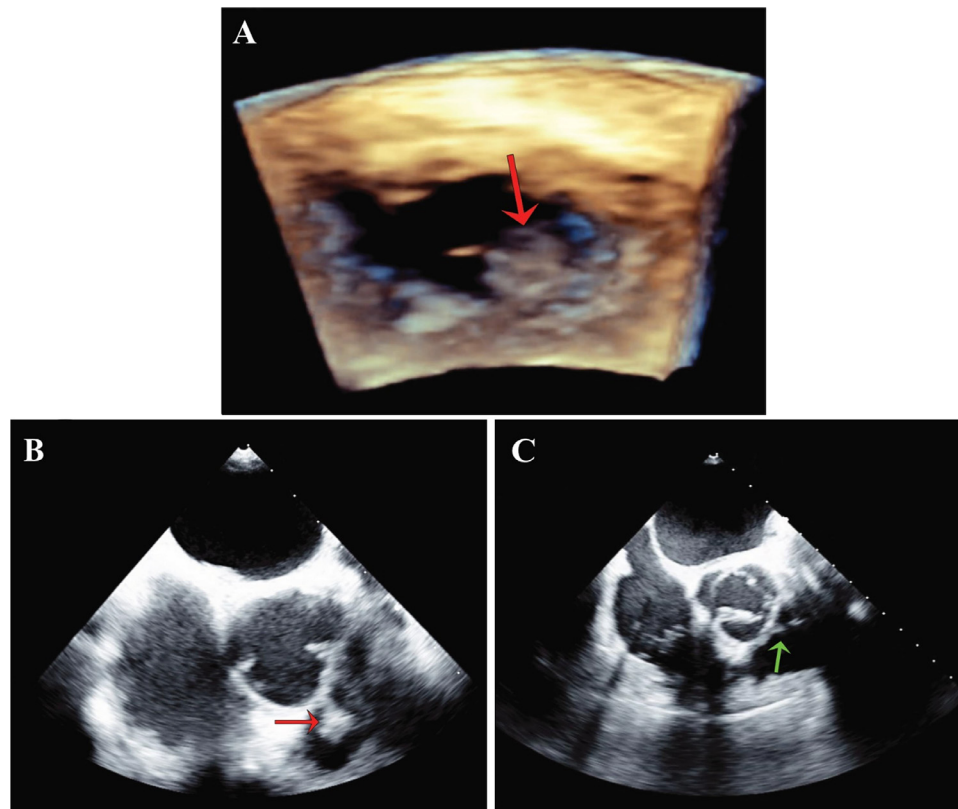


Fig. 1 – 3-D rendering (A) and 2-D image (B) of pulmonic valve papillary fibroelastoma (red arrows) as obtained by TEE. Mass was resected (green arrow) and valve remained competent (C).

tential to fragment into the bloodstream, facilitate thrombus formation, and restrict blood flow [2]. Due to these risks, careful resection of the mass is recommended for symptomatic patients and asymptomatic patients if the tumor is large (>1 cm), mobile, or on left-sided valves [3,4]. Here we present a case of an incidental finding of a pulmonic valve papillary fibroelastoma in a 65-year-old male by transesophageal echocardiography (TEE) during a coronary artery bypass graft (CABG) procedure.

Case presentation

A 65-year-old male with a past medical history significant for coronary artery disease (CAD), supraventricular tachycardia (SVT), chronic obstructive pulmonary disease (COPD), peripheral vascular disease, hypertension, hyperlipidemia, and tobacco-use presented to the hospital for an elective CABG due to an abnormal stress test and coronary computed tomography angiography (CTA). Patient had a single-photon emission computerized tomography (SPECT) nuclear stress test in 2014 for angina evaluation and was found to have a small amount of septal ischemia. Following these findings, he had a left heart catheterization (LHC) and underwent a percutaneous coronary intervention (PCI) to the left anterior descending coronary artery (LAD). Transthoracic echocardiogram (TTE) at that time showed mild concentric left ventricular hypertro-

