Original Article

Cardiac Tumours : A Clinico-Pathological Analysis of 112 Cases in a Tertiary Heart Care Centre

S. Sathyavathi[1,*], B. N. Krishnamurthy[1], Rashmi B. K[1]

[1]Department of Pathology, Sri Jayadeva Institute of Cardiovascular Sciences & Research, Bengaluru, India

***Correspondence:** sathyavathikiran@gmail.com

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Abstract

Objective: Cardiac tumours are rare amongst which 80 % are myxomas. Cardiac myxomas are the most common benign cardiac tumors having endocardial origin. The study was conducted among 112 patients clinically diagnosed as cardiac myxomas in a tertiary care centre among the south Indian population to analyse the clinical, radiological and pathological findings which aid in better diagnosis.

Methods: Clinical presentation, Echocardiographic and radiological findings with histopathological analysis of 112 cardiac tumour cases were done and reviewed retrospectively.

Results: Among 112 patients, 42 were males and 70 females presenting with an average age at diagnosis ranging from 58 to 65 years. Breathlessness was the most common presenting feature. Cardiac myxoma was the most common tumour found among the patients presented with cardiac mass. Myxomas were commonly located in the left atrium followed second by the right atrium with 1 being biatrial in location. 6 cases were recurrent in this study.

Conclusion: Cardiac myxomas are the most common benign primary tumours of the heart and occasionally may have a varied presentation with serious embolic complications. So early recognition and surgical resection with histopathological confirmation of the resected specimen is suggested.

Keywords: cardiac tumours; cardiac myxomas; left atrium; stellate cells

Introduction

Cardiac tumours are rare having a worldwide incidence of 0.0017 to 0.30 % [1]. 75% of the cardiac tumours are benign in nature with cardiac myxoma accounting for nearly 80%. Cardiac myxoma is an uncommon benign mesenchymal tumour of the heart usually arising in the left atrium supposedly near the fossa ovalis [1]. It mostly affects the adults between the third and sixth decades of life. Cardiac myxomas usually develop in the atrium with 75 to 80% arising from the left atrium and 20% from the right atrium [2] and 1-3% are biatrial [3]. Due to its varying location, the tumour size and its histogenesis, they have a varied clinical presentation like breathlessness, emboli, stroke and other constitutional symptoms [2] like fatigue, fever, myalgia, rashes and is associated with abnormal laboratory investigations like decreased hemoglobin, increased erythrocyte sedimentation rate (ESR) and raised C reactive protein (CRP) levels [2].

About 90% of cardiac myxomas are sporadic in nature with 10% being familial having autosomal dominant transmission (Carney syndrome). Carney syndrome is seen in younger patients with greater risk of recurrence and is characterized by multiple cardiac and extracardiac (skin) myxomas with spotty skin pigmentation, endocrine overactivity, schwannomas and epithelioid blue nevus [1].

Cardiac myxomas are multipotent in origin arising from mesenchymal cells or from the cardiac myocyte progenitor cells or subendothelial cells [1]. It is speculated to arise from subendothelial vasoformative reserve cells or primitive cells that later differentiate along the lines of the endothelium. Macroscopically they are of two types, solid pedunculated and papillary [1, 2] with papillary cardiac tumours presenting with embolic presentation and solid tumours presenting with cardiac failure symptoms [1] like breathlessness. Microscopically the myxomas are identified by the presence of Stellate cells arranged singly or in small clusters floating in a myxoid gelatinous stroma [1, 2]. As the cell of origin is still a mystery, Immunohistochemistry (IHC) using S-100, NSE, Vimentin and Calretinin positivity is considered to confirm the diagnosis [4].

We present a retrospective study conducted among south Indian population in a tertiary cardiac care centre, Bengaluru to analyse the clinical, radiological and pathological features of cardiac myxomas.

Material and Methods

This study is a retrospective analysis of 112 surgically resected clinically diagnosed cardiac tumour specimens at Sri Jayadeva institute of cardiovascular sciences and research, Bengaluru, South India between 2009 to 2021. Of the 112 cases, 107 were cardiac myxomas, 2 thrombus, 1 case was of mitral valve having myxomatous degeneration, 1 Lipoma and 1 was a metastatic tumour, primary being Renal cell carcinoma. The age at presentation had a wide range, the youngest being 12 years of age and the oldest 78 years with an average between 58 to 65 years. Female preponderance was noted in our study.

