Clinical Investigation

Primary Cardiac Tumors

A Clinical Experience of 12 Years

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From January 1980 through December 1992, we performed operations on 31 patients with primary cardiac tumors. There were 12 male and 19 female patients; the ages ranged from 7 to 71 years (mean, 47.7 ± 18.3 years); 29 were adults and 2 were children. Twenty-six (83.9%) of the tumors were benign, and 5 were malignant. The most frequently encountered benign tumor was myxoma (22), and the most frequently encountered malignant tumor was fibrosarcoma (4). The most common symptoms at clinical presentation were those associated with cardiac insufficiency or embolization. Definition of the tumor was accomplished in all patients by echocardiography; cardiac angiography was performed in only 4 patients. All patients survived the operative procedure and were discharged from the hospital, but 4 patients with malignant disease died within 30 days. Follow-up for myxoma patients ranged from 16 to 151 months (mean, 68.5 ± 36 months). Among the 26 patients with benign tumors, there was 1 death, 72 months after the surgical procedure, of right-heart failure due to recurrence of the myxoma. ***(Texas Heart Institute Journal 1993;20:223-30)***

Primary cardiac tumors are rare: their prevalence ranges from 0.0017% to 0.28% in various autopsy series, and they are up to 20 times less frequent than are secondary tumors of the heart.[1-4] The prevalence of primary cardiac tumors other than benign myxomas is of course even lower. In 52,500 patients who underwent cardiac operations during a 22-year period, Reece[5] found only 71 cases of primary tumors, 51 of which were myxomas.

Patients, Methods, and Results

From January 1980 through December 1992, 31 patients underwent operation for primary cardiac tumors at our institution (Table I). Twelve of these patients were male and the rest female. The patients' ages ranged from 7 to 71 years (mean, 47.7 ± 18.3 years); there were 29 adults and 2 children. In 26 patients, these primary tumors were benign, and in 5 they were malignant. We present our data on the symptoms, diagnostic methods, histologic types of cardiac tumor, types of operation, postoperative course, and follow-up.

Benign Tumors

There were 26 cases of benign tumors: myxoma was the most frequent, with 22 cases, followed by lipoma, fibroma, papillary fibroelastoma, and interatrial cyst, with 1 case each (Table I).

Myxomas. Myxomas were found in 6 males and 16 females ranging in age from 16 to 71 years (mean, 52.7 ± 15.7 years); only 2 of these patients were under 21 years of age. Concomitant cardiac conditions (discounting atrial fibrillation) were observed in 3 myxoma patients:

1) In Patient 11, a 63-year-old woman complaining of exertional dyspnea lasting 1 year, the 2-dimensional echocardiogram revealed a left atrial myxoma, high-grade tricuspid regurgitation, and elevated pulmonary pressures; these data were confirmed by cardiac catheterization.

2) A 65-year-old man (Patient 19), who 8 years earlier had sustained an inferior myocardial infarction, was completely asymptomatic for angina but complained of palpitations. The electrocardiogram showed atrial fibrillation, and coronary angiography revealed a proximal occlusion of the right coronary artery; the other vessels were normal. Two-dimensional echocardiography showed a left-atrial mass.

Key words: *Fibrosarcoma; heart atrium/surgery; heart neoplasms/mortality/surgery/ultrasonography; lipoma; lymphoma; myxoma*

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Table I. Patients with Primary Cardiac Tumors: Characteristics, Surgical Procedures, and Outcomes