phy (LVH) with an ejection fraction (EF) of 55-60%, as well as trivial mitral, tricuspid, and aortic valve regurgitation. However, the pulmonic valve was not well visualized. In 2021, he was evaluated for shortness of breath with another TTE which showed mild LVH with an EF of 55%, mild LV diastolic dysfunction, and that the pulmonic valve was normal in appearance. In 2022, he started to have anginal equivalent chest pain and a subsequent stress test was abnormal. Coronary CTA was done and showed significant calcification of the left main, LAD, first diagonal, obtuse marginal (OM1), and right coronary arteries suggestive of significant stenosis. Total coronary artery calcium score was significantly elevated at 2445 Hounsfield units (HU) (reference range: < 300). He then underwent another LHC which showed 60%-70% stenotic lesion on the left main coronary artery and 70%-75% stenotic lesion of the Left Circumflex (LCx). Given the multivessel disease, he was admitted to the hospital for an elective CABG.

Upon admission, his vitals and physical exam were unremarkable with no murmurs. EKG showed normal sinus rhythm. His laboratory workup which included a CBC, CMP, INR, and PT/PTT were all unremarkable except a sodium level of 132 mmol/L (ref. range: 136-145 mmol/L) and a glucose level of 117 mg/dl (ref. range: 74-106 mg/dL) with an A1c of 5.1% (ref. range: < 5.7). Following admission, a pre-op transesophageal echocardiogram (TEE) showed a mass attached to the base of the right cusp of the pulmonic valve (Figs. 1A and B). The pulmonic valve had normal leaflet thickness with no evidence of valvular stenosis or regurgitation. In terms of differential

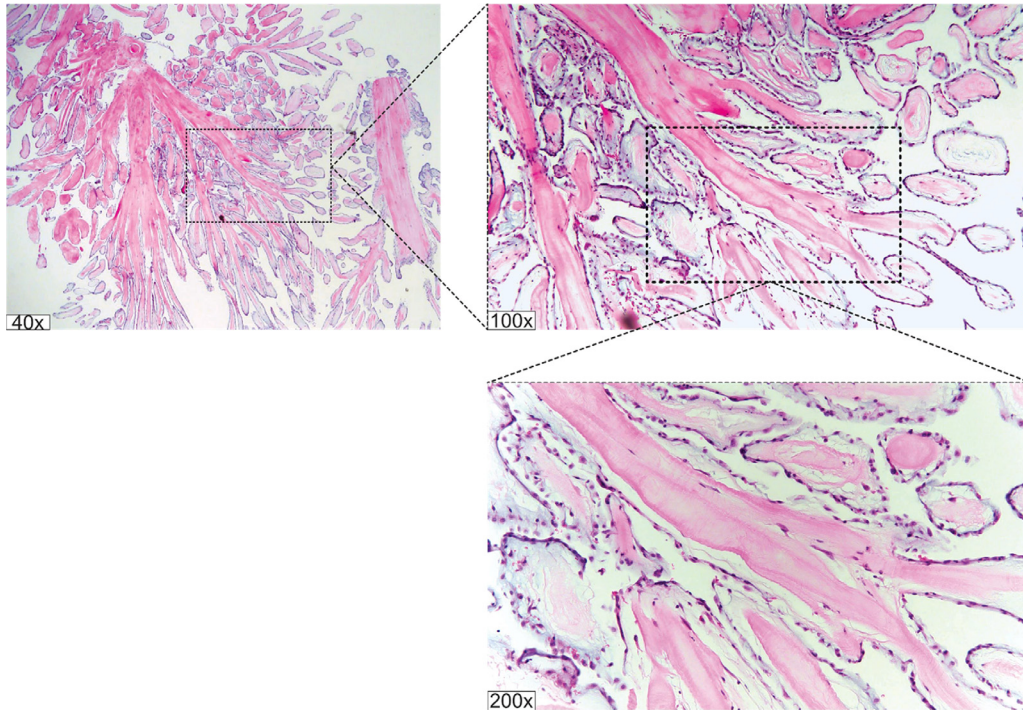


Fig. 2 – Mass was tan-colored soft tissue with multiple villous projections (2.2 × 1.8 × 0.4 cm in aggregate) and identified as a papillary fibroelastoma after histopathological analysis with H&E staining. 40× magnification (A), 100× (B), and 200× (C). Dotted lines indicate area used for magnification.

diagnoses, PFE, Lambl's excrescences, myxoma, infective vegetation, and Libman Sacks endocarditis were considered. Considering the patient's normal bloodwork, location and morphology of the mass, and no history of autoimmune disease, PFE was the most likely diagnosis pending confirmation with histology.

