

Female with Atypical Chest Pain: not an Acute Coronary Syndrome, but an Aortic Valve Fibroelastoma

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Abstract

A 50-year-old woman was evaluated at the clinic after complaining of atypical chest pain, dyspnea, dizziness and palpitations after one year of evolution. Cardiologist did, a 24 hour holter monitoring, carotid doppler, EKG, transthoracic echocardiogram and Left heart catheterization, all of them were within normal limits, except the transthoracic echocardiogram that presented a valvular mass versus valvular vegetation. Transesophageal echocardiogram described the presence of a pedunculated mass at the left coronary cusp. Patient was evaluated by a cardiothoracic surgeon who performed open thoracotomy resecting a mass of 0.7cm x 0.7 cm x 0.4 cm in size. Histopathology results revealed Papillary Fibroelastoma of the aortic valve. Microscopically a papillary proliferation with characteristic avascular branching papillae, composed of collagen covered by endothelium.

Keywords: Chest pain; Papillary fibroelastomas; Cardiac Papillary fibroelastoma; Acute Coronary Syndrome

Introduction

Papillary fibroelastomas is the third most common primary benign cardiac tumor after myxoma and lipoma, representing 7.9 to 10%. [1] Usually papillary fibroelastoma affects cardiac valves, can occur at any age but the median age is 60 years and is more common in men. [1] It can be asymptomatic, found incidentally, but when symptoms are present they can vary from dizziness, chest pain, dyspnea and can cause a thromboembolic stroke. [2] In recent years with the implementation of cardiac Magnetic Resonance Imaging, multi- slice spiral Computed Tomography, transthoracic and transesophageal echocardiogram have been more recognized. [2] Recommended treatment is surgery as it can lead to cardiac and vascular complications if not treated.

Case report

A 50-year-old woman patient with a past medical history of chronic sinusitis, arterial hypertension, lifetime nonsmoker who came for evaluation of atypical chest pain, dizziness, dyspnea, and palpitations of one year of evolution. She described

chest pain as retrosternal, stabbing, non-radiated, that worsened with exertion, the reason for which unstable angina was suspected. An EKG was found with NSR, non-ischemic changes, and a 24-hours holter monitor did not demonstrate any atrial or ventricular arrhythmia. Routine labs were unremarkable including cardiac enzymes. A transthoracic echocardiogram reported aortic valvular mass versus valvular vegetation. For this reason, blood cultures were taken and came back negative for infection. Left heart catheterization was unremarkable for coronary artery disease.

Additionally, a transesophageal echocardiogram described the presence of a pedunculated mass of the left coronary cusp (figure 1). Case was consulted to a cardiothoracic surgeon and open thoracotomy was performed, resecting a mass of 0.7 cm x 0.7cm x 0.4 cm in size. Histopathology reported a papillary proliferation with characteristic avascular branching papillae, composed of collagen covered by endothelium (figure 2). During hospitalization was stable, tolerated the procedure without complications and in one follow up she reported complete resolution of her symptoms.

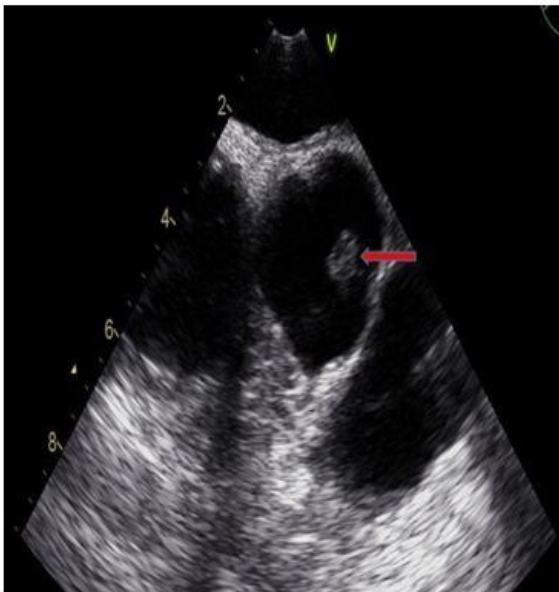


Figure 1:

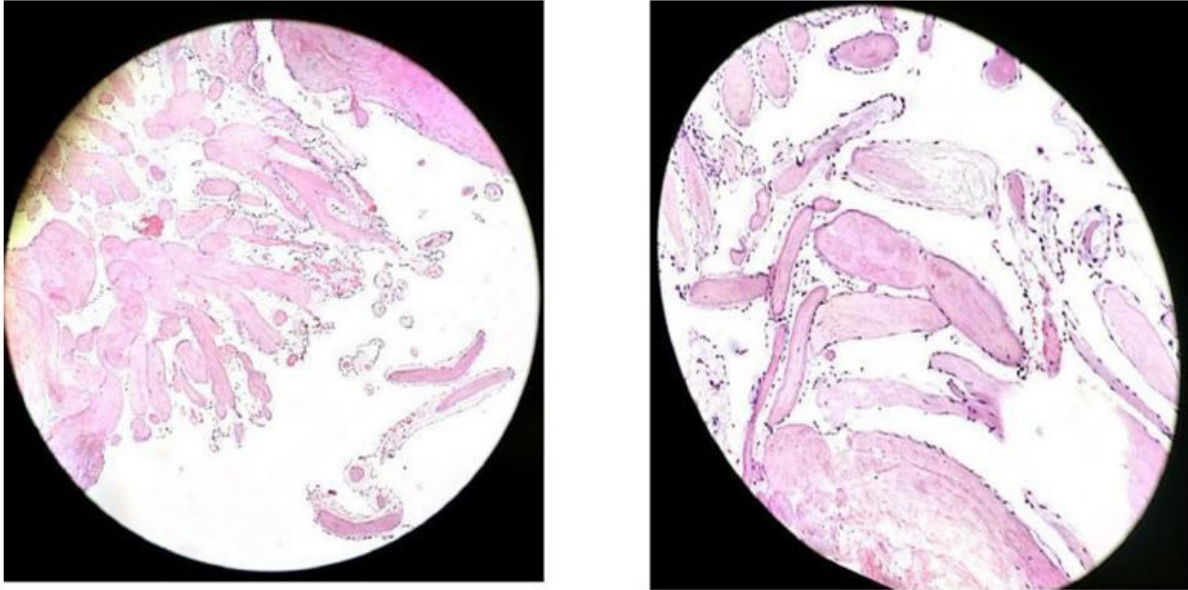


Figure 2:

Discussion

Cardiac Papillary fibroelastoma (CPF) is a benign primary endocardial tumour with valvular predominance arising from the endocardium in 80 %. [3] They account for approximately 10% of all cardiac tumours. Typically affecting the aortic valve followed by the mitral valve, tricuspid, and finally pulmonary valve. [4] Cardiac tumours are classified as benign, intermediate or uncertain behavior and malignant according to the aggressiveness of the tumours. Grossly, CPF have a sea anemone appearance with a gelatinous surface and a stalk with multiple papillary projections. Surgical approach generally involves resection only, with sparing of underlying valve tissue. It is most commonly seen in adults, of more than 60 years of age with a male preponderance (57.1%). Complications include thromboembolic events, usually in the form of syncope or transient ischemic attacks, ventricular arrhythmias, and prolapse into the coronary ostia leading to chest pain. [4] There are cases reported in the literature of secondary myocardial infarction and sudden cardiac death. [5]

Besides the neurological symptoms secondary to thromboembolic stroke, it may be presented with chest pain, dyspnoea and palpitations. Our patient had atypical vague chest pain, dyspnoea, and palpitations. The vague chest pain could be associated with the partial, intermittent obstruction or limitation of blood flow through the right coronary artery orifice in the aortic root due to the strategic location and variable mobility of the

tumour mass during various phases of the cardiac cycle. Therefore, this could be considered as an angina-like symptom simulating myocardial infarction. Since multiple conditions could be presented with these symptoms, recognition of CPF is difficult.

Recognition of CPF in the last couple of decades has increased since more imaging of the chest is studied with better resolution, especially on cardiac structures. Transesophageal echocardiography is more sensitive than transthoracic echocardiography in detecting cardiac tumors. Diagnosis is usually made by 2- dimensional or transesophageal echocardiography. Recently, 3-D echocardiography, magnetic resonance imaging and multi-slice spiral computed tomography have also been used for better assessment and characterization of the tumor component and localization within the heart. [6] Most of the time Cardiac echocardiography shows a small (1-4 cm) pedunculated or sessile valvular or endocardial mobile mass, with a pedicle attached to the valve or endocardial surface and a frond-like appearance with or without multifocal involvement into the cardiac chambers. Echocardiographically, papillary fibroelastomas appear speckled with echo lucencies near the edges. In the review of Gowda et al, it was postulated that tumor mobility was the only independent predictor of CPF related death or nonfatal embolization. The contrast CT image typically shows a filling defect in the aortic root adjacent to the origin of the coronary artery. Differential diagnosis includes cardiac myxoma, Lambli's excrescences, bacterial vegetations, and thrombi.

Treatment varies from medical to surgical, including the use of anticoagulation since tumor fragments can embolize different organs. Complete excision is the only definitive way to eliminate the source and potential recurrence of embolization. [7] Also, preservation of the valve is recommended. In case of valve insufficiency or defect after removal of the tumor it should be repaired, otherwise a valve replacement is warranted. Symptomatic patients who are not surgical candidates can be offered long-term oral anticoagulation for stroke prevention; however, no data strongly validate the efficacy of this approach. [7] These tumors carry a good prognosis and low recurrence rate; otherwise the patient will need cardiologist surveillance.

This case is important in order to educate internal medicine residents, general physicians and primary care physicians. They should include cardiac tumors or masses in the differential diagnosis of patients complaining of chest pain. Also they can learn about diagnostic tools available to identify these kinds of cardiac masses.

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