

## Case Report

## Transesophageal Echocardiography for Detection of a Papillary Fibroelastoma of the Aortic Valve

KONSTANTINOS CHRYSAGIS<sup>1</sup>, ARIS LIANGOS<sup>1</sup>, FRANK WESTHOF<sup>1</sup>, GERHARD BATZ<sup>2</sup>, ANNO DIEGELER<sup>1</sup>

<sup>1</sup>Department of Cardiovascular Surgery, <sup>2</sup>Department of Anaesthesiology, Rhoen Klinikum, Bad Neustadt, Germany.

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Papillary fibroelastoma (PF) is a benign cardiac tumor, typically attached to the cardiac valves. It is usually found incidentally at autopsy or surgery. It is rarely symptomatic, but can cause myocardial infarction, cerebral infarction and systemic embolism, even in young patients, and sometimes results in sudden death. We report on a case of a PF attached to the *nodulus arantii* of the left coronary cusp in a 56-year-old woman. She was referred to our institution due to a non ST-elevation myocardial infarction diagnosed by troponin positive-test. No other embolic source was identified. Diagnosis was obtained by two-dimensional echocardiography and confirmed by transesophageal echocardiography. After complete surgical removal of the tumor, we reconstructed the left coronary cusp with autologous pericardium. The histology revealed the diagnosis of a PF. Intraoperative transesophageal echocardiography confirmed complete restoration of aortic valve cusp coaptation. The clinical course was uneventful.

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Address:  
Konstantinos Chryssagis

Department of  
Cardiovascular Surgery  
Rhoen Klinikum  
Bad Neustadt  
Salzburger Leite 1  
97616 Bad Neustadt,  
Germany  
e-mail:  
[KChryssagis@t-online.de](mailto:KChryssagis@t-online.de)

**P**apillary fibroelastoma (PF) is a rare benign cardiac tumor, originating in the endocardium and including the heart valves. It is common in older subjects and accounts for 7% of cardiac tumors. It is usually found incidentally at autopsy or surgery and is rarely symptomatic, but it can cause myocardial infarction, cerebral infarction and systemic embolism, even in young patients, and sometimes results in sudden death. For this reason, despite the benign nature of the tumor, surgical removal is necessary. Two-dimensional echocardiography and transesophageal echocardiography are essential in the diagnosis and management of this disease.

We describe the clinical and echocardiographic findings from a middle-aged woman with an aortic valve PF who presented with a non ST-segment elevation myocardial infarction (NSTEMI).

### Case presentation

A 56-year-old woman was referred to our institution after an NSTEMI. On admission we saw the patient in good condition without physical distress. There was no cardiac murmur, her blood pressure was 130/80 mmHg, the heart rate was 53 /min and her body temperature was not elevated. The blood examination data were within normal ranges. No abnormalities were found on chest radiography or on the resting electrocardiogram.

Cardiac catheterization showed low flow in the proximal left anterior descending coronary artery, a slight diffuse coronary atherosclerosis without relevant stenoses and normal left ventricular systolic function. There was considerable aortic valve insufficiency.

Transthoracic two-dimensional and transesophageal echocardiography (General Electric GE vivid i ultrasound device with a

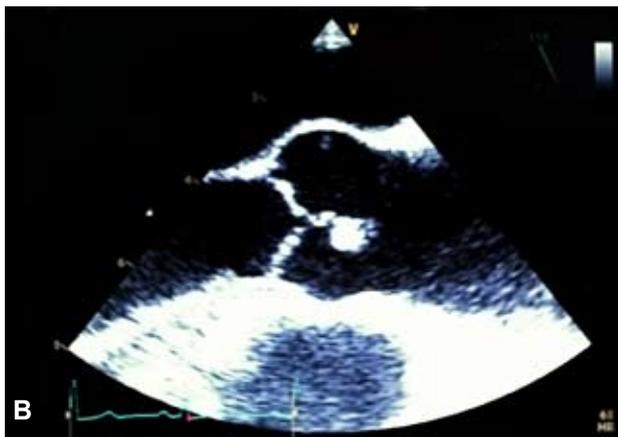
GT-RS TEE-probe) showed a 1 cm long, pedunculated, floating, shaggy structure attached to the aortic valve (Figures 1 & 2). The patient was referred for surgery.

After median sternotomy a pericardial patch, 6 × 8 cm, was excised and fixated in 0.6% glutaraldehyde solution. Cardiopulmonary bypass was instituted in standard fashion using cold crystalloid cardioplegic cardiac arrest.

The ascending aorta was opened and a rounded, pale yellowish, soft friable mass 10 mm in diameter was found, attached to the *nodulus arantii* of the left coronary cusp in an otherwise normal aortic valve. The tumor was excised *in toto* by a triangular resection of the central part of the valve cusp. The defect was replaced by the autologous pericardial patch, which was tailored to match the defect and sutured to the valve cusp with interrupted 7 × 0 Prolene sutures (Figure 3).

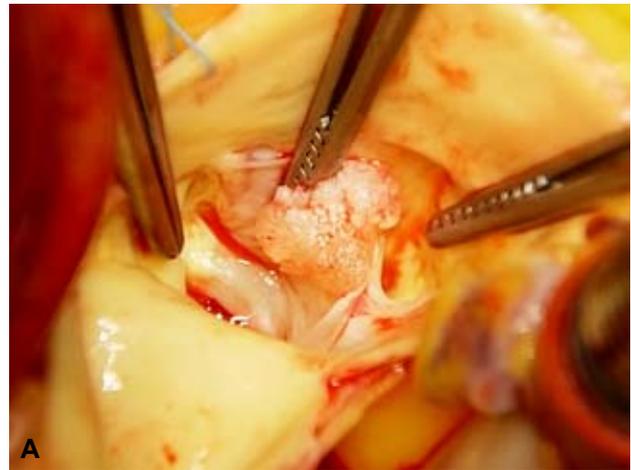


**Figure 1.** Mid-esophageal short-axis view of the aortic valve with the tumor on the left coronary cusp.

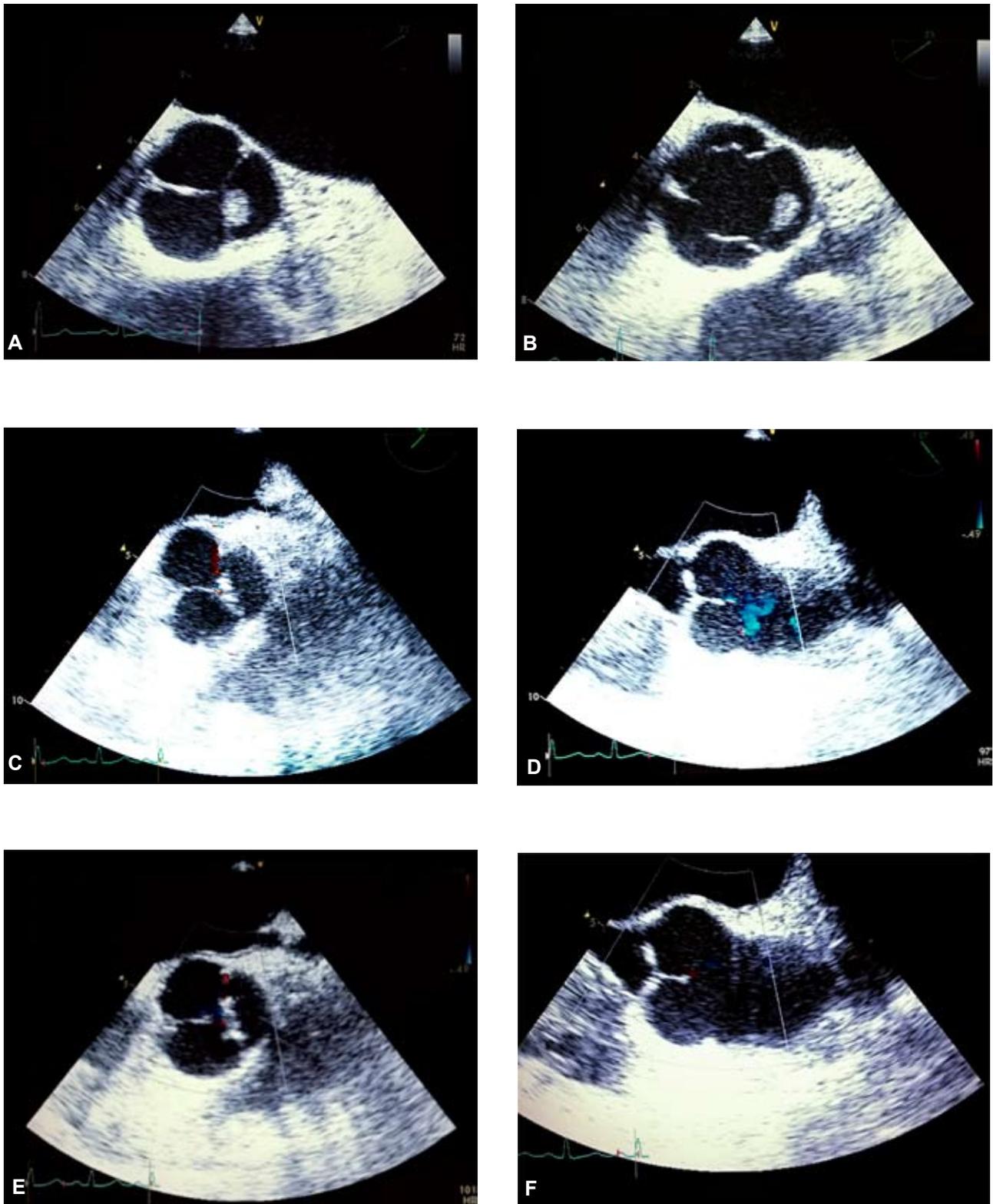


**Figure 2.** Mid-esophageal long-axis view with the aortic valve and ascending aorta showing the tumor.

After closure of the aortotomy and de-airing of the heart and reperfusion the patient was in spontaneous stable sinus rhythm. She was re-warmed and



