

An Unexpected Finding on Echocardiography in a Patient with Fatigue: A Two-Centimeter Papillary Fibroelastoma

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Abstract

Cardiac papillary fibroelastomas (CPFs) are rare primary cardiac tumors. The majority of these tumors are located on valvular surfaces. Although most patients are asymptomatic, some patients may be complicated by stroke, myocardial infarction and even sudden cardiac death. The diagnosis of CPFs can be very challenging.

An 83-year-old male patient presented to the outpatient clinic with the complaint of fatigue and a 2 cm lesion with soft tissue echo pattern was detected in the left atrium. Among differential diagnosis of the lesion based on both echocardiogram and cardiac magnetic resonance images (MRIs) were thrombus, myxoma, Lambli's excrescence and vegetation. Pathological examination after prompt surgery revealed papillary fibroelastoma. We planned to present this case with both echocardiographic and MRI findings.

Keywords: Heart, neoplasms, mitral valve

INTRODUCTION

The incidence of primary cardiac neoplasms is rare and the most common three tumors are myxomas, lipomas and papillary fibroelastomas in order of decreasing frequency. Cardiac papillary fibroelastomas (CPFs) may be found on the endocardial tissues, with a predilection for valve structures. Clinical presentation of CPFs can vary with majority of patients being asymptomatic with incidental findings on echocardiography. Tumors on both left and right sides of the heart can present with embolic events because of their fragility and high affinity for platelet aggregation.

Echocardiography is the major cardiac imaging modality for the diagnosis of cardiac tumors, but the diagnosis based upon

imaging modalities is not definitive most of the time. Cardiac masses differ in size and shape making it difficult to diagnose a tumor versus a thrombus based on imaging findings and clinical presentation. The sensitivity and specificity are around 90% when using transthoracic echocardiography for the diagnosis of CPF especially if tumor size is more than 2 mm.¹ We present a patient who underwent echocardiographic examination that revealed a mass in close proximity to mitral valves, which turned out to be CPF after surgery.

CASE PRESENTATION

An 83-year-old male patient admitted to cardiology outpatient clinic with the complaint of fatigue. His medical history

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revealed long lasting primary hypertension, chronic obstructive pulmonary disease and benign prostatic hyperplasia. He was on valsartan 160 mg tb 1x1, aspirin 75 mg tb 1x1, tamsulosin 0.4 mg cps 1x1, and inhaled bronchodilators. On physical examination, the sole pathological finding was 2/6 ejection type systolic murmur that was heard over aortic area. Blood pressure was normal. ECG was normal. On echocardiogram (GE Vivid S5, GE Healthcare, USA), aortic valves were calcified and Doppler findings were compatible with mild aortic stenosis, mitral valves, left ventricular ejection fraction and pericardium were all normal and a 2.0x2.5 cm round-shaped, well-circumscribed, highly mobile, echogenic solitary mass was realized in the left atrium with close proximity to mitral valves (Figure 1). With continuous wave Doppler imaging, mean mitral valve gradient was 4.0 mmHg indicating an obstruction of the left ventricular inflow. Differential diagnose were primary cardiac tumors, thrombus, vegetation and secondary cardiac tumors. Patient's chest X-ray and thoracoabdominal computerized tomography (CT) scan were found to be normal. Blood biochemistry including tumor markers were normal. For more detailed imaging of the mass, transesophageal echocardiogram was done at another hospital that could not reveal further information, so cardiac magnetic resonance imaging (MRI) was planned. Cardiac MRI was not done properly and prematurely stopped since the patient could not tolerate the procedure, and thus, contrast medium could not be injected and tissue characterization could not be done. However, MRI (Siemens Aera, 1.5T, Germany) showed a 1.8x2.0 cm nodular mass attached to the laterobasal portion of posterior mitral valve leaflet with a broad base (Figure 2). Since the mass was obstructing left ventricular inflow and the presence of risk of stroke surgical intervention was planned. The patient was operated by heart surgeons with minimally invasive approach. Through right thoracotomy the mass attached to posterior mitral valve leaflet was excised. The pathological examination revealed cardiac papillary fibroelastoma (CPF) with narrow, elongated and branching papillary fronds consisting of central avascular collagen and variable elastic and collagenous connective tissue, surrounded and covered by acid mucopolysaccharide which were lined by endothelial cells (Figure 3). The patient was discharged from the hospital 5 days after the surgery.

