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Surgery for Cardiac Papillary Fibroelastoma: A 12-Year Single Institution Experience

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Background: We reviewed our clinical experience with cardiac papillary fibroelastoma from 2005 to 2017. The objective of this study was to investigate the clinical and operative data, as well as the early survival rate and immediate postoperative complications.

Material/Methods: We performed a retrospective analysis of 11 patients (eight males and three females) who underwent resection of cardiac papillary fibroelastoma in our institution.

Results: Mean age at tumor diagnosis was 60±14 years. The mean dimension of the tumor was 14±11 mm. The most common symptoms were dyspnea, palpitation, and angina pectoris, while one patient had recurrent fever attacks and another patient had a transient ischemic attack. Two patients had concomitant malignant tumors (cervical and colon carcinoma) and another two had concomitant benign neoplasms (liver cyst and thyroid adenoma). Bypass and cross clamp times were 77±32 minutes and 54±18 minutes, respectively. The tumors were found predominantly on cardiac valves (n=7). In eight cases, only tumor extirpation was performed, whereas in the other three cases, the valves had to be replaced. The mean intensive care unit length of stay was 1.1±0.3 days and there was no in-hospital mortality. All patients were alive at one-year follow-up and the survival rate was 91% in the mean follow-up period of 4.15 years.

Conclusions: The surgical treatment of cardiac papillary fibroelastoma was curative and safe. Thus, potential complications such as embolization or mechanical irritation of the valves can be avoided without high surgical risk.

MeSH Keywords: Heart Neoplasms • Patient Outcome Assessment • Surgery Department, Hospital

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Background

In 1931, Yater described valvular tumors for the first time and a few years later, Campbell and Carling reported on a patient with sudden death probably related to a tumor located on the aortic valve [1]. Cheitlin et al. introduced the term "cardiac papillary fibroelastoma" in 1975 [2].

Cardiac papillary fibroelastoma is a rare benign tumor; however, it is the second most common type in adults that mostly attacks heart valves, predominantly the aortic valve, following by the mitral valve. This type of cardiac tumor may cause peripheral and cerebral thromboembolism or mechanical damage of the valves [3–11].

The incidence of cardiac papillary fibroelastoma is higher than commonly diagnosed; in fact it may be the most common cardiac tumor [3,4,5,8,12]. In an autopsy series, the prevalence was estimated at 10% of all cardiac tumors [3]. Nowadays, echocardiography has enabled a more frequent clinical diagnosis of cardiac papillary fibroelastoma [1,13–16].

Elbardissi et al. showed that cardiac papillary fibroelastoma could be linked to previous cardiac surgery and other cardiac diseases such as hypertrophic obstructive cardiomyopathy. Based on these findings, it can be suggested that cardiac papillary fibroelastoma is a response of pressure-induced or surgical trauma [5,8].

The male gender predominates in most case series [1,3,17]. Most tumors occur as solitary lesions on the left side of the heart and are ≤1.5 cm in diameter [1,14,17–19]. The clinical presentation of cardiac papillary fibroelastoma varies from asymptomatic to severe embolic complications [18]. The most common symptoms are cerebral embolic events and chest pain [1,20]. Sudden death and myocardial infarction are described in the literature in cases of tumor prolapse into coronary ostia or occlusion of large coronary branch [1,6,9,10,15,21]. Furthermore, cardiac papillary fibroelastoma may mimic infective endocarditis [13]. For that reason, other differential diagnosis should be considered such as other heart tumors, thrombi, vegetations, and valvular calcification [1,14].

Although cardiac papillary fibroelastoma is histologically a benign tumor, surgical intervention is indicated based on a high embolization tendency [8,13,17]. Nevertheless, the management of asymptomatic cardiac papillary fibroelastoma remains controversial [12,19]. However, based on potential serious complications, symptomatic lesions should undergo surgery [1,19]. The surgical treatment has been reported to be curative and safe [1,12,14]. The alternative treatment for asymptomatic or elderly patients is long-term oral anticoagulation [1,16]. Tamin et al. reported increased mortality and a high stroke rate in the non-operative study group, which was significant higher compared to the surgical group (6% and 13% versus 2% and 8% at 1-year and 5-years, respectively) [16]. For these reasons, surgical therapy should be recommended for cardiac papillary fibroelastoma in symptomatic as well as asymptomatic patients [8,16,19,21].

In most cases, cardiac papillary fibroelastoma can be resected with preservation of cardiac valves [12,19]. If an extensive resection is needed, valve replacement may be necessary [14]. Furthermore, the long-term postoperative prognosis is excellent with low recurrence rate [1,11,12,18].

