

Association between Aortic Valve Fibroelastoma and Acute Myocardial Infarction

Thiago Augusto Vieira, Sandra de Barros Cobra Negreiros, Diogo Wagner da Silva de Souza

Hospital de Base do Distrito Federal, Brasília, DF – Brazil

Introduction

Cardiac tumors are rare and diagnosis depends mainly on the degree of clinical suspicion, since its prevalence in autopsies is 0.02%¹. Primary heart tumors include myxoma as the one with the highest incidence (40% to 70%), followed by lipomas and papillary fibroelastoma (PFE)².

PFE is a benign tumor that is present in about 10% of the cases, mostly of related series³. These are commonly single moving masses, pedunculated and well delimited, which especially affect the heart valves. They are usually asymptomatic, but have great potential to promote embolic phenomena^{2,4-6}.

Diagnosis is often incidental during transthoracic echocardiography tests for other clinical indications⁴.

This is a clinical case for which the diagnosis of PFE was suggested during cardiac workup in a coronary artery disease setting. Echocardiography plays a role in the diagnosis of primary cardiac tumors, especially PFE, either incidentally, as in most occasions, or in the investigation of embolic events.

Case Report

Black, male, retired farmer, 52 years old. The patient was admitted to the emergency department of the Cardiology Unit of Hospital de Base do Distrito Federal – Brasília in March 2015. His initial complaint was chest pain, and the pain irradiated to the neck and mandibular area. He had personal medical history of acute myocardial infarction (AMI) six years before, when he underwent coronary artery bypass grafting surgery (CABG). Seven months before, he had been admitted to the same service for inferior wall AMI and underwent coronary angiography (Figure 1). At the time, the test showed severe atherothrombotic obstruction of the right coronary artery. The assisting team chose to conduct percutaneous treatment with non-pharmacological stent. He was later discharged for outpatient monitoring, asymptomatic.

Keywords

Heart Neoplasms/complications; Heart Valve Diseases; Myocardial Infarction; Embolism; Coronary Artery Disease.

Mailing Address: Thiago Augusto Vieira •

SMAS, trecho 1, lote C, bloco H, Apto. 905, Condomínio Living Zona Industrial. Postal Code: 71218-010. Guará, DF - Brazil

E-mail: thiago_escs@yahoo.com.br

Manuscript received June 26, 2015; revised manuscript August 8, 2015; accepted August 26, 2015.

DOI: 10.5935/2318-8219.20150035

A few weeks after discharge, he complained of chest pain on small and moderate stress. To investigate acute ischemia, exercise testing was requested and revealed clinical and electrocardiographic criteria suggestive of acute ischemia. A new catheterization at the hospital in March 2015 showed injury of approximately 90% in the right coronary artery at the stent site and a new obstruction of 70% in the most distal portion of the artery.

During the complementary investigation for coronary artery disease, Doppler echocardiography was performed. Among other findings, mobile echodense image attached to the valve face of the non-coronary leaflet of the aortic valve was found. Transesophageal echocardiography (TEE), in turn, enabled a more detailed imaging (Figure 2), which consisted of a mobile and heterogeneous mass with regular borders measuring approximately 1.2 x 1.2 cm in its largest diameters. Papillary fibroelastoma was then suspected.

The patient was then referred to contrasted magnetic resonance imaging (MRI) of the heart and the finding was: small aortic valve mass attached to the commissural pillar between the non-coronary and coronary cusps of the right aortic face, compatible with papillary fibroelastoma (Figure 3).

Discussion

This case demonstrates that echocardiography is an inexpensive test that can be an extremely useful in diagnostic suspicion of embolic events originated in the heart. PFE is, most often, an incidental finding. Usually as a single lesion which in 90% of the cases affects the heart valves, without causing valve failure.

The aortic valve is the most affected valve (44%), followed by the mitral, tricuspid and pulmonary valves and may involve the papillary muscles and tendinous chordae. Other sites are more rare, especially the left ventricle, left atrium, atrial septum and right atrium².

