Cardiac Papillary Fibroelastoma. Experience of an Institution

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Primary intracardiac tumors are rare, with prevalence between 0.0017% and 0.19% from non-selected autopsy studies. Approximately 75% are benign and almost half of them are myxomas. The remaining tumors are divided among rabdomyomas, lipomas and fibroelastomas. Myxomas are the most common intracardiac tumors in adult age and rabdomyomas the most common among pediatric population.

Papillary fibroelastoma (PFE) is a relative rare benign heart tumor, corresponding to approximately 8% of intracardiac tumors. They most commonly manifested in cardiac valves[1]. In the past, they either consisted of necropsy findings or were found in surgical procedures at random. In vivo diagnosis was sporadic[2]. With the improvement of echocardiography techniques, PFE has been more frequently diagnosed. They are usually described as a movable, pedunculate, well-delimited mass and with predilection for valve endocardium. Therapeutic proposal, when they are pedunculate, is surgical resection, preventing cerebral, pulmonary, coronary or peripheral embolic phenomena[1,3]. Five cases diagnosed in our institution, in the period from August 1995 to June 2004, will be presented.

Case Reports

Case 1 **-** A 27-year-old female patient, under Turner Syndrome follow-up, sent due to echocardiographic finding of tumoral, pedunculate, movable in tricuspid valve topography image. She did not show symptoms and her physical exam was normal, except for bodily marks from the syndrome itself. Echocardiogram evidenced a round echodense movable image, located at right atrium, adhered to apical portions of tricuspid valve septal leaflet, with signs of left ventricular filling obstructions. The 1.8 cm x 1.2 cm mass was submitted to surgical exeresis (fig. 1). Microscopic exam confirmed papillary fibroelastoma diagnosis.

Case 2 **-** Female patient, 67 years old, clinically asymptomatic, submitted to physical examination in which discreet systolic cardiac murmur was detected. Transesophageal echocardiogram showed pedunculate movable mass in aortic valve, which moved towards the aorta (fig. 2). Magnetic nuclear resonance imaging evidenced the referred mass in aortic valve leaflet (fig. 3). She was submitted to tumor exeresis. Diagnostic confirmed papillary fibroelastoma.

Case 3 **-** A 63-year-old female diabetic patient, with mitral commissurotomy history due to rheumatic mitral stenosis and chronic atrial fibrillation. She made use of oral anticoagulant with coumarinic and showed recurring episodes of transitory ischemic attack (TIA).

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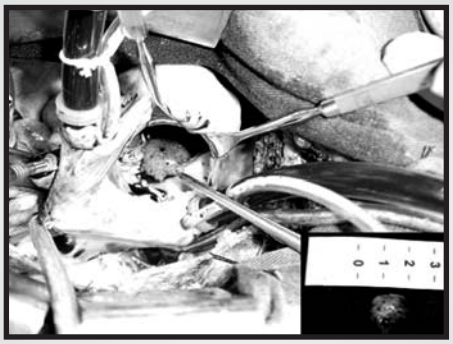

Fig. 1 - Intraoperative photo of tumoral mass in tricuspid valve.

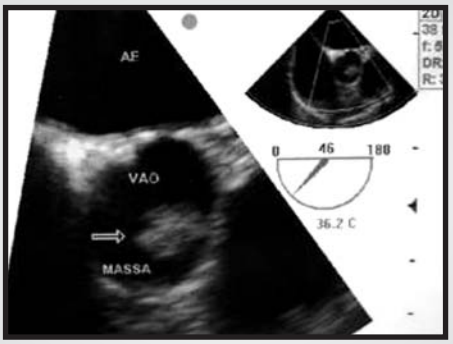

Fig. 2 - Transesophageal echocardiogram demonstrating pedunculate tumoral mass adhered to aortic valve.

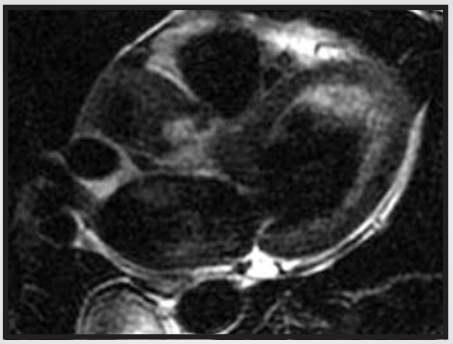

Fig. 3 - Magnetic nuclear resonance imaging evidencing pedunculate tumoral mass in aortic valve leaflet.

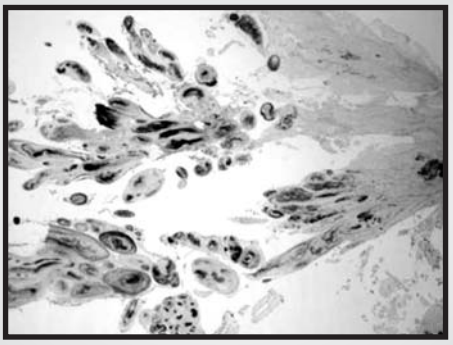

Fig. 4 - Histopathological exam evidencing elastic fibers with Morat coloring.

Echocardiogram evidenced mitral stenosis with valve area of 1.4 cm^2 and anomalous echoes of 1.8 cm x 1.8 cm in tendinous chord, suggestive of vegetation. There were left atrium thrombus signs. She was sent to surgical treatment and, as a finding, a characteristic tumoral formation with a diameter of, 2 cm, which extended to the papillary muscle, was evidenced in mitral valve anterior leaflet. The exchange of mitral valve through biological prosthesis and tumor exeresis were performed. Microscopic exam confirmed papillary fibroelastoma diagnosis (fig. 4).