This retrospective study describes a cohort of patients who were treated surgically due to cardiac papillary fibroelastoma from 2005 to 2017. The analysis included a significant number of patients from our institution (Department of Cardiac Surgery, University of Heidelberg) high-volume centers in recent literature.

Material and Methods

Study population

From 2005 to February 2017, we evaluated all patients undergoing cardiac surgery due to cardiac papillary fibroelastoma at our institution (Department of Cardiac Surgery, University of Heidelberg). Clinical histories, operative data, histopathologic findings, and survival rates were reviewed. The diagnosis of cardiac papillary fibroelastoma was made by echocardiography and confirmed by histopathologic examination. Figures 1–3 show echocardiographic examples of cardiac papillary fibroelastoma in our patient population.

Surgical techniques

In two cases (18%), the operation was undertaken on an urgent basis because of acute left ventricular heart failure. All patients were operated using cardiopulmonary bypass (CPB). In six cases (55%), bicaval cannulation was performed. In all cases, complete surgical excision was achieved; in three patients (27%), the involved valve was replaced in consequence of extensive excision. In all patients, transesophageal echocardiography was performed intra-operatively for assessment of valvular function after excision.

Statistical analysis

Statistical analysis was performed using SPSS 24.0 (IBM Corp.). Categorical data are described as percentages and continues data as mean ± standard deviation. Survival rates were described by Kaplan-Meier curve.

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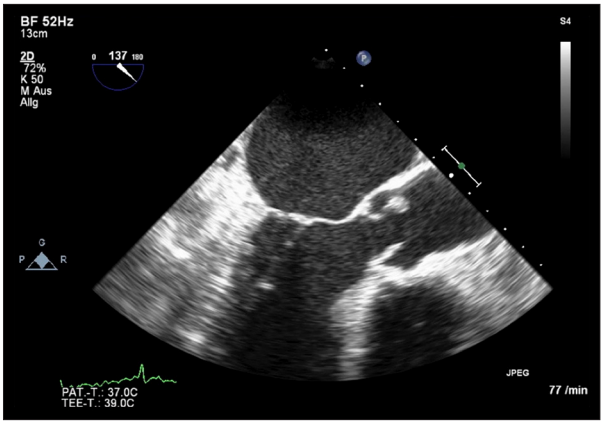

Figure 1. Fibroelastoma of the aortic valve; long axis view transesophageal echocardiography.

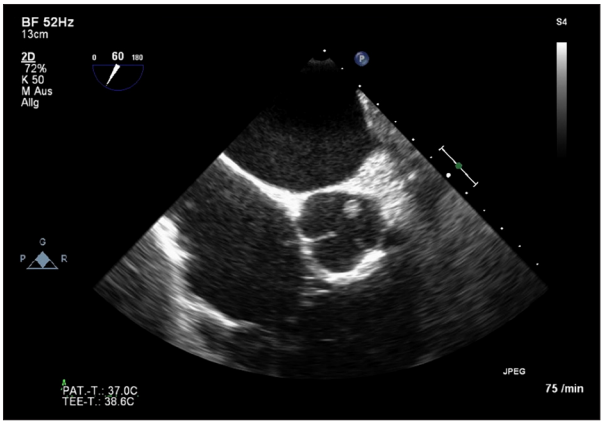

Figure 2. Fibroelastoma of the aortic valve; short axis view transesophageal echocardiography.

Results

Patients characteristics

The clinical and demographic characteristics are showed in Table 1. A total of 11 patients with a mean age of 60±14 years (range, 33 to 76 years) were included in the study. Eight patients (73%) were male. Suspected cardiac tumor was the indication for surgery in 10 cases (91%); in one case infective endocarditis was suspected. The majority of patients were symptomatic (9 of 11); the most common presenting complaints were dyspnea, palpitation, and angina pectoris in seven cases (64%). One patient had fever attacks and another patient had TIA preoperatively. In one patient with mitral valve tumor, TIA was the clinical presentation. Cardiovascular risk factors included hyperlipidemia in 64% (n=7), hypertension in 72% (n=8), diabetes mellitus in 36% (n=4), smoking in 73% (current smoker n=6, ex-smoker n=2) and obesity with a mean body mass index of 26.2±2.8 kg/m^2.

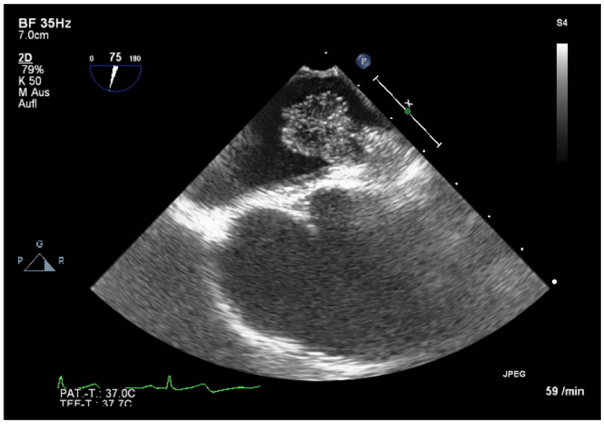

Figure 3. Fibroelastoma at the atrial septum; transesophageal echocardiography

Table 1. Patients' demographics and preoperative baseline characteristics.