**Figure 3.** A. Intraoperative view of the tumor. B. Left aortic valve cusp reconstruction, after excision of the tumor, with an autologous pericardial patch. C. Complete aortic valve cusp coaptation after reconstruction.



**Figure 4.** Intraoperative detection of the tumor by transesophageal echo. A. Mid-esophageal short-axis view of the aortic valve with the tumor on the left coronary cusp. B. Mid-esophageal short-axis view of the aortic valve in the open position with the tumor on the left coronary cusp. C. Same view as in A, postoperative aspect after removal of the tumor and reconstruction of the aortic valve. D. Mid-esophageal long-axis view of the postoperative result showing good coaptation of the cusps. E, F. Mid-esophageal short-axis and long-axis views of the corrected valve with color flow Doppler imaging showing an excellent result without any aortic regurgitation.

weaned from cardiopulmonary bypass without difficulties. Intraoperative and postoperative transesophageal echocardiographies (Figure 4) confirmed a normal aortic valve without aortic regurgitation and complete restoration of aortic valve cusp coaptation.

The pathological examination found that the tumor was 7 x 8 x 9 mm in size and resembled a sea anemone (Figure 5). Microscopic examination revealed a papillary fibroelastoma. The surface of the tumor was covered by endocardial cells.

The patient's postoperative course was uneventful and she was discharged on the seventh postoperative day. Six months after the operation the patient had a good quality of life without any complaints.

## Discussion

PF is a rare benign cardiac tumor with multiple papillary fronds resembling a sea anemone and usually attached by a short pedicle. It accounts for about 7% of primary cardiac tumors.<sup>1,2</sup> Most patients are older than 50 years of age, but in some cases this tumor may also develop in infants.<sup>3</sup> Diagnosis has often been made accidentally, either during open heart surgery or at autopsy.<sup>3</sup>

The most frequently affected regions are the aortic and pulmonary valves, although this type of neoplasm may develop at various locations of the mural endocardium with no predilection for the right or left side of the heart.<sup>4,5</sup>

PFs are usually solitary tumors, but more than one intracardiac location in a single patient has been described.<sup>4,6,7</sup> Although most PFs are asymptomatic, severe complications resulting from arterial embolization, such as myocardial infarction, stroke and periph-

eral arterial embolizations are possible. The emboli can be either tumor mass or thrombotic deposits from its surface. Transient occlusion of coronary arteries with a pedunculated piece of the tumor mass can cause temporary myocardial ischemia.<sup>8-12</sup> Surgical resection of the tumor is mandatory in most cases.

The differential diagnosis includes, besides endocarditic vegetation, a fibroma, a myxoma, a rhabdomyoma, a lipoma or a giant Lambl's excrescence.<sup>5,11,13,14</sup>

In our case, the tumor was attached to the left coronary aortic cusp at its *nodulus arantii*, causing temporary myocardial ischaemia with NSTEMI. After confirming the diagnosis using transesophageal echocardiography, we excised the tumor and preserved the aortic valve. Using autologous pericardium we restored the triangular gap at the left aortic cusp. Intraoperative and postoperative transesophageal echocardiography demonstrated complete coaptation and normal appearance of the aortic valve cusps. Therefore, based on our experience, the aortic valve should be repaired rather than replaced in such cases.

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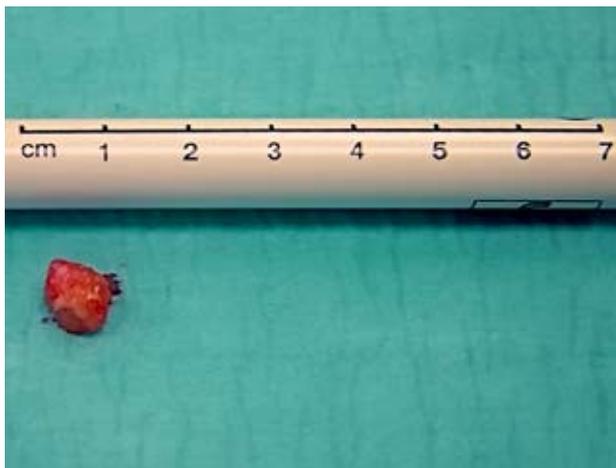


Figure 5. The excised tumor.

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