CASE PRESENTATION

Multimodality imaging of a papillary fibroelastoma of the tricuspid valve - case report

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Abstract: **Introduction –** Papillary fibroelastoma is the most common cardiac tumor with valvular localization, involving more frequently the aortic or mitral valve and much rarely the tricuspid or pulmonary valve. It is the second most common primary cardiac tumor, after myxoma. Many patients are asymptomatic, but the tumor represents a potential source of embolism which explains its clinical importance. **Case presentation –** We report the case of a 60-year-old woman with nonspecific symptoms in whom a mass attached to the tricuspid valve was found during a routine transthoracic echocardiography, as part of her cardiac evaluation for an elective orthopedic surgical procedure. Multimodality imaging including transesophageal echocardiography and cardiac MRI was afterwards performed. Since the patient had a high global risk of pulmonary embolism, the tumor was surgically removed with preservation of the tricuspid valve and an uneventful postoperative course. The histopathological examination confirmed the diagnosis of papillary fibroelastoma. **Conclusion –** This case emphasizes the importance of echocardiography as a first-line imaging modality in the diagnosis of cardiac tumors. The embolic risk for papillary fibroelastomas is high and even if the discovery of the tumor is incidental, excision is advisable as surgery has excellent long-term results.
Keywords: cardiac tumor, embolism, papillary fibroelastoma, echocardiography

Rezumat: **Introducere –** Fibroelastomul papilar este cea mai frecventă tumoră cu localizare valvulară, interesând în special valva aortică și mitrală și mult mai rar valva tricuspidă și pulmonară. Fibroelastomul papilar ocupă locul doi ca frecvență între tumorile cardiace primare, după mixom. De multe ori pacienții sunt asimptomatici, însă tumora are potențial emboligen crescut, de aici derivând importanța sa clinică. **Prezentarea cazului –** Raportăm cazul unei paciente în vârstă de 60 de ani cu simptomatologie nespecifică la care s-a diagnosticat prezența unei formațiuni tumorale la nivelul valvei tricuspide cu ocazia unei ecocardiografii transtoracice efectuate în cadrul evaluării cardiologice pentru o intervenție chirurgicală ortopedică electivă. Explorarea imagistică a fost completată ulterior cu ecocardiografie transesofagiană și rezonanță magnetică cardiacă. Riscul global de embolie pulmonară al pacientei fiind mare, s-a decis excizia chirurgicală a formațiunii tumorale, reușindu-se prezervarea valvei tricuspide, cu evoluție postoperatorie favorabilă. Examenul histopatologic a confirmat diagnosticul de fibroelastom papilar. **Concluzii –** Acest caz subliniază importanța ecocardiografiei ca metodă imagistică de primă linie în diagnosticarea tumorilor cardiace. Cu toate că descoperirea este incidentală, pentru fibroelastomul papilar riscul emboligen este ridicat, de aceea este mai bine să fie excizat, chirurgia având rezultate excelente pe termen lung.
Cuvinte cheie: tumoră cardiacă, embolie, fibroelastom papilar, ecocardiografie

INTRODUCTION

Primary cardiac tumors are rare. Their incidence is 0.1% on pathological studies, significantly lower compared to secondary cardiac tumors (metastases) that affect the heart 20 times more frequently[1]. Cardiac tumors can be symptomatic or may be discovered incidentally by various imaging techniques (echocardiography, MRI, CT) during a routine evaluation. Symptoms are nonspecific and may mimic other cardiovascular diseases[1,2].

Most primary cardiac tumors are benign. 10% of them are represented by papillary fibroelastoma, which is the second most common primary cardiac tumor in adults, after myxoma, but the most common tumor with valvular localization, accounting for 85% of cardiac valvular tumors[3,4]. Papillary fibroelastoma can occur at any age but is most commonly seen after the fourth decade of life, usually the diagnosis is made at an average age of 60 years. It can be found everywhere on the endothelial surface, but in 80% of the cases is

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situated on the valves, more frequently on the valves of the left heart. The location on the tricuspid valve is relatively rare and according to literature involves 17% of the cases[3,4,5].