It is usually identified as a moving mass with dimensions that may vary from 2 to 28 mm, but in rare cases (1%) it reaches dimensions larger than 20 mm^{2,5}. In this case, the dimensions described were even smaller.

Its pathogenesis is not well defined and various hypotheses are discussed: thrombotic, traumatic or neoplastic origin; however, the most widely accepted is that it originates from the Lambl's excrescences⁷.

It occurs at any age and is more common after age 40. In most cases it is asymptomatic, but may be accompanied by nonspecific conditions such as chest pain, dyspnea, syncope and, more rarely, sudden death⁸.

Our case demonstrates an epidemiological profile similar to that of the literature, both in its location and in its form of presentation^{2,9}.

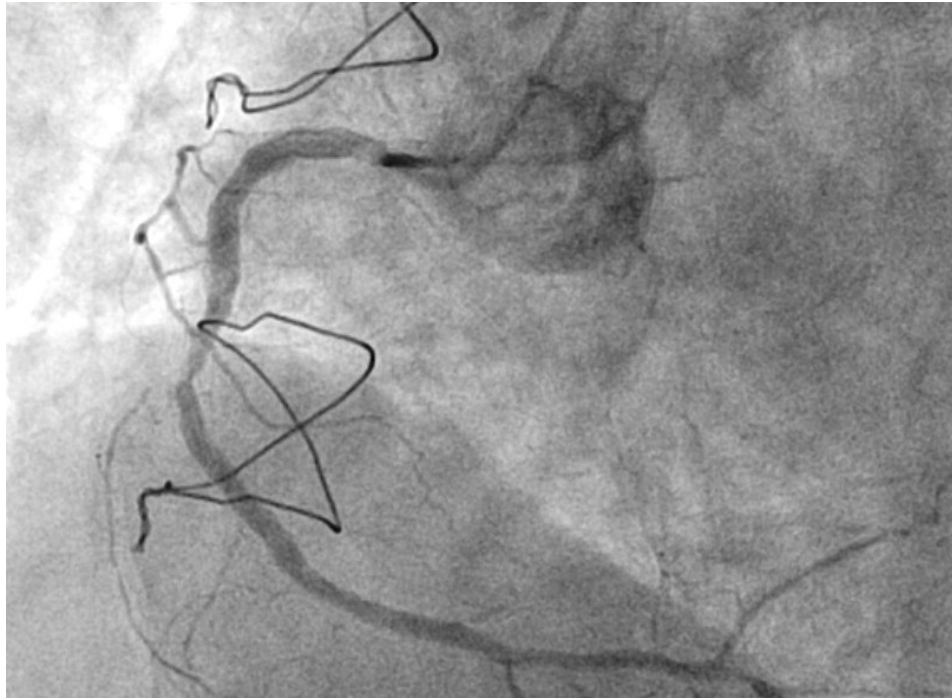


Figure 1 – Coronary angiography of the right coronary artery.

