

A Common Disease, an Uncommon Location: Left Ventricle Papillary Fibroelastoma

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Abstract

Cardiac papillary fibroelastoma (PFE) is a rare benign cardiac tumor that is usually related to cardiac valves. Non-valvular, left ventricular (LV) cavity papillary fibroelastoma (PFE) is extremely rare with a few reported cases in the literature. Herein, we report a 75-year-old man who first presented in 2018 with exertional chest pain and was referred to our tertiary-care hospital for further evaluation. On echocardiogram, the patient was found to have a left ventricle lesion related to the papillary muscle. Assessment with cardiac MRI revealed an enhancing mobile lesion related to the LV anterolateral papillary muscle with initial diagnostic possibility of a myxoma. The patient underwent surgical resection of the lesion, and the histopathology examination revealed the diagnosis of PFE.

Keywords: MRI; Cardiac Tumor.

Introduction

Cardiac tumors are rare in general, with a reported prevalence of 0.002-0.3%, according to the autopsy report. Most cardiac tumors (75%) are benign, with myxoma being the most common benign cardiac tumor.¹ The clinical presentation of cardiac tumors is usually not specific and influenced by the size, location, and embolism tendency.² PFEs is the second most common benign cardiac tumor and the vast majority of PFEs are related to cardiac valves. However, rarely PFEs can arise from the left ventricle (LV) endocardium. Herein, we report a rare case of a left ventricular mass that was detected incidentally on an echocardiogram. Further assessment with CMR showed an enhancing lesion related to the LV anterolateral papillary muscle. The differential diagnosis included LV myxoma and lipoma. The patient underwent excision of the LV mass and histopathology confirmed the diagnosis of PFE. The information provided in this article aims to increase awareness of physicians and radiologists about this rare entity and to consider including it in the differential diagnosis of LV intracavity masses. To the best of the author's knowledge, this is the first case of left ventricular papillary fibroelastoma detected on cardiac MRI to be reported in Oman.

Case report

A 75-year-old man, known to have diabetes mellitus, was admitted to our tertiary hospital with a history of exertional chest discomfort for further evaluation. His electrocardiogram showed sinus rhythm. Further evaluation with echocardiogram revealed severe aortic valve stenosis with concentric left ventricle hypertrophy and normal left ventricle systolic function, ejection fraction (EF) was 59%. There was an incidental mobile lesion related to anterolateral papillary muscle of the left ventricle measuring 1 x 0.6 cm. Coronary angiography showed normal coronary arteries. Cardiac Magnetic Resonance (CMR) was performed for further evaluation and showed a small mobile mass related to the anterolateral papillary muscle of the left ventricle, measuring 0.8 x 0.5 cm. The lesion was hyperintense in T2-weighted images and hypointense in T1-weighted images, with homogenous enhancement on post contrast sequence (Figure1). The impression from MRI was a left ventricle (LV) myxoma.