Papillary fibroelastoma can cause symptoms, but up to a third of patients are asymptomatic and the tumor is an incidental finding. The clinical manifestations depend on the location of the tumor and most commonly symptoms are due to embolic events; only in rare cases symptoms are caused by interference of the mass with cardiac function. The papillary fibroelastomas of the right heart are usually asymptomatic, but sometimes, as they may generate tricuspid regurgitation, arrhythmias, intermittent obstruction of the right ventricular outflow tract or pulmonary embolism, they can cause chest discomfort, exertional dyspnea, palpitations, syncope, right heart failure, sudden death[3,6,7].

We present a case of papillary fibroelastoma of the tricuspid valve in a woman admitted to our medical centre in whom the tumor was diagnosed during preoperative cardiac evaluation for a noncardiac surgical procedure.

CASE PRESENTATION

A 60- year-old woman was reffered to our hospital for cardiological assessment prior to undergoing an elective orthopedic surgery (bilateral hip arthroplasty). The patient had multiple cardiovascular risk factors (active smoking, hypertension, dyslipidemia). She complained of palpitations and exertional dyspnea. Her exercise capacity was also severely limited by the coexistence of the orthopedic pathology. No significant abnormalities were found on the physical examination.

As underlined by the guidelines, because the patient was symptomatic we started the evaluation of her perioperative risk with the performance of a 12-lead ECG and a transthoracic echocardiography. The main focus was the assessment of the left ventricular function, as left ventricular systolic dysfunction represents the most important predictor of major cardiac events in patients undergoing noncardiac surgery. The ECG was normal and contrary to initial expectations, transthoracic echocardiography did not reveal any significant changes in the left heart (the left ventricle was mildly hypertrophied with normal ejection fraction), but described instead the presence of an 13/14 mm-size homogeneous echogenic mass on the anterior leaflet of the tricuspid valve; the mass had gelatinous structure, was pedunculated, mobile and did not cause valvular dysfunction (Figures 1, 2).

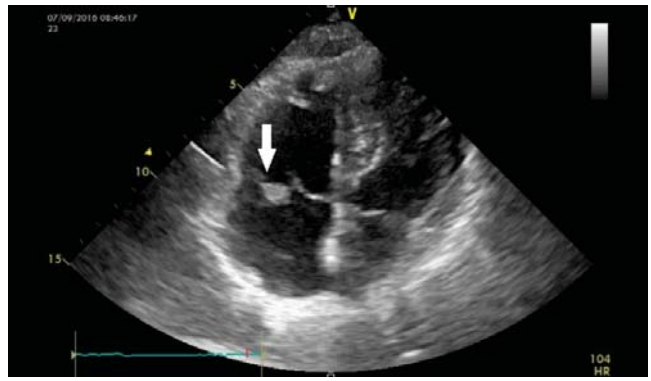

Figure 1. The transthoracic echocardiography (apical 4 chambers view) identifies an echo dense mass (arrow), on the anterior cusp of the tricuspid valve.

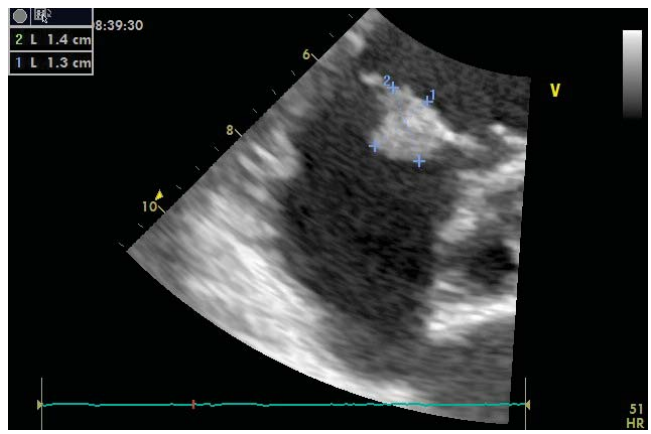

Figure 2. Transthoracic echocardiography (parasternal short axis view) with measuring of the echo dense mass, sized 13/14 mm.

At this point, the differential diagnosis between a cardiac tumor and valvular vegetation was taken into consideration, but in the absence of clinical and biological evidence of infection, we thought that the described mass most likely represented a primary cardiac tumor.

