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Atrial Myxomas: A Single Unit's Experience in the Modern Era

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ABSTRACT

Background: Although an atrial myxoma is the commonest cardiac tumor, it is still relatively rare, with an annual incidence of approximately 0.5 per million. In our unit, which performs 1000 major cardiac procedures per year, this equates to approximately 3 patients annually. We therefore sought to evaluate our experience of managing this type of tumor over the last 5 years.

Methods: A retrospective review was performed of prospectively collected data from the departmental database. We analyzed consecutive patients who were operated upon between 2002 and 2007. Three patients with a papillary fibroelastoma on histological examination were excluded from this study.

Results: We have performed excision of atrial myxoma in 18 patients. Twelve patients (66%) were female; the median age was 64 years (range, 35-80 years), and the median logistic euroSCORE was 5.22% (range, 1.51-27.82%). Fifteen patients (83%) were deemed urgent, 2 elective, and 1 emergency. Sixteen tumors (89%) were left sided. Symptoms attributable to the tumor were found in 16 of the 18 patients (embolic, n = 9; chest pain, n = 3; palpitations, n = 2; incidental finding, n = 2, others n = 4), and the mean time from diagnosis to operation was 3 days (range, 0-22 months). The median cardiopulmonary bypass time was 87 minutes (range, 28-228 minutes), with the median aortic cross clamp time being 61 minutes (16-175 minutes).

The approaches used were transeptal via right atriotomy (n = 8), biatrial/Dubost (n = 4), left atrial (n = 4), and right atrial (n = 2); the interatrial septum was involved in 14 patients. The resultant defect was closed using a pericardial (n = 8) or prosthetic patch (n = 5) or directly sutured (n = 5). Concomitant procedures were performed in 8 patients (coronary artery bypass graft [CABG], n = 4; mitral valve replacement [MVR], n = 2; valve + grafts, n = 2). All tumors were completely excised.

Postoperatively there were no deaths within 30 days of the procedure. Indeed, only 2 patients have died at 4 and 25 months postoperatively, respectively, both of unrelated causes. Median intensive therapy unit (ITU) stay was 2 days (range, 1-9 days), and median hospital stay was 10 days (range, 5-20 days). A permanent pacemaker was required in only 1 patient, and median blood loss was 340 mL (range, 140-1760 mL). Atrial fibrillation was the commonest complication affecting 6/18 patients (33%).

Conclusions: Excision of atrial myxoma can be performed using a variety of intraoperative approaches and closure techniques, all with acceptable postoperative morbidity and low mortality rates. To date, no recurrences have been found at median 2-year follow-up.

INTRODUCTION

Atrial myxoma is the commonest primary cardiac tumor, although it is still relatively rare, with an annual incidence of approximately 0.5 per million population [MacGowan 1993]. The presentation of this tumor is usually sporadic, with symptoms ranging from fever/malaise to embolic phenomena and occasionally syncope due to obstruction of the mitral valve [Walkes 2008]. The pathology of the tumor and its relationship with the left atrium and mitral valve is well documented. In recent years, with increasing experience of mitral valve surgery and approaches to the left atrium, we sought to evaluate our unit's experience of managing this type of tumor and examine any changes in practice. Our unit performs approximately 1000 major cardiac operations per year and serves a population of about 2 million people. We therefore calculated that we should encounter 2 to 3 atrial myxomas per year.

PATIENTS AND METHODS

A retrospective review was performed of all prospectively collected data from the departmental database since its inception. All patients who underwent atrial myxoma excision between January 2001 and December 2007 were included in the study. A total of 18 patients underwent excision of atrial myxoma during the 7-year study period. An additional 3 patients presented with atrial masses suspected to be