DISCUSSION

Primary cardiac tumors are rare with an incidence of <0.1%. Around 75% of them are benign and CPF is the most common of the primary valvular tumors.² Appearance of CPFs resembles sea anemones and their size vary from 0.2 to 7 cm. More than 80% originate from valvular surfaces with the most commonly affected valve being the aortic valve.³

Risk factors of CPFs are unknown and diagnosis is reached en passant by echocardiography. Echocardiogram usually

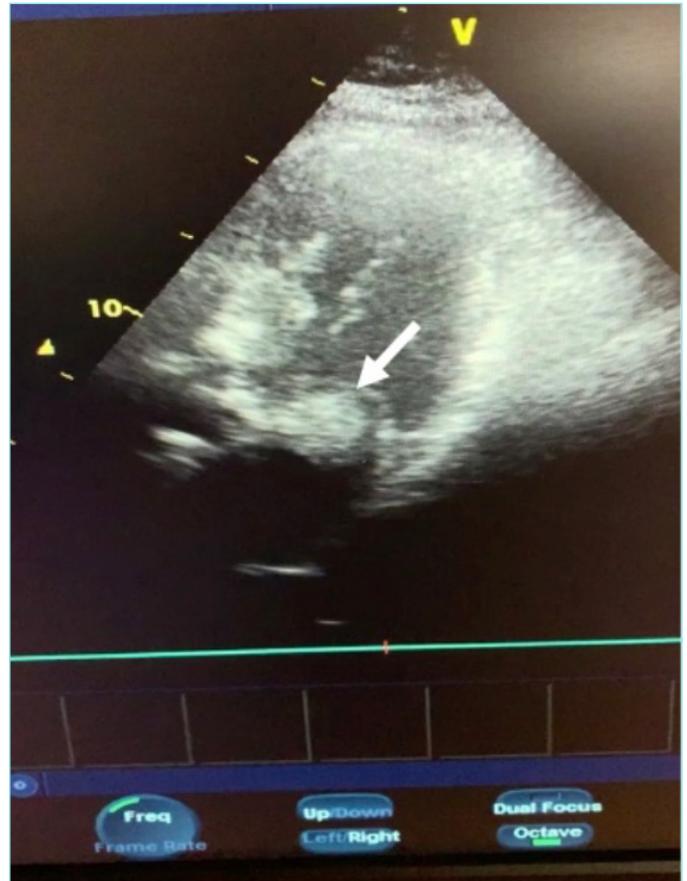


Figure 1. Doppler imaging findings

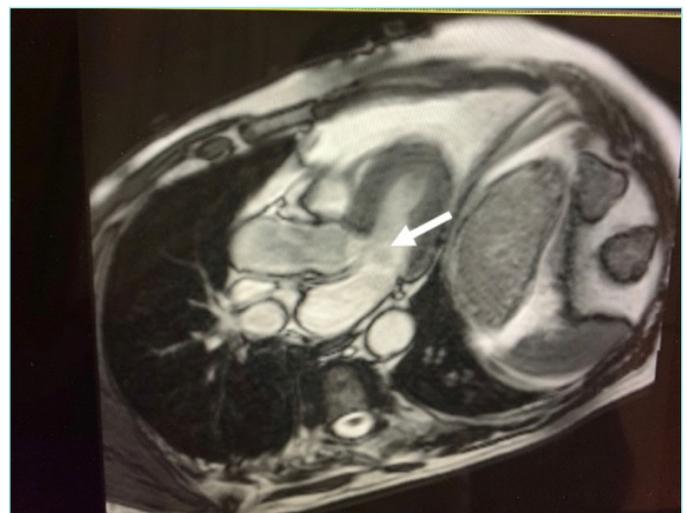


Figure 2. MRI findings

MRI: Magnetic resonance imaging

reveals a mobile, pedunculated or sessile mass, which flickers into the cardiac chambers. On cMRI, CPF typically appears as a small, round, homogeneous mass, usually bound to valvular structures and signal intensity is isotense on T1 and hyperintense or isoense on T2-weighted fast spin echo

