CASE REPORT

Resection of papillary aortic fibroelastoma with valve preservation

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Papillary fibroelastoma is the second most common cardiac tumor and the most common valvular one. The case of a 70-year-old female with a history of fainting is presented. Given the progressive increase in the size of the lesion, surgical resection was performed with valve preservation. This tumor is usually asymptomatic, but 20% present chest pain, arrhythmias, or embolisms. Mortality is 50% without resection due to embolization and outflow tract obstruction.

***Key words:** Cardiac tumor; Embolism; Papillary fibroelastoma.*

El fibroelastoma papilar es el segundo tumor cardíaco más frecuente y el tumor valvular más frecuente. Se presenta el caso de una mujer de 70 años de edad con antecedentes de desmayo. Dado el progresivo aumento del tamaño de la lesión, se realizó resección quirúrgica con preservación valvular. Este tumor suele ser asintomático, pero el 20% presenta dolor torácico, arritmias o embolias. La mortalidad es del 50% sin resección por embolización y obstrucción del tracto de salida.

***Palabras clave:** Tumor cardiaco; Embolismo; Fibroelastoma papilar.*

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Papillary Aortic Fibroelastoma (PAF) is the second most common cardiac tumor. It is found in less than 10% of all primary cardiac tumors examined at autopsy. It usually has a benign course; however, due to the embolic risk, surgical resection should be carried out [1, 2].

The case of a 70-year-old woman who had a history of syncope, palpitations and dyspnea on moderate exertion is presented. Within case management, 3 filiform masses were found in the aortic valve corresponding to PAF.

CLINICAL CASE

We present here a 70-year-old female patient with a history of hypertension, type 2 Diabetes Mellitus, Obstructive Apnea Syndrome and controlled primary hypothyroidism. She presented to our unit with palpitations, atypical angina and dyspnea with moderate exertion, 3 years prior to diagnosis. A transthoracic echocardiogram (TTE) found 3 filiform masses of 8 mm in their maximum diameter on the aortic valve. A transesophageal echocardiogram (TEE) confirmed a mass of 8 mm by 4 mm and 8 mm by 1 mm in the right coronary cusp, and 7 mm by 1 mm in the non-coronary cusp corresponding to aortic PAF. During the analysis of the case, a diagnosis of atrial fibrillation and microvascular angina was made. Surveillance was decided due to good functional capacity and not initially considered a candidate for surgery. At 3-years of follow-up, a new TEE reported an increase in the size of the lesions up to 13 and 10 mm in the right coronary cusp and in the non-coronary cusp of 9 mm in its maximum diameter (**Fig. 1**). Due to the risk of embolic phenomena, with a diameter greater than 10 mm, the lesions were surgically resected without complications. Adequate valve function was confirmed with intraoperative TEE. She had a favorable postoperative course with suspension of vasopressors and early weaning of the ventilator. The pathology sample was consistent with PAF (**Fig. 2**).

COMMENT

Three quarters of primary cardiac tumors are benign, the majority being atrial myxomas. The PAF comprise less than 10% of all primary cardiac tumors at autopsy and is the second most common primary cardiac tumor. It occurs predominantly in men and there is an increased incidence after the fourth decade. They are the most common valvular tumor; however, they do not usually cause regurgitation and therefore it is possible to preserve the valve [1,2].

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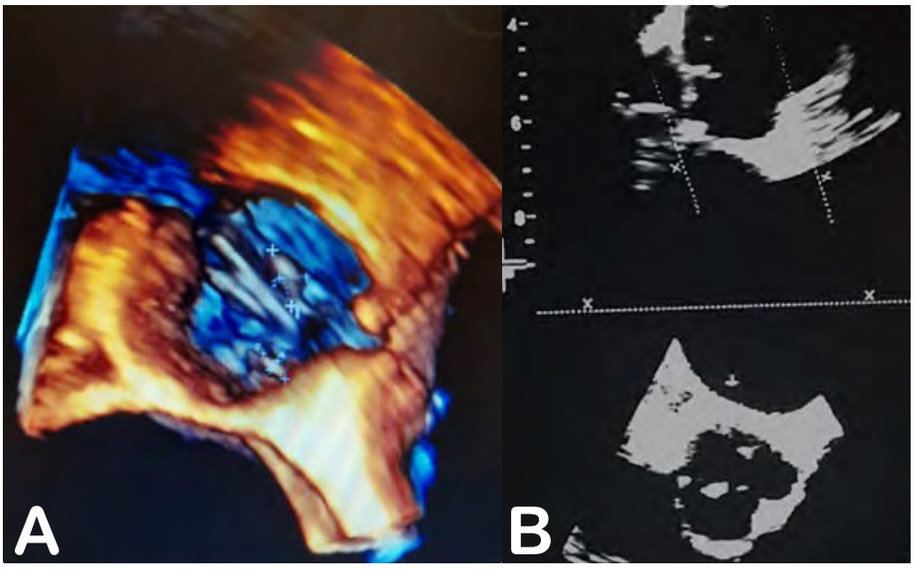

Figure 1. Aortic fibroelastomas located in the right coronary cusp (13 mm by 3 mm) and left coronary cusp (10mm by 3 mm) observed in transesophageal echocardiogram in 3D (A) and in 2D (B).

