

# Late Onset Takotsubo Following Surgery of Tricuspid Valve Fibroelastoma

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Takotsubo syndrome (TTS) often develops following stressors such as cardiac surgery. It may be fatal in rare cases. Our 70-year-old female patient presented with a rare case of tricuspid valve papillary fibroelastoma, which was complicated by fatal TTS after successful resection. The patient had a right atrial mass that was investigated with computed tomography and transesophageal echocardiography (TTE). She was scheduled for surgery. 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 TTS. The patient died one hour after her admission despite optimal medical therapy.

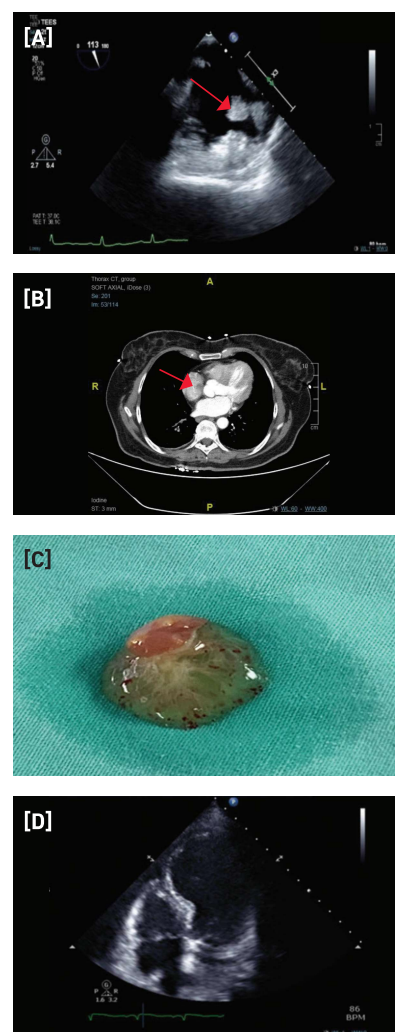
## PATIENT DESCRIPTION

A 70-year-old female with past medical history of hypertension and well-controlled type 2 diabetes mellitus was evaluated for chest pain and progressive dyspnea the year before admission. Her physical examination was unremarkable with no cardiac murmurs or jugular venous distension. During TTE, a rounded mass was demonstrated in the

right atrium with no valvular pathology and with normal systolic left ventricular function [Figure 1A]. Transesophageal echocardiography (TEE) showed a mobile mass (1 × 1 cm), attached to the base of the septal leaflet of the tricuspid valve with no evidence of tricuspid valve regurgitation [Figure 1B]. Chest computed tomography (CT) revealed that the mass was restricted to the tricuspid valve with no extension to the vena cava [Figure 1C]. Differential diagnosis included right atrial myxoma and right sided papillary fibroelastoma. After discussion with our cardiothoracic team, a decision was made to proceed with surgery for mass excision. Coronary angiography showed patent coronary arteries with no obstructive disease. A 1-cm mass was separated from the septal leaflet of the tricuspid valve [Figure 1D] with no complications. The patient was discharged 5 days later in good clinical condition with no signs of heart failure. Echocardiography at discharge was within normal limits with good systolic function, no valvular pathology, no pericardial effusion, and no wall motion abnormality. Three weeks later, she was admitted with weakness, agitation, tachycardia 100 beats per minute, and blood pressure of 70/30 mmHg with diffuse ST segment changes. The patient was started on intravenous noradrenaline and fluids with no hemodynamic improvement. Echocardiography revealed apical ballooning with basal hypercontractility and left ventricle outflow obstruction without

**Figure 1.** The tricuspid fibroelastoma after excision

**[A]** Transesophageal echocardiography, **[B]** Computed tomography, **[C]** After excision, **[D]** Typical echocardiograph of Takotsubo in the fourth chamber view



pericardial effusion [Figure 1E]. TTS was highly suspected based on the typical echocardiographic appearance and the normal coronary angiography performed 3 weeks earlier. Noradrenaline was stopped, and she was treated with beta-blockers without any clinical or hemodynamic response. The patient died one hour after admission.

**COMMENT**

Papillary fibroelastoma is a benign cardiac tumor that is typically located on the aortic valve, and less commonly on the other valves and cardiac structures [1,2]. A multi-imaging approach is recommended for diagnosis, using TEE and cardiac computed tomography. Surgical resection is recommended to prevent embolic events and is usually definitive as the recurrence rate is very low [2,3]. One of the rare complications of cardiac surgery is TTS, which is characterized by left ventricular apical ballooning and basal segment hypercontractility. TTS is an acute reversible type of cardiomyopathy that mimics acute coronary event with similar presentation, ECG changes, and elevated cardiac troponin [4]. In this case, the most reasonable trigger for TTS was the cardi-

ac surgery with the accompanied stress and pain during the postoperative days. The previous normal angiogram and the complete normal echocardiography on the day of discharge made coronary etiology less reasonable. Moreover, the absence of fever in the postoperative course and on the day of readmission, as well as normal inflammatory markers levels, sterile blood, and urine cultures (obtained later) practically excluded infectious cause. Last, the right ventricle was not dilated and with preserved contractility, findings that exclude massive pulmonary embolism. Treatment of TTS depends on the clinical condition and echocardiographic features. Hemodynamically stable patients may benefit from beta blockers and angiotensin converting enzyme inhibitors. Mechanical circulatory support may be needed in cases of cardiogenic shock, which may be caused by ventricular arrhythmia, severe left ventricular dysfunction, right ventricular involvement, and left ventricular outflow tract obstruction with secondary mitral regurgitation.

**CONCLUSIONS**

TTS should always be included in the differential diagnosis of patients with

unexplained shock following cardiac surgery. Although most cases of TTS exhibit a benign course, fatal complications may occur and should be identified and managed urgently.

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**Capsule**

**Effectiveness of a meningococcal group B vaccine (4CMenB) in children**

In September 2015, the four-component, protein-based meningococcal serogroup B vaccine (4CMenB; Bexsero) became available for private purchase in Spain. **Castilla** and colleagues conducted a nationwide matched case control study to assess the effectiveness of 4CMenB in preventing invasive meningococcal disease in children. The authors compared 306 case patients (243 [79.4%] with serogroup B disease) with 1224 controls. A total of 35 case patients (11.4%) and 298 controls (24.3%) had received at least one dose of 4CMenB. The effectiveness of complete vaccination with 4CMenB (defined as receipt of at least two doses, administered in accordance with the manufacturer's recommendations) was 76% (95% confidence interval

[95%CI], 57–87) against invasive meningococcal disease caused by any serogroup, and partial vaccination was 54% (95%CI 18–74) effective. Complete vaccination resulted in an effectiveness of 71% (95%CI 45–85) against meningococcal serogroup B disease. Vaccine effectiveness with at least one dose of 4CMenB was 64% (95%CI 41–78) against serogroup B disease and 82% (95%CI 21–96) against non-serogroup B disease. With the use of the genetic Meningococcal Antigen Typing System, serogroup B strains that were expected to be covered by 4CMenB were detected in 44 case patients, none of whom had been vaccinated.

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