Recurrent Myocardial Infarction in a Patient with Papillary Fibroelastoma

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This report describes a case of a 62 year-old man admitted for recurrent myocardial infarction and normal coronary arteries, caused by coronary embolism from aortic papillary fibroelastoma. Other conditions causing acute coronary syndrome and normal coronary arteries are discussed. A careful evaluation by transthoracic and transesophageal echocardiography is required in this clinical setting. Surgical excision of the tumor is safe and curative.

Introduction

Myocardial infarction with normal coronary arteries is a syndrome resulting from a number of conditions that may include coronary embolism, coronary spasm, coronary artery anomalies, coronary dissection, hyper-coagulating states and imbalance of blood flow and supply. One of the rare sources of coronary embolism is cardiac papillary fibroelastoma, which is the third most common primary heart tumor, which most commonly affects the cardiac valves[1]. We report a case of aortic valve papillary fibroelastoma causing recurrent myocardial infarction, detected by echocardiography.

Case report

A 62 year-old man with history of hypertension, hypercholesterolemia and paroxysmal atrial fibrillation (not on oral anticoagulation) was admitted to our hospital with complaints of chest tightness with sudden onset at rest. The electrocardiogram (ECG) on admission showed atrial fibrillation with rapid ventricular rate and normal ST-segment and T waves. Physical examination was unremarkable but there was elevation of cardiac biomarkers, with troponin I peak of 1.84 ng/ml and CK-MB mass of 13.4 ng/ml. The initial diagnosis was non-ST elevation acute myocardial infarction and the coronary angiography showed no significant coronary stenosis. The echocardiogram revealed normal left ventricle function without regional wall motion abnormalities; the valves had no significant morphological or functional changes and the pericardium was normal. Having a $CHADS_2$ score of 1, the patient was discharged on Aspirin 100, ramipril 5, bisoprolol 5 and rosuvastatin 10.

Three weeks later, the patient started complaining of epigastric pain irradiating to the chest. ECG showed atrial fibrillation with rapid ventricular rate and negative T waves in leads V4-V6. Troponin I and CK-MB mass rose to a peak value of 13.4 ng/ml and 52.7 ng/ml, respectively. Coronary angiography revealed no evidence of coronary stenosis.

To rule out the possibility of an embolic source, transesophageal echocardiogram (TEE) was performed revealing a highly mobile 13 mm long filamentous mass with a nodular edge, attached to the aortic valve right coronary cusp, proximal to the right coronary artery ostium (figure 1). This exam also showed intense spontaneous echo contrast in the left atria, but no images of thrombi.

The patient underwent surgical excision of the lesion, and a 3 mm tumor arousing from the comissure between the right and left coronary ostia was removed. Atrial fibrillation surgery was simultaneously performed, consisting of left atrial appendage excision, left atria denervation and isolation of pulmonary veins.

Histopathological examination confirmed the diagnosis of papillary fibroelastoma (figure 2). The patient was discharged from hospital on the ninth day after surgery and remained asymptomatic at three-month follow-up.

Discussion

Although most acute coronary syndromes are caused by atherothrombosis, they may occur in patients with coronary arteries that appear normal in angiography. Myocardial infarction (MI) and angiographically normal coronary arteries have been described in the literature for more than 30 years but they are always a challenge due to unclear pathophysiology, management and prognosis. Indeed, even when alternative etiologies for myocardial ischemia are sought, they can be detected in only one third of the cases[2]. The overall prevalence rate of MI with normal coronary arteries ranges from 1 to 12% and seems to be more frequent in female and younger patients[3]. Microvascular disease (X-syndrome) and

Keywords

Myocardial Infarction; Heart Neoplasms; Embolism.

