Case Series

Cardiac Papillary Fibroelastoma:

Single-Institution Experience with 14 Surgical Patients

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In general, treatment for symptomatic and asymptomatic cardiac papillary fibroelastoma is surgical resection—particularly of left-sided lesions, because of the risk of systemic embolization. However, few institutions have enough experience with these tumors to validate this approach. We present our institutional experience with papillary fibroelastoma and discuss our current approach.

We searched our institution's cardiac tumor database, identified all patients diagnosed with cardiac papillary fibroelastoma from 1992 through 2014, and recorded the clinical and pathologic characteristics of each case. We found 14 patients (mean age, 60.5 ± 12.3 yr) who had 18 lesions. Eleven patients (79%) were symptomatic; however, we could not always definitively associate their symptoms with a cardiac tumor. Most lesions were solitary and ≤1.5 cm in diameter; half involved the left side of the heart. All 18 lesions were surgically excised. There were no operative or 30-day deaths, and no patient needed valve replacement postoperatively. There was one late death; at one year, another 3 patients were lost to follow-up, and the others were alive without tumor recurrence.

Because of the embolic risk inherent to intracardiac masses and our relatively good postoperative outcomes, we recommend the surgical resection of all left-sided papillary fibroelastomas in surgical candidates, and we discuss with patients the advisability of resecting right-sided lesions. ***(Tex Heart Inst J 2016;43(2):148-51)***

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Primary cardiac tumors are rare, with an incidence of 0.003% in early autopsy series.[1] Of these neoplasms, 75% are benign, and 75% of these are myxomas. Papillary fibroelastomas (PFEs), which constitute approximately 10% of all primary cardiac neoplasms,[2] are the second most prevalent cardiac tumor. They can occur anywhere on the endocardium. They are reported most often in the left side of the heart and in valvular structures. The noncoronary leaflet of the aortic valve is most often affected.[3,4] Most PFEs are incidentally found on echocardiograms and are clinically silent; however, symptoms can occur after embolization and give rise to a wide variety of presenting features, including neurologic events (such as transient ischemic attack [TIA], stroke, amaurosis fugax, and spinal cord infarction),[5-7] acute coronary syndrome,[8,9] and distal thromboembolism.[10] Very few systemic or comprehensive study results have been published to validate a single medical or surgical approach to rarer primary cardiac tumors like PFE, so we reviewed our institution's surgical experience with PFE during a 22-year period. The surgical resection of PFE, especially left-sided lesions, has been recommended to prevent embolic events. Regardless, many reported PFEs have been diagnosed only after an embolic event. No one knows how many asymptomatic fibroelastomas exist and contribute to stroke.

We present our findings and discuss our current treatment approach.

Patients and Methods

We searched our institution's cardiac tumor database to find records of all patients who had a diagnosis of cardiac PFE from 1992 through 2014. We recorded relevant information, including demographics, symptoms, lesion characteristics, pathologic progression, treatment, and follow-up data. Our institutional review board approved this retrospective study.

Results

We identified 14 patients (mean age, 60.5 ± 12.3 yr) who had undergone the surgical excision of one or more cardiac PFEs from 1992 through 2014 (Table I). Of the

14, 11 were white (78.6%), 4 were women (28.6%), and 8 were older than 60 years of age (57.1%). The presumptive diagnosis of PFE, made with use of transthoracic echocardiography (TTE) or transesophageal echocardiography (TEE) (Fig. 1), was confirmed after histopathologic analysis. The patients' most prevalent comorbidities were hypertension, coronary artery disease, hyperlipidemia, and diabetes mellitus.

The 14 patients had 18 lesions (13 valvular and 5 muscular) (Table II). Ten patients had solitary lesions (71.4%). Most PFEs were 1 to 1.5 cm in diameter (38.9%). The tricuspid valve was most often involved, and then the mitral valve.

TABLE I. Characteristics of the 14 Patients with Cardiac Papillary Fibroelastoma

Variable	No. of Patients (%)
Sex	
Male	10 (71.4)
Female	4 (28.6)
Age (yr)	
25–40	1 (7.1)
41–60	5 (35.7)
61–80	8 (57.1)
Race	
White	11 (78.6)
Black	2 (14.3)
Other	1 (7.1)
Cardiac comorbidities	
Hypertension	10 (71.4)
Hyperlipidemia	8 (57.1)
Diabetes mellitus	8 (57.1)
Coronary artery disease	7 (50)
Atrial fibrillation	3 (21.4)
Mitral regurgitation	2 (14.3)
Mitral stenosis	2 (14.3)
Aortic stenosis	1 (7.1)
Heart failure	1 (7.1)