	Date of Operation (Month/Year)	Age/Sex	Presenting Symptoms	Diagnostic Methods	Diagnosis	Location of Tumor	Concomitant Condition	Surgical Procedure	Outcome/ Follow-up
1	05/80	9Y/M	Syncope, tamponade	Echo	Fibrosarcoma	RA		Tumor resection; pericardial patch on RA free wall	Dead in 6 months
2	06/80	32Y/M	Tamponade	Echo	Fibrosarcoma	RA,RV		Biopsy	Dead in 1 month
3	11/80	40Y/F	Asymptomatic	Echo	Myxoma	LA (fossa ovalis), introflected towards MV		Biatrial approach	AF on postoperative day 3, treated with direct-current shock; recurrence of AF in 1990
4	12/82	55Y/F	Stroke, syncope, dyspnea	Echo	Myxoma	LA (fossa ovalis)		Biatrial approach	NYHA class I
5	05/83	57Y/F	Syncope	Echo	Myxoma	LA (fossa ovalis)		Biatrial approach	AF; sinus rhythm obtained with amiodarone; 1986 recurrence of AF → CHF; direct-current shock → sinus rhythm; 1988 recurrence of AF; NYHA class III
6	04/84	44Y/F	Dyspnea, fatigue	Echo	Myxoma	LA (fossa ovalis)		Biatrial approach	AF; sinus rhythm obtained with amiodarone; NYHA class I
7	11/84	68Y/F	Syncope, ankle edema	Echo	Myxoma	LA anulus, posterior leaflet		Biatrial approach	NYHA class I
8	12/84	16Y/F	Palpitations	Echo	Myxoma	LA, introflected towards MV		Biatrial approach	NYHA class I
9	01/85	44Y/F	Dyspnea	Echo	Myxoma	LA (septum); LV (PPM)		Biatrial approach	Recurrence; died 6 years later; no redo
10	08/85	65Y/M	Syncope	Angiography; echo	Myxoma	LA (fossa ovalis)		Biatrial approach; GORE-TEX® patch on septum	TIA on postoperative day 5; NYHA class I
11	10/85	63Y/F	Dyspnea, fatigue	Echo	Myxoma	LA (fossa ovalis)	Tricuspid regurgitation	Biatrial approach; tricuspid valve annuloplasty	NYHA class I
12	10/85	60Y/F	Dyspnea, palpitations, syncope, JVD	Echo	Myxoma	RA (fossa ovalis), introflected towards TV		Right atrial approach	Mediastinitis; NYHA class II
13	04/86	71Y/M	Dyspnea	Echo	Myxoma	LA (fossa ovalis), introflected towards MV		Biatrial approach	NYHA class I
14	09/86	65Y/M	Dyspnea, ankle edema, JVD	Venography of SVC,IVC; CT	Lymphoma	RA	Thrombosis to SVC, RIV, LIV, IVC, LSV	Tumor resection; thrombectomy; 12-mm GORE-TEX® shunt	Dead in 1 month
15	03/87	67Y/F	Stroke, syncope	Echo	Myxoma	LA (fossa ovalis)	AF	Biatrial approach	AF; NYHA class II

16	02/88	58Y/M	Dyspnea, palpitations, diaphoresis	Echo	Myxoma	LA (fossa ovalis)		Biatrial approach	NYHA class II
17	03/88	67Y/F	CHF, dyspnea	Echo	Myxoma	LA (fossa ovalis)		Biatrial approach	AF on postoperative day 3; direct-current shock → sinus rhythm; NYHA class II
18	10/88	43Y/M	Dyspnea, fatigue	Echo	Fibrosarcoma	RA,RV		Partial tumor resection; TV annuloplasty	Dead in 1 month
19	02/89	65Y/M	Palpitations	Echo	Myxoma	LA	AMI 8 years earlier; AF	Biatrial approach	AF; NYHA class II
20	03/89	40Y/M	Palpitations, diaphoresis	Echo; CT; MRI	Fibroma	LV wall		Partial resection	NYHA class I; no echo changes
21	11/89	50Y/F	Asymptomatic	Echo	Myxoma	LA (fossa ovalis)		Biatrial approach	NYHA class I
22	02/90	34Y/F	Dyspnea	Echo	Myxoma	LA anulus, anterior leaflet		Biatrial approach; cryoablation of tumor at implant site	NYHA class I
23	06/90	42Y/M	Dyspnea, fatigue	Echo	Fibrosarcoma	RA,RV		Biopsy	On list for heart transplantation; died 1 month later
24	06/90	7Y/F	Asymptomatic	Echo	Interatrial septal cyst	RA (septum, posterior leaflet TV)	Interatrial defect	Right atrial approach; defect closure	NYHA class I
25	09/90	25Y/F	Asymptomatic	Echo; CT; MRI	Lipoma	RV (inferior wall adhesion)		Right atrial approach	NYHA class I
26	11/90	52Y/M	Dyspnea, palpitations	Echo	Myxoma	LA (fossa ovalis), introflected towards MV		Biatrial approach	AF; NYHA class II
27	11/90	38Y/F	Asymptomatic	Echo	Myxoma	LA (fossa ovalis)		Biatrial approach	NYHA class I
28	12/90	64Y/F	CHF, palpitations, syncope, TIA	Echo	Myxoma	LA (fossa ovalis)	AF	Biatrial approach	AF; NYHA class II
29	05/91	18Y/M	Angina	Echo	Myxoma	RA, posterior aspect of TV anulus	WPW	Epicardial approach to cryoablation of accessory pathway; right atrial approach to tumor	NYHA class I
30	08/91	64Y/F	Palpitations, fatigue, syncope	Echo	Myxoma	LA, multiple locations: anulus, anterior leaflet MV, posterior medial commissure	Dextroposition aorta diagnosed 19 years earlier	Biatrial (superior transseptal) approach; GORE-TEX® patch of the septum	NYHA class II
31	06/92	53Y/M	TIA, syncope	TEE	Papillary fibroelastoma	Aortic valve, left coronary cusp		Aortotomy	NYHA class I