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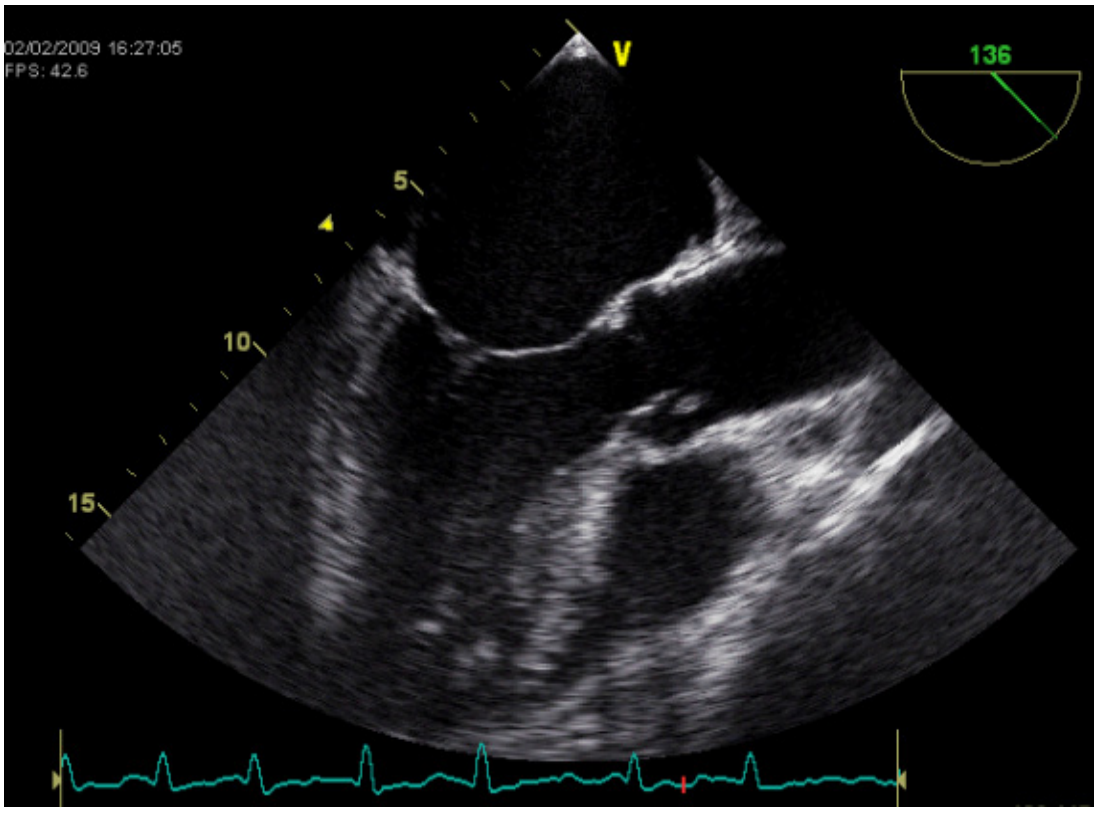

Figure 1 - *TEE shows a nodular mass (arrow) attached to the aortic right coronary cusp by a short pedicle.*

