

A rare case of tricuspid valve fibroelastoma complicated by late onset takotsubo syndrome after surgery

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Abstract

Background Takotsubo syndrome (TTS) often develops following various types of stressors including cardiac surgery, and may be fatal in rare cases. Case summary We report a rare case of right atrial papillary fibroelastoma that was complicated by fatal takotsubo syndrome after successful resection. The patient was evaluated for chest pain and right atrial mass that was confirmed by computed tomography and trans-esophageal echocardiography. Pathology findings were consistent with papillary fibroelastoma of the tricuspid valve. Three weeks after successful surgery, the patient was admitted due to cardiogenic shock with echocardiographic findings of apical ballooning and left ventricular outflow tract obstruction, consistent with takotsubo syndrome. Patient died one hour after her admission despite optimal medical therapy.

1 Introduction

Papillary fibroelastoma is a benign cardiac tumor which is typically located on the aortic valve, and less commonly on the other valves and cardiac structures (1,2). Multi-imaging approach is recommended for diagnosis, using trans-esophageal echocardiography and cardiac computed tomography. Surgical resection is recommended in order to prevent embolic events, and is usually definitive as the recurrence rate is very low (3). One of the rare complications of cardiac surgery may be takotsubo syndrome, which is characterized by left ventricular apical ballooning and basal segment hypercontractility. Takotsubo syndrome is considered acute reversible type of cardiomyopathy; however, life threatening cardiogenic shock may complicate the course of this disease.

2 Case report

A 70-year- old female with past medical history of hypertension and well-controlled diabetes mellitus was evaluated for chest pain and shortness of breath during the last year. Her physical examination was unremarkable with no cardiac murmurs or jugular venous distension. Echocardiography revealed normal systolic left ventricular function and the presence of mobile mass in the right atrium (**Fig. 1A**). Transesophageal echocardiography revealed a mobile mass in the right atrium, attached to the base of the septal leaflet of the tricuspid valve with no evidence for tricuspid valve regurgitation (**Fig.1B**). Chest computerized tomography showed a 1 cm mass in the right atrium with no extension to the vena cava (**Fig. 1C**). Differential diagnosis included right atrial myxoma vs. right sided papillary fibroelastoma. After discussion with the cardiac surgery team, a decision for mass excision was taken. Coronary angiogram before the surgery showed patent coronary arteries with no obstructive disease (**Fig. 2**). The patient was referred for cardiac surgery, and 1 cm mass was resected from the right atrium (**Fig. 3**), with no complications during the surgery or the post-operative course. She was discharged five days later in good clinical condition with no signs of heart failure. Three weeks later, the patient was admitted with weakness, agitation, tachycardia 100 beats

per minutes and blood pressure of 70/30 mmHg with diffuse ST segment changes in the ECG (**Fig. 4A**). The patient was started on intravenous noradrenaline and fluids, without any hemodynamic improvement. Echocardiography revealed severe apical ballooning with left ventricle outflow obstruction (**Fig.4B**). TTS was highly suspected based on the typical echocardiographic appearance and the normal coronary angiography three weeks earlier. Noradrenaline was hold and she was treated with beta-blockers, however, without any clinical or hemodynamic response. The patient died one hour after admission.

Discussion

TTS, also known as the 'broken-heart' syndrome is a unique form of transient acute left ventricular dysfunction that predominantly affects post-menopausal women. It mimics acute coronary event in that it presents with chest pain, ECG changes and elevated cardiac biomarkers. Most patients report an emotional or physical trigger shortly before the presentation, however, about 30% of cases lack any obvious trigger (4, 5). The pathophysiology is not fully understood, but several theories have been proposed to explain the underlying mechanisms. High levels of plasma catecholamines and neuropeptides have been reported in patients with TTS (5). Other possible mechanisms include coronary endothelial dysfunction, various cytokines activation, impaired fatty acid metabolism in cardiac tissue, and multi-vessel spasm (6). Post-operative TTS has been previously reported after cardiac surgery such as mitral valve replacement and atrial myxoma resection and may be explained by the high catecholamine levels after surgery (7, 8). In addition, delayed onset TTS has been documented several weeks following mitral valve surgery, epileptic seizures, or even after routine exercise stress test (9, 10). Although delayed onset TTS is usually described 24-48 h after physical or emotional trigger, a wide time range has been reported. In one series of 25 epileptic patients, post seizure TTS was detected 5-288 hours post-ictally (11). In our case, the most probable trigger for TTS is the previous surgery with its accompanied stress and pain during the post-operative days. No other etiology could explain her dramatic presentation, given the normal angiogram that was performed three weeks earlier and the lack of fever or any inflammatory markers in her blood tests. In addition, her echocardiography demonstrated preserved systolic function of the right ventricle without dilation, findings that actually exclude massive pulmonary embolism.

