

Multiple Cardiac Papillary Fibroelastoma and Transient Left Ventricular Apical Ballooning Syndrome in an Elderly Woman: Case Report

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An elderly Japanese woman presented with evidence of a myocardial infarction. Emergency angiography showed no significant atherosclerotic disease, but the anterior and anteroseptal walls were akinetic, with 'ballooning' of the apex. She was suspected to have transient left ventricular apical ballooning syndrome (TLVABS). Two months later, transthoracic echocardiography showed normal left ventricular wall motion and function, together with a 5-mm cardiac papillary fibroelastoma (CPF) attached to the aortic valve. Transesophageal echocardiography showed a stalked CPF on the aortic side of the left coronary cusp, and a smaller CPF on the right coro-

Transient left ventricular apical ballooning syndrome (TLVABS) may represent an unusual manifestation of transient cardiac ischemia. Whilst TLVABS has been mainly reported in Japanese patients, more recent studies have suggested that other populations might also be affected.

Case report

An 81-year-old Japanese woman presented with a 2-h history of central chest pain suggestive of myocardial infarction. Other than a history of hypertension, she had no other risk factors for coronary artery disease. Emotional distress was not a precipitating factor. The patient's blood pressure was 140/80 mmHg and the heart rate 100 beats per min, and regular. Electrocardiography showed ST elevation in the anterolateral leads (V2-V6, I and aVL). The initial troponin I level was 0.3 ng/ml, but this had peaked at 4.2 ng/ml by the next morning. The patient was not in cardiogenic shock or heart failure, nor did she develop

nary cusp. It was hypothesized that the CPF caused the TLVABS through myocardial stunning. This may occur as a result of transient dynamic ostial occlusion by the fibroelastoma, or because of emboli from the fibroelastoma which then subsequently spontaneously lysed. This syndrome may represent an unusual manifestation of transient cardiac ischemia. Whilst TLVABS has been mainly reported in Japanese patients, more recent studies have suggested that other populations might also be affected.

The Journal of Heart Valve Disease 2005;14:137-139

any arrhythmias. Emergency coronary angiography showed an absence of any significant coronary artery disease, but there was akinesia of the entire apex of the left ventricle with 'ballooning' (Fig. 1). The patient was treated with metoprolol, low molecular-weight heparin and aspirin. She had no further chest pain and was discharged four days later.

Transthoracic echocardiography performed two months later showed normal left ventricular (LV) function and wall motion. The clinical picture was consistent with that of TLVABS, but the echocardiogram also showed a small 5-mm stalked mass attached to the left coronary cusp (Fig. 2) which was suggestive of a cardiac papillary fibroelastoma (CPF). Transesophageal echocardiography subsequently showed a 7×7-mm

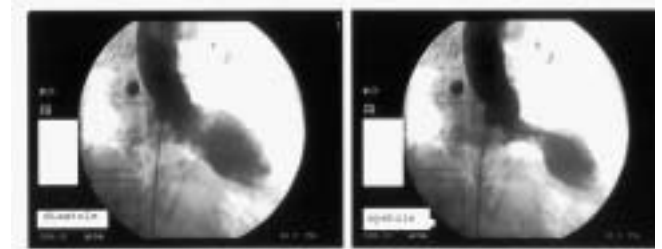


Figure 1: Left ventriculogram during angiography, showing apical ballooning during systole.

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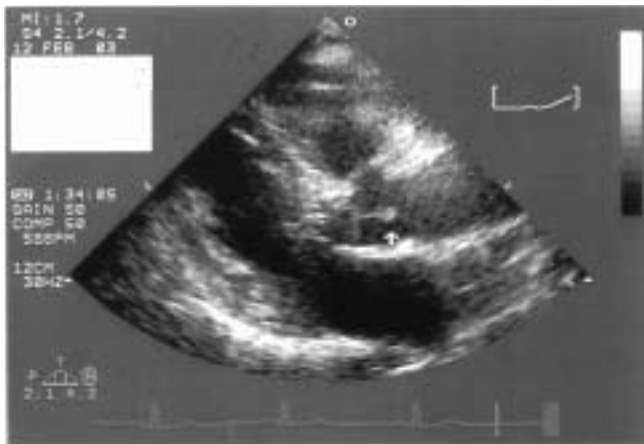


Figure 2: Transthoracic echocardiogram showing a stalked papillary fibroelastoma attached to the aortic valve.

pedunculated CPF attached to the aortic side of the left coronary cusp (Fig. 3), and another smaller CPF attached to the right coronary cusp. In addition, the LV wall motion and function remained normal. The patient declined surgery, citing advanced age as a reason.