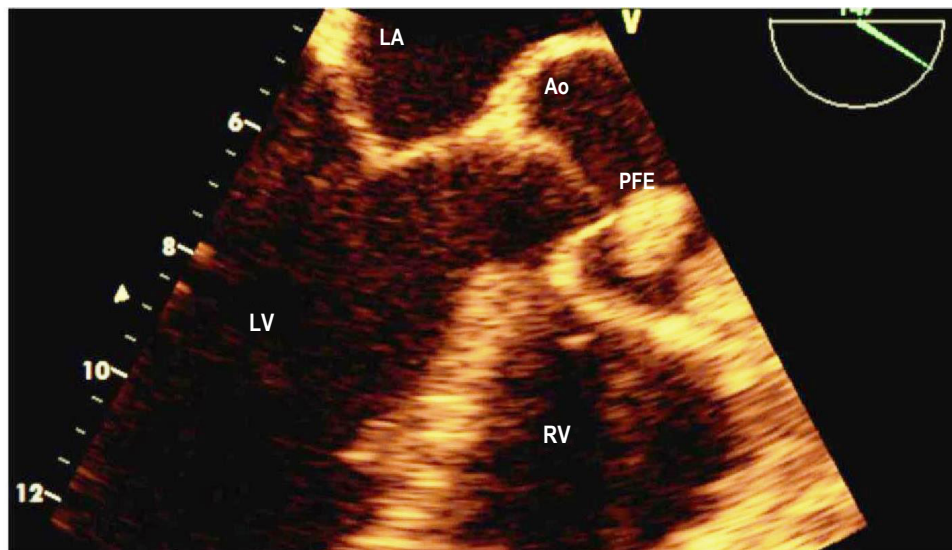


Figure 2 – Transesophageal echocardiography. LA: left atrium; Ao: aorta; LV: left ventricle; RV: right ventricle; PFE: papillary fibroelastoma.

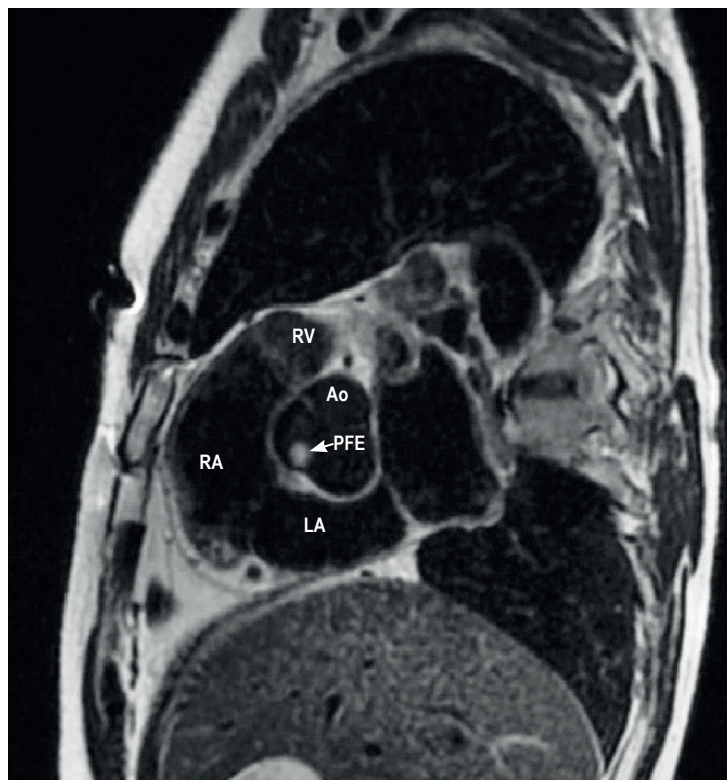


Figure 3 – Magnetic resonance imaging. LA: left atrium; RA: right atrium; RV: right ventricle; Ao: aorta; PFE: papillary fibroelastoma.

The complications described are a source of concern about the finding of PFE, particularly those related to embolic phenomena, whether peripheral or central. Such events depend on its size, location and mobility, in addition to the platelet aggregation triggering potential^{2,5}, and include: AMI, strokes, IIA, mesenteric and renal ischemia.

Specifically in tumors localized in the aortic valve, complications such as sudden death and AMI are observed². Other nonspecific findings may also be present, such as atrial arrhythmias and increased cardiac chambers.

Echocardiographic diagnosis has a sensitivity of 88.9% and specificity of 87.8%^{2,6,8}, especially in cases of larger tumors. The characteristic finding is a mass similar to a “sea anemone,” with multiple ramifications held by a pedicle to the endocardium. Transesophageal echocardiography may be an option when the size or location cannot be properly defined by transthoracic method.

Other diagnostic methods include MRI, which allows better morphological definition of the tumor and its anatomical relations, increasing the degree of suspicion when the enhancement technique is used¹⁰. Coronary angiography is an additional method used for stratifying the coronary lesions associated, and intracoronary ultrasound allows the distinction of atherosclerotic disease from other thrombotic disorders. There are no imaging techniques that allow a

definitive diagnosis, which is determined by histopathological analysis of the tumor.