AF = atrial fibrillation; AMI = acute myocardial infarction; CHF = congestive heart failure; CT = computed tomography; Echo = transthoracic 2-dimensional echocardiography; IVC = inferior vena cava; JVD = jugular vein distension; LA = left atrium; LIV = left innominate vein; LSV = left subclavian vein; LV = left ventricle; MRI = magnetic resonance imaging; MV = mitral valve; PPM = posterior papillary muscle; RA = right atrium; RIV = right innominate vein; RV = right ventricle; SVC = superior vena cava; TEE = transesophageal echocardiography; TIA = transient ischemic attack; TV = tricuspid valve; WPW = Wolff Parkinson White Syndrome

3) In Patient 29, an 18-year-old boy complaining of chest pain, we found electrocardiographic patterns of Wolff-Parkinson-White syndrome. This patient underwent an electrophysiologic study that showed an accessory pathway in the anteroseptal region, and 2-dimensional echocardiography revealed a 2- x 1.2-cm globular mass adherent to the posterior aspect of the tricuspid valve. The patient underwent a combined procedure of accessory pathway cryoablation through an epicardial approach and removal of the myxoma through a right-atrial approach (Fig. 1). The postoperative course was normal, and the patient was dismissed on the 5th postoperative day.

Symptoms exhibited by the myxoma patients were dyspnea (10 cases), syncope (8), palpitations (7), fatigue (3), dizziness (2), congestive heart failure (2), stroke (2), transient ischemic attack (1), diaphoresis (1), angina (1), ankle edema (1), and jugular vein distension (1) (Table I). In 2 patients, the electrocardiogram showed atrial fibrillation, and in 2 other cases (Patients 5 and 8) a cardiac murmur was heard. Three patients were completely asymptomatic, and the myxoma was discovered by chance during a routine check-up that included echocardiography. The diagnosis was always obtained through echocardiography; in 4 patients, angiography was also performed. Symptoms were present in the myxoma patients for a mean of 24.7 ± 36.1 months before operation (range, 1 month to 12 years); the tumors were located in the left atrium in 20 cases and in the right atrium in 2. Left-atrial myxomas were located in the interatrial septum at the level of the fossa ovalis in 16 cases; at the anterior aspect of the mitral annulus in 1 case; at the posterior aspect of the mitral annulus in 1 case; and at multiple locations (the septum, the anterior aspect of the mitral annulus, and the posterior commissure) in 1 case. There was 1 patient (number 9) in whom the myxoma from the septum and the posterior wall of the left atrium was in continuity with the posterior papillary muscle through the mitral valve.

