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Cardiac Myxomas: A Single-Center Case Series of 15 Patients Over A 10-Year Period Study

Briki, J[1*], Benlefqih, C[1], Idrissa, M[1], Saadouni, Y[1], Eledrissi, A[1], Bouhdadi, H[1], Amelal, M[1], Rhissassi, J[1], Sayah, R[1], Laaroussi, M[1]

[1]Cardiovascular Surgery Department A, Ibn Sina University Hospital, Rabat, Morocco

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***Corresponding author:** Briki, J
Cardiovascular Surgery Department A, Ibn Sina University Hospital, Rabat, Morocco

Abstract **Original Research Article**

Background : Myxomas are the most common primary cardiac tumors, most frequently located in the left atrium, where surgical treatment is the gold standard for good outcomes. ***Methods and Results :*** This is a retrospective study aiming to analyze clinical and intraoperative caracterestics from cardiac myxoma cases in the cardiac surgery departement of the last 10 years. The study population was 15 cardiac surgical patients. 87,6% of cases had the myxoma located at left atrium and, 86,6% of all patients fully recovered. ***Conclusion :*** The availability of transthoracic echocardiography and improvements in other cardiac Imaging techniques have made it possible to diagnose cardiac myxomas rapidly, allowing them to be managed surgically without delay and, as result, improving their outcome and preventing complications, which can be fatal.

Keywords : Cardiac myxoma**,** Transthoracic echocardiography, left atrium, surgery.

INTRODUCTION

The prevalence of cardiac tumors at autopsy varies from 0.001% to 0.3%. Over 72% of primary cardiac tumors in adults are benign, and over 50% of benign cardiac tumors are myxomas. In 7% of cases, the tumor is of genetic origin, corresponding to an inherited disorder with certain clinical manifestations [1, 2].

The myxomatous cell is thought to derive from residual pluripotent or multipotent mesenchymal stem cells [1, 3, 4]. Embryonic remnants of the latter persist during septation and differentiate into endothelial cells, smooth muscle cells and other mesenchymal smooth cells. This explains the prevalence of myxomas in the interatrial septum [1], as well as the wide morphological variation.

Histologically, these tumors are composed of scattered cells within a mucopolysaccharide stroma [2, 3]. Macroscopically, typical myxomas are pedunculated and gelatinous in consistency; the surface may be smooth, villous or friable [4].

Tumors vary widely in size, ranging from 1 to 15 cm in diameter and weighing between 15 and 180 g. Around 35% of myxomas are friable or villous, and tend to have emboli. Larger tumors are more likely to have a smooth surface and to be associated with cardiovascular symptoms [4].

The clinical expression of cardiac myxomas is highly variable and non-specific, related to tumor size, location and mobility [1, 5, 6].

In some cases, the tumour may be revealed by embolic complications or signs of heart failure. Patients may be asymptomatic, particularly those with small tumors [1, 7].

Transthoracic echocardiography (TTE) provides an excellent morphological study of the myxoma and its impact on the heart [8, 9]. It remains the reference examination, given its accessibility and low cost.

PATIENTS AND METHODS

The aim of this study is to review our experience with cardiac myxomas over a 10-year period by analyzing and comparing the demographic and intraoperative data of patients who underwent cardiac surgery for myxoma excision at a single-center, in order to document the variation of these data through the years.

This is a retrospective, descriptive study of 15 cases of cardiac myxoma, operated on in the Cardiovascular Surgery Department A of the Ibn Sina University Hospital in Rabat, over a period from January 2013 to January 2023, representing a frequency of 0,6 % of all patients operated on.

Patients whose data were incomplete were excluded.

Pre- and postoperative data were recorded from medical records and operative and extracorporeal circulation reports.

A longitudinal median sternotomy was performed in all patients, with extracorporeal circulation via cannulation of the aorta and both superior and inferior vena cava. Cardioplegia with cold blood solution was administered via the anterograde route, at the level of the root of the initial aorta.