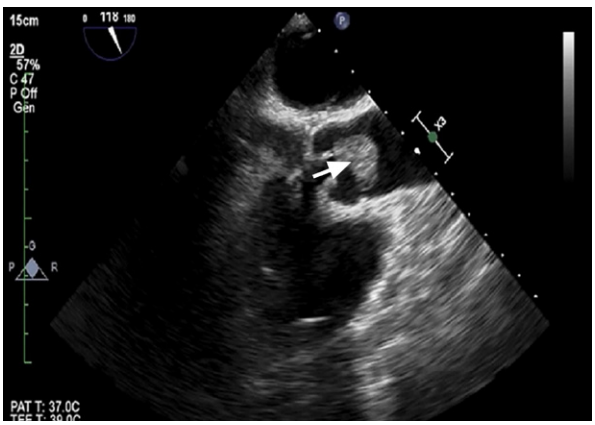

Fig. 1 *Transesophageal echocardiogram shows a 2 × 2-cm immobile mass (arrow) arising from the noncoronary cusp of the aortic valve.*

Table III summarizes the patients' presenting symptoms on the basis of tumor location. Eleven patients were symptomatic (78.6%), typically with chest pain (5 patients, 45.5%), although direct causation by the tumor could not be definitively established except

TABLE II. Characteristics of the 18 Cardiac Papillary Fibroelastomas and Operative Data

Variable	No. of Patients (%)
Lesion location	
Valve	13 (72.2)
Tricuspid	7 (53.8)
Mitral	5 (38.5)
Aortic	1 (7.7)
Muscle wall	5 (27.8)
Left ventricle	4 (80)
Left atrium	1 (20)
Lesions per patient	
1	10 (71.4)
2	4 (28.6)
Diameter (cm)	
0.5	1 (5.6)
>0.5 to 1	5 (27.8)
>1 to 1.5	7 (38.9)
>1.5	5 (27.8)
Surgical access	
Median sternotomy	11 (78.6)
Minithoracotomy	2 (14.3)
Mini-J sternotomy	1 (7.1)
Operation	
Excision only	5 —
Excision + repair	10 —
Excision + repair + other	3 —
Coronary artery bypass grafting	2 —
Maze procedure	1 —

TABLE III. Presenting Symptoms and Associated Fibroelastoma Location

Symptoms	Patients (n)	Tumor Site
Asymptomatic	3	2 TV 1 MV + LV
Palpitations	3	1 TV 1 MV 1 MV + LA
Chest pain	2	2 TV
Chest pain and dyspnea	3	1 TV 1 AV 1 MV + LV
Congestive heart failure	2	1 TV 1 MV + LV
Transient ischemic attack	1	1 LV

AV = aortic valve; LA = left atrium; LV = left ventricle; MV = mitral valve; TV = tricuspid valve

in the case of neurologic symptoms. Twelve patients (85.7%) had PFEs on either the tricuspid or mitral valve. The patient who presented with TIA had a PFE in the left ventricle (LV). The tumors were surgically approached through a median sternotomy in 11 patients (78.6%), a minithoracotomy in 2 (14.3%), and a mini-J sternotomy in 1 (7.1%).

In addition to PFE excision, 3 patients underwent concomitant surgical treatment for their symptomatic disease (coronary artery bypass grafting in 2, and the maze procedure in 1).

Three patients were asymptomatic. One had a tricuspid valve PFE that was seen on an echocardiogram before the patient's hip surgery. The 2nd patient had a long history of coronary artery disease, and an echocardiogram as part of that evaluation showed a tricuspid valve PFE. The 3rd patient's PFE, on the mitral valve, was discovered during evaluation of mild mitral stenosis. There was no obvious correlation between PFE size and the development of symptoms: the 3 asymptomatic patients had relatively large tumors (>1.5 cm in 2, and ~1.5 cm in 1).

In gross appearance, all 18 PFEs were soft, friable, yellow to yellow-white masses (Fig. 2). Some had a thin stalk. Some looked like fronds or cauliflower. Microscopically, they had papillary projections or architecture with a hypocellular matrix and were lined with endothelial cells. All the pathologic specimens were confirmed to be PFE.

The valvular tumors arose from leaflet tissue. All were resected along with a small base of valvular tissue and were repaired with use of interrupted suture. No valves needed replacement, and no new valvular regurgitation was noted on intraoperative TEE.

The mean follow-up period for all patients was 32 ± 52 months (range, 1 mo–15 yr). On all 14 postoperative TTEs, no new valvular dysfunction was noted. The 30-day survival rate after resection was 100%. One patient died 49 days after discharge from the hospital, of urinary tract sepsis, at another institution. At one year, another 3 of the 14 patients were lost to follow-up, and the others were alive without tumor recurrence.

