Journal of Cardiovascular Disease Research

ISSN:0975 -3583,0976-2833 VOL14, ISSUE 12, 2023

Case Series

Atrial Myxoma - Clinicopathologic Profiles and Review of Literature - A Tertiary Care Centre Study in Kerala – (Case Series)

[1]Dr. Aswathi R., [2]Dr. Austin Raj R. S., [3]Dr. Vanesa John T.

[1]Assistant Professor, Department of Pathology, Jubilee Mission Medical College and Research Institute, Thrissur, Kerala, India.

[2]Assistant Professor, Department of Cardiovascular & Thoracic Surgery, Jubilee Mission Medical College and Research Institute, Thrissur, Kerala, India.

[3]Assistant Professor, Department of Pathology, Jubilee Mission Medical College and Research Institute, Thrissur, Kerala, India.

Corresponding Author

Dr. Aswathi R., Assistant Professor, Department of Pathology, Jubilee Mission Medical College and Research Institute, Thrissur, Kerala, India.

ABSTRACT

Introduction

Atrial myxoma is the most common benign primary cardiac neoplasm.Myxomas can arise in any of the cardiac chambers although 75% occur in left atrium.Clinical presentation mimics multiple cardiac diseases.

Material and Methods

During the 2017–2023 study period, eleven cases of cardiac myxomas at the Cardiac Surgery Department were chosen for study. Microsoft Windows SPSS software was used to analyze the data.

Results

Out of 11 cases of cardiac myxomas 9 were females (81%).Median age is 58 yrs (Range 34-68yrs).Most common site was left atrium(63%).Majority of patients presented with non specific cardiac symptoms such as breathlessness, unstable angina and generalized tiredness. Echocardiography could establish pre resection diagnosis in all cases. Gross examination of specimen showed tumor diameter ranged from 2.5-7.5 cm. Histopathological diagnosis was based on characteristic spindle or stellate cells with eosinophilic cytoplasm and ovoid nucleus, embedded in amorphous myxoid matrix. All cases showed hemosiderin laden macrophages in background. Gamnagandy bodies were noted in 2 cases (18%)and calcification was noted in one case.

Conclusion

The present case review showed the clinical and pathological profiles of cardiac myxomas. Inflammatory cells and hemosiderin deposits were common findings. Longterm follow up is excellent with no surgical recurrences noted in 11 cases.

Keywords

Cardiac Neoplasms; Immunohistochemistry; Local; Myxoma; Neoplasm Recurrence; Pathology, Surgical.

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INTRODUCTION

The most prevalent primary cardiac neoplasm is atrial myxoma. More than 72 percent of heart tumors are benign. It has a genetic origin and manifests clinically in 7% of cases. It is a component of a hereditary illness. At autopsy, cardiac tumors range in frequency from 0.001% to 0.3%, with myxomas accounting for more than 50% of cases.[1,2] Additionally, it is believed that they originate from primitive cells found in the endocardium and around the fossa ovalis, cardiomyocyte progenitor cells, or endocardial sensory nerve cells.[3-6]

Certain immunohistochemical and morphological characteristics might be connected to the clinical manifestations. Burke discovered that embolic myxomas were more likely to be thrombosed, widely myxoid, and had an uneven surface resembling fronds than nonembolicmyxomas. They were also less likely to be fibrotic. Older people were more likely to have fibrotic and non-thrombosed tumors, and their mean duration of clinical symptoms was longer. Younger women had higher rates of recurrent, numerous, and familial myxomas, which were also more likely to have uneven surfaces and histologically myxoid characteristics.[7]

PRKAR1A mutations are present in two thirds of cardiac myxomas linked to the Carney Complex. PRKAR1A mutations have not been reported in isolated (nonsyndromic) cardiac myxomas, although they are present in both familial and sporadic types of CNC. While there are no consistent genetic changes in sporadic myxomas, activating mutations in the GNAS1 gene or null mutations in PRKAR1A are present in family diseases linked to myxomas.[8] Echocardiography and MRIare the important non invasive diagnostic tools. Early diagnosis and surgical resection remain the treatment of choice to prevent complications.

A wide range of myxomas have been documented in earlier research; yet, the clinicopathologic classifications have received little attention, and the myxoma cells' origin is still up for debate.[9] Hence we present a cases series of 11 cases during 2017-2023 to find the clinical and histological features of cardiac myxoma.

MATERIAL AND METHODS

During the 2017–2023 study period, eleven cases of cardiac myxomas at the Cardiac Surgery Department were chosen. Prior to surgical resection, the diagnosis was made in each and every instance. The Institutional Review Board gave their approval for this project. Information on clinical presentation, diagnostic workup, histopathologic diagnosis, and follow-up was gathered from medical records.

In every instance, the maximal dimensions and location of the tumor were assessed. Following their fixation in 10% buffered formalin and paraffin embedding, all specimens underwent immunohistochemical and hematoxylin and eosin (H&E) staining procedures.

Alcian blue-Periodic acid Schiff (AB-PAS) staining was used on the last ten cases in order to identify and quantify the carