Papillary fibroelastoma: Rare but important finding

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The term "papillary fibroelastoma" (PFE) was first used in a case report in 1957 [1]. PFE is, next to the myxoma, the most frequent benign primary cardiac tumor and is usually positioned on cardiac valves [2]. Morphologically it resembles a sea anemone with frond-like arms and a stalk. Histologically it consists of endothelial cells surrounding the connective tissue core [3].

This case report relates to a 68-year-old woman who was referred to the cardiology department due to resting chest pain. Her past medical history was positive for arterial hypertension and paroxysmal atrial fibrillation treated with oral anticoagulant. On admission the patient was stable, the chest pain had already resolved, and markers of myocardial necrosis were normal with no electrocardiogram abnormalities. Auscultation of the heart did not reveal any murmurs.

Transthoracic echocardiography showed a 9 × 8 mm, oval-shaped, mobile mass on the left coronary cusp of the aortic valve (Figure 1A). No other abnormalities were detected, and the mass did not cause any hemodynamic abnormalities. A transesophageal echocardiogram confirmed the presence of a pedunculated, ballotable mass attached to the left coronary cusp next to the L-N commissure (Figure 1B–D). No thrombus was present neither in the cardiac chambers nor in the left atrial appendage.

Differential diagnoses included thrombus, bacterial vegetation, and primary cardiac tumors. Due to active anticoagulation, no clinical signs of endocarditis, normal levels of C-reactive protein and procalcitonin, the hypothesis of tumor seemed to be the most plausible.

After a consultation with the Heart Team, a decision was made to pursue catheterization of the coronary arteries before the final decision. Due to the potential risk of embolization, a neuroprotective filter was used. The examination revealed no significant lesions, but after the procedure, a small amount of thrombus was detected in the filter.

The patient was qualified for surgical treatment. The valve-sparing surgery was successfully performed through mini-sternotomy under cardio-pulmonary bypass with concurrent closing of the atrial appendage using the AtriClip system. The mass was fragile and easily fragmented during removal. Histopathology confirmed the primary diagnosis of fibroelastoma (Figure 1E–F).

The postoperative period was uncomplicated. Control transthoracic echocardiography showed no significant regurgitation or stenosis of the aortic valve. The patient was discharged home on the eighth day after surgery in good condition.

Three-dimensional echocardiography images can more accurately define the entire volume of a tumor, its anatomical relationships, and the influence on the valves, which is crucial for differential diagnosing and optimal therapeutic decisions, particularly surgical planning.

Although there are no strict guidelines for the management of PFE, a consensus based on studies [4, 5] recommends surgical excision for symptomatic patients and asymptomatic patients with large (>1 cm), mobile tumors. In patients who do not undergo tumor excision, the risk of cerebrovascular accident is increased due to the possible detaching of PFE fragments or thrombi mobilization.

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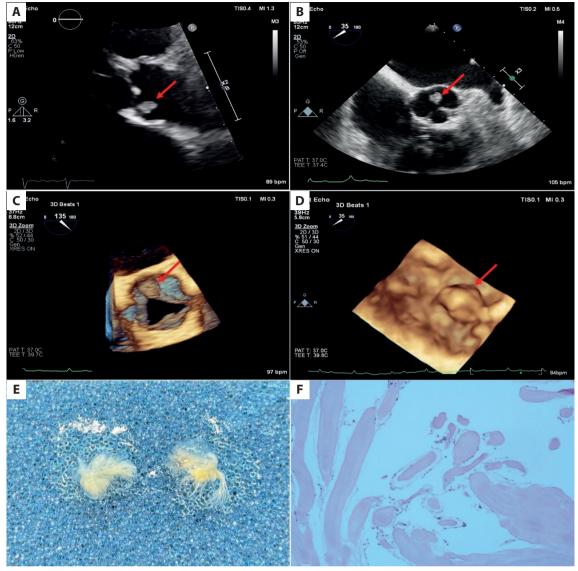


Figure 1. A. A mass on the aortic valve cusps (arrow) in transthoracic echocardiography parasternal long axis view. **B.** The mass in transoesophageal echocardiography mid-oesophageal short axis view (arrow). **C.** Transoesophageal echocardiography 3D view of the mass attached to the L-N commissure (arrow). **D.** Magnified 3D view of the mass (arrow). **E.** Tumor excised during surgery. **F.** Histology specimen with hematoxylin-eosin staining

Anticoagulation has been suggested for poor surgical candidates.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/polish heart journal.

Article information

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