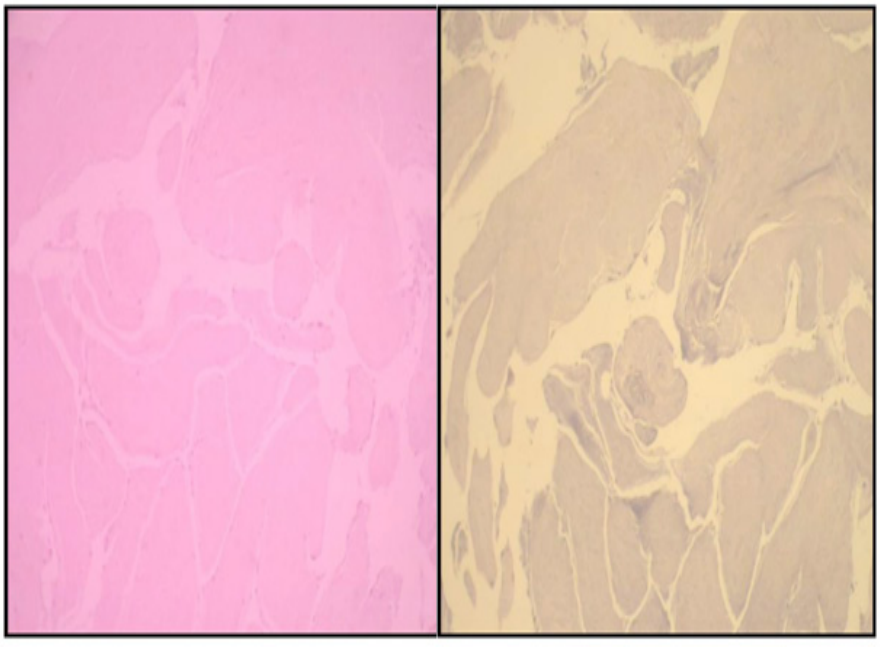

Figure 2 - *Microphotograph demonstrates that the tumor is composed of a number of papillary fronds which consist of a central core of dense, hyalinised, acellular collagen, and a single layer of endothelial cells lining the papillae (hematoxylin-eosin stain on left). Elastic fibers were demonstrable within the fronds in the elastic van Gieson stained section on right.*

inbalance between oxygen demand and supply are frequent causes of MI without coronary stenosis. Coronary artery spasm is a diagnosis to bear in mind, especially when associated with cocaine or smoking in young patients[4]. Coronary embolism is an unusual cause of myocardial ischemia and accounts for 5 to 10% of all paradoxical embolisms[5], although there are other sources of embolization such as left atrial thrombus, thrombosed prosthetic valves, left heart-sided endocarditis and left-sided cardiac and valve tumors. Coronary artery dissection and coronary artery anomalies[6] such as myocardial bridges and origin from the opposite sinus are rare causes of myocardial ischemia. Hyper-coagulating states have also been hypothesized as possible causes in this context. In other conditions such as primary myocardial disorders, congestive heart failure with diastolic dysfunction, amyloidosis, hypothyroidism[7] and "tako-tsubo" cardiomyopathy have been described as leading to an increase in cardiac troponins in patients without epicardial coronary artery disease.

In this case report, recurrent MI was caused by coronary embolism due to a papillary fibroelastoma (PFE) of the aortic valve. This is the third most common cause of primary cardiac tumors, accounting for 7% of reported cardiac tumors[1]. Out of these, 80% arise from the valvular endocardium and the aortic valve is the most commonly involved[8]. The majority of patients with PFE are asymptomatic and, as a result, most of these tumors are found incidentally on autopsy or cardiac surgery. Nevertheless, it can result in life-threatening complications such as coronary and cerebral embolism or acute valvular dysfunction. Left-sided PFE and highly mobile tumors are more prone to embolization and embolic fragments may originate from the tumor itself or from surface formation of platelet and fibrin thrombi[8]. Acute MI may also result from direct occlusive effects on the coronary arteries. Transthoracic echocardiography is the initial exam for evaluation of suspected PFE, although transesophageal echocardiography is often necessary for a more accurate assessment. On echocardiogram, PFEs are hyper-echogenic, highly mobile, spherical or ovoid, usually homogeneous and with sharply delimited masses. Identification of PFE is important because it represents a correctable cause of embolic events. Surgical excision is curative and the long-term post-operative prognosis is excellent. In asymptomatic patients, the mobility of the tumor is considered a determining factor for surgery decision because it is an independent predictor of death or non-fatal embolization[8].

This case raises the important issue of MI with normal coronary arteries and the need to seek its etiology. This clinical setting led us to consider a possible embolic source, especially in a patient with atrial fibrillation who was not on oral anticoagulation. Actually, this patient was found to have two potential embolic sources and even though no atrial thrombi were found, there was intense spontaneous echo contrast and low left atrial appendage velocities. Moreover, there are reports of acute coronary syndromes secondary to thromboembolism, in which atrial fibrillation is the most frequent risk factor[9]. However, the highly embolic pattern of PFE in addition to the discrepancy between tumor size assessed by echocardiography and during surgical resection strengthen the hypothesis of embolization of tumor fragments to the coronary arteries.

In conclusion, this diagnosis must be suspected in the presence of MI with normal coronary arteries. A careful evaluation by echocardiography should always be performed in this clinical setting. Surgical excision of the tumor is safe and curative.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any post-graduation program.

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