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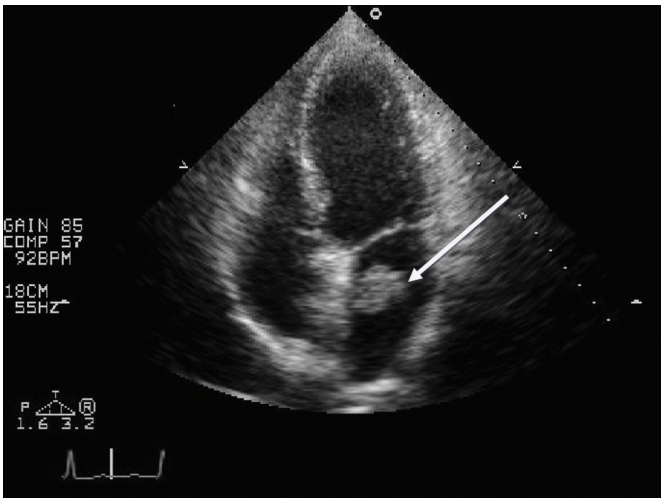

Figure 1. Transthoracic echocardiogram (4-chambered view) showing left atrial myxoma (arrow). Courtesy of Northern General Hospital, Sheffield.

myxomas, but histological examination confirmed papillary fibroelastoma, and thus these patients were excluded.

All data are presented as median and range unless otherwise stated. Proportions are expressed as actual number and percentage of the population.

The patients' demographic details, symptoms, investigations, preoperative, perioperative, and postoperative data were reviewed. The majority of patients were female (n = 12 [66%]), with a median age of 64 years (range, 35-80 years). The median time from diagnosis to operation was 3 days (range, 0-22 months). One patient presented with features of mitral valve obstruction and underwent an emergency procedure. Two patients were operated upon electively—1 of the tumors was found during routine investigation for atrial fibrillation with no other symptoms, and the other patient was treated as a left atrial thrombus secondary to atrial fibrillation and anticoagulated. When the "thrombus" failed to resolve, the patient was referred to our unit, and the mass promptly excised.

Symptoms attributable to the tumor were found in 16 of 18 patients. Interestingly, half of the patients (9/18 [50%]) presented with embolic phenomena. The myxoma was found incidentally in 6 patients (33%) during investigation for atrial fibrillation, and only 3 patients (17%) were diagnosed with features of mitral valve obstruction.

RESULTS

The median logistic euroSCORE of the 18 patients undergoing resection of their atrial myxoma was 5.22 (range, 1.51-27.82). In accordance with the published literature 16 (89%) of the tumous were left-sided, and the inter-atrial septum was involved in 77% of cases (n = 14). Median cardiopulmonary bypass time was 87 minutes (range, 28-228 minutes), with the aortic cross clamp time being 61 minutes (range, 16-175 minutes). The median intraoperative blood loss was 340 mL (range, 140-1760 mL).

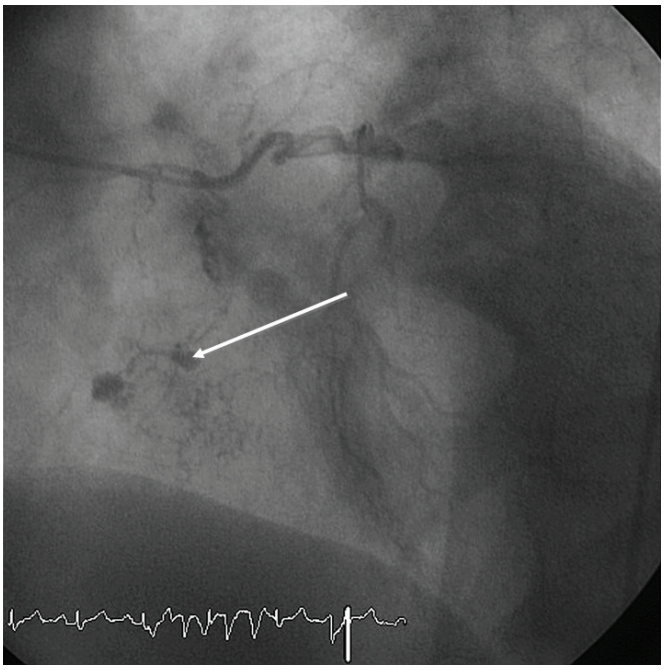

Figure 2. Coronary angiogram: right anterior oblique view demonstrating tumor "blush" of the same atrial myxoma seen in Figure 1. Courtesy of Northern General Hospital, Sheffield.

