
Benign Cardiac Neoplasms: the Experience at the Cardiovascular Center of Puerto Rico and the Caribbean

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Introduction: Neoplasms of the heart are extremely rare and difficult to diagnose. In this study, we intend to describe the experience at the Cardiovascular Center of Puerto Rico and the Caribbean (CCPRC) dealing with benign cardiac neoplasms.

Methods: A retrospective review of the archival material at our institution was conducted using the 2007 ICD-9-CM code 212.7. A total of 77 cases with a diagnosis of Benign Heart Neoplasm were found from 1992 to 2008 but only those with an official pathologic report (n = 43) were considered. The clinical charts provided data related to age, sex, presenting symptoms, non-invasive and invasive characteristics, surgical procedures, and pathological findings. Data was compared using paired and unpaired Student's t-test, χ^2 , and Fisher's Exact Test when appropriate. For interpretation of results, a p-value < 0.05 was considered significant.

Results: The mean age for diagnosis was 51.49 years. The most common presenting symptom by which

these patients sought medical attention was congestive heart failure (35%), followed by chest pain (18%) and neurologic symptoms (14%). The most common location of the mass was the left atrium (81%) and overall, the most common heart neoplasm in this study was myxoma (83.7%). The rate of surgical complications was 30% (n = 13), with the cross clamp time and bypass time found to be the only factors influencing length of stay. Mortality rate was 7% (n = 3).

Conclusions: This study of a total of 43 pathologically confirmed benign cardiac neoplasms admitted to the Cardiovascular Center of Puerto Rico and the Caribbean from 1992 to 2008 validates the fact that heart neoplasms represent a fairly rare diagnosis. It is also consistent with previously published series that establish myxoma as the most common of these benign neoplasms. The main clinical findings as well as the results of surgery were presented.

Key words: Cardiac myxoma, Benign heart neoplasm, Surgical complications

The diagnosis of primary neoplasms of the heart may be elusive in some cases for two reasons: for one, they tend to mimic other cardiac conditions; and two, their occurrence is extremely rare with an estimated incidence of 0.1% (1). Out of these, nearly 75% are benign (2-6), myxomas being the most primary cardiac neoplasm. Cardiac myxomas tend to have a preference for the left side of the heart, as nearly 75-80% of them develop within the left atrium, the remainder mostly localized within the right atrium (7-10).

Clinical manifestations will depend mostly upon the localization of the tumor (7, 11). Neoplasms arising in the left atrium may obstruct appropriate circulation through the mitral valve, producing symptoms of heart

failure such as dyspnea, orthopnea, paroxysmal nocturnal dyspnea, cough, and fatigue. On the other hand, neoplasms arising in the right atrium may produce symptoms of right-sided heart failure and tricuspid valve stenosis. Ventricular neoplasms, whether right- or left-sided, interfere with ventricular filling and/or cause outflow obstruction, leading to syncope and symptoms of heart failure. Invasion of the myocardium may result in impaired contractility as well as conduction defects. Regardless of the localization of the tumor, embolization of thrombi -or even tumor fragments- may result in neurologic manifestations (12), pulmonary (8), and systemic emboli. Though cardiovascular manifestations are found in most patients with cardiac myxomas, these tumors are capable of producing various inflammatory cytokines and growth factors (13-14), which are believed to be responsible for constitutional symptoms observed in about one-third of the patients (15). Laboratory abnormalities, including thrombocytosis, hypergammaglobulinemia, elevated erythrocyte sedimentation rate, and C-reactive protein may also occur.

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Diagnosis is usually established by echocardiography, although cardiac magnetic resonance imaging (MRI) or computerized tomography (CT) scan may also be helpful identifying the presence of a cardiac tumor. Typically, myxomas are pedunculated and have a gelatinous consistency (16). Histologic examination reveals scattered cells within a mucopolysaccharide stroma (17).

Because of the risk of embolization and sudden cardiac death associated with these tumors, prompt resection is recommended once a diagnosis of myxoma has been established (9, 18-19). Surgical resection results are usually good, though up to 5% of patients may develop recurrent myxoma (16, 20).