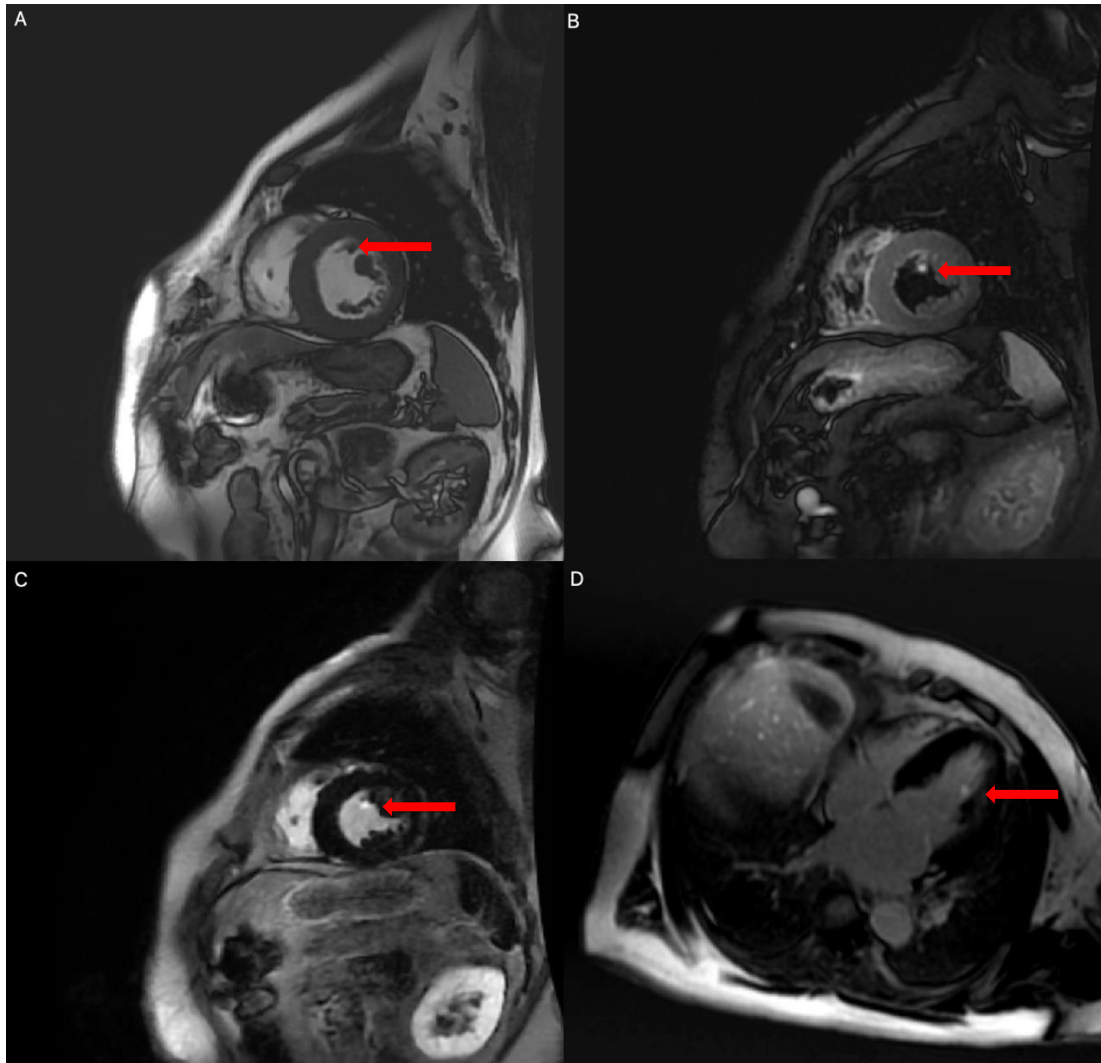


Figure 1: Steady State Free Precision short-axis oblique (A) and T2-wighted images (B) showing a small lesion related to the left ventricle anterolateral papillary muscle 9. Late gadolinium enhancement short-axis oblique (C) and 4-chamber (D) views showing an enhancing lesion related to the left ventricle anterolateral papillary muscle 9.

The patient underwent open cardiac surgery for aortic valve replacement and excision of LV mass. Microscopic examination of the excised lesion showed branching papillary fronds composed of avascular fibroblastic cores surfaced by a single layer of bland endocardium (Figure 2). Histopathology findings of the excised mass were consistent with papillary fibroelastoma. Consent was taken from the patient for the publication of this case report.