The mass was completely resected, and the valve stayed competent (Fig. 1C). After the mass was successfully resected, he underwent a CABG in which the LAD and OM were grafted (LIMA-LAD and GSV-OM). Histopathological examination of the pulmonic valve mass showed multiple tan, villous portions of soft tissue measuring 2.2×1.8×0.4 cm in aggregate resulting in a final diagnosis of papillary fibroelastoma (Fig. 2). The patient recovered well from the surgeries and was later discharged from the hospital.

Discussion

Primary cardiac neoplasms, or tumors, are a rare finding, with a prevalence around 0.02%, and are benign in approximately 75% of cases [1]. Primary cardiac tumors include papillary fibroelastomas (PFE), myxomas, hemangiomas, and lipomas, of which fibroelastomas and myxomas are the most common [5]. We diagnosed our patient's PFE when performing a pre-CABG TEE, however other imaging modalities, such as TTE, magnetic resonance imaging, positron emission tomography, and computed tomography scans could be used. Our patient was asymptomatic, and the finding was incidental. Given the advancements in cardiac imaging and their increasing availability, the prevalence of PFE will likely increase.

PFEs can be found anywhere in the endocardium, but they are predominantly found on the valvular surface. In a study of 725 cases in 2001, PFEs were most found on the left-sided valves with prevalence rates of 44% for aortic valves and 35% for mitral valves. For right-sided PFEs, the tricuspid valve was the most common location at a prevalence of 15%, and the pulmonary valve was the least common at 8%. Overall, PFEs are most commonly found on the left-sided heart valves (aortic and mitral) (95%) [3].

PFEs have the appearance of a flower or sea anemone that contains multiple branching fronds with a short pedicle connecting it to the endocardium [5,6]. It consists of avascular, collagenous papillary fronds covered by a layer of endothelium cells with a fibroelastic core [7,8]. Due to this structure, thrombi can form in the spaces between the fronds, especially in the presence of von Willebrand Factor from the endothelial cells or exposed collagen fibrils. As such, it has been suggested that patients with papillary fibroelastomas should be placed on anticoagulants if surgery is not immediately available [8,9].

Differential diagnoses

In addition to PFE, Lambl's excrescences, myxomas (rarely), infective vegetation, and Libman Sacks endocarditis appear as mass projections from the heart valves. Lambl's excrescences are small, thin, multiple, filiform structures (thread-like strands) that form at the contact points of the heart valves, and they are usually found on the mitral valve [10]. Myxomas are typically found on the right or left atrium, and they are rarely found on the valves [11]. In contrast, PFEs have a pedunculated and frond-like appearance on echocardiogra-

phy, and they commonly present on the midportion of valve leaflets [10]. Considering that our patient's mass originated from the heart valve as a single, large mass, Lambl's excrescences and myxoma were eliminated as a potential diagnosis, pending further confirmation by histological analysis. Our patient did not have the risk factors or other symptoms that would indicate the presence of an infective vegetation. Additionally, he did not have a history of autoimmune disorders, such as lupus or antiphospholipid syndrome, and had normal bloodwork, so Libman Sacks endocarditis was unlikely [12].

Treatment standards

Resection is the gold standard of treatment for PFE, and it is indicated for symptomatic or high-risk asymptomatic patients, such as those with a large, left-sided mobile mass. Conservative management with serial echocardiography, and potentially anticoagulant medications, can be considered if the tumor is small, non-mobile, and located on right-sided valves. However, surgical resection is still the preferred treatment method.

Conclusion

PFE is a rare occurrence and pulmonary PFE is even more so. Diagnostic imaging, such as a TEE in our case, often finds PFEs incidentally. Mass resection is the standard treatment of choice for PFEs, especially in symptomatic patients or asymptomatic patients with a high-risk, mobile mass, to prevent future embolic events.

Patient consent

This case report was written with consent from the patient.

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