After initial clinical diagnosis and imaging confirmation, all 112 cases underwent surgical resection. The specimens were fixed in 10% Neutral buffered formalin, grossing was done and the tissue bits after tissue processing were embedded in paraffin wax. The sections taken were stained with routine Hematoxylin & Eosin (H&E) staining.

Special stains like Periodic Acid Schiff (PAS) and Alcian blue was done to identify the stellate cells and the myxoid stroma. IHC staining like Vimentin, Calretinin, S-100 and Non specific enolase [NSE] was done in few cases to confirm the histopathological diagnosis.

Results

The cases were collected, studied and analysed retrospectively between the year 2009 and 2021. Out of the 112 cardiac tumour cases suspected clinically as myxomas, 107 cases correlated with clinical diagnosis of Myxoma on histopathology examination, 2 cases were thrombus, 1 was Mitral valve with myxomatous degeneration, 1 Lipoma and another was a Metastatic tumour. The male: female ratio was 1:1.6 with female being the most commonly affected. The cases ranged between 12 to 78 years of age with a mean of 50.8 years and nearly half the cases being in the fifth and sixth decade.

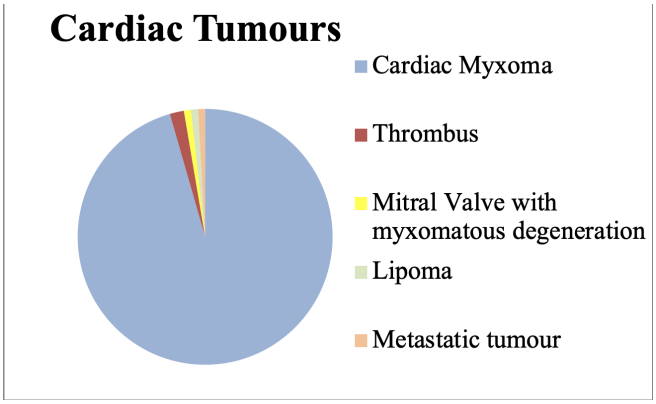

Figure 1: Types of cardiac tumours according to histopathology.

Among the 112 cardiac tumour cases, 88 % of the cases were arising from the left atrium, 12% from right atrium and 1 case was bilateral [0.8%]. 6 cases [5.8 %] had history of recurrence, 2 being familial. Breathlessness [60%] was the commonest presenting feature of all cardiac tumours followed by Embolism [10%] which was mostly seen in tumours arising from the right atrium. On physical examination 79 cases [72.5%] presented with a systolic murmur on auscultation and 7 cases with tumour plop [6.5 %].

Electrocardiography (ECG) was normal in 70 % of the cases with very few having atrial fibrillation, tachycardia and right ventricular hypertrophy. 2D Echocardiography (ECHO) was performed in all the 112 cases to determine the tumour location, size and shape. Few cases who underwent Computerized Tomography (CT Scan) also had similar diagnosis confirming the Echocardiographic (ECHO) findings.

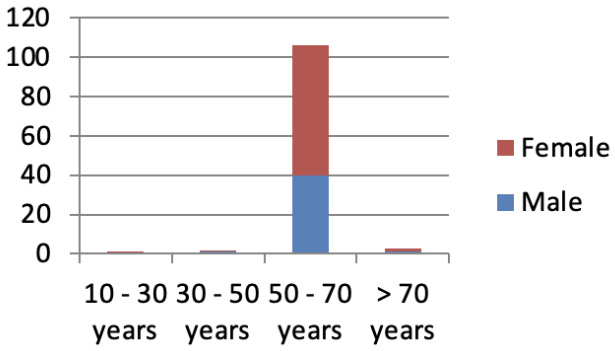

Figure 2: Age and sex distribution.

Table 1: Distribution of cardiac tumours according to location of origin.

Location	No. of Cases	%
Left Atrium	98	88 %
Right Atrium	13	12 %
Bi - atrial	1	< 1%
Total	112	100%

All the patients were subjected to surgical resection of the primary cardiac tumours.