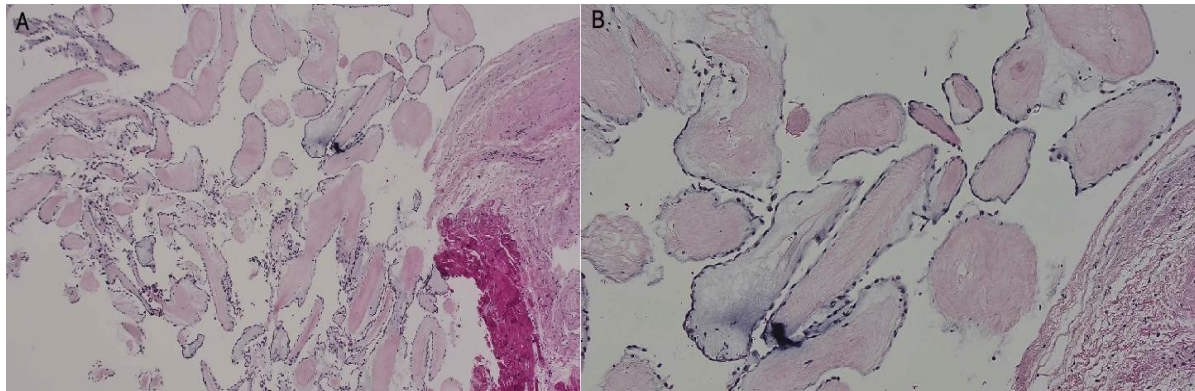


Figure 2: The lesion is composed of many papillary fronds, (HE x 40) (A). The papillary fronds show avascular hyalinized cores and are lined by a single layer of endocardial cells (HE x100), (B).

Discussion

Papillary Fibroelastoma (PFE) is a rare, small, benign endocardial tumor. The most common location is cardiac valves but can arise from anywhere in the heart. PFEs are the most common primary benign valvular tumors accounting for <10% of overall primary cardiac tumors.^{3, 4} Historically, PFE believed to be the second most common of overall benign primary cardiac tumors after cardiac myxoma; however, a study by Tamin et al. published in 2015 to describe the frequency and clinical course of patients with surgically removed PFE and suspected PFE on echocardiography identified 511 cases of cardiac myxoma and PFE from 1995 to 2010 in Mayo Clinic. The study showed that PFEs are more common than cardiac myxomas.⁴⁻⁶

Patients with PFEs have a mean age of 60 years at the presentation.^{4, 5, 7} However, PFEs can be seen at any age and have no gender predominance, although some papers showed slight increased predilection in men.^{4, 5, 7, 8} A study by Cianciulli et al. which assessed 54 cases with PFE showed increased prevalence in the 6th and 8th decades, like our case.⁵

The most common location of PFE is the cardiac valves, about 90% of the time. The other locations include left ventricle or atrium, right ventricle or atrium, atrial septum, the papillary muscle, chordate tendinea, Eustachian valve and Chiari network.⁷ The nonvalvular right sided PFE are more common than the left side.⁶

PFEs are usually detected incidentally during echocardiogram, cardiac surgery, or autopsies.⁵ Although most of the patients with PFEs are asymptomatic, some can present with acute stroke related to embolization, syncope, angina, myocardial infarction, and even sudden cardiac death.^{4, 7} The clinical presentation depends on the size, location, growth rate and mobility of the tumor. Because of its embolic potential, PFE are clinically important to identify and treat.^{4, 6} PFEs are thought to be an acquired rather than an inherited lesions.^{4, 5} PFE have left heart location predominant, particularly aortic valve, therefore, the symptoms related to embolization are more common.^{4, 5, 9} The most common presenting symptom is neurological event either transient ischemic attack, TIA, or stroke.⁶ Gowda et al. in a meta-analysis of 725 PFE in 2003 reported that the most common presenting symptom related to PFE is TIA or stroke in 44% of patients and the second most common symptom is angina by 18%.⁸ The remainder of the symptoms are less common. In our case, the patient presented with exertional chest pain which prompts further evaluation with echocardiogram. No prior study discussed the most frequent symptoms related to the left ventricular PFE. A case report by Hyun et al. of a patient with left ventricular PFE presented with chest pain for 3 days.⁷ Another case report of PFE in left ventricle by de Klerk et al., the presenting symptom was dyspnea.⁵

PFEs are composed of fibroelastic tissue surrounded by endocardium. Grossly, PFE are flower-like with multiple papillary avascular fronds covered by a gelatinous membrane and connected to a central stalk which is attached to the endocardium.^{7, 8} The gross appearance may be altered by the thrombi attached to its surface.⁸ Microscopically, the villus-like projections are avascular and composed of a dense collagen fibers core with matrix consists of proteoglycans, elastin fibers, and rarely spindle cells resembling smooth muscle cells or fibroblasts.^{7, 8} The surface epithelial lining is continuous with the endocardium.⁷ The hallmark of PFE is the presence of elastic fibers in variable distribution making the demonstration of these fibers difficult.⁸ The Different

pathological processes have been proposed as predisposing factors for the development of PFEs including epithelial hyperproliferation from hemodynamic damage to the endothelium, trauma, and organized thrombus.⁵