In this study, we intend to describe the experience at the Cardiovascular Center of Puerto Rico and the Caribbean (CCPRC), a tertiary specialty hospital serving a large Hispanic population, dealing with benign cardiac masses.

Methods

Patient Selection

A retrospective review of the archival material at our institution was conducted. Using the 2007 ICD-9-CM code 212.7, a total of 77 cases with a diagnosis of Benign Heart Neoplasm were found from 1992 to 2008. This study considered only those records with an official pathologic report (n = 43). The clinical charts provided data related to age, sex, presenting symptoms, non-invasive and invasive characteristics, surgical procedures, and pathological findings. A clinicopathological comparison was made between those patients who had any type of complications during and after surgery with those who had no complications at all.

Statistical Analysis

For statistical analysis, categorical variables were reported as percentage and continuous variables were reported as mean. All calculations were conducted using the Statistical Package for the Social Sciences (SPSS) software. Data was compared using paired and unpaired Student's t-test, χ^2 , and Fisher's Exact Test when appropriate. For interpretation of results, a *p*-value < 0.05 was considered significant.

Results

Population Demographics

Out of the 77 reported cases of benign heart neoplasms between 1992 and 2008, a sample of 43 was included in statistical analysis. The mean age for diagnosis was 51.49 years. The majority of the patients (72%) were women (Table 1). The most prevalent medical conditions in

these patients were arterial hypertension (44%), diabetes mellitus (23%), atrial fibrillation (19%), coronary artery disease (12%), chronic kidney disease (9%), and smoking (9%). Only one patient (2%) had previous history of myxoma resection.

Table 1. Population Demographics

Characteristic	Result
Age (years)	51.49
Sex	
Male	12 (28%)
Female	31 (72%)
Weight (lbs)	149.15
Height (in)	61.3
Diabetes Mellitus	10 (23%)
Arterial Hypertension	19 (44%)
Dyslipidemia	3 (7%)
Chronic Kidney Disease	4 (9%)
Coronary Artery Disease	5 (12%)
Heart Failure	2 (5%)
Arrhythmias	8 (19%)
Smoker	4 (9%)
Bronchial Asthma	2 (5%)
Previous Myxoma Resection	1 (2%)
Cerebrovascular Events	1 (2%)

Initial Diagnosis

The most common presenting symptom by which these patients were diagnosed was congestive heart failure (CHF, i.e. shortness of breath, leg edema, dyspnea upon exertion, etc), found in 35% of the cases. Chest pain (18%) and neurologic symptoms including ischemic stroke and transient ischemic attacks (14%) were the second and third most common complaints (Figure 1). The diagnosis was