After the operation, patients were transferred to the cardiovascular intensive care unit and then to conventional hospitalization.

Follow-up TTE was performed regularly by cardiologists in the CCVA department.

RESULTS

Women represented 66.6% of the cases studied, the mean age of patients was 57.53 years, with extremes ranging from 11 to 82 years.

Concerning the circumstances of discovery, 53% of patients had presented with progressively worsening dyspnea and 20% with ischemic stroke. In only 2 cases (13%) was the myxoma discovered incidentally during a standard workup. Altered general condition was noted in two patients (13%). Other clinically relevant characteristics are summarized in (Table 1).

Sinus tachycardia on the electrocardiogram was noted in 53% of cases, 40% were in cardiac arrhythmia due to atrial fibrillation. Standard radiographe showed isolated cardiomegaly in 26% of patients, cardiomegaly associated with bilateral hilar overload in 20%, and unremarkable radiography in 34%.

The left atrium was the most frequent location in 73% (Figure 1). of cases, and the right atrium in 27%. There were 3 cases of double localization at the level of the inter-atrial septum and the mitral valve (Table 2). Myxoma size ranged from 13/9 mm to 70/54 mm.

Nine patients had concomitant valvular disease, including 1 case of isolated mitral insufficiency, 1 case of mitral valve disease and associated tricuspid insufficiency, and 7 cases of isolated tricuspid insufficiency. Hypertension was present in 58.3% of patients on Trans thoracic echocardiography.

All patients underwent surgery under extracorporeal circulation. Cardiotomy was performed via the biatrial transeptal approach in all cases studied. Total surgical excision of the tumor (Figures 2 & 3), and its base of implantation was performed, combined with mitral plasty in 1 case, mitral and tricuspid plasty in 1 patient, tricuspid valve plasty in 7 patients, and pericardial patch reconstruction of the septal defect in two patients.

The average length of stay in the intensive care unit was 2.4 days, and the average hospital stay was 10 days. The immediate post-operative course was straightforward, except for one patient who was operated on for a mitral leak deemed severe, and underwent mitral annuloplasty. Two cases of death were recorded following hemodynamic instability with bi-ventricular dysfunction in the first case and septic shock in the second, giving a post-operative morality rate of 13%.

Postoperative L-ETT was normal in all the other cases studied.

Table 1: Epidimiogical and clinical caracterisctics

Masculin\ Feminin	**10\5**
Age (mean value)	57,35
Symptoms (n)	
incidental	2
Dyspnea	8
Systemic embolic events	3
Palpitation	2
Chest pain	3
	2
NHYA classification	
Class I	5
Class II	1
Class III	2
Class IV	0

Table 2: Insertion site of cardiac myxomas

Insertion site	**n of patients**
Left auricle	11
Right auricle	4
Atrial septum	2
Mitral valve	1

