

A Huge Papillary Fibroelastoma Case Presenting with Recurrent Acute Pulmonary Edema

Tekrarlayan Akut Akciğer Ödemi ile Prezente Olan Dev Bir Papiller Fibroelastom Vakası

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Abstract

Primary intra-cardiac tumors are rare and reported to be less than 0.3% in autopsy series. Approximately 50% of them are cardiac myxomas. Papillary fibroelastomas are the second most common primary cardiac tumors following myxomas and they generally involve and damage the cardiac valves. The clinical presentation of these tumors vary from asymptomatic cases to cases with severe clinical presentation like sudden cardiac death. In this case, we described a 77 years old male patient presenting with recurrent acute pulmonary edema and diagnosed as mitral posterior valve papillary fibroelastoma. (*Sakarya Med J* 2016, 6(2):110-114)

Keywords Papillary fibroelastoma, mitral posterior valve, acute pulmonary edema

Öz

: Primer intrakardiyak tümörler klinik pratikte nadir görülür ve otopsi serilerinde sıklığının %0.3'ten daha az olduğu bildirilmiştir. Bunların yaklaşık %50'si kardiyak miksomalardır. Papiller fibroelastomlar, kardiyak miksomalardan sonra ikinci en sık görülen primer kardiyak tümörlerdir ve genellikle kalp kapaklarını tutar ve onlara zarar verir. Bu tümörler asemptomatik seyredebileceği gibi ani kardiyak ölüm gibi ciddi sonuçlarla da ilişkili olabilirler. Biz bu vakada, tekrarlayan akut pulmoner ödem ile prezente olan ve mitral arka kapakta papiller fibroelastom tanısı alan 77 yaşında bir erkek hastayı sunduk. (*Sakarya Tıp Dergisi* 2016, 6(2):110-114)

Anahtar Kelimeler: Papiller fibroelastom, mitral arka kapak, tekrarlayan akut pulmoner ödem

INTRODUCTION

Primary intra-cardiac tumors are rare with a prevalence ranging from 0.0017 to 0.28% in various autopsy series and account for an only 0.3% of all open heart operations¹. Papillary fibroelastomas (PFE) account for less than 10% of all cardiac tumors, representing the most common valvular and the second most common cardiac benign tumor following myxomas². PFE can cause thromboembolism or mechanical interference with valvular function. Here, we presented a rare case of PFE arising from the posterior leaflet of the mitral valve, thus mimicking mitral annular calcification and leading to recurrent acute pulmonary edema due to the mechanical interference.

CASE PRESENTATION

A 77-year-old man was referred to our department because of night sweating, dyspnea with a New York Heart Association functional class of 2, weight loss (10 kilograms) and recurrent acute pulmonary edema attacks in the last two months. Past history revealed hypertension, coronary artery disease and hyperlipidemia. Cardiovascular examination and routine blood laboratory tests were unremarkable. Electrocardiogram showed normal sinus rhythm with a rate of 75 beats per minute and left anterior fascicular block. Transthoracic echocardiography (TTE) demonstrated a round hyperdense, immobile mass with a diameter of 29x20 mm attached to the posterior leaflet of the mitral valve (Figure 1). There was mild mitral stenosis with a mean gradient of 4 mmHg. Due to the location of the mass and the older age of the patient, at first look we suspected from the diagnosis of a huge mitral annular calcification (MAC). However, to get a definite diagnosis and avoid a misdiagnosis, transesophageal echocardiography (TEE) was planned. TEE revealed a mobile, round mass of 22x27 mm at the ventricular site of the mitral posterior leaflet (Figure 2). To further characterization and diagnose differentially with metastasis of the mass, ECG-gated magnetic resonance imaging (MRI) was performed. This showed an approximately 30x20 mm solid mass just below the posterior leaflet of the mitral valve (Figure 3A) that is mobile accompanying posterior leaflet and sometimes obstructing the left atrial flow. After intravenous gadolinium administration (DTPA), mass lesion with homogenous intense opaque uptake was seen on cine cardiac MRI (Figure 3B). The combination of information ga-

ined by multimodality imaging allowed for definite diagnosis of PFE and subsequent planning of the surgical approach. Subsequently, the patient was referred to department of cardiovascular surgery for the operation.