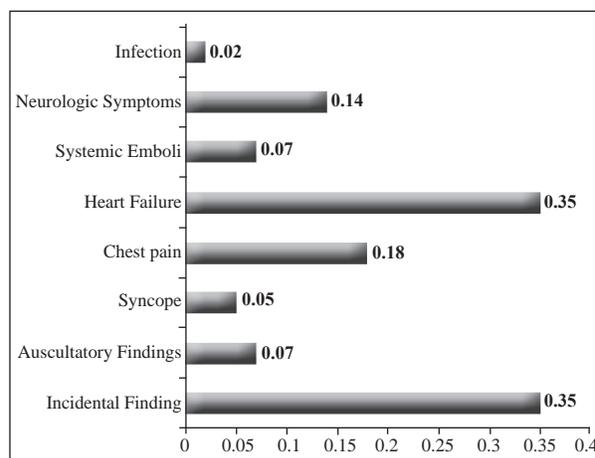


Figure 1. Clinical Presentation

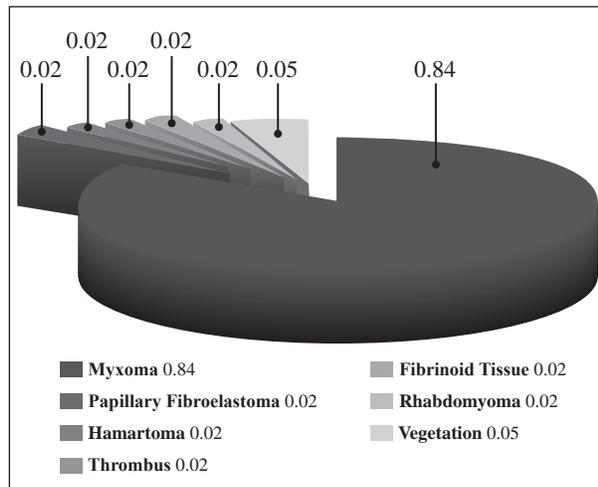


Figure 2. Pathologic Diagnosis

an incidental finding during echocardiography for some other reason in 35% of patients, and auscultatory findings seem to have a low sensitivity with only 7% of patients falling under this category. The majority of the patients were diagnosed with a conventional two-dimensional echocardiogram (80%).

The most common location of the mass was the left atrium (81%), followed by the right atrium (14%) and the left and right ventricles (2% each). Overall, the most common heart neoplasm in this study was the myxoma, seen in 83.7% of cases (Figure 2), with 91.7% of these located in the left atrium. Two patients were found to have large infective vegetations mimicking a cardiac mass.

Surgery and Complications

Thirty five percent (n = 15) of the patients had complex surgery with left atrial mass and atrial septal defect repair being the most common (16%, n = 7). The average cross clamp and bypass times were 35.79 and 55.85 minutes, respectively. The average length of stay was 9.11 days. The rate of complications was 30% (n = 13) with the most common complications being anemia (15%, n = 3), transient ischemic attacks (15%, n = 3), and death (15%, n = 3). Regarding the technicalities of the surgery itself (Table 2), cross clamp time was 31.6 min in the group without complications vs. 54.0 min in the other group (p 0.0017). Similarly, bypass time was 51.4 min in the group free of complications vs. 65.7 min (p 0.029). Length of stay was significantly higher in patients who suffered surgical complications, with an average stay of 13.4 vs. 7.2 days (p 0.007). There were no major differences between the location of the mass or the type of surgery between those that developed and did not develop complications.

Table 2. Surgical Complications

Characteristic	No Complications (n = 30)	Complications (n = 13)	p-Value
Cross Clamp (min)	31.6	54	0.0017
Bypass time (min)	51.4	65.7	0.0290
Length of stay (days)	7.2	13.4	0.0070

Discussion

This study validates the fact that heart neoplasms represent a fairly rare diagnosis and is consistent with previously published series that establish cardiac myxoma as the most common of these benign neoplasms. Regarding surgical treatment, patients with prolonged surgical times had more complications than those with lower surgical times. The emergence of complications was not influenced by the patient's pre-existing medical conditions, the location of the mass, or histology of the neoplasm. As expected, surgical complications nearly doubled hospital stay, which roughly translates into increased healthcare costs.

The mortality for intra-cardiac masses in this series was 7% (n = 3). Two of these deaths were unexpected. However, one patient was over 80 years old and weighed 160 pounds. The third death was an hemodialysis patient in septic shock with multiple systemic emboli. If we only take into account the cardiac myxomas cases, the mortality decreases to 5.5%, which is consistent with what is reported in the literature (< 5%). This demonstrates that our findings are in compliance with that of an ample review of literature on cardiac myxomas.

Resumen

Los tumores intracardiacos son extremadamente raros y difíciles de diagnosticar. En este estudio se describe la experiencia en el manejo de masas benignas intracardiacas en el Centro Cardiovascular de Puerto Rico y el Caribe. Se realizó un estudio retrospectivo donde se analizaron los expedientes médicos de aquellos pacientes admitidos con un diagnóstico de tumor benigno del corazón en el periodo del 1992 al 2008. Se analizaron un total de 43 casos con reporte oficial de patología. Los hallazgos clínicos principales así como los resultados de las intervenciones quirúrgicas fueron descritos y analizados.

