

# Fifteen-year experience in the surgical management of right atrial myxoma

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## Abstract

**Objective:** This study presents our experience with the clinical presentation, tumor characteristics, surgical management and post-operative outcomes of patients operated for right atrial myxoma.

**Methods:** A retrospective analysis of all patients diagnosed with right atrial myxoma in our center from years 2000 to 2015 was conducted. All data were gathered from medical records following board approval. Twelve cases were identified, eight of which were female. Data is reported as frequencies, percentages, averages and ranges.

**Results:** Clinical presentation proved very heterogeneous amongst our group. The most common finding was right-sided heart failure in 2 patients (17%). The average tumor size was 30mm and the most common attachment site was the inter-atrial septum. Myxomas were resected through median sternotomy or right thoracotomy. Post-operative morbidity includes dysrhythmias and tricuspid disease despite morphologically normal tricuspid structure in most cases. There was no perioperative mortality. Follow-up averaged 39 months and showed no recurrence.

**Conclusions:** While surgical results for right atrial myxoma are satisfactory, its inconsistent presentation and rarity continue to pose a diagnostic challenge to clinicians who encounter it. Our data and review of literature identified right-sided heart failure, tricuspid regurgitation and pulmonary embolism as common findings in right atrial myxoma and lead us to suggest that clinicians consider it in their differential diagnosis in situations where such findings go unexplained.

**Abbreviations:** RAM: right atrial myxoma; ECC: extracorporeal circulation; TR: tricuspid regurgitation; TTE: trans-thoracic echocardiography

## Perspective statement

Right atrial myxomas (RAM) are exceedingly rare, and literature describing them is limited. This study contributes a large center's experience in the management of RAM and offers guidelines aimed at helping clinicians recognize situations in which it should figure amongst their differential diagnosis. Outcomes and complications are discussed to help surgeons make informed decisions when dealing with RAM.

## Central message

Surgical excision remains the mainstay of therapy for right atrial myxoma as it provides excellent outcomes.

## Introduction

Primary cardiac tumors are very rare, with a reported incidence averaging 0.02% throughout various autopsy studies [1]. Myxomas account for the majority of these and most commonly originate from the left atrium [2,3]. Right atrial myxoma (RAM) is a rare benign tumor with an often-asymptomatic presentation [2,4]. Echocardiography plays an important role in its diagnosis while surgical excision remains the only curative treatment [4]. Few data describing this condition are currently available. The objective of this study is to document the clinical presentation, surgical management and post-operative

outcomes of right atrial myxoma in hopes of helping clinicians better recognize and manage this condition.

## Methods

A retrospective analysis of all patients who underwent surgical excision of right atrial myxoma (RAM) at a single institution from years 2000 to 2015 was conducted after receiving board approval. Twelve cases were identified. The patient population was composed of 8 women (67%) and 4 men (33%) with a mean age of  $55 \pm 17$  years (range 19 to 87). All patients had submitted to trans-thoracic echocardiography (TTE) and magnetic resonance imaging preoperatively. All diagnoses were confirmed by pathology. Information on presenting symptoms, surgical interventions, early outcomes and follow-up details were gathered from medical records and defined according to the Society of Thoracic Surgery guidelines for reporting mortality and morbidity. Nominal data are reported as frequencies and percentages while numerical data are reported as a mean value and range. No supplementary statistical analysis was performed.

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**Key words:** right atrial myxoma, surgical management, outcomes, complications

**Received:** December 05, 2017; **Accepted:** December 27, 2017; **Published:** December 30, 2017

## Results

### Presentation

Symptoms at presentation were very variable in our series. At time of discovery, 8 patients (67%) were asymptomatic. Others presented with right-sided heart failure (2 patients, 17%), isolated tachycardia (1 patient, 8%) and atypical chest pain (1 patient, 8%). One patient's right atrial myxoma was discovered during the excision of a left atrial myxoma. All patients were in sinus rhythm at the time of diagnosis. Left ventricular ejection fraction was preserved (>55%) in all but one patient. Nine patients (75%) were in NYHA functional class I while the remaining three were in classes II and III. Additional preoperative findings include pulmonary embolism in the patient with atypical chest pain and pleural effusion in another. Comorbidities consisted mainly of cancers of the colon (2 patients, 17%) and lung (1 patient, 8%), as well as a combination of testicular neoplasm and follicular lymphoma, possibly in the setting of Carney complex (1 patient, 8%). One patient suffered from moderate tricuspid regurgitation (TR) and stable coronary artery disease preoperatively. Family history for cardiac myxoma was positive in two (17%).

### Tumor characteristics

Tumor size ranged from 20 to 80mm in the longest axis, averaging 30mm. The most frequent implantation site was the inter-atrial septum (9 tumors, 75%) while the others emerged from the atrial free wall (3 tumors, 25%). All tumors were macroscopically described as polylobate and friable. All diagnoses were confirmed by pathology. Tumor characteristics and clinical presentations are summarized in Table 1.

### Surgical management

All surgeries were performed with the use of extracorporeal circulation (ECC) under general anesthesia. Ten (83%) were approached by way of median sternotomy under full cardiopulmonary bypass. The other two (17%) were approached by way of right thoracotomy on a pump-assisted beating heart. Aortobicaval cannulation was used in 9 of the 10 surgeries performed via sternotomy, while both thoracotomies and one sternotomy were performed utilizing an aortic, superior vena cava and right femoral vein cannulation approach. The approach chosen depended on both tumor location and surgeon experience. Average ECC and cross-clamp times were 45 minutes (range 14 to 90) and 23 minutes (range 0 to 56) respectively. Tumors were removed in a single piece as much as possible in an effort to prevent the shedding of tumor debris into the heart cavity. In two cases surgeons noted that the tumor was unusually friable, one of which was in the patient who developed this series' only case of pulmonary embolism preoperatively.