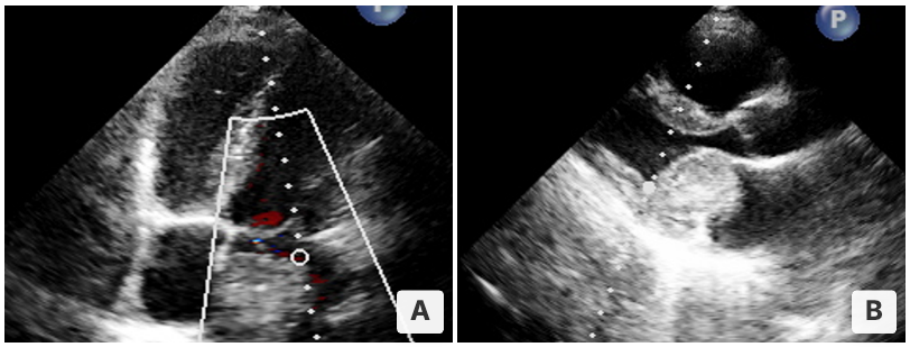

Figure 3: A- Echo showing myxoma measuring 3 x 3 cms situated in left atrium, B- echo showing left atrial myxoma abutting the mitral valve.

Histopathology

Gross

The resected tumour specimens received in our laboratory ranged in size from 2 to 8 cms having no difference depending on its location of occurrence. 68 cases [60.1%] out of 112 cases were pedunculated and on cut section was solid with gelatinous appearance, rest 41 cases [36.6%] were of the papillary type having variable appearance and on cut section showed glistening gelatinous surface with focal areas of haemorrhage with or without calcification.

Microscopy

The cardiac tumour was confirmed microscopically as myxoma by the presence of stellate cells floating in myxoid stroma. The stellate cells were arranged singly (85%) in most tumours, few in cord like pattern (10%) and occassional tumour had glandular arrangement (5%). Focal collection of thin walled blood vessels, haemorrhage, hemosiderin laden macrophages and necrosis was noted in few myxomas. Inflammatory cells like lymphocytes were seen very frequently in the stroma, calcification was noted in few (10 cases, 8.9%) and 2 myxomas showed gamna gandy bodies.

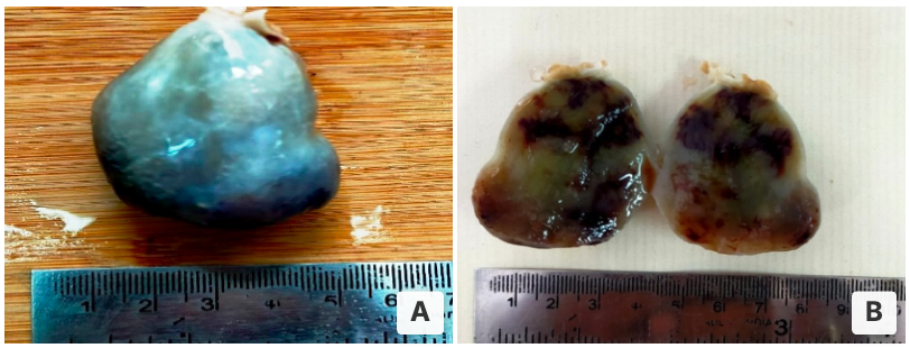

Figure 4: A- Gross specimen of cardiac myxoma measuring 3 x 3 cms. B- Cut section of myxoma showing large areas of hemorrhage.

Recurrence

6 cases of the total 112 cases [5%] had history of recurrence and mostly seen in patients of younger age group.

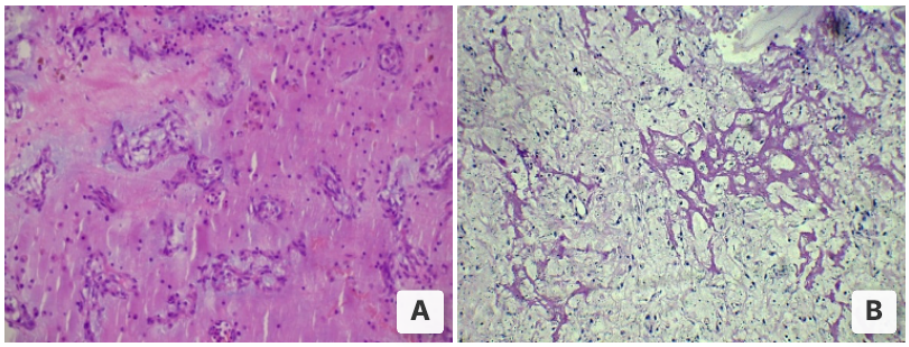

Figure 5: A- Microscopic findings show stellate cells arranged around vascular channels in pools of mucin (H&E stain 10 X). B- Microscopic findings showing pools of PAS positive mucin between the tumor nests (PAS stain 10 X).

The neoplastic cells are arranged individually, and in nests, and are oriented in single or multiple layers around vascular channels. The neoplastic cells are immunoreactive for Vimentin, Calretinin, S100, Nonspecific Enolase, factor VIII, CD31 and CD34.

