We would like to present an interesting case operated on in our department and discuss the international bibliography about this issue. We also present some interesting images of this case. Our material is composed from a 68-year-old woman treated by the authors. She presented with a small murmur in the auscultation while she was asymptomatic and then she diagnosed with a tumor on the left coronary cusp of the aortic valve with the characteristics of papillary fibroelastoma. On the basis of the potential embolic risk either of the mass itself or of associated thrombus and the possibility of further enlargement, the patient although asymptomatic at the time of diagnosis was referred for elective surgical excision of the mass. She underwent on median sternotomy and through extracorporeal circulation the mass has been excised with the preservation of the well-functioning valve. Through this case, we would like to discuss the bibliography for the decision making in these cases. Hence, the aim of our study is that we have to keep in mind that this kind of friable mass may be the cause of embolism, stroke or coronary artery occlusion and must be excised in a conservative setting, sparing the aortic valve.

**Key words:** Aortic insufficiency; Aortic valve; Cardiac tumor; Heart tumor; Heart valve disease; Myocardial infarction; Myxoma; Papillary fibroelastoma; Stroke; Valve surgery

**INTRODUCTION**

Cardiac neoplasms are rare, constituting only 0.001–0.28% of autopsy cases. They are divided into primary cardiac tumors, metastatic cardiac tumors, and intracardiac tumors originated from infradiaphragmatic organs. Primary cardiac tumors can be either benign or malignant neoplasms arising from the endocardium, heart valves or myocardium. Papillary fibroelastomas account for <10% of all cardiac tumors, representing the most common tumor of the valves and the second most common cardiac benign tumor, following myxomas. The clinical presentation of papillary fibroelastoma varies from asymptomatic to severe embolic complications with stroke or myocardial infarction. Cardiac fibroelastomas occur sporadically, exhibit a wide distribution of age, with predominance between the fourth and eight decades of life, and it is more frequent in the male sex. They are generally small, friable, and slow growing tumors with multiple avascular papillary fronds made of collagen, elastic fibers, and proteoglycans. Their pathogenesis remains yet uncertain. Although histologically benign, papillary fibroelastomas have the potential to lead in life-threatening complications from embolization of fragments into the coronary arteries, systemic circulation, and pulmonary circulation depending on their dimension, mobility, and location. Cerebral and retinal
arteries are typically affected through embolization of fibrin thrombus or parts of the tumor giving genesis to the most common presenting symptoms of neurologic impairment ranging from transient ischemic attack to real stroke. Papillary fibroelastomas can also cause angina, myocardial infarction or even sudden death through direct occlusion of the coronary arteries. It can also cause embolization of visceral and peripheral vessels.[9,10] Surprisingly, although commonly valvular, they are rarely associated with valvular dysfunction.[9]

We would like to present a case diagnosed incidentally with papillary fibroelastoma and underwent surgical excision of the tumor with the use of the extracorporeal circulation.

CASE REPORT

A 68-year-old asymptomatic woman was referred to our hospital after an aortic valve mass was detected on transthoracic echocardiography after a heart murmur was found on routine auscultation. Her medical history was free. At transthoracic echocardiography, a mass 10.5 mm × 4.6 mm attached to the left coronary cusp was detected. The mass was a pedunculated, mobile, round, echo dense, and stipple in texture structure, with well-demarcated borders, features typical of a fibroelastoma. It projected into the arterial lumen of the aorta without causing any aortic insufficiency while the rest echocardiographic examination revealed an estimated left ventricular ejection fraction of 0.50 without other valvular abnormalities [Figure 1]. Because of the gross appearance and the location of the lesion, a working diagnosis of papillary fibroelastoma was made. On the basis of the potential embolic risk either of the mass itself or of associated thrombus and the possibility of further enlargement, the patient although asymptomatic at the time of diagnosis was referred for elective surgical excision of the mass in our department. This is confirmed by the international bibliography[9] other than our experience. A median sternotomy and a standard cardiopulmonary bypass were established. After the aortotomy a pedunculated flower-like tumor was found [Figure 2]. A valve-sparing technique with simple shave excision of the tumor was undertaken with particular care in avoiding embolization and ensuring that no remnants from fragmentation of this friable tumor were left behind both locally on the cusp and in the vicinity of ascending aorta and left ventricle. The resected lesion had a flower-like appearance with frond-like projections deriving from the tumor. The lesion was histologically diagnosed as a papillary fibroelastoma [Figure 3]. The patient’s postoperative course was uncomplicated. She discharged the 6th postoperative day in optimal clinical conditions. A follow-up echocardiogram at 2 weeks did not demonstrate any tumor recurrence or aortic regurgitation or dysfunction. The patient will be followed-up in 3 months with repeat echocardiography.
CONCLUSIONS

Therapeutic decisions should take into consideration not only the presence of symptoms but also the potential of life-threatening complications. Although histologically benign, papillary fibroelastomas have the potential to lead in life-threatening complications from embolization of fragments into the coronary arteries, systemic circulation and pulmonary circulation depending on their size, mobility, and location. Cerebral and retinal arteries are typically affected through embolization of fibrin thrombus or parts of the tumor giving origin to the most common presenting symptoms of neurologic impairment ranging from stroke to transient ischemic attack. Papillary fibroelastomas can also cause angina, myocardial infarction, or even sudden death through direct occlusion of the coronary arteries or embolization to a coronary vessel as well as heart failure and peripheral embolization.[9,10] Surprisingly, although commonly valvular, they are rarely associated with valvular regurgitation or dysfunction.[9] It has also been described multiple left and right ventricle papillary fibroelastomas other than on tricuspid valve localization.[7-10] The left side and mainly the location on the aortic valve are the lesion more dangerous for embolic events.[8-10] The transthoracic, as well as transoesophageal echocardiography it is generally the only imaging, examination required preoperatively except for the cases where the presence of coronary disease must be evaluated. Coronary angiography is associated with an added risk because the catheter may dislodge a fragment of the tumor and lead to embolism coronary or systemic embolization.[8-9] Patients with localization of the tumor in the aortic valve will probably need a noninvasive modality like computed tomography angiography or scintigraphy to evaluate the coronary anatomy and avoid the high cardioembolic risk. This in accordance with the bibliography.[9] Therapeutic decisions should take into consideration not only the presence of symptoms but also the potential of life-threatening complications. There is a general consensus that symptomatic patients should be referred for curative surgical excision of the tumor. In addition, asymptomatic patients with large (>1 cm) mobile masses especially left sided, as in our case, should also be considered candidates for curative surgical excision due to the increased risk of cardiovascular complications from embolization and sudden cardiac death.[9] On the other hand, according to the bibliography,[2-6] asymptomatic patients with small left sided nonmobile lesions are being closely followed up with echocardiography until symptoms develop or tumors enlarge and become mobile. In this way, echocardiography proves to be an invaluable tool for decision making. Patients not candidates for surgical treatment should be treated with long-term anticoagulation, despite the lack of guidelines to support this strategy.

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Conflicts of interest
There are no conflicts of interest.

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