A point mutation of KRAS exists in 79%. However, no clear risk factor for the development of this tumor has been found 80% are valvular in location, most common in the aortic valve, followed by the mitral valve [2, 3].

Most of them are asymptomatic. Nonetheless, 20% may present chest pain and embolic phenomena. Embolization has been associated with transient ischemic attack (17%), unstable angina (7%), acute myocardial infarction (3.8%), sudden death (3%), syncope (1.6%), and blindness (1%) [1]. Arrhythmia and outflow tract obstruction may also occur. The diagnosis of atrial fibrillation and PAF coincides in the case presented, an association not previously described to our knowledge. In a multivariate analysis tumor mobility (p=0.0001) was the only predictor of mortality or nonfatal embolization [3].

The patient's clinical manifestations of palpitations and syncope, with resolution after resection, can be explained by the PAF. Being an unusual form of presentation.

Evolution is slow, with reported growth from 6 months to 15 years. In the exposed case there was an average growth of 2 mm/year [2]. The differential diagnosis of PAF includes other tumors, vegetations, thrombi, valvular calcifications, Libman-Sacks endocarditis and Lambl's excrescences [4].

The diagnostic modality of choice is TTE, where they are seen as small filiform tumors, generally less than 12 mm, homogeneous and with a visible stem in 50% of cases [4]. However, the sensitivity of TTE is only 61% compared to TEE which is 76% [1]. Histologically they are avascular, with multiple papillary sheets in hyaline stroma [5, 6].

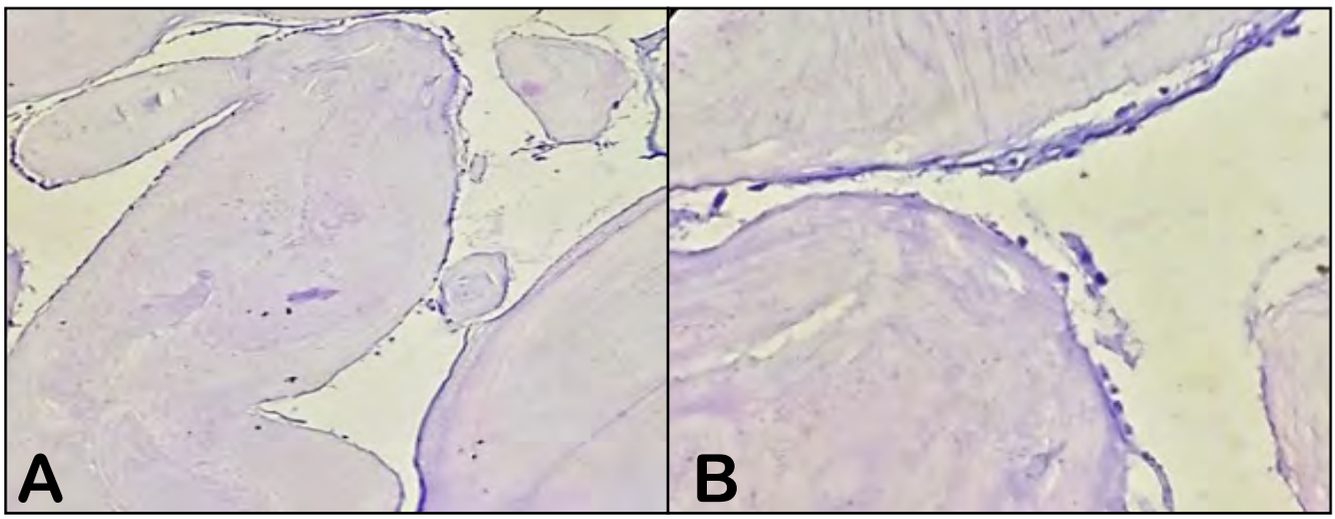

Figure 2. H&E histological sample showing fibroelastic avascular tissue with papillary projections limited by an endothelial layer, consistent with aortic fibroelastoma. View at 10x (A) and 40x (B).

According to some authors, anticoagulation should be started indefinitely at the time of diagnosis, though it is controversial because <3% may present thrombus on the surface of the tumor [3]. Due to the history of atrial fibrillation, our patient was already under anticoagulation, which could have influenced the absence of embolic phenomena and only clinical manifestations due to obstruction.

For tumors larger than 1 cm and mobile, due to the risk of embolization, surgical removal is the only curative treatment [7].

The prognosis after surgery is excellent with no recurrence reported in follow-ups of up to 11 years. On the other hand, large series of more than 600 cases document a 50% mortality in patients who did not undergo resection for embolization or LV outflow tract obstruction [3, 8].

As a conclusion, PAF is the second most common benign cardiac tumor and the most common of valvular location. It usually has an asymptomatic course, nevertheless with embolic potential and occlusion of the ostium and the outflow tract of the left ventricle. Mobility is the only predictive variable for mortality and nonfatal embolization. The treatment of choice is surgical resection, which is curative.

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