Concomitant procedures for associated cardiac lesions include tricuspid valvuloplasty and coronary artery bypass grafting, in the same patient. In six patients it was necessary to use either Dacron or autologous pericardial patches to repair three atrial wall defects and three atrial septal defects, of which one was congenital. In one case surgeons were surprised to find a residual mass in the inferior vena cava measuring 1.5 x 1.0 cm on post-ECC trans-esophageal echocardiography. This segment of the tumor was presumably hidden by the cannula inserted into the inferior vena cava throughout the procedure. Considering the small size of the residual mass, it was decided to leave it in place to avoid cannulating the heart a second time.

### Outcomes

Average ICU and hospitalization times were respectively 1.5 days (range 1 to 2) and 5 days (range 4 to 7) with no perioperative mortality. Early complications include delirium (1 patient), pleural effusion (1 patient), supraventricular tachycardia (1 patient) and sinus node disease that required permanent pacemaker implantation (1 patient). One patient was reoperated for excessive bleeding.

One patient underwent concomitant tricuspid valvuloplasty but showed persistence of his TR despite the surgery. His valve was mildly thickened. Another patient developed moderate TR on early post-operative TTE after excision of a particularly large myxoma, as did a third patient during the follow-up period. Tricuspid structure was normal in these patients. Average follow up was 39 months (range 13 to 72) and showed no mortality or recurrence. The surgical management and associated outcomes are summarized in Table 2.

### Discussion

Right atrial myxoma is a rare benign tumor with a highly variable and non-specific presentation [4-6]. It rarely figures amongst a clinician's differential diagnosis and continues to be discovered by chance on routine investigations in many cases [5]. In our experience and others [5,7-10], findings in patients with RAM tend to include right-sided heart failure, tricuspid regurgitation, pleural effusion and pulmonary embolism.

We recommend that the otherwise unexplained presence of such findings in an adult patient lead a clinician to consider right atrial myxoma as a potential diagnosis and to evaluate the necessity for further investigations, most probably by way of TTE.

It is of interest to note that patients who developed right-sided heart failure had two of the largest myxomas of this series, each measuring 50mm and 80mm. This finding leads us to consider obstruction of the tricuspid valve by a voluminous myxoma to be the likely cause of isolated right heart failure in these patients, as has been found in a similar case report [11].

Once diagnosis is made, treatment should be initiated promptly to avoid eventual local or embolic consequences. Locally these included tricuspid regurgitation and right-sided heart failure in our series. Embolic consequences most frequently are limited to pulmonary emboli (8% at presentation in this series), although cases of stroke due to RAM have been reported in a patient with atrial septal defect secundum [12]. Interesting of note is that the presence of both local and embolic findings at presentation seemed to correlate positively with tumor size in our study. Our population however was not large enough to establish a statistically meaningful correlation. Such associations could perhaps be made on large-scale study.

**Table 1.** Presentation and characteristics of right atrial myxoma

	None	8	67%	
Clinical manifestation of RAM <sup>1</sup>	Right-sided heart failure	2	17%	
	Atypical chest pain <sup>2</sup>	1	8%	
	Isolated achycardia	1	8%	
	Size (mm)	20 to 39	7	58%
40 to 59		4	33%	
60 to 80		1	8%	
Tumor characteristics	Implantation	Interatrial septum	9	75%
		Right atrial free wall	3	25%

<sup>1</sup>RAM=right atrial myxoma, <sup>2</sup>The authors attribute this symptom to the patient's known stable coronary artery disease

**Table 2.** Surgical management and associated post-op complications

Associated surgical procedures	Tricuspid valvuloplasty	1	8%
	CABG	1	8%
Patch closure	Atrial free wall	3	25%
	Atrial septum	3	25%
	Not required	6	50%
Complications	Tricuspid regurgitation	3 (2 early, 1 late)	25%
	Dysrhythmia	2	17%
	Bleeding (requiring surgical intervention)	1	8%
	Mortality	None	-

Surgery proved curative in 100% of cases in our study with no recurrence and no mortality during follow-up. Post-operative complication rates are passable, although surgeons should be aware of the risk of damaging cardiac conduction tissue during excision of RAM, in particular those attached to the inter-atrial septum. Low mortality, recurrence and complication rates continue to be reported [5].

We look forward to large scale studies to help better outline investigation guidelines for right atrial myxoma in clinical practice. Recent literature continues to highlight the excellent results achieved with surgical excision of RAM. Alternative treatment strategies have yet to be defined for patients with an unacceptable operative risk.

## Conclusion

Right atrial myxoma is a rare myocardial tumor with a favorable prognosis. Although diagnosis is difficult, treatment is usually curative and helps prevent the development of serious complications such as embolization and valvular disease.

## Ethics approval

Approval for this study was obtained from the Montreal Heart Institute Ethics Committee.

## Consent for publication

All authors of this study express their consent for the publication of this work.

## Conflict of interest

None to declare.

## Funding

No funding was received for this work.

## Author's contributions

- Hassane Abdallah: Data collection, production of text, data analysis.
- Justin Michetti: Production of text, data analysis.
- Philippe Demers: Data collection and analysis.
- Raymond Cartier: Data collection and analysis.

## Acknowledgements

Not applicable

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