Characteristics	Value
Demographic data	
Number of patients	11
Age (years)	60±14
Male	8 (73%)
Body mass index (BMI) [kg/m^2]	26.2±2.8
Myxoma as suspected diagnosis	8 (73%)
Comorbidities	
Arterial hypertension	8 (72%)
Hyperlipidemia	7 (64%)
Diabetes mellitus	4 (36%)
Chronic obstructive pulmonary disease (COPD)	3 (27%)
Status post ischemic stroke	2 (18%)
Transient ischemic attack (TIA)	1 (9%)
Current smoker	6 (55%)
Ex-smoker	2 (18%)
Preoperative symptoms	9 (82%)
Dyspnea, palpitation, angina pectoris	7 (64%)
Fever	1 (9%)
TIA	1 (9%)

All patients were initially diagnosed with transthoracic echocardiography. The diagnosis was, in every case, verified preoperatively and intra-operatively with transesophageal echocardiography.

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Table 2. Patients' surgical data, postoperative adverse events and outcome.

Characteristics	Value
Surgical data	
Median sternotomy	9 (82%)
Mini J-sternotomy	1 (9%)
Anterolateral thoracotomy	1 (9%)
Bicaval cannulation	6 (55%)
Cardiopulmonary bypass time [min]	77±32
Aortic cross clamp time [min]	54±18
Valve replacement	3 (27%)
Mean tumor dimension [mm]	14±11
Postoperative adverse events	
Thromboembolic events	0%
Acute renal failure	0%
Rethoracotomy due to mediastinal bleeding	9% (n=1)
Pacemaker necessity	0%
Outcome	
Intensive care unit length of stay [days]	1.1±0.3
In-hospital mortality	0%
One-year survival	11 (100%)
Survival in mean follow-up time of 4.15±4.2 years	91%
Tumor recurrence	0%

One female patient had undergone a previous cardiac surgery for aortic valve replacement six years before.

Preoperatively, the suspected diagnosis was cardiac myxoma in eight patients (72%) and endocarditis in one patient (9%).

In seven cases (64%) the cardiac valves were involved. In four cases (36%), the aortic valve was the most common tumor site, and the left coronary cusp was frequently affected. In three cases the mitral valve was involved. Three tumors were found in the right atrium and one tumor in the left atrium. The mean dimension was 14±11 mm.

Surgical data

Surgical data, postoperative adverse events and outcomes are summarized in Table 2.

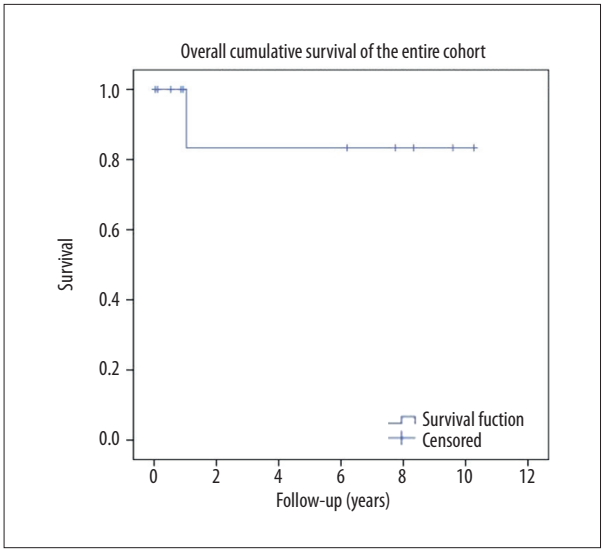

Figure 4. Kaplan-Meier survival estimate for patients with cardiac papillary fibroelastoma who underwent surgical treatment between 2005 and 2017.

All patients had surgery using extracorporeal circulation with mean CPB time 77±32 minutes and cross clamp time 54±18 minutes (in 9 of 11 patients). Two patients (18%) with right atrial tumors were operated on via beating-heart. Nine patients (82%) were operated via median sternotomy, one patient had a mini-J-sternotomy and one patient had an anterolateral thoracotomy.

Depending on the tumor location, various heart chambers were opened. In three cases (27%) the exposition of the left atrium and mitral valve was through transseptal approach and in one case the left atrium was opened directly. Four patients (36%) had only aortotomy and three patients (27%) had direct atriotomy for the right atrium.

