

# Six Years Clinical Experience and Surgical Considerations in Management of Cardiac Myxoma in Iraqi Center for Heart Disease—Single Center Experience

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### Abstract

Background: Cardiac myxomas are the most frequently encountered benign cardiac tumors that if left untreated are inexorably progressive and potentially fatal. Surgery is the only way of treatment, and if not treated with the right surgical technique recurrence occurs. Objectives: In this single center study we documented the patterns of presentation, localization, surgical approaches and outcome of cardiac myxomas. Methods: This is a retrospective study of 20 patients who underwent surgical removal of atrial myxoma from January 2010 to December 2015. All patients underwent general investigations, and echocardiography was performed on all patients and surgery was done using extracorporeal circulation and mild hypothermia. Results: The ages of the patients ranged from 14 years to 71 years, with a mean of 51.45 years. Most myxomas (75%) originated from left atrium, 20% from right atrium and biatrial in 5% of cases. The male-to-female ratio was 1:2.3 (14 females and 6 males). Myxomas were more common in blood group A+ and B+. Chief complaints were dyspnea (70%) and palpitation (50%). The majority of masses were attached to the interatrial septum (65%) and four of cases (20%) arose from the lateral wall. Right atrial trans-septal incision was used in 55% of cases. No recurrence was recorded in our study. Six patients had postoperative complications, mainly in the form of arrhythmia (3 cases), bleeding (0ne case) and renal failure (one case) which resulted in the death of the patient.

**Conclusions:** Cardiac myxoma excision account for a very small percentage of cardiac procedures. Immediate surgical treatment is indicated because of high risk of embolization and sudden death. Cardiac myxomas can be excised with a low rate of mortality and morbidity. Follow-up examination, including echocardiography, should be performed regularly.

### **Keywords**

Atrial Myxoma, Cardiac Tumors, Cardiac Surgery

# **1. Introduction**

The most frequent tumor of the heart is an intra-cardiac myxoma, which is thought to occur 0.5 times per million people annually [1]. Myxomata make up 15% of benign cardiac tumors in children and 50% of benign cardiac tumors in adults [2]. They are benign. They have been documented in both sexes and in all age categories, but they most frequently affect women between the ages of 30 and 60. Although myxomas are typically sporadic, at least 7% of cases are associated with an autosomal dominant condition. In the latter case, the myxoma is a part of a more extensive disease known as the Carney complex. Myxomas are connected to patchy skin pigmentation and endocrine hyperactivity in the Carney complex [3]. Any cardiac tumor, but especially myxomas, may exhibit systemic symptoms and be mistaken for collagen vascular disorders [4]. Myxomas typically grow from the endocardium and enter a heart chamber [3]. They originate from the interatrial septum in the left atrium close to the fossa ovalis around 75% of the time [5]. About 15 - 20 percent of the rest will develop in the right atrium; they can come from valve surfaces and the walls of other heart chambers [3]. These tumors are pedunculated and have a gelatinous nature under the microscope. The surface may be smooth (65%), villous, or friable [6]. These tumors can range in size from 1 to 15 cm in diameter, but they are often around 5 cm in diameter [3] [7]. The most frequent presenting symptoms are produced by restricted cardiac blood flow, and the tumor's size, location, and degree of mobility all have an impact on how severe the symptoms are [8]. The blockage progresses as is typical [9]. The mitral valve is the most often occluded location; this form of blockage causes clinical indications resembling those of mitral valve disease. The symptoms of a mobile tumor are frequently intermittent and may alter in intensity when the patient shifts positions. Embolic events happen when tissue fragments from the tumor break off and enter the bloodstream or when thrombi on the tumor's surface detach to generate emboli. As emboli move from the systemic circulation into the cerebral circulation, the majority of myxoma-related embolic events are transitory ischemia episodes or cerebrovascular accidents. The coronary, retinal, iliac, and femoral arteries can also be affected by myxoma-related emboli or tumor pieces [8] [10] [11]. A wide range of constitutional symptoms, including fever, myalgia, and arthralgia, are reported by almost all myxoma patients. Leukocytosis, increased erythrocyte levels and sedimentation rate, thrombocytopenia, and raised C-reactive protein may also accompany these complaints [2]. About one-third of cases have hemolytic anemia, which is most common when a calcified myxoma is present. This hemolytic anemia and the thrombocytopenia that occasionally occurs are likely caused by the mechanical destruction of produced blood components. With tumor removal, these characteristics can be reversed [9]. A myxoma infection is a rare condition that manifests clinically as infected endocarditis. An infected myxoma requires prompt surgical resection because infection raises the risk of systemic embolization [2]. The most important diagnostic tool, echocardiography, can inadvertently discover myxomas. The only effective therapy option for individuals with cardiac myxoma is surgical resection, which should not be postponed because up to 8% of patients waiting for surgery may die from embolization or obstruction to flow within the heart. Usually, a median sternotomy technique is used, along with ascending aortic and bi-caval cannulation. Due to the known embolic tendency and friability of myxomas, manipulation of the heart prior to the start of cardiopulmonary bypass is minimized. To prevent additional embolization and provide the patient enough time to stabilize their brain for cardiopulmonary bypass, the tumor should be removed seven days after the incident if there was preoperatively known cerebral embolization without hemorrhage. The tumor is removed in one piece. It is possible to remove tumors from generally well-defined pedicles without completely removing an atrial wall button. However, a lot of surgeons favor excising a section of the atrial wall, particularly when the tumor is broad-based or originates from the interatrial septum [3].