The approaches used for the 16 left sided tumors were transeptal via right atriotomy in 8 of 18 cases (45%), bi-atrial/Dubost in 4 of 18 cases (22%), and left atriotomy in 4 of 18 cases (22%). The 2 right atrial tumors (2 of 18, 11%) were operated on via right atriotomy. The resultant defects were closed using a pericardial patch in 8 of 18 patients (44%), prosthetic Dacron patch in 5 of 18 patients (28%), or directly sutured in 5 of 18 patients (28%). Histological examination confirmed that all tumors were completely excised.

Concomitant procedures were performed on 8 patients, coronary artery bypass grafting (CABG) was performed in 4 patients, 3 of them single saphenous vein grafts, 1 a triple bypass. Two patients had a mitral valve replacement, 2 patients had valve replacement and CABG (1 aortic, 1 mitral). One further patient underwent percutaneous coronary intervention (PCI). A permanent pacemaker was required by only 1 patient.

The median ITU stay was 2 days (range, 1-9 days), and hospital stay 10 days (range, 5-20 days). Atrial fibrillation was the most common postoperative complication, affecting 6 of 18 patients (33%).

Postoperatively, there were no deaths within 30 days of the procedure. Indeed, only 2 patients have died at 4 and 22 months postoperatively, respectively, both of noncardiac causes.

All patients are reviewed annually with echocardiographic surveillance. To date, no recurrences have been found at a median 2-year follow-up. This ongoing study will be extended to ascertain long-term recurrence and mortality data.

Summary of the Most Recent Case Series Involving Atrial Myxomas

	This Study	Lijoi 1993	Meyns 1993	Keeling 2002	Stevens 2003	Yu 2006
Number of cases, n	18	26	32	49	58	74
Female patients, %	66	54	72	75	66	72
Mode of presentation, %						
Obstruction	17	54	66	61	18	60
Embolization	50	31	16	35	31	11
Incidental	33	23	16	—	12	14
Location of tumor, %						
Left atrium	89	85	97	96	91	95
Right atrium	11	8	3	4	7	5
Surgical approach, %						
Left atrial	22	68	—	—	41	14
Bi-atrial	22	23	—	65	59	54
Right atrial/trans-septal	44	—	—	—	—	—
Right atrial	11	8	—	—	0	37
Defect closure, %						
Suture	28	87	94	94	72	27
Patch	72	13	6	6	28	70
Concomitant procedure, %	44	—	6	18	29	13
ITU stay, d	2	—	2	—	—	2
Hospital stay, d	10	—	12	—	—	12
Postoperative complications, %						
Atrial fibrillation	33	35	3	23	39	3
In-hospital mortality	0	4	3	2	2	0

DISCUSSION

Atrial myxomas can be complex cardiac problems that require urgent treatment. They are the most common cardiac tumor, but are still relatively rare. They annual incidence is approximately 0.5 per million population [MacGowan 1993], or approximately 1 in every 500 cardiac operations [Walkes 2008]. Certainly the incidence of atrial myxomas in our study mirrors that found in other larger series [Tazelaar 1992]. They account for 40% to 50% of all cardiac tumors [Walkes 2008] and rarely metastasize or recur if excision is complete [Castells 1993]. So far, we have encountered no patients with recurrence. Most cases of atrial myxoma are sporadic, with approximately 10% being familial—these are inherited in an autosomal dominant fashion. Multiple tumors occur in approximately 50% of these familial cases.

The majority (75%) of sporadic myxomas are located in the left atrium, with 10% to 20% in the right atrium and the remainder located in the ventricle. Many of these tumors involve the inter-atrial septum and are characteristically described as having a pedunculated and polypoid appearance on macroscopic examination, with a microscopic appearance of polygonal cells encased in an acid muco-polysaccharide matrix [Reynen 1995].