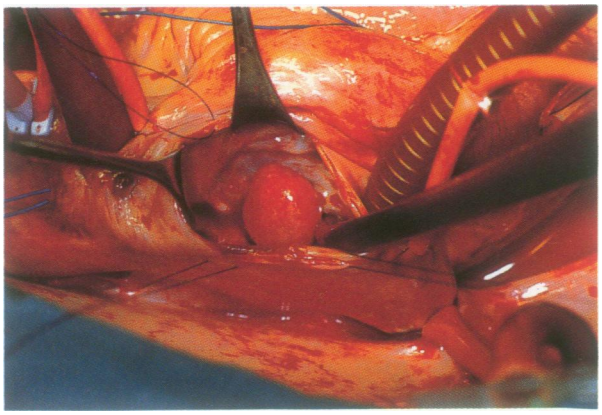

Fig. 1 *Intraoperative photograph of a right atrial myxoma in an 18-year-old patient with Wolff-Parkinson-White syndrome. (Patient 29)*

All left-atrial myxomas were removed through a biatrial approach: a traditional posterior transseptal approach in 19 patients and a superior transseptal approach in Patient 30. Patients were connected to extracorporeal circulation via 2 venous caval cannulae. After cardiac arrest had been induced by infusing cold cardioplegia into the aortic root, the right atrium was opened and the interatrial septum was incised at the level of the fossa ovalis. The left atrium was explored, and the myxoma was located. In all cases of septal implantation, we excised the full thickness of the septum in the region of the tumor's attachment. In only 2 cases was it necessary to close the resulting atrial septal defect with a GORE-TEX® patch; a primary closure was possible in the other cases.

One right-atrial myxoma (in Patient 12) was implanted in the septum between the fossa ovalis and the coronary sinus, and the other (in Patient 29) was attached at the level of the posterior aspect of the tricuspid annulus.

There was no operative mortality. The postoperative course was uneventful in 16 cases, and the postoperative hospital stay for myxoma patients ranged from 3 to 7 days. Eight myxoma patients had postoperative atrial fibrillation; in 2 of these, sinus rhythm was obtained with an infusion of amiodarone. In the others, cardiac defibrillation was necessary; their hospital stay was between 6 and 13 days. One 65-year-old man (Patient 10) sustained a transient ischemic attack on the 5th postoperative day, but was discharged on the 7th day. Mediastinitis prolonged to 3 months the hospitalization of Patient 12, a 60-year-old woman.

The mean duration of follow-up was 68.5 ± 36 months (range, 16 to 151 months). Patient 9, a 44-year-old woman whose myxoma from the septum was in continuity with the posterior papillary muscle, died 72 months after the procedure: she sustained a cerebral embolus 68 months after surgery, and an echocardiogram showed a recurrence that was judged inoperable at another institution. Thirteen myxoma patients (59%) were completely asymptomatic at long-term follow-up, although several of these NYHA functional class I patients underwent transient episodes of atrial fibrillation during the immediate postoperative period. Thirty-six months after the operation, Patient 5 experienced an acute episode of atrial fibrillation that was treated successfully by means of electrical defibrillation; nevertheless, she developed congestive heart failure and presented again 2 years later with atrial fibrillation, which was well tolerated. Now in her 9th postopera-

tive year, she is in New York Heart Association functional class III.

Benign Nonmyxomatous Tumors. Two male and 2 female patients, ranging in age from 7 to 53 years, underwent surgery for resection of benign tumors that were not myxomas (Table I). Presenting symptoms included transient ischemic attacks in a 53-year-old man with papillary fibroelastoma of the aortic valve (Patient 31) and diaphoresis and palpitations in a 40-year-old man with fibroma (Patient 20). The other 2 patients were asymptomatic: chest radiography aroused suspicion in the case of Patient 25, who had a right-ventricular lipoma; and echocardiography before appendectomy revealed the septal cyst of the right atrium in a 7-year-old girl (Patient 24). In Patient 31, with papillary fibroelastoma, the tumor was visualized by transesophageal echocardiography (Fig. 2). Patient 20, with fibroma of the left ventricle, was the only patient in this group who underwent partial resection of the tumor, and this was due to a large intraparietal extension. After 42 months, he remains in good condition, with an unchanged echocardiogram.