Case 4 - Male patient, 59 years old, with history severe mitral regurgitation due to myxomatous degeneration, made use of coumarinic in force of chronic atrial fibrillation and showed a TIA episode. He was under a follow-up due to functional class III heart failure (NYHA). He was submitted to surgical treatment, in which chord rupture was evidenced and quadrangular resection of posterior cuspid was carried out, followed by raffia and posterior annuloplasty with a bovine pericardium strip. Besides chronic valvopathy with fibrosis and myxoid material deposit, tumorgenicity compatible with papillary fibroelastoma was evidenced in microscopic exam.

Case 5 - A 49-year-old male patient, with history of rheumatic mitral lesion, under follow-up due to functional class III (NYHA). He was submitted to mitral valve exchange for biological prosthesis. Histopathological finding of material sent for study was compatible with papillary fibroelastoma, located in one of tendinous chords.

Patients evolved without immediate complications and free from reincidences in all five cases.

Discussion

Papillary fibroelastoma is a low-prevalence benign tumor, with tendency to valvar involvement[2,4,5]. Concerning frequency, it corresponds to the third most common primary intracardiac tumor, preceded by myxomas and lipomas[6]. It represents less than 10% from all primary intracardiac tumors, either those studied in autopsy or after resection[1,7,8].

Approximately 90% of PFE undertakes cardiac valves, usually as a single lesion, on the atrial face of atrioventricular valves or on any one of the sides of semilunar valves[3,4]. They rarely occur as multiple lesions[1,6].

Approximately 44% of PFE is found in aortic valves, followed by the undertaking of mitral valve in 35% of the cases, in 15%, in tricuspid valves and in 8%, in pulmonary ones[9]. Reports of cases on those tumors have demonstrated undertaking of all endocardial surfaces, including papillary muscles, tendinous chords, the septum or free walls from any of cardiac chambers[1,5,8,10].

The size of described PFE varied from 0.1 to 4 cm, and most of them were smaller than 1cm of diameter[4]. Their genesis remains controversial. Real neoplasias until

hamartomas, organized thrombi, reactive responses to mechanic trauma, to surgical or radiotherapy damage[1,3,4,8,9] have been considered. Prevalence is unknown due to the group of non-diagnosed silent tumors[4].

The age of patients is variable, from cases in neonates to well-advanced age patients[1,4,7]. Most of it is described in adults, over 50 years of age, and there is no difference between sexes[1,4,9].

PFE is an incidental finding in most cases, although among symptomatic patients the clinical presentation is variable and dependent on location, motility and size of tumor[4,9]. As most of it originates from left chambers (more than 95% of the cases), the most feared complication is the systemic embolization, especially for cerebral or coronary circulation[4,9]. It is not clear whether the embolus is tumor- or platelet-origin and if systemic anticoagulation could prevent from such events[2,4,9]. The most common clinical presentation described was stroke or transitory ischemic attack (TIA). Other described manifestations were: angina, myocardial infarction, sudden death, heart failure, syncope, pulmonary embolism, blindness, peripheral embolism of renal infarction[9]. In aortic valve tumor patients, sudden death and myocardial infarction were the most common manifestations. In rebuttal, stroke was the prevailing presentation[9] in mitral valve tumors. Tumoral motility is the only independent mortality and non-fatal embolization predictor[9].

Electrocardiographic findings are non-specific, as atrial arrhythmias may occur. Thoracic radiography may demonstrate signs of increase of cardiac chambers, pulmonary hypertension or congestion, especially if the tumor is occluding the mitral valve. Transthoracic echocardiogram is the ideal method for tumor diagnosis and characterization, as it usually demonstrates the mass with its varied proportions, motile or not, well-delimited, pedunculate or sessile, of round, oval or irregular shape[2]. They are mostly small (99% smaller than 2.0 cm)[2]. In a case-control study, the sensitivity and specificity of echocardiogram was 88.9% and 87.8%, respectively[2]. Magnetic resonance imaging demonstrates the mass in valve leaflet or cardiac chamber, and the presence of enhancement with gadolinium in tumoral mass, increases the suspicion level[9]. Cardiac catheterization does not contribute for the diagnosis. In coronary angiography it is possible to visualize total occlusions of coronary arteries, as well as aneurysmatic dilatations and secondary narrowing to tumoral emboli[1,9].

PFE has characteristic appearance, resembling a sea anemone, with multiple ramifications held by a pedicle to endocardium. At histological exam, it consists of an endothelium coat, which covers a connective tissue matrix with variable amounts of collagen, smooth muscle cells and elastic fibers[2,4].

For symptomatic patients, surgical exeresis is the choice for treatment, by trying to always preserve valve tissue and its function[7]. Among asymptomatic individuals, surgical procedure is controversial, as tumoral motility is the determining factor of surgical indication, for being an independent embolization and death predictor. Surgery is curatory and there is no report on reincidences[4,9]. Follow-up of asymptomatic patients who were not submitted to surgery must include anticoagulation, although its efficacy in protection against embolic phenomena is controversial[2]. Management before a left side isolated lesion includes surgical removal, when the mass is big and/or movable or in the presence of patent ovale foramen due to the possibility of paradoxical embolism[2].

In the last years, PFE has progressed from an autopsy incidental finding to an *in vivo* diagnosed disease with potential injurious complications, which require proper diagnosis and treatment. Due to its rareness, data on the treatment and follow-up are mostly derived from accounts from patients and experiences as those described in our work.

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