# 2. Patients and Methods

This is a retrospective single-center study of 20 patients (14 females and 6 males) with atrial myxomas who were admitted to and operated upon in the Iraqi Center for Heart Disease (ICHD), Baghdad, Iraq in a 6-year period ending at 31<sup>st</sup> December 2015. The study was ethically approved by the Council of Arabic Board for Medical Specializations/Scientific Council of Cardiac Surgery and the ICHD while patients' consents were not relevant due to the retrospective design of the study.

The medical records of all patients diagnosed as atrial myxoma and surgically treated in our center during the above period were reviewed. On admission, a thorough history was taken and physical examination was performed noting the presenting symptoms and eliciting the relevant physical signs. Thereafter, a diagnostic work up including general and specific investigations was carried out. Apart from routine hematological and biochemical tests, specific work up included a chest X-ray (CXR) in PA and lateral views, electrocardiography (ECG) and trans-thoracic echocardiography (TTE). TTE was the main diagnostic tool whereas coronary angiography wasn't performed unless the patient was above the age of 40 or had chest pain suspicious of ischemic nature.

Surgery was performed shortly after the diagnosis of atrial myxoma to avoid the potential life-threatening complications such as valvular obstruction or embolic phenomena. All patients were approached via median sternotomy with the aid of cardiopulmonary bypass (CPB) and mild hypothermia. Standard aortic and separate vena caval cannulation was used. Myocardial protection was achieved by cold crystalloid cardioplegia. Cardiac manipulation was avoided until the aorta was cross-clamped to avoid tumour fragmentation and systemic embolization. In myxomas of the right atrium (RA), the main pulmonary artery was cross-clamped as well to prevent possible embolization of tumour fragments into the pulmonary arteries.

Intra-cardiac access was achieved through left atrial, right atrial, bi-atrial and right atrial trans-septal approaches according to the tumour location. Methods of tumour resection included simple excision, excision and cauterization, excision and suture, excision and patch closure of the resultant atrial septal defect (ASD). Besides the involved cardiac chamber, other chambers were thoroughly explored for additional masses. After tumour removal, copious irrigation of the atria and ventricles with cold saline was performed to eliminate any loose tumor fragments that might have been dislodged during tumor removal.

After surgery, the patients were admitted to the intensive care unit (ICU) and usually extubated within a few hours and transferred to the ward after 48 hours. The patients were discharged home within several days provided no early complications developed. Routine follow up visits were scheduled to assess the patients clinically and by echocardiography. The retrieved data was analyzed and statistically tested.

## 3. Results

Of the twenty atrial myxoma patients enrolled in this study, 14 (70%) were females and 6 (30%) were males with a female to male ratio of 2.3 to 1. The age ranged from 14 to 80 years with a mean of 51.4. The highest incidence was in the age interval 41 - 60 years (n = 9, 45%) as shown in **Table 1**.

The demographic data is shown in **Table 2**.

Table	1. Age and sex distribution	•
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Age (years)	Males	Females	Total
11 - 20	1	0	1
21 - 30	0	0	0
31 - 40	1	3	4
41 - 50	1	3	4
51 - 60	2	3	5
61 - 70	1	4	5
71 - 80	0	1	1
Total	6	14	20

ID	Age & Sex	Bl. gr.	Site	ID	Age & Sex	Bl. Gr.	Site
1	70, F	A+	LA	11	60, M	B+	LA
2	70, F	A+	RA	12	54, M	O+	RA
3	32 F	A+	LA	13	63, M	B+	LA
4	71, F	B+	LA	14	48, F	A+	LA
5	58, F	B+	LA	15	50, F	A-	RA
6	52, F	A+	LA + RA	16	57, F	B-	LA
7	31, M	B+	RA	17	35, F	O+	LA
8	39, F	B+	LA	18	42, F	AB-	RA
9	14, M	A+	LA	19	48, M	O-	LA
10	70, F	B+	LA	20	65, F	A+	LA

 Table 2. The demographic dat.

It is evident that 14 (70%) myxomas were located in the left atrium, 5 (25%) in the right atrium and one (5%) in both atria. The 2 most frequent blood groups of the patients in this study were A+ (7, 35%) and B+ (7, 35%).

The mode of clinical presentation is shown in **Table 3**.

Most of the patients presented with symptoms related to the cardiovascular and respiratory systems such as SOB, palpitation, chest pain, orthopnea, easy fatigability, peripheral oedema, syncope, pleural effusion and limb ischemia. Constitutional symptoms such as weight loss, myalgia and fever were less frequent while neurological symptoms were infrequent. None of the patients had a family history of atrial myxoma.

The echo findings are shown in **Table 4**.

In regard to the site of origin of the atrial myxomas, TTE showed that 13 (65%) myxomas arose from the interatrial septum, 4 (20%) were attached to the lateral atrial wall and 3 (15%) were attached to the anterior leaflet of the mitral valve. Moreover, a prolapse of the tumour mass through an atrioventricular valve was shown in 9 (45%) cases.

The different cardiac approaches and methods of resection of atrial myxoma are shown in Table 5.

The commonest approach utilized in this series was the right atrial trans-septal approach (55%), followed by the right atrial (20%) and left atrial (20%) while the bi-atrial approach was used once (5%). Following tumour resection, no more action is needed in 50% of cases, tumour origin was cauterized in 20% of cases and atrial septal defect was sutured in 25% of cases. In one case (5%), a patch of pericardium was used to close the defect in the atrial septum following resection of a cuff of the interatrial septum with the atrial myxoma. On average, the tumour size was  $3 \times 5$  cm in 80% of cases and  $5 \times 7$  cm in the remainder 20% of cases. The smallest one was  $2.7 \times 3.5$  cm and the largest one was  $6 \times 7.5$  cm in size.

Furthermore, 3 (15%) patients had concomitant surgeries including mitral valve repair (1), mitral valve replacement (1) and coronary artery bypass grafting (CABG; LIMA to LAD) (1).

Symptom	No (%)
Dyspnea	14 (70)
Palpitation	10 (50)
Chest pain	7 (35)
Orthopnea	7 (35)
Fever	6 (30)
Easy fatigability	6 (30)
Myalgia	5 (25)
Weight lose	4 (20)
Peripheral edema	3 (15)
Syncope	3 (15)
Pleural effusion	3 (15)
Neur. Symptom	1 (5)
Limb ischemia	1 (5)

 Table 3. The clinical presentations.