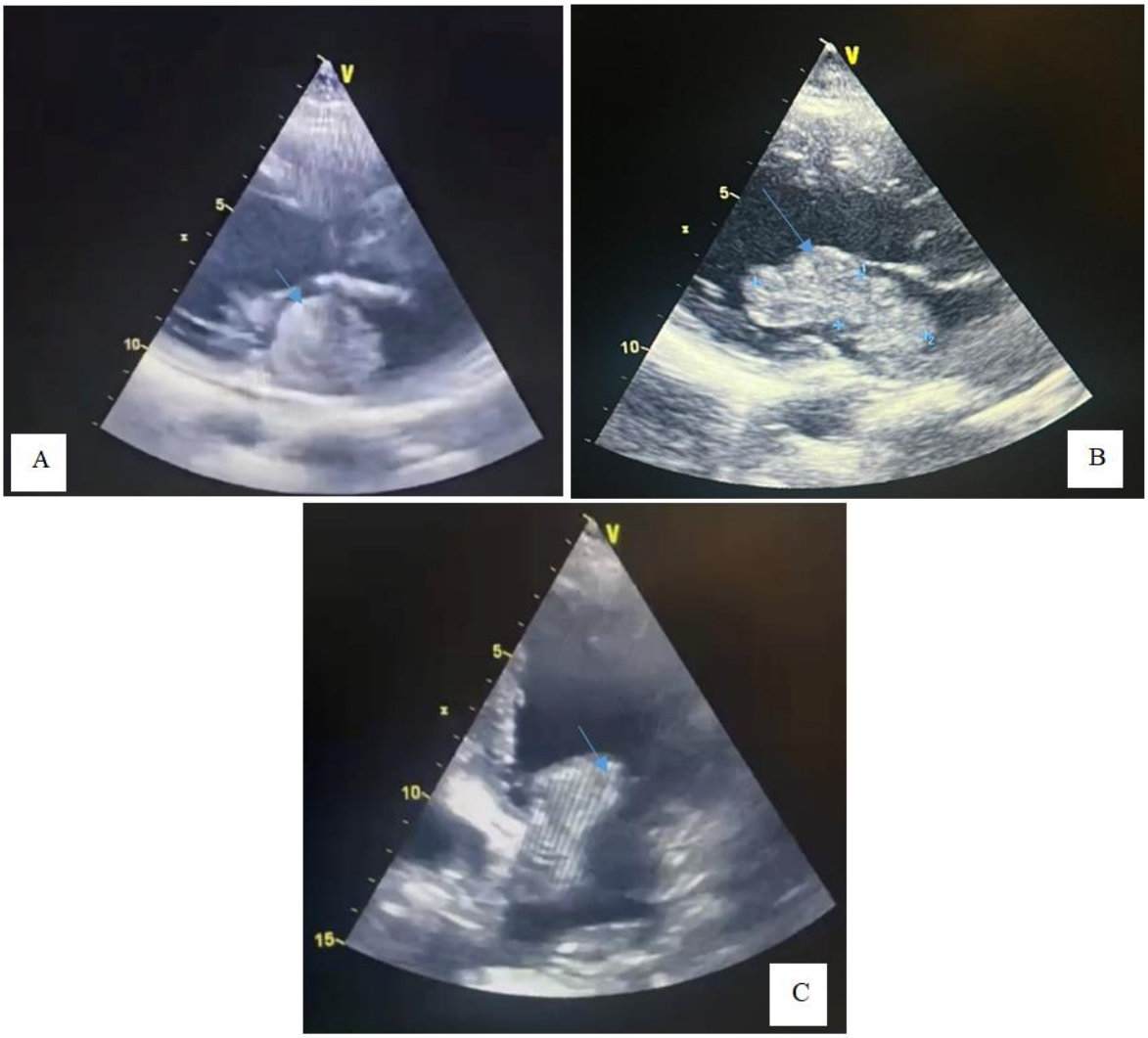

Figure 1: Imaging studies from patients with left atrial myxoma. (A, B): Parasternal long-axis echocardiogram reveals the myxoma protruding into the left ventricle during diastole; (C): 4-cavity echocardiogram section showing a cardiac myxoma in the left atrium, prolapsed through the mitral valve