The patient described has evolved with successive events of AMI associated with thrombotic complications, which should be related to atherosclerotic disease and cannot, however, rule out the possible contribution of PFE in the etiology of these events, considering the close relationship of the tumor with the ostium of the coronary arteries. Definitive diagnosis, however, was not possible, as the assisting medical team opted for conservative treatment, taking into account the clinical conditions of the patient.

The differential diagnosis of PFE is extensive and includes other cardiac tumors (myxoma, lipoma), vegetation, thrombus, valve degeneration tissue and the Lambl's excrescence.

The treatment of choice for symptomatic patients with associated complications is surgical resection. As for asymptomatic patients, the treatment of choice is still undefined, but it should be based on the size, location and especially on mobility, which is an independent factor for severe complications^{2,6,8}.

The procedure is curative in most cases, with resection made to the base of implant to prevent recurrence; or valve replacement if the repair is not possible. Patients not undergoing surgery should remain in full anticoagulation, although of uncertain efficiency^{6,8}.

Conclusion

Papillary fibroelastoma is an extremely rare medical condition in our community, of often incidental diagnosis. Although benign in nature and predominantly asymptomatic, its clinical importance is based on the occurrence of potential embolic complications due to the high morbidity and mortality associated. In this context, it turns out to be a hypothesis to be investigated in patients with coronary and/or cerebrovascular events of unknown etiology.

Although it lacks confirmation by invasive coronary and pathological methods, we report the occurrence of PFE in patients with previous episodes of AMI, emphasizing the importance of the association of these events. Surgical treatment is the preferred option, although it is not performed in this case by joint decision of the multidisciplinary team.

References

1. Reynen K. Frequency of primary tumors of the heart. *Am J Cardiol.* 1996;77(1):107.
2. Gowda RM, Khan IA, Nair CK, Mehta NJ, Vasavada BC, Sacchi TJ. Cardiac papillary fibroelastoma: a comprehensive analysis of 725 cases. *Am Heart J.* 2003;146(3):404-10.
3. Fabricius AM, Heidrich L, Gutz U, Mohr FW. Papillary fibroelastoma of the tricuspid valve chordate with a review of the literature. *Cardiovasc J S Afr.* 2002;13(3):122-4.
4. Sun JP, Asher CR, Yang XS, Cheng GG, Scalia GM, Massed AG, et al. Clinical and echocardiographic characteristics of papillary fibroelastomas: a retrospective and prospective study in 162 patients. *Circulation.* 2001;103(22):2687-93.
5. Law KB, Phillips KR, Cusimano RJ, Butany J. Multifocal "tapete" papillary fibroelastoma. *J Clin Pathol.* 2009;62(12):1066-70.
6. Mariscalco G, Bruno VD, Borsani P, Dominici C, Sala A. Papillary fibroelastoma: insight to a primary cardiac valve tumor. *J Card Surg.* 2010;25(2):198-205.
7. Saxena P, Konstantinov IE, Lee A, Newman MA. Papillary fibroelastoma of aortic valve: early diagnosis and surgical management. *J Thorac Cardiovasc Surg.* 2007;133(3):849-50.
8. Oliveira SFM, Dias RR, Fernandes F, Stolf N A C, Mady C, Oliveira SA. Cardiac papillary fibroelastoma: experience of an institution. *Arq Bras Cardiol* 2005;85(3):205-7.
9. Chryssagis K, Liangos A, Westhof F, Batz G, Diegeler A. Transesophageal echocardiography for detection of a papillary fibroelastoma of the aortic valve. *Hellenic J Cardiol.* 2010;51(2):170-4.
10. Fujita N, Caputo GR, Higgins CB. Diagnosis and characterization of intracardiac masses by magnetic resonance imaging. *Am J Card Imaging.* 1994; 8 (1):69-80.

Authors' contributions

Research creation and design: Vieira TA, Negreiros SBC, Sousa DWS. Data acquisition: Vieira TA, Negreiros SBC, Sousa DWS. Data analysis and interpretation: Vieira TA, Negreiros SBC, Sousa DWS.

Potential Conflicts of Interest

There are no relevant potential conflicts of interest.

Sources of Funding

This study had no external funding sources.

Academic Association

This study is not associated with any graduate program.