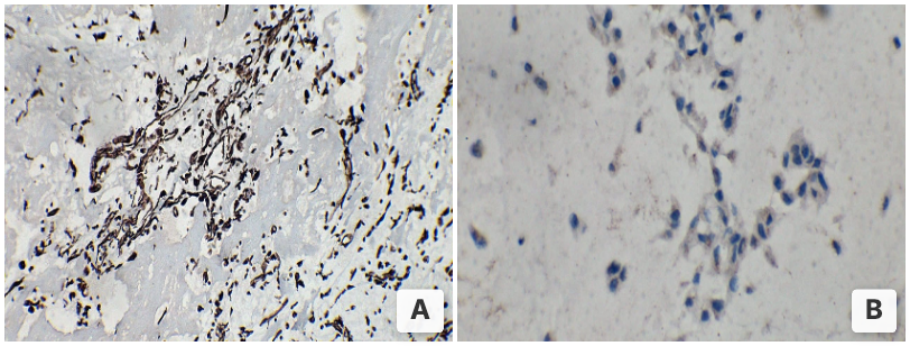

Figure 6: IHC staining of cardiac myxoma. A: Cardiac myxoma showing strong cytoplasmic positivity for Vimentin (10 X) [V9 clone from Diagnostic Biosystem company with ready to use dilution]. B: Cardiac myxoma showing mild cytoplasmic positivity for Calretinin (10 X) [H5 clone from Diagnostic Biosystem company with ready to use dilution].

Discussion

The most common primary cardiac tumours occurring are benign in nature and nearly 80% of the excised tumours are cardiac myxomas [5]. The present study is a retrospective analysis of 112 cardiac myxoma cases among the South Indian population in a tertiary care cardiac institute.

The tumour usually occurs in adult population ranging from 12 to 80 years of age [3, 6, 7]. The youngest in our study was a 12 year old and the oldest 78 years of age. Females were most commonly affected which was similar to the other studies compared [1, 3, 5]. 90% of cardiac myxomas occur in the atrium with left atrium accounting to nearly 78.57%

[96/112cases] and 13 cases [11.6%] in right atrium [1], one case was biatrial myxoma and none of cases studied in our study had ventricular location [7]. Multilocular myxomas are rare and in our study we reported a single case with biatrial myxoma [8]. Recurrence is rare and are usually associated with familial myxomas and seen in younger individuals [1]. In our study we reported 6 cases having recurrence [1]. Even though the prognosis is excellent with surgical resection of the tumour but periodic regular follow up is advised to identify and treat the recurrent tumours early [9].

Clinically cardiac myxomas have a varied diverse presentation and is related to the tumour size and location [1, 3, 6, 7]. The most common clinical feature is breathlessness [60 %] which could be attributed to the atrioventricular valvular obstruction by the tumour [1, 10]. Morphologically myxomas are friable tumours with gelatinous appearance having papillary surface causing the soft tissue to fall off and result in embolism [3, 6]. The commonest sign elicited is a systolic murmur on auscultation [3].

Electrocardiography (ECG) showed non-specific ST-T changes with very few cases showing arrhythmias like Atrial fibrillation. Echocardiography (ECHO) is the most widely used imaging method for identifying cardiac tumours and many patients are incidentally detected during routine echocardiography. Through ECHO the size and localization of the tumor, as well as its shape and mobility are determined [10]. ECHO was the most sensitive means of diagnosis [3, 6, 11] followed by Transesophageal echocardiography to confirm the location and size of the tumour both pre-op and post op. CT and Magnetic resonance imaging helps in confirmation of the tumour diagnosis and in differentiating myxomas from other intra cavitary masses [8, 12].

Conclusion

Cardiac myxomas, the most common primary cardiac tumours are asymptomatic many a times and are incidentally detected on routine cardiac evaluation. Clinical suspicion of myxomas is essential as it can lead to serious complication like embolism and sudden cardiac death due to the obstruction to the cardiac outflow. Early detection and timely intervention by surgical resection of the myxomas with histopathological confirmation is essential for the benefit of the patient.

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Ethical Committee Clearance: Taken

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Abbreviations: CRP: C Reactive Protein
CT: Computerized Tomography
ECG: Electrocardiography
ECHO: Echocardiography
ESR: Erythrocyte Sedimentation Rate
H&E: Hematoxylin & Eosin
IHC: Immunohistochemistry
NSE: Non Specific Enolase
PAS: Periodic Acid Schiff

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