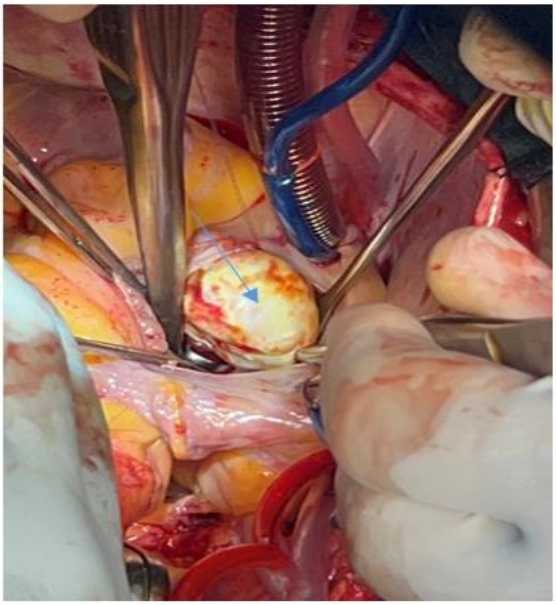

Figure 2: Operative view showing the mass in the left atrium

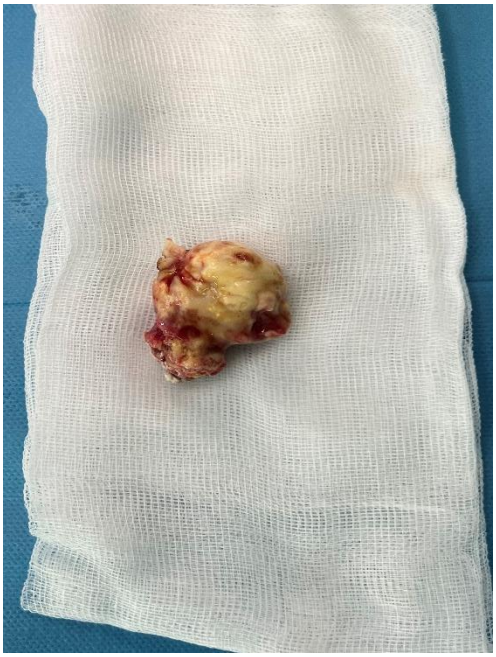

Figure 3: Operative view showing the entire mass excised

DISCUSSION

Primary tumors of the heart are rare, with an approximate prevalence of 0.02% in autopsy series [10]. In our study we found a higher rate (0,6%), which can probably be explained by the interruption of planned surgical activity in our department during the covid19 period and for which the surgical management only concerned emergencies, in particular cardiac myxomas. 75% of cardiac tumors are benign, with myxoma accounting for 50% and rhabdomyoma 20% of lesions [11].

The etiopathogenesis of cardiac myxomas is not clearly elucidated; they may arise from intracardiac embryonic mesenchymal remnants capable of differentiation [12]. However, some authors suggest that intracardiac sensory nervous tissue may be the origin of myxomas [13, 14].

Predominantly female [9, 15, 16], as was also found in our study with a rate of 66,6%, myxomas are most often diagnosed between the ages of 30 and 60 [17].

The clinical manifestations of cardiac myxoma are highly variable. Intracardiac obstruction is associated with large or mobile myxomas that interfere with ventricular filling, closure of the atrioventricular valve or cause valvular lesions [18]. Symptoms include dyspnea, cough, recurrent pulmonary eodema or features of right heart failure, or features of congestive heart failure [19, 4]. Complete and sudden obstruction by a tumor prolapsing into the valvular orifice can lead to syncope or even sudden death [19, 20].

Direct invasion of the myocardium may impair myocardial contractility, causing arrhythmias, or pericardial effusion with or without tamponade [4]. Invasion of the adjacent lung may be responsible for pulmonary symptoms. Finally, tumors of the left atrium can cause embolic events, usually systemic, through the release of tumor fragments or thrombi, which can reveal the pathology in 30% of cases; in our study, ischemic events were revealing in 20% of cases.

The growth rate of myxomas is not clearly defined in the literature, as they are mainly treated surgically, and are only very rarely managed medically due to contraindications to surgery [21].

The left atrium is the most frequent location in around 75% of cases, and the right atrium in 20%, and only in 5% of cases are myxomas found in the ventricles [22], in our analysis the left atrium was the most frequent location in 73% of cases, followed by the right atrium, with no other location outside these two cavities.

The aim of the initial evaluation is to confirm the presence or absence of a cardiac tumor, and to determine whether it is malignant or benign, as far as possible. TTE is a simple, non-invasive technique, and remains the gold standard given its accessibility and low cost. With a sensitivity of 95%, it can generally confirm the presence or absence of a mass.

In addition, echocardiography can provide information on any other associated lesions, including valve obstruction by protrusion of the cardiac mass, identify the likelihood that this mass may be the origin of an embolic event, and also diagnose associated valvulopathy [23].