Discussion

Cardiac papillary fibroelastomas, although rare, are the second most common primary cardiac tumor and the most common valvular cardiac tumor. CPFs are avascular tumors derived from normal components of the endocardium. They have been described as 'sea anemone' because they consist of multiple frond-like projections; at echocardiography they appear as a small, well-delineated, pedunculated mass with a predilection for valvular endothelium (1-3).

Published case series from The Cleveland Clinic (1), Mayo Clinic (2) and a more recent literature review (4) have suggested that most tumors (~40-50%) are found on the aortic valve. Complications have included transient ischemic attack/cerebrovascular accident (most common), angina, myocardial infarctions, sudden death, pulmonary embolism, blindness, mesenteric

emboli and peripheral embolic. The disease was most prevalent during the eighth decade of life. In the retrospective analysis by Gowda et al. (4), tumor mobility was the only independent predictor of CPF-related death or non-fatal embolization. These authors considered that symptomatic patients should be treated surgically because of the excellent postoperative prognosis.

However, case reports of myocardial infarction resulting from the CPFs have suggested different mechanisms (5-11). One study reported isolating emboli from occluded coronary vessels (9), while another observed dynamic ostial obstruction of the aortic valve coronary ostium resulting in myocardial ischemia (11).

Transient left ventricular apical ballooning syndrome is characterized by acute-onset, transient (i.e. reversible) LV apical wall motion abnormalities with chest pain, electrocardiographic changes (ST elevation, depression, abnormal Q waves) and minimal myocardial enzyme release mimicking myocardial infarction in patients without angiographic stenosis on coronary angiography (12). This syndrome is of unclear etiology.

Most of the patients reported have been elderly females in their seventh decade of life. Precipitating events have included emotional disturbances, exacerbations of existing medical problems and surgical procedures (12,13). In published case series, coronary vasospasm was found to be uncommon (12,13). This disease has been reported mainly in the Japanese literature but a recent case series from Belgium suggested that other populations might also be at risk (14).

Acute complications are similar to that of true myocardial infarctions and include arrhythmias, cardiac failure and cardiogenic shock.

The present case report is interesting because the patient had both CPFs and features consistent with TLVABS. It would be extremely rare for both conditions to occur unrelated in the same patient. Because the pathophysiology and etiology of TLVABS remains unclear, it is hypothesized that the CPFs might have resulted in emboli to the coronary arteries or caused dynamic ostial obstruction with resultant myocardial stunning that manifested as TLVABS. This case suggests that TLVABS may have more than one causative precipitating factor. It is conceivable that the syndrome simply represents an unusual manifestation of transient cardiac ischemia.

However, if the underlying mechanism of the LV apical ballooning were due to ischemia, there must be other reasons for the LV apex being particularly susceptible and ballooning. Postulated factors included: (i) an easy loss of elasticity of the LV apex after excessive expansion; (ii) the LV apex being at the border zone of the perfusion area of the major coronary arter-

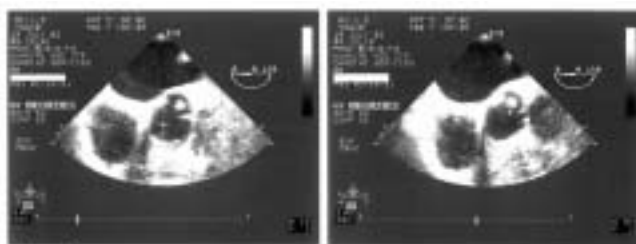


Figure 3: Transesophageal echocardiogram showing the papillary fibroelastoma.

ies; and (iii) there being a delay in functional recovery from global dysfunction (12). Hence, TLVABS might represent a pathological end-point for a variety of precipitating causes including emboli, dynamic ostial obstruction, thrombus or arterial spasm, all of which can result in ischemic and myocardial stunning.

Currently, there is no explanation available for the predilection of this disease in elderly Japanese females. However, with new data provided by Desmet et al. (14), clinicians should begin to consider the possibility of this diagnosis in patients of other ethnic groups.

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