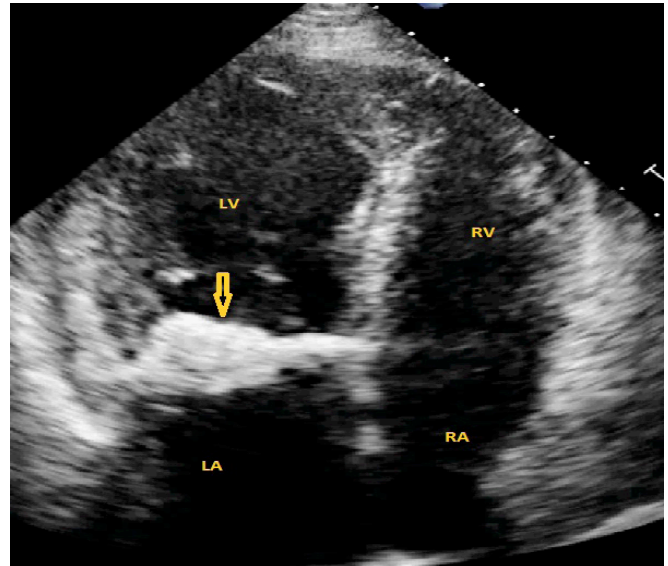


Figure 1. Transthoracic echocardiography four chamber (A) and two chamber (B) view showed a round mass with an irregular borders of 22x29 mm attached at mitral valve posterior leaflet (arrow).

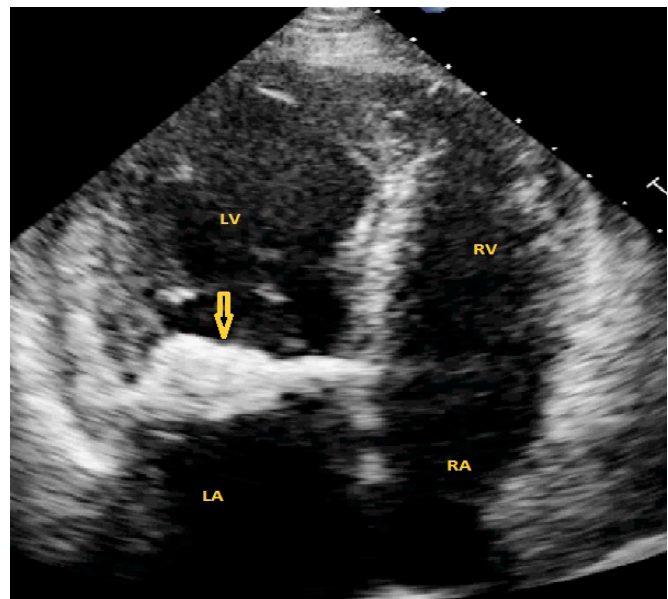


Figure 2. Transesophageal echocardiography showed a bulbous mass of 22x29 mm attached at mitral valve posterior leaflet (arrow).

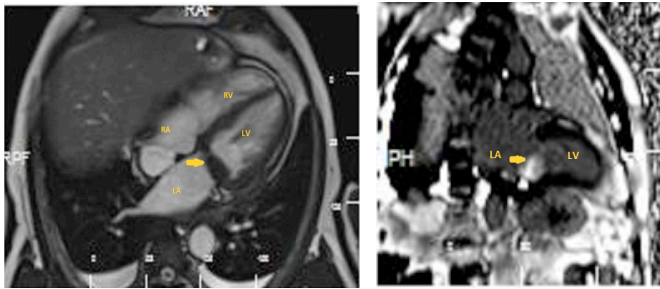


Figure 3. Cine cardiac magnetic resonance imagings

DISCUSSION

Papillary fibroelastomas are the second most common cardiac benign tumors² and typically arise from the valvular endocardium. PFE usually develop on cardiac valves, but many arise anywhere else in the heart. They most often affect the aortic valve (52%), followed by the mitral valve (16%), tricuspid valve (6%), and pulmonary valve (2%)³. They occur occasionally, usually in middle-aged and older patients, with a slightly higher incidence in males⁴. Most PFEs are asymptomatic and found incidentally at the time of routine physical examination, echocardiography, cardiac catheterization, cardiac surgery or autopsy.