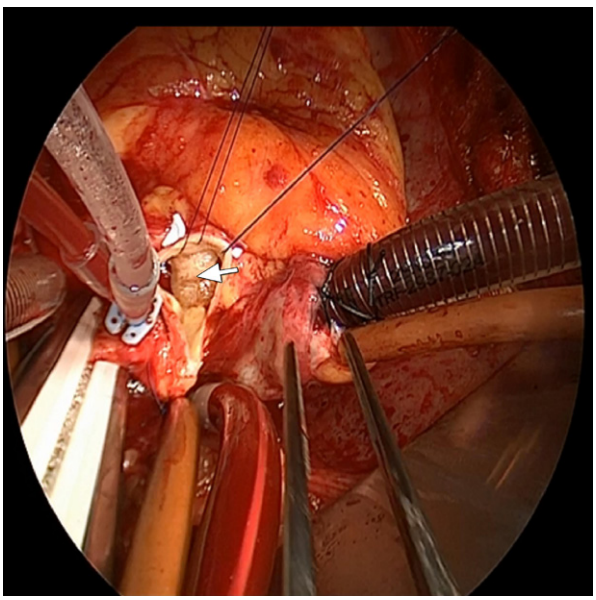

Fig. 2 *Intraoperative photograph shows a soft, friable mass (arrow) arising from the aortic valve.*

Discussion

Papillary fibroelastomas have been described as neoplasms, hamartomas, organizing thrombi, or posttraumatic tumors. The organizing-thrombi hypothesis is supported by the presence of fibrin, hyaluronic acid, and laminated elastic fibers within the fronds.[11,12] The presence of miniature tendinous chords, as well as the association of congenital PFE with other congenital cardiac anomalies, is the basis of the hamartoma hypothesis.[13,14] Another proposed association is the relationship between PFE generation and iatrogenic trauma that is related to hemodynamic changes from mechanical alterations or from direct endocardial damage after radiation exposure.[15,16]

Papillary fibroelastomas most often arise from the valvular endocardium. Gowda and colleagues[4] found that PFEs were on valvular surfaces in 84% of cases; however, PFEs can also appear over the papillary muscles, chordae tendineae, ventricular septum, or endocardial surface.[17] Multiple lesions can occur in the same location or in different places. The authors of one case report documented as many as 40 PFEs in both ventricles.[18]

In our patients, 50% of the PFEs arose from the left side of the heart, and 13 of the 18 involved heart valves. None of the 6 patients whose tumors involved the mitral or aortic valve needed valve replacement, and no patient had postoperative valvular insufficiency.

A PFE can be discovered incidentally on echocardiograms or magnetic resonance images (MRIs), during open-heart surgery, or at autopsy. When symptomatic, PFEs typically cause cardiovascular symptoms. Of our 11 symptomatic patients, 5 presented with chest pain, although causation by the tumor was difficult to establish. Presentation is also highly variable in patients with PFE; according to Klarich and colleagues,[19] embolic events can occur in up to 35% of cases. Only one of our patients (with the LV tumor) presented with embolic stroke, including <24 hours of right-sided weakness with facial numbness, drooping, and slurred speech. An MRI did not reveal a corresponding lesion. However, left-sided heart tumors can cause angina pectoris, myocardial infarction, TIA, and stroke—and our patient with TIA symptoms was found to have a PFE in his

LV. We therefore recommend the surgical excision of left-sided PFEs.

Most right-sided tumors are reportedly asymptomatic until they become large enough to form a surface thrombus that can obstruct intracardiac blood flow or cause pulmonary embolism. We discuss the risk with patients, in contemplation of resection.

When symptomatic patients present with PFE, and particularly with a history of embolic episodes, surgical resection is appropriate, absent major contraindications. Complete excision is the only definitive way to eliminate the source and potential recurrence of embolization. Symptomatic patients who are not surgical candidates can be offered long-term oral anticoagulation for stroke prevention; however, no data strongly validate the efficacy of this approach. The real debate concerns asymptomatic patients with incidentally discovered PFEs. Because these pedunculated tumors are unpredictable, many surgeons recommend elective resection in asymptomatic patients—as we do for all left- and some right-sided lesions. Embolic fragments can originate from the tumor itself because of its friable texture, and perhaps from the surface formation of platelet and fibrin thrombi.[3]

In Gowda and associates' analysis of 725 reported cases,[4] only tumor mobility was an independent predictor of PFE-related nonfatal embolization—tumor size was not. Klarich and co-authors[19] found that 22% of patients with PFE who were not surgically treated sustained neurologic events, and 4% had peripheral embolic events during the follow-up period (vs none in the authors' surgical cohort). We had only one patient (with the LV tumor) who presented with neurologic symptoms, and these resolved completely after resection. The unclear reason for this low number of patients might relate to our status as a cardiac tumor referral center for numerous patients whose clinically silent cardiac masses were discovered incidentally.

This study is limited by the inherent bias of a retrospective study. However, given the rarity of PFE, there have been no longitudinal follow-up studies and thus there is little information about the natural history of PFE. The senior author (MJR) has monitored various patients whose right-sided tumors were not surgically excised; however, these instances and their outcomes were not recorded in our surgical database.

In conclusion, we safely resected the PFEs in all 14 of our patients with no 30-day deaths, no valve replacements, and no postprocedural valvular insufficiency.

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