References

- Lam KY, Dickens P, Chan ACL. Tumors of the Heart: A 20-Year Experience with a Review of 12,485 Consecutive Autopsies. Arch Pathol Lab Med 1993;117:1027-1031.

2. Molina JE, Edwards JE, Ward HB. Primary Cardiac Tumors: Experience at the University of Minnesota. *Thorac Cardiovasc Surg* 1990;38:183-191.
 3. Tazelaar HD, Locke TJ, McGregor, CGA. Pathology of Surgically Excised Primary Cardiac Tumors. *Mayo Clin Proc* 1992;67:947-954.
 4. Larrieu AJ, Jamieson WR, Tyers GF, et al. Primary Cardiac Tumors: Experience with 25 Cases. *J Thorac Cardiovasc Surg* 1982;83:339-348.
 5. Odum J, Reehal V, Laks H, et al. Surgical Pathology of Cardiac Tumors: Two Decades at an Urban Institution. *Cardiovasc Pathol* 2003;12:267-270.
 6. Kamiya H, Yasuda T, Nagamine H, et al. Surgical Treatment of Primary Cardiac Tumors: 28 Years' Experience in Kanazawa University Hospital. *Jpn Circ J* 2001;65:315-319.
 7. Vander-Salm TJ. Unusual Primary Tumors of the Heart. *Semin Thorac Cardiovasc Surg* 2000;12:89-100.
 8. Kuon E, Kreplin M, Weiss W, Dahm, JB. The Challenge Presented by Right Atrial Myxoma. *Herz* 2004;29:702-706.
 9. Keeling IM, Oberwalder P, Anelli-Monti M, et al. Cardiac Myxomas: 24 Years of Experience in 49 Patients. *Eur J Cardiothorac Surg* 2002;22:971-977.
 10. Jelic J, Milicic D, Alfirevic I, et al. Cardiac Myxoma: Diagnostic Approach, Surgical Treatment and Follow-up: A Twenty Years Experience. *J Cardiovasc Surg* 1996;37:113-117.
 11. Colucci WS, Schoen FJ. Primary Tumors of the Heart. In: Braunwald, E; Zipes, DP; Libby, P (Eds). *Heart Disease*, 6th ed, WB Saunders, 2001.
 12. Lee VH, Connolly HM, Brown RD. Central Nervous System Manifestations of Cardiac Myxoma. *Arch Neurol* 2007;64:1115-1120.
 13. Sakamoto H, Sakamaki T, Kanda T, et al. Vascular Endothelial Growth Factor is an Autocrine Growth Factor for Cardiac Myxoma Cells. *Circ J* 2004;68:488-495.
 14. Seino Y, Ikeda U, Shinada K. Increased Expression of Interleukin-6 mRNA in Cardiac Myxomas. *Br Heart J* 1993;69:565-568.
 15. Maisch B. Immunology of Cardiac Tumors. *Thorac Cardiovasc Surg* 1990;38(Suppl 2):157-161.
 16. Pinede L, Duhaut P, Loire R. Clinical Presentation of Left Atrial Cardiac Myxoma: A Series of 112 Consecutive Cases. *Medicine* 2001;80:159-162.
 17. Ferrans VJ, Roberts WC. Structural Features of Cardiac myxomas: Histology, Histochemistry, and Electron Microscopy. *Hum Pathol* 1973;4:111-146.
 18. Selkane C, Amahzoune B, Chavanis N, et al. Changing Management of Cardiac Myxoma Based on a Series of 40 Cases with Long-Term Follow-up. *Ann thorac Surg* 2003;76:1935-1938.
 19. Cina SJ, Smialek JE, Burke AP, et al. Primary Cardiac Tumors Causing Sudden Death: A Review of the Literature. *Am J Forensic Med Pathol* 1996;17:271-281.
 20. Centofanti P, Di Rosa E, Deorsola L. Primary Cardiac Tumors: Early and Late Results of Surgical Treatment in 91 Patients. *Ann Thorac Surg* 1999;68:1236-1241.
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