For a more comprehensive and accurate assessment, multimodality imaging was performed. The transesophageal echocardiography confirmed the feature of the mass described transthoracically and highlighted a mild tricuspid regurgitation, unrelated to the presence of the tumor on the valve as the cooptation of the tricuspid leaflets was not influenced by the punctiform insertion of the tumor's pedicle (Figure 3). We continued the evaluation with MRI which offers in a single examination the assessment of morphology, anatomy and functional impact of a cardiac tumor, with a unique ability of tissue characterization, compared to 3D-Echo and cardiac CT. The contrast-enhanced MRI described on the anterior leaflet of the tricuspid valve a 10/6 mm well defined nodular structure, with inter-

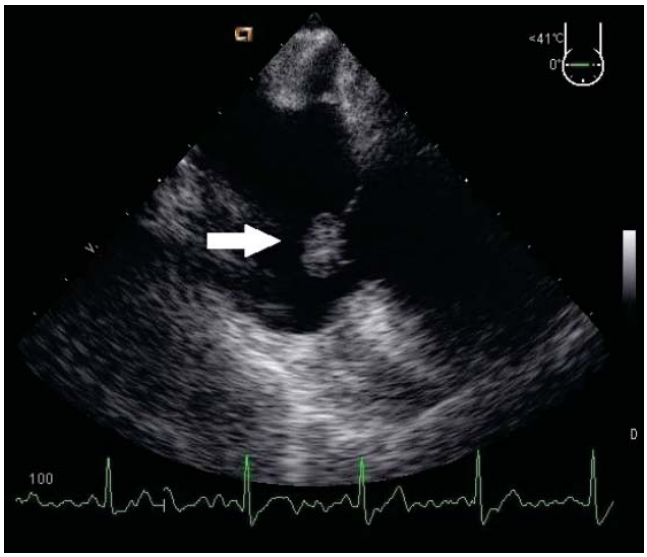

Figure 3. Transesophageal echocardiogram: the mass (arrow) was round, with well-demarcated borders, echo dense, with a homogenous structure, pedunculated, features typical of a papillary fibroelastoma. It was confirmed the presence of the tumor on the anterior cusp of the tricuspid valve.

mediate signal in the T1- and T2- weighted sequences and intense enhancement in the delayed phase. The mass was located at the free edge of the valve, on its atrial side, was mobile with no impact on the closure or opening of the valve (Figures 4, 5). The characteristics of the tumor were suggestive of papillary fibroelastoma. There were no signs of pulmonary embolism (D-dimer levels were normal, there were no clinical, ECG or echocardiographic signs suggestive of pulmonary embolism and no thrombus was described in the main pulmonary arteries on MRI).

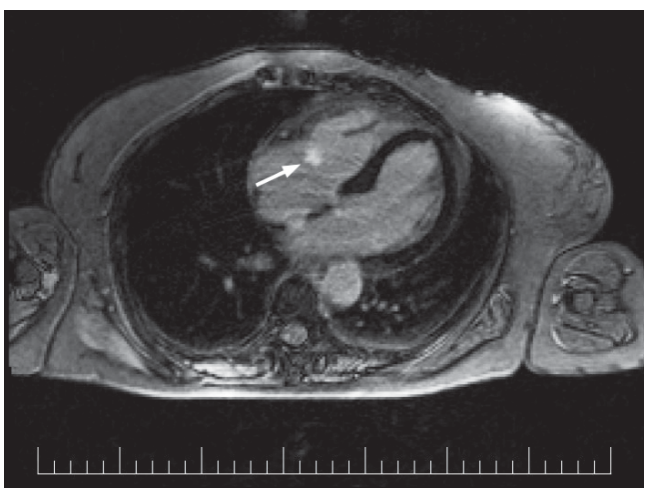

Figure 4. MR examination, T1-weighted sequence with phase sensitive inversion recovery (PSIR) in transversal view: one endocavitary mass (arrow) can be identified on the tricuspid valve, small, well-outlined lesion.