Malignant Tumors

Five patients underwent operative resection for malignant cardiac tumors: 4 fibrosarcomas and 1 lymphoma. All 5 patients were male, and their ages ranged from 9 to 65 years. The most common symptoms in these patients were those associated with tumor obstruction, but in 2 cases there was cardiac tamponade, symptomatic of pericardial effusion. The average duration of symptoms before diagnosis was 35 days (range, 10 days to 3 months). The tumor was suspected preoperatively in all patients, and was visualized by echocardiography or computed tomography. In 3 patients, we performed an incomplete resection of the tumor mass. In 1 of these (Patient 14, with superior vena cava syndrome caused by a lymphoma), it was necessary to graft a GORE-TEX® shunt between the innominate vein and the right atrium to ensure venous drainage. In the other 2 patients, we performed a biopsy alone, due to the extension of the neoplastic mass. Patient 23, with a fibrosarcoma that infiltrated both chambers of the right heart, was considered for heart transplantation because there was no sign of metastasis, but he died

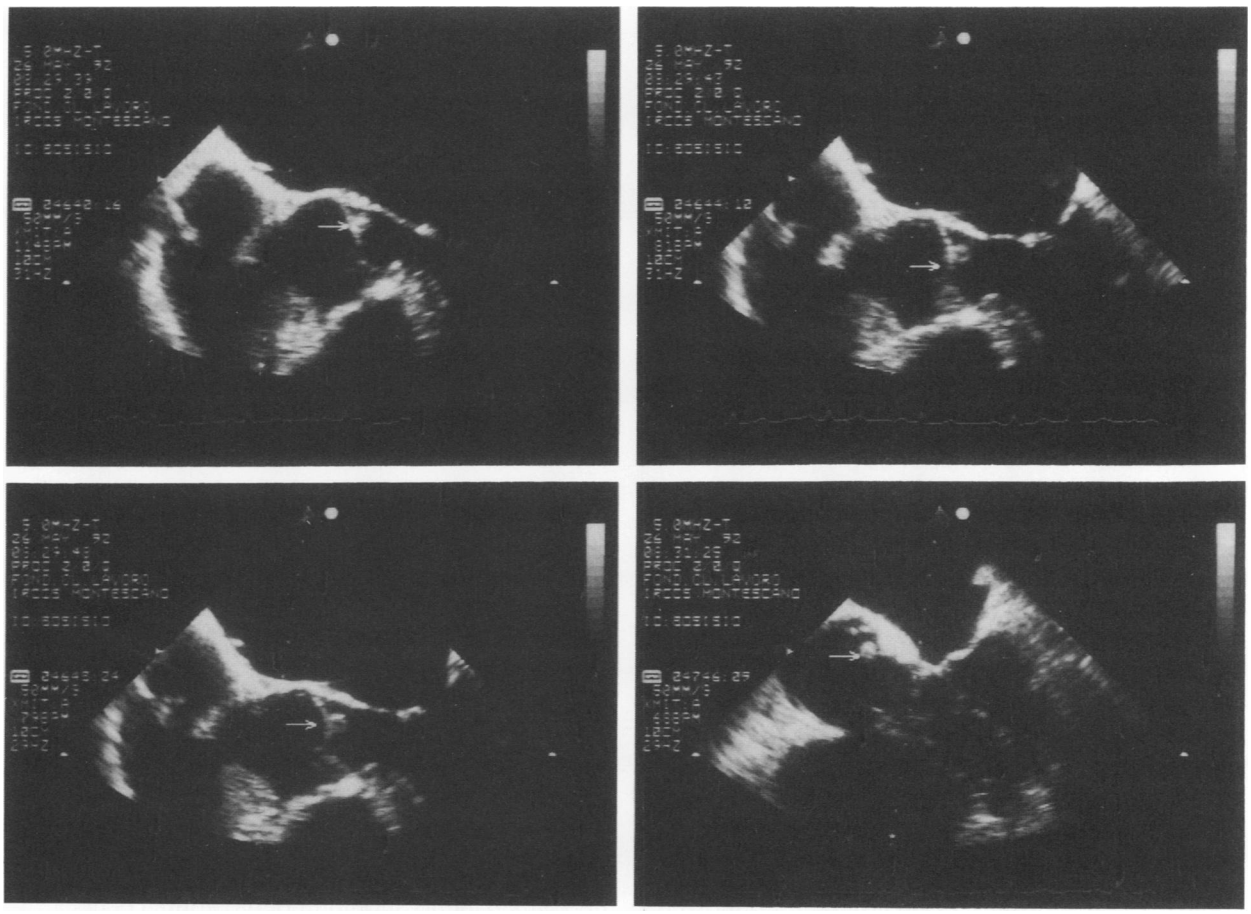

Fig. 2 *With the aid of transesophageal echocardiography, it is possible to visualize a small mass, only 5 mm in diameter, adherent to the coronary cusp of the aortic valve (arrows). (Patient 31)*

of right-heart failure 1 month after exploratory surgery. All 5 patients with malignant tumors survived operation and were discharged from the hospital. Survival after operation ranged from 1 to 6 months; all deaths were attributable to local recurrence.

Discussion

Primary cardiac tumors are rare. About 75% are benign, and 25% malignant. In most reports, myxomas (the most common primary tumors) comprise 40% of benign tumors, while sarcomas comprise 75% of malignant lesions.[1-5] In this study, the rate of malignant tumors (16%) was lower than might be expected, while the rate of myxoma occurrence among patients with benign lesions (84.6%) was much higher than usual.

Although myxomas are considered benign for lack of aggressive mitotic activity and metastasis, these tumors can cause cardiac insufficiency by interfering with atrioventricular or valvular function. Cardiac insufficiency can be found in 70% of patients with myxoma.[3]