Multimodality imaging is essential for the evaluation of cardiac mass including PFEs. The diagnosis of cardiac tumors is based on the location, mobility, size, and imaging features. On transthoracic echocardiogram, TTE, PFEs appear typically as a round mobile homogenous echogenic lesion that has a stalk connecting to endocardium.^{4, 10} It can be non-mobile. The transesophageal echocardiogram, TEE, is done after the TTE and shows higher sensitivity, especially for the smaller lesions.^{4, 8}

MRI gives better details of the tumor tissue characteristics due to its high contrast resolution and multi-sequential assessment.^{11, 12} The MRI typically shows a mass on the endocardial surface of the cardiac structure, commonly the valve leaflets.^{3, 8} On T1 and T2 weighted images, PFE demonstrates a well-circumscribed homogenous intermediate signal intensity to the myocardium, which represent fibroelastic tissue. The cine gradient echo, GRE, sequence may show the tumor mobility and perilesional flow artifact due to turbulence blood flow.^{3, 8, 9, 13} PFE shows diffuse homogenous late gadolinium enhancement, LGE, which will differentiate the tumor from thrombus, a well-known tumor mimicker.^{8, 13} Also, the thrombus shows susceptibility artifact on GRE sequence due to methemoglobin and hemosiderin content.¹²

On imaging given overlapping characteristics, the differentiation between PFE and myxoma can be difficult, especially when PFE arises from a non-valvular cardiac structure, like in our case.⁹ The prior case report by de Klerk et al. of the left ventricular PFE was also reported as myxoma on imaging before resection.⁴

Rhabdomyoma is another benign cardiac tumour that is typically seen in paediatric patients; on CMR, it appears isointense on T1-weighted images, hyperintense to the myocardium on T2-weighted images and shows minimal or no contrast enhancement on post-contrast examination. Cardiac lipomas are benign cardiac tumours that have homogenous high signal intensity on T1-weighted images with complete suppression on fat suppression sequences. Cardiac lipomas show no enhancement on post-contrast sequences due to their poor vascularity.¹⁴

The description of PFE on electrocardiogram gated computed tomography, ECG-gated CT, is limited. The thin sections and better anatomical details on CT can show the PFE small stalk that is difficult to demonstrate on MRI.^{4, 11} ECG-gated CT is better in the assessment of calcifications within the cardiac masses, which is more often found in case of cardiac myxoma.^{4, 12} Contrast enhanced CT demonstrates enhancement of the PFE.¹¹

Both MRI and CT can assess the integrity of cardiac valves, if the tumor arising from cardiac valve, and the extent of tumor.¹² The presence or absence of valvular destruction helps in differentiating infective endocarditis vegetation from PFE, although infective endocarditis is generally clear clinically.^{9, 13}

The treatment options include curative surgical removal versus follow-up. Surgical excision is the treatment of choice in symptomatic patients.⁸ Both short and long-term prognosis after surgery is excellent with no reported recurrence.^{4, 7}

In asymptomatic patients with mobile PFE, surgical removal should be considered if the tumor is mobile due to increased risk of embolization. Tumor mobility is an independent predictor of embolization and death.^{4, 8}

Follow-up is an option for asymptomatic patients with non-mobile PFE. As reported by Tamin et al. in the follow-up group of 317 patients, the risk of cerebrovascular accident was 6% and 13% at 1 and 5 years, respectively.⁶

Conclusion

Cardiac papillary fibroelastoma are rare, benign cardiac tumours that are typically related to the cardiac valves. However, papillary fibroelastoma can be seen with the left ventricle cavity. Patients with cardiac papillary fibroelastoma are frequently asymptomatic; however, some patients can be symptomatic depending on the size and location of the papillary fibroelastoma. Awareness of the typical CMR findings of cardiac papillary fibroelastoma allows pre-operative diagnosis with a high level of confidence.

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