#### Table 4. The echocardiographic findings.

Findings	No (%)	Findings	No (%)
Valvular	3 (15)	L. atrial dilatation	5 (25)
Tumor prolapse through A-V valve	9 (45)	Mitral insufficiency	4 (20)
Pedunculated L.A clot	2 (10)	Mitral stenosis	3 (15)
Tumor attached to ant. M.V leaflet	3 (15)	Pulmonary artery hypertension	4 (20)
Bi-atrial tumor	1 (5)	Pericardial effusion	1 (5)
Attached to the septum	13 (65)	Tricuspid Insufficiency	4 (20)
Attached to the lateral wall	4 (20)	Tricuspid Stenosis	1 (5)

Table 5. Types of cardiac incisions and methods of resection of atrial myxomas.

Cardiac Incision	%	Methods of Resection	%
Right atrial trans-septal approach	55	Excision	50
Right atrial approach	20	Excision with cauterization	20
Left atrial approach	20	Excision with direct suture of ASD	25
Bi-atrial approach	5	Excision with patch closure of ASD	5
Total	100		100

The postoperative course was uneventful in most patients. A few patients had some complications as shown in **Table 6**.

One patient (5%) died due to acute renal failure despite hemodialysis and there was no tumour recurrence during the follow up period.

Complication	Number	Management
Bleeding	1	Re-exploration
Arrhythmia	3	Medication
Pericardial Effusion	1	Aspiration
Wound Infection	1	Rewiring
Low cardia output	2	Inotropes and prolonged intubation
Renal failure and Death	1	dialysis

Table 6. The postoperative complications.

# 4. Discussion

This is the study of 20 patients of cardiac myxoma, operated upon in Iraqi Center for Heart Disease between January 2010 and December 2015.

Myxoma represents most common primary cardiac tumors. In the 3220 operations done in Iraqi center for Heart Disease, cardiac myxoma incidence was 0.6% of all surgical cardiac patients, the incidence in our study compared with similar studies which was 0.5% in Ibn Al Betar hospital Hameed, A. *et al.* [12], 0.81% done Wu Xingli *et al.* [13] in China.

Although they have been reported in both sexes, they most often occur in women [3], in the present study male to female ratio was 1:2.3 compared with similar study which was male to female ratio 1:3.07 Włodzimierz Kuroczyński *et al.* [14], in contrast to other study done in Pakstan male to female ratio was 1.2:1 Muhammad Aamir K *et al.* [15]. Cardiac myxoma can occur in any age group, but its rare in infancy and elderly, the peak incidence is between third and sixth decade of life [2]. in our study mean age group is 51.4, is more common between fourth and sixth decade of life, with agreement to national and international study done by Włodzimierz Kuroczyński *et al.* in Germany [14] and Maryam Sotoudeh A *et al.* Iran [16], which mean age group 50, 57.9 and 54 respectively. While in the other international study done for example in India cardiac myxoma most common in young age group, mean age group 36.05 Amit Mishra *et al.* [17].

Myxomas most commonly occur in the atria. Approximately 75% arise in left atrium, 15% - 20% arise from right atrium, the reminder of myxomas are located in ventricle [3], in our study 75% were from left atrium, 20% from right atrium and 5% bi-atrial, we found similar result in national (2003) and international studies done in India by Amit Mishra *et al.*, G Samanidis *et al.* in Greece and Maciej Rachwalik *et al.* in Poland, [17] [18] [19].

In the present study we found cardiac myxoma more common in blood group A+ and B+ followed by blood group O+, compared to national study cardiac myxoma more common in blood group A+ followed by blood group O+, Siddiq, A. *et al.* [20].

The classical clinical presentation of a myxoma is intra-cardiac obstruction with congestive heart failure, sings of embolization, systemic or constitutional symptoms of fever, weight loss and fatigue, and immunologic manifestations of myalgia, weakness and arthralgia [2]. In our study main chief complain was dyspnea followed by palpitation, chest pain similar to international studies done by G Samanidis *et al.* in Greece, Maryam S A *et al.* in Iran, Reyaz Ahmed Lone *et al.* in India and Maciej Rachwalik *et al.* in Poland [16] [18] [19] [21], while in other study like Stavros Siminelakis *et al.* in Greece [22], emboli was main chief complain.