Embolism, either of thrombus or of tumor fragments, is encountered in up to 40% of myxoma cases. In more than half these cases the embolism is cerebral;[6,7] alternatively, it can be localized to the coronary arteries,[8] or there can be wide dispersion of emboli from a right-sided tumor to the pulmonary vascular tree.[9] In our study, 13.6% of myxomas were associated with embolic episodes: 2 cases of stroke (Patients 4 and 15) and 1 case of transient ischemic attack. Among patients with benign nonmyxomatous tumors, only Patient 31 (with papillary fibroelastoma of the aortic valve) experienced an embolic episode, which was cerebral in nature. Cerebral embolism is a frequent indication of papillary fibroelastoma.[10]

Myxomas often can be asymptomatic: in 130 patients, McAllister and Fenoglio found that 12% had no symptoms.[11] In our series of 22 myxoma patients, we discovered 3 (13.6%) without symptoms.

An important aspect of our study is that the tumor was defined in all patients by echocardiography; cardiac angiography was performed in only 4 cases. Coronary angiography, however, should be performed routinely in older patients, who may have incidental coronary disease.

Transesophageal echocardiography can generate high-resolution images not obtainable by transthoracic echocardiography:[12,13] the direct contact with cardiac structures enables study of both atria and precision in finding the point of tumor attachment. In Patient 31 with papillary fibroelastoma of the aortic valve, transesophageal echocardiography showed a small, 5-mm mass adherent to the ventricular aspect of the left coronary cusp. Therefore, transesophageal echocardiography always should be considered when the results of transthoracic echocardiography are confusing.[14]

Magnetic resonance imaging can be of considerable value in differentiating the type of tumor.[15] In our study, densitometric analysis enabled a preoperative diagnosis of lipoma of the right ventricle (in Patient 25).[16] Because of its high tissue discrimination, computed tomography has the advantage in determining the degree of tumor extension.

Since Crafoord's 1st attempt to remove a left atrial myxoma in 1954,[17] surgical excision has become routine. We excised left atrial myxomas through a biatrial approach during total cardiopulmonary bypass support with bicaval cannulation, and removed the full thickness of the septum. We believe that the biatrial approach enables accurate exploration of all 4 cardiac chambers and avoids excessive tumor manipulation, reducing the risk of embolization.