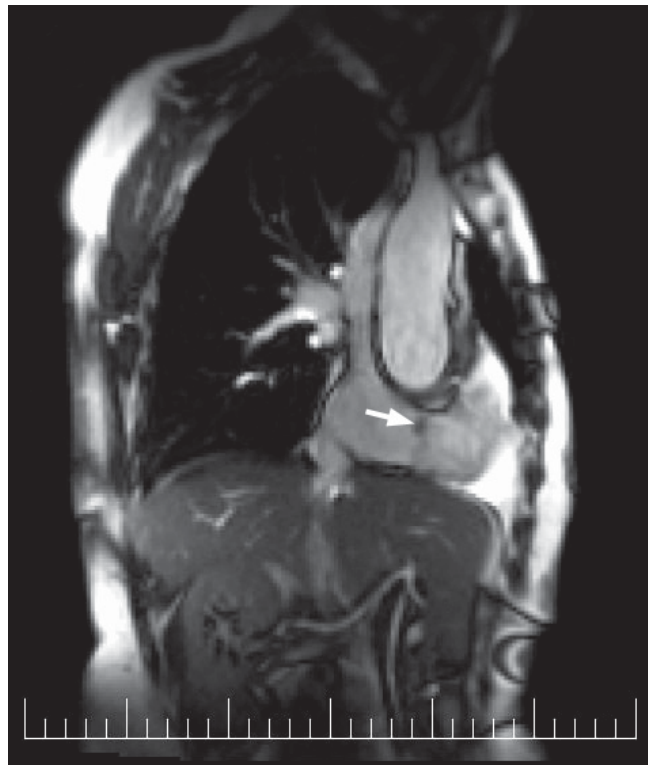

Figure 5. MR examination, T2-weighted sequence in sagittal view: papillary fibroelastoma (arrow) originating in the anterior cusp of the tricuspid valve.

The patient had a high global risk of pulmonary embolism, determined on the one hand by the orthopedic pathology (arthroplasty of the lower limb requiring prolonged bed rest) and on the other hand by the intrinsic embolic risk of the tumor (mobile pedunculated mass around 1 cm-size localized on the free edge of the leaflet) combined with the possibility of thrombus formation on the surface of the tumor. For these reasons we recommended surgical removal. No coronary artery disease was found on the preoperative coronary angiography.

The excision of the mass was performed followed by the repair of the anterior leaflet of the tricuspid valve (Figure 6). At the macroscopical histopathological examination, the tumor measured 1.3/1 cm and resembled a sea-anemone after its placing in saline solution (Figure7). Microscopically the tumor was composed of multiple dense, hyaline branches formed by crowded elastic laminae; at the surface the branches were covered by CD 31-positive endothelial cells and there was a reduced inflammatory infiltrate composed of lymphocytes, plasmocytes, rare neutrophils localized at the implantation base of the tumor (Figure 8 and 9). Histological and immunohistochemical characteristics confirm the diagnosis of papillary fibroelastoma.

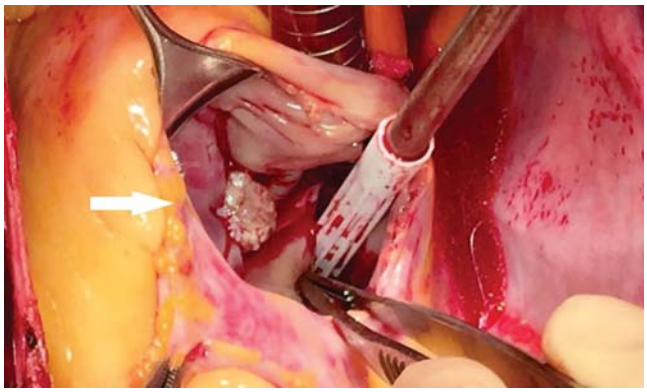

Figure 6. Intraoperative finding shows the pedunculated mass and with multiple papillary fronds (arrow) attached to the anterior cusp of the tricuspid valve.