Treatment of TTS depends on the patients' clinical condition. Hemodynamically stable low risk patients with preserved (or mildly reduced) systolic left ventricular ejection fraction and without significant secondary mitral regurgitation may benefit from beta blockers (12). Cardiogenic shock in TTS may be caused by several mechanisms including ventricular arrhythmia, left ventricular dysfunction, right ventricular involvement, and left ventricular outflow tract obstruction with secondary mitral regurgitation, mechanical circulatory support may be needed in such cases (13).

4 Conclusion

Takotsubo syndrome should always be included in the differential diagnosis among patients with acute chest pain or unexplained shock following cardiac surgery. Although rare, fatal complications such as left ventricular outflow obstruction with secondary severe mitral regurgitation may occur and should be identified and managed urgently.

Conflict of interest : The authors declare no conflicts of interest.

Ethics

Informed consent was obtained from the family for publication of the paper.

A waiver from the local ethical committee in the hospital was also obtained.

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Figure legends

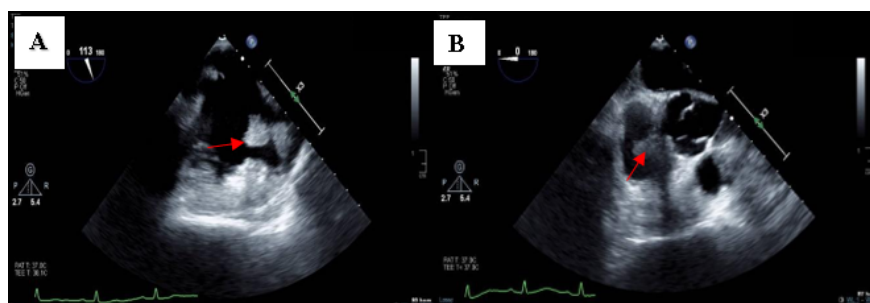
Figure 1(A-C): Transesophageal echocardiography showing mobile mass in the right atrium in the bicaval (A) and short axis (B) views (arrows). Chest CT (C) showing the same mass in the right atrium (arrow). The mass is attached to the septal leaflet of the tricuspid valve. The differential diagnosis was between right sided atrial myxoma and fibroelastoma.

Figure 2: Coronary angiography showing patent left system (left) and right coronary artery (right) with no evidence for atherosclerotic obstructive lesions.

Figure 3: The resected mass from the right atrium. The pathology findings were consistent with fibroelastoma. This is a rare location of fibroelastoma as the most common sites for these tumors are the ventricular side of the aortic valve followed by the atrial side of the mitral valve.

Figure 4(A-B): ECG showing diffuse ST segment changes with minimal ST elevation in the anterior leads

(A), finding that has been reported in many patients with TTS. The initial ECG before surgery was completely normal. Apical four chambers view (B) showing prominent apical ballooning typical for takotsubo. Cine echocardiography showed basal hypercontractility with dynamic mitral regurgitation secondary to left ventricular outflow tract obstruction.

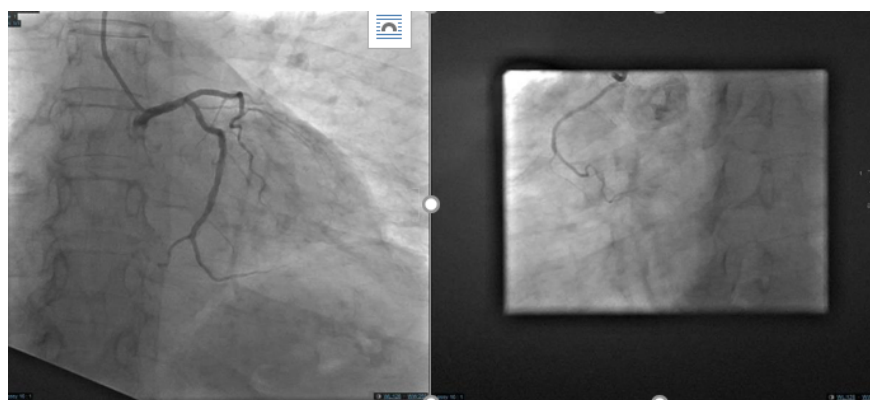


TEE: Bicaaval view

TEE: Short axis view



Computed tomography



Coronary angiography