In our series, the region of the myxoma's attachment was always resected, usually creating an interatrial septal defect that was repaired in 2 patients with a GORE-TEX® patch and in other patients by direct suture. Some authors[1,2,4] have recommended simple excision of the tumor attachment and have observed the patient for periods of 4 to 10 years without evidence of recurrence. Others[3,4,18] have performed a wide excision with patch repair in order to prevent recurrence. The rate of recurrence is less than 5%, and the main causes are inadequate tumor resection, intraoperative implantation, tumor embolization, and multiple myxomas (some undiscovered at surgery).[19] Again, we believe that the biatrial approach is best for exploration of all cardiac chambers.

Another important consideration is the prospect of familial myxoma. Often the tumor is diagnosed in a young patient (<20 years), and may be accompanied by a complex of conditions that can include lentigines, nevi, and adrenal and pituitary tumors. When the patient is young, it is critical to extend echocardiographic testing to the other members of the family. In our study, only 2 patients with myxoma were under 20 (Patients 8 and 29), and their relatives underwent echocardiography without the discovery of other tumors.

We had only 1 recurrence: in Patient 9, a myxoma arising from the interatrial septum and the posterior wall of the left atrium was in continuity with the posterior papillary muscle. As we said earlier, this recurrence was diagnosed 68 months after the procedure, and the patient died 4 months later of cardiac insufficiency. Because recurrence has been recorded even 10 years after the procedure,[20] all myxoma patients should undergo annual echocardiography.

Arrhythmias and other conduction alterations are frequent either in the immediate postoperative pe-

riod or later.[21] In our experience, atrial fibrillation after myxoma resection appeared in the immediate postoperative period in 4 patients (numbers 3, 5, 6, and 17). Sinus rhythm was obtained with amiodarone infusion in Patients 5 and 6 and with direct-current shock in Patients 3 and 17. The analysis of follow-up showed 4 other patients with atrial fibrillation (Patients 15, 19, 26, and 28); of these, Patients 15 and 28 had the arrhythmia before the operative procedure. Bateman and colleagues[21] have reported that arrhythmias can be caused by trauma to the specialized conduction tissue during myxoma resection and also by atrial distention during the biatrial approach.[22]

The prognosis in the other nonmalignant tumors is highly related to their resectability. Unresectable fibromas and lipomatous hypertrophy of the interatrial septum carry a poor prognosis, since they often result in arrhythmias and sudden death.[4] However, our Patient 20, with a partially resected atrioventricular fibroma, is asymptomatic 3½ years later; and his echocardiogram, done yearly, has not shown tumor growth. Murphy[3] reported that 7 children who did not undergo complete resection of cardiac fibroma were in good condition, with an average follow-up of 7.3 years; 1 of these underwent incision biopsy alone, and was alive and asymptomatic 9 years later. Therefore the operation can be postponed if resection is not feasible or if there are no symptoms indicative of obstruction or arrhythmias. On the other hand, Yamaguchi and coworkers[23] have said that it is mandatory to perform tumor resection in the newborn when congestive heart failure and dangerous arrhythmias are present.

The prognosis in patients with malignant cardiac tumors is poor: death often occurs within 6 months, regardless of the histologic diagnosis.[11] In our series of 5 patients with malignant tumors, the longest survival after surgery was 6 months, in the 9-year-old boy with fibrosarcoma (Patient 1). In the literature, there are 2 reported cases of heart transplantation for cardiac fibroma,[3] and autotransplantation was performed by Cooley in 1985[24] in order to resect a large pheochromocytoma of the posterior wall of the left atrium. Palliative surgery can improve the quality of life, while chemotherapy and radiation therapy can sometimes extend survival from 1 to 3 years.[25-27]

Our data, in general, support the case for aggressive surgical intervention. A cure can be achieved in patients with benign tumors, and palliation can be extended both to patients with malignant lesions and to those with unresectable or partially resectable benign tumors. However, improvement in adjuvant therapy will be necessary in order to increase survival times. In conclusion, the advancement in diagnostic techniques and the routine practice of echocardiography have enabled earlier detection of cardiac tumors and doubtless have improved the prognosis for many patients.

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