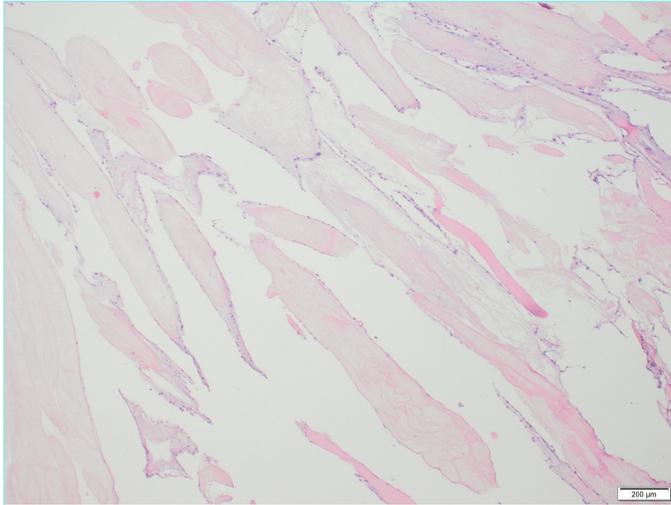


Figure 3. Pathological examination

images. However, T1 and T2 weighted images reflect their fibroelastic composition with uniform intermediate signal intensity similar to myocardium. Steady-state free-precession (SSFP) cine is also useful for the mobility of the mass. CPFs present as low signal, well-circumscribed mobile valve nodule on SSFP with peri-tumoral flow artefact on MRI.³⁻⁵ Vegetation is associated with valve destruction unlike CPFs with typically spared valve function.

Most CPFs are asymptomatic or associated with vague symptoms as in our patient but systemic embolization is frequent because more than 95% of CPFs are located on the left side of the heart and they are fragile and have high affinity for platelet aggregation.^{1,3}

Among differential diagnosis of CPFs are other cardiac tumors, vegetations and thrombus. The tumors, in general, are rarely present on valves. Thrombus can be differentiated by an irregular or lobulated shape, laminated appearance, microcavitations and absence of a pedicle.⁶ With cMRI, fat-saturation sequences may help differentiate fibroelastomas from lipomas.³ cMRI with gadolinium can also be used to distinguish thrombi from tumor.⁷ Furthermore, cMRI provides an unrestricted spatial approach, high temporal resolution (30–50 ms) and noninvasive tissue characterization. cMRI, however, may have limitations for evaluating small (<10 mm) and mobile masses like fibroelastomas.⁸ Unfortunately, our patient could not tolerate MRI procedure so we could not benefit from its advantages.

In conclusion, we presented a patient with unexpected mobile mass in left atrium which turned out to be CPF. Whatever the histopathological nature of mobile mass in heart chambers is, it is important to act fast and reach true diagnosis before a detrimental complication occurs. In this study, we could reach the final diagnosis after prompt surgical resection of the mass despite the use of advanced imaging modalities.

MAIN POINTS

- Echocardiographic examination even in patients with subtle symptoms is important and necessary.
- Whatever the histopathological nature of mobile mass in heart chambers is, it is important to act fast and reach true diagnosis before a detrimental complication occurs.
- Management of cardiac masses especially in the elderly is very challenging. In this study, we could reach the final diagnosis after prompt surgical resection of the mass despite the use of advanced imaging modalities.

ETHICS

Informed Consent: It was obtained.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: R.M.B., Design: R.M.B., Data Collection and/or Processing: R.M.B. Analysis and/or Interpretation: R.M.B., Ç.Z., S.B., Ö.T., M.M., Literature Search: R.M.B., Ç.Z., S.B., Ö.T., M.M., Writing: R.M.B.

DISCLOSURES

Conflict of Interest: The authors have no conflicts of interest to declare.

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