A classification of large myxomas has been proposed by Charuzi, Y et al according to their size and relationship to the mitral valve:

- Class I - small and prolapsed through the mitral valve.
- Class II - small and not prolapsing.
- Class III-large and prolapsed.
- Class IV-large and not prolapsing [24].

Trans esophageal echocardiography is useful in cases of small myxomas, with 100% sensitivity, or to assess the presence of thrombus in contact with the myxoma [25].

Differential diagnosis is mainly with intracardiac thrombus; CT, MRI and trans esophageal echocardiography are relevant investigations for diagnosis and differentiation with intracardiac thrombi.

However, the definitive diagnosis of cardiac myxoma is established by anatomopathological examination of the excised specimen following curative surgical treatment.

Treatment:

The diagnosis of a cardiac myxoma confirmed by imaging, whatever its location, represents an absolute indication for surgical management, which must be performed without delay once the diagnosis is accepted [26-28], Surgical resection is the gold standard in the treatment of cardiac myxomas. This indication was respected in all the cases in our study who underwent emergency surgery.

This aggressive approach is justified by the constant threat of systemic or pulmonary embolization and cardiovascular complications, including sudden death.

The results of surgical resection are generally very good, with most series reporting an operative mortality rate of less than 5% [6]. In our study, the mortality rate was 13% (2 cases). This high rate was essentially due to a postoperative respiratory infection caused by a resistant germ, complicated by septic shock.

The chosen approach should allow better exposure for the surgeon and also minimize manipulation of the myxoma to reduce the risk of embolism.

The choice of the ideal approach is the subject of controversy, with some authors, such as DiSesa *et al*., [28], preferring a posterior incision in the interatrial groove. Bortolotti et al find that the transeptal approach after right atriotomy is extremely practical and effective. After complete myocardial relaxation, achieved by cardioplegic arrest, the wide incision in the fossa ovalis provides access to all cardiac cavities, enabling inspection. This approach guarantees complete and rapid excision of the tumour, and exclusion of any coexisting masses. This is the recommended approach for all intracardiac tumours [29].

In our series, the approach was exclusively tanspetal, which we feel is the preferred route for best exposure and careful exploration of all cavities.

The aim of surgical treatment is not only complete resection of the entire mass, but also its attachment base. This is achieved by wide excision of the inter-atrial septum with patch reconstruction when the tumour is attached, as in most cases, to the oval fossa. Or by excision of the surrounding endocardium and part of the underlying myocardium when the tumour is attached elsewhere [30]. All our patients benefited from wide resection of the myxoma with its base of implantation, which in two patients required reconstruction of the septal defect with an autologous pericardial patch.

Postoperative recovery is generally rapid. However, atrial arrhythmias or atrioventricular conduction abnormalities have been observed postoperatively by Bateman et al, the origin of which is probably related to surgical lesions of the conduction voices intraoperatively [31].

As with any neoplastic pathology, tumor recurrence is considered a potential concern in cardiac myxoma, with a recurrence rate of 5.6% [13].

Although there is no concrete theory regarding the cause of recurrence, numerous hypotheses have been proposed, including multifocality, intraoperative implantation or embolization, DNA ploidy, inadequate resection and malignant transformation [32, 33, 34].

Other risk factors for tumor recurrence have been proposed by Iskhan, K et al in their retrospective analysis of 194 cases of cardiac myxoma, including young age, small tumor size, and ventricular location. Hence the recommendation of regular annual ultrasound surveillance for the first 10 years postoperatively [35].

In familial forms, recurrence is more frequent, occurring in 10-21% of cases [36]. These forms are also characterized by their atypical localization [37] and may recur several times in the same patient [35], even reaching five recurrences [38, 20]. In our study, no case of recurrence was noted.

CONCLUSION

Early diagnostic and therapeutic management of cardiac myxomas leads to the best results, but in view of a non-negligible risk of recurrence, follow-up by TTE is recommended.

Conflicts of Interest: There are no conflicts of interest.

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