Papillary fibroelastomas present with a great variety of symptoms, especially cardiovascular in nature⁵. Although they are benign tumors; they can result in life-threatening complications such as myocardial infarction, cardiac arrest and stroke⁶⁻⁸. Interestingly, valve dysfunction is rarely described, although the tumor is usually located on the valves. Therefore, for early diagnosis and prevention of severe complications, and due to their infrequent occurrence, each case is interesting to report in order to improve the diagnosis and management of this uncommon tumor. We presented a very rare case of papillary fibroelastoma attached to the posterior leaflet of the mitral valve and caused acute pulmonary edema attacks due to blockage of mitral inflow.

Diagnosis is usually made with the use of echocardiography. In patients with a cardiac mass, the echocardiographic differential diagnosis includes myxoma, vegetations, and mural thrombus. Features that distinguish PFE from other intracardiac masses are suggestive, rather than definitive: a small lesion less than 1 cm in diameter, but perhaps as large as 3 to 4 cm;

and a highly mobile mass, with a pedicle attached to the valve or endocardium.

In our case, these distinguishing features of PFE were absent on TTE. A large lesion 3 cm in diameter, immobile without a pedicle attached to the mitral posterior leaflet was observed at TTE. Therefore, we got far away from the diagnosis of PFE and assumed the lesion as being a huge MAC. However, we kept in mind that, it can be a rare cardiac mass mimicking MAC. So that, firstly we performed TEE which demonstrated that the lesion was mobile and attached to the ventricular surface of the mitral posterior valve, not on the valve itself like MAC and moving along with the posterior leaflet.

Although TTE and TEE are important modalities in the diagnosis, newer imaging techniques, such as cardiac MRI, can help to better delineate cardiac masses. Therefore, cardiac MRI was performed and definite diagnosis of PFE was reached. However, PFE are usually not observed on CT or MR images as they are small (<1,5 cm in diameter) and are attached to the moving valves⁹. MRI typically demonstrates the presence of a mass on a valve leaflet or on the endocardial surface. These tumors can create turbulence in the blood flow, which might be demonstrated with the use of cine MRI. For assessment of PFE with MRI, characteristics include: pedunculated, mobile, endocardial, or valvular mass. Other signal characteristics have not been well defined.

Surgical intervention is the definitive treatment for cardiac fibroelastoma and should be done in all symptomatic or asymptomatic patients with mobile lesions for prevention of complications¹⁰. Asymptomatic patients with nonmobile tumor smaller than 1 cm can be closely followed-up with echocardiography until symptoms develop or tumors enlarges and becomes mobile, and after that tumor could be surgically removed¹¹. Patients who from other reasons are not candidates for surgical treatment should be treated with long-term anticoagulation therapy.

In this paper, we report the uncommon location of a PFE with mechanical interference. Initially, the mass was evaluated as mitral annular calcification in view of the mass location, and echogenicity upon TTE and TEE. However, eventually PFE was

detected on the assesment with cardiac MRI that planned for further examination. And then the patient was referred for operation.

CONCLUSIONS

We described a rare case of papillary fibroelastoma attached to the posterior leaflet of the mitral valve, thus mimicing mitral annular calcification and presenting with recurrent acute pulmonary edema due to the blockage of mitral inflow. We anticipated that raising of sample and numbers of case presentations such as our patient, provide to keep in mind for PFE on the similar cases. Because, approach, treatment and clinical courses of MAC and PFE differ significantly, utilization of multimodal imaging techniques to reach a definite diagnosis of such a MAC-like mass is crucial.

Conflict of interest: None of the contributing authors have conflict of interest, including specific financial interests or relationship and affiliations relevant to the subject.

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