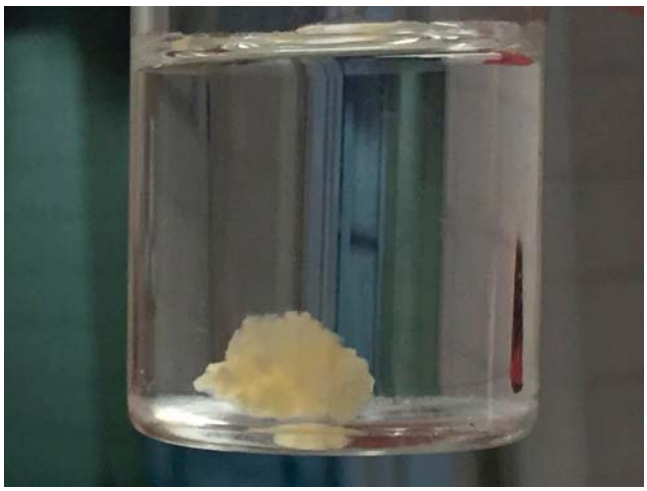

Figure 7. Gross view of the tumor with characteristic frond-like appearance and resemblance to a sea anemone, after immersion in saline solution.

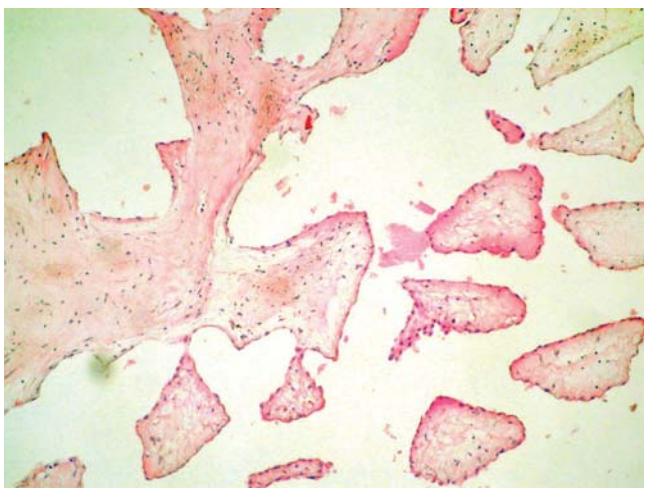

Figure 8. The histological specimen of the papillary fibroelastoma (hematoxilin-eosin stain, magnification x10): multiple dense, hyaline branches covered by endothelial cells.

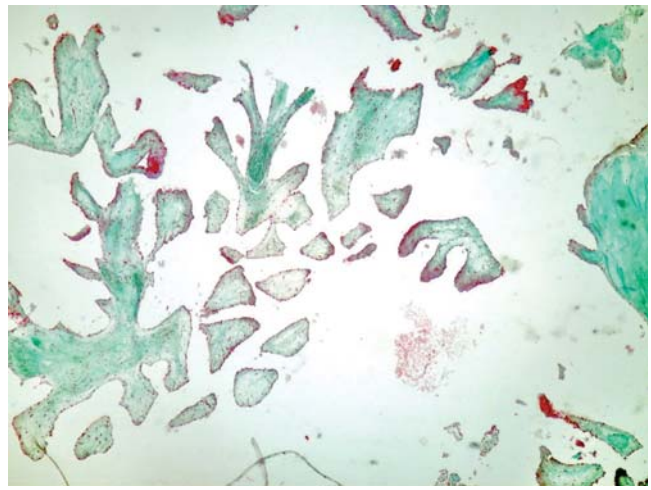

Figure 9. The tumor was histologically diagnosed as a papillary fibroelastoma covered with CD31-positive endothelial cells (immunohistochemistry CD31 staining, magnification x10).

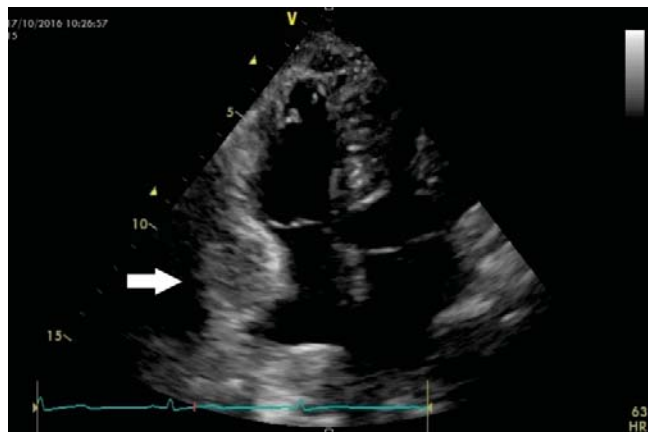

Figure 10. Postoperative transthoracic echocardiogram (apical 4 chambers view): tricuspid valve with normal appearance, pericardial effusion located next to the right atrium (arrow).