The location of tumors in our series of patients appears to corroborate the published data.

Atrial myxomas can present in a number of ways depending upon which chamber of the heart is involved. The majority of cases present with features suggesting obstruction to blood flow such as dyspnea [Goodwin 1968], palpitations, and syncope [St John Sutton 1980]; up to 30% of patients may present with features of embolism [Veitch 2006]. Myxomas have also been diagnosed as an incidental finding in patients who present with constitutional features such as fever, weight loss, and lethargy [Walkes 2008], or even Raynaud's phenomenon [Skanse 1959]. Our patient group, however, appears to differ in that half of our patients presented with embolic features, and only one-third described obstructive symptoms. Whether this is due to small numbers of patients studied or the population from which they were selected is unclear.

The gold standard for investigation of an atrial mass is echocardiography (Figure 1), whether it is transesophageal echocardiography (TOE) or transthoracic echocardiography (TTE) [Engberding 1993; Daniel 1995]. Echocardiography has 100% sensitivity for diagnosing the size, site, mobility, local relationships, and attachments of the tumor [Engberding 1993]. Intraoperative TOE can also assess the posterior wall of the atria as well as the intra-atrial septum and right

atrium, which are often not well visualised on TTE. In our hospital, all patients had intraoperative TOE because it is an easily accessible and cheap investigation that provides the surgeons with clear views of the tumor and its local relations, thus helping to plan operative strategy.

Atrial myxomas have a peak incidence between the third and sixth decades of life [Walkes 2008]. In these patients, concomitant cardiac conditions such as coronary artery disease would not be entirely unexpected. We feel that coronary angiography is an essential component of the preoperative work-up of these patients [Singh 1984; Janas 2006]. Not only can it diagnose any associated coronary artery stenoses, but it can also visualize the vascular supply to the tumor (Figure 2). This may also modify the operative strategy [Janas 2006] such that failure to ligate a large arterial branch supplying the tumor may result in excessive intraoperative bleeding, or even a "steal syndrome," whereby blood may be directed from the coronary arteries into the left atrium causing myocardial ischemia [Janas 2006]. In our unit, we perform angiography in all but the most urgent of patients undergoing resection of their atrial myxoma.

Differential diagnoses of an atrial mass include thrombus or vegetation [MacGowan 1993]. The delay from diagnosis to procedure in 1 patient of 22 months immediately raises concern. It should be noted that this is the time from initial detection of a left atrial mass to surgery. Unfortunately, the mass was initially diagnosed as a thrombus, and the patient was anticoagulated. When the mass failed to resolve, she was then referred to our institution, whereupon the mass was promptly excised. Despite the urgency with which all procedures in our series were performed following referral, due to the variability of the presenting signs and symptoms, a long lapse of time between these and the time of diagnosis is not unusual [Alvarez-Sabín 2001] and has been reported to be up to 7 years [Knepper 1988]!

In recent years, there has been a trend toward minimal access cardiac surgery, particularly in aortic and mitral valve repair/replacement—the Cleveland Clinic, for example, performing 90% of their aortic valve replacements using this technique [Gillinov 2000]. The theoretical benefits include better cosmesis [Christiansen 1999; Mächler 1999], less need for ventilation [Mächler 1999], less pain, and quicker return to normal activities [Chitwood 2003]. Reports of atrial myxomas being resected by minimally invasive techniques are few [Walkes 2008]. In our series, all procedures were performed by full sternotomy and utilized cardiopulmonary bypass.

A summary of the most recent case series involving atrial myxomas is shown in the Table [Lijoi 1993; Meyns 1993; Keeling 2002; Stevens 2003; Yu 2006].

In summary, resection of atrial myxoma can be performed safely using a variety of intraoperative approaches and defect closure techniques. We experienced very few postoperative complications and zero mortality, despite a high calculated risk of mortality and morbidity in some cases. This again reflects current worldwide surgical practice [Christiansen 1999; Mächler 1999].

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