In three patients (27%), valve replacement was necessary because of extensive excisions. Pericardial patch was used in one patient. No patients had undergone concomitant procedures.

Clinical outcome

There were no deaths within 30 days after surgery. There were no embolic events or acute renal failure. One patient experienced a rethoracotomy due to mediastinal bleeding. No patient developed respiratory insufficiency or sternal wound infection. We did not observe a pacemaker necessity.

One female patient with an aortic valve prosthesis implanted before the tumor resection, developed an aortic and mitral valve endocarditis six years after the tumor resection. Another male patient underwent a redo surgery in consequence of degenerative valve failure three years after the tumor surgery.

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The follow-up period ranged from 14 days to 10.3 years, during which time one patient died of consequences of colon cancer. The one-year survival was 100% and the survival rate during a mean follow-up time of 4.15±4.2 years was 91%. During the follow-up, no embolic events or recurrences of cardiac papillary fibroelastoma occurred. A Kaplan-Meier survival curve is shown in Figure 4.

Discussion

Primary cardiac tumors are a rare disease affecting people in all age groups; however, predominantly affecting adults between the fourth and eighth decade of life, with an incidence of 0.02% [6,8,13]. Within this rare tumor-type, papillary fibroelastoma comes second after myxoma with an incidence of 10% of all benign cardiac tumors [1,3]. It is usually asymptomatic and the diagnosis is mostly incidental. While the diagnosis can be done with computerized tomography (CT) and magnetic resonance imaging (MRI), echocardiography is useful in identifying the degree of tumor mobility and possible accompanying valve dysfunction [1,8,19,20].

Surgery is the best therapeutic option for primary cardiac tumors with excellent results [6]. Although the natural history of cardiac papillary fibroelastoma is largely unknown, surgery is indicated relatively urgently because of the risk of embolization, in contrast to myxomas which cause hemodynamically complications mostly due to size and location [22,23]. Nevertheless, small asymptomatic right-sided fibroelastoma can be treated conservatively [1,6,11].

Similar to the findings in the literature, the incidence of fibroelastoma in our institution was extremely low with less than one case a year. Also, patient characteristics did not deviate from those reported by other groups. However, in our patient collective, the majority were symptomatic with only one having TIA as the first manifestation of the tumor.

There have been associations between primary cardiac tumors and extracardiac neoplasms [3], however, this did not prove true in our case series.

Of note, one of our patients was thought to have infective endocarditis, which confirms the reputation of papillary fibroelastoma as a great mimicker [13]. Although the size of the tumors was not exceedingly large (mean 14 mm), the most common presenting symptom was dyspnea, suggesting that even small tumors may lead to impairment of hemodynamics. However, this is countered by the fact that many of the tumors involved cardiac valves.

Although typical echocardiographic features of papillary fibroelastoma are well-documented [20], most of the patients underwent surgery due to myxoma as the suspected diagnosis. This was most probably governed by the fact that myxoma is much more common and comes to mind as a possible differential diagnosis for an intracardiac mass. Based on location, size, and tumor mobility, cardiac fibroelastomas and myxomas can be differentiated echocardiographically. Cardiac myxomas arise usually from the interatrial septum and appear heterogeneous, whereas cardiac fibroelastomas can be easily detected by echocardiography. Echocardiography shows cardiac fibroelastoma often as a small size (usually <1.5 cm), mostly pedunculated and mobile, valvular or endocardial masses [1,5,13,24].

Nevertheless, preoperative differentiation is not of much importance, as surgical indications are quite similar in both cases.

The aim of surgery is complete excision of the tumor, as recurrence, though quite rare, has been reported [16]. The single operation-related complication in our study was postoperative bleeding, which was probably not related to the tumor or the tumor resection, meaning that fibroelastoma resection *per se* was associated with low complication rate in our small case series.

Moreover, during the complete follow-up period of up to 10 years, no cardiac deaths occurred. We also did not see any recurrence; in the literature, the recurrence rate is lower than 2% [11,16,21].

Conclusions

In conclusion, we present a short case series of cardiac papillary fibroelastomas. We demonstrated excellent results with a very favorable prognosis after mid-term follow-up. This study reported our experience with excision of cardiac papillary fibroelastoma. We demonstrated excellent outcomes and survival after surgery. Nevertheless, our study was limited by the retrospective design and lack of a control group. The small number of patients was the most important limitation of this study. We believe that surgical treatment should be recommended in all cases of cardiac papillary fibroelastoma. We need prospective studies to investigate the effectiveness of non-surgical treatment compared with surgical resection.

Conflict of interest

None.

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