The postoperative course was uneventful. Transthoracic echocardiography described a normal tricuspid valve and a small intrapericardial hematoma located next to the right atrium with no hemodynamic impact (Figure 10) which disappeared on follow-up echocardiographic exams. Clinical outcome was favorable, so the patient was discharged 14 days later. The patient is scheduled for echocardiographic reevaluation 6 months later and annually afterwards.

DISCUSSIONS

Papillary fibroelastoma was first described by Yater in 1931. It is an avascular structure composed of dense connective tissue lined by endocardium. It is the most common primary tumor of the heart located on the valves, mostly involving the aortic (29% of cases) and the mitral valve (25% of cases), rarely the tricuspid

valve (17% of cases) and the pulmonary valve (13% of cases). Its size may range from a few mm up to 5 cm, the average size is 1 cm, as was the case in our patient. It is usually a solitary tumor (90% of cases), but occasionally can have multiple location, on the same valve, on multiple valves or on any endocardial surface (mural endocardium, papillary muscles, chordae tendineae)[4,8]. Papillary fibroelastoma rarely causes significant valvular dysfunction, but it is a potential source of embolism which explains its clinical importance. Emboli may come directly from the tumor, but more commonly from thrombus attached to its surface. The tumor itself is slow growing, but can be a very good substrate for deposition of a large quantity of thrombotic material in a relatively short period of time and can cause life threatening major embolic complications. In our case, this was the main reason we considered it necessary to excise the tumor[9,10].

The surgical treatment of symptomatic papillary fibroelastoma is widely accepted in literature, but asymptomatic tumor excision, especially for those located in the right heart, is controversial[11,12,13]. Once discovered, many authors recommend excision of left-sided fibroelastomas, regardless of tumor characteristics (size, mobility), especially if the patient is a good surgical candidate (*Society of Thoracic Surgeons* score <1%), because there is no imagistic characteristic based on which the embolic risk of the tumor can be determined with certainty (although, according to some observations, the embolic risk is higher for large, mobile and pediculated tumors)[4,14,15]. This attitude is supported by the fact that the surgery in experienced centers has excellent results, with high likelihood of valve preservation (in 98% of cases)[14,16]. The recurrence rate of the tumor is small (1.6%), but it is not null as reported in previous studies and patients require lifetime echocardiographic follow-up[14]. Surgical excision of asymptomatic right-sided fibroelastomas is not unanimously recommended in the literature. We believe that the decision should be individualized for each case, taking into account the global embolic risk (which was considered high for our patient), the surgical risk and not least the patient's preferences[1,11,12,13]. If non-surgical approach is chosen, some authors recommend treatment with anticoagulant or antiplatelet agents in order to lower the risk of thrombotic material overlay on the surface of the tumor and consequently its embolic potential[14].

Finally we would like to emphasize the importance of echocardiography as a first-line imaging modality in the diagnosis of cardiac tumors. Echocardiography can provide valuable information on tumor location and characteristics (size, shape, mobility), on its hemodynamic impact and can guide to a certain point the differential diagnosis (for papillary fibroelastomas the differential diagnosis should include valvular vegetations, Lambl's excrescence, valvular strands, other types of valvular tumors, such as myxomas with valve location,)[10,17,18,19]. The sensitivity of echocardiography for detecting papillary fibroelastomas is 62% for the transthoracic examination and 77% for the transesophageal examination[8,20], explained by the fact that the tumor is generally small and may go unnoticed, especially at a superficial examination or in patients with poor acoustic windows. In the case of our patient, the echocardiographic finding of a papillary fibroelastoma changed the entire initial therapeutic strategy and surgical removal of the heart tumor was considered necessary before the surgical treatment of orthopedic pathology.

CONCLUSIONS

The papillary fibroelastoma of the tricuspid valve may be an incidental finding during echocardiographic examination performed for a completely different reason. Its embolic risk is high, so if the patient is not a high risk surgical patient, the surgical excision of the tumor is recommended as surgery has excellent long-term results.

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