The majority of masses in our study arise from interatrial septum, and some of them showed itself as a mass prolapsing through atrioventricular valve with agreement to national Siddiq, A. *et al.* and international study done by Reyaz Ahmed Lone *et al.* in India,Maryam S A *et al.* in Iran and Maciej Rachwalik *et al.* in Poland [16] [19] [21].

Regarding the size of the masses in our study most of them about  $3 \times 5$  cm in 80% of cases with agreement to study done by Hamdy D. Elayouty *et al.* in Egypt [23].

There still argument regarding the most appropriate surgical approach to a chief complete excision of intra cardiac myxoma. The left atrium approach gives a direct and fast access to the myxoma although not to its attachment.

Bi-atrial approach not only gives good exposure but also facilitates the easy extraction of the tumor with minimum handling and also permits good examination of all the cardiac chambers but the technique is more radical.

The trans-septal approach through right atriotomy suggested by Chitwood is more practical. It gives good access to the myxoma with minimal handling and allows inspection of all cardiac chambers. The RA and RV myxomas are approached through right atrium. RA myxomas demand more care during cannulation.

In our study most LA myxoma were successfully excised by RA trans-septal approach. In four cases LA myxoma was approached through LA and in other four patients, RA myxomas were approached through the right atrium, bi-atrial technique used just in one case, with agreement to other studies done by Włodzimierz Kuroczyński *et al.* in Germany, Amit Mishra *et al.* in India and Reyaz Ahmed Lone *et al.* in Kishmer [14] [17] [21]. But in comparing with other study LA myxoma approached through the left atrium as in national, Siddiq, A. *et al.* [20] and international study Chekir Selkane *et al.* [24].

Simple excision was performed in 50% of patients followed by excision with cauterization 20%, excision and suture 15%. With agreement to similar national study by Siddiq, A. *et al.* [20] and international study, Chekir Selkane *et al.* and Alvaro D.B *et al.* in Portugal [24] [25]. Three of our patients required further cardiac surgery, mitral valve replacement, one mitral valve repair and one coronary artery bypass graft, similar to studies done by Siddiq, A. *et al.* and Alvaro D.B *et al.* [20] [24]. In the present study postoperative complication was within acceptable range similar to national study by Hameed, A. *et al.* [12], arrhythmia was the main postoperative complication 15%, bleeding 5%, pericardial effusion 5%, wound infection 5%, renal failure 5%, death 5% and there is no recurrence

reported in our study, in comparative with similar study, present study has recorded less postoperative complication than them, Reyaz Ahmed Lone *et al.*, Alvaro D.B *et al.*, Włodzimierz Kuroczyński *et al.* [14] [21] [25].

# **5.** Conclusion

Cardiac myxoma is the most common primary cardiac tumor in all age groups, most commonly find in left atrium but it may occur in other cardiac chambers. We should consider cardiac myxoma in the differential diagnosis in cases of fever of unknown origin. Cardiac myxoma is considered as an emergency in cardiac surgery, because of the risk of life threaten conditions like embolic events or obstruction of the mitral orifice. Echocardiography is a main diagnostic tool. Surgical excision of cardiac myxoma carries a low operative risk and gives excellent results.

The recurrence rate is rare when occurring sporadically while is high when it occurs in familial or as a part of syndrome such as carney complex.

# Disclosure

This article is inspired by a thesis entitled (Six-years clinical experience and surgical considerations in management of cardiac myxoma in Iraqi Center for Heart Disease-Single center experience) prepared by Dr. Hozhan Hussein Blbas (MBChB) under the supervision of Assistant Professor Dr. Abdulameer Mohsin Hussein (FICMS, FACS) and submitted to the Arabic Council for Medical Specializations as a partial fulfillment of the degree of fellowship of Arabic Council for Medical Specializations in Cardiothoracic Surgery in 2018.

# **Conflicts of Interest**

The authors declare no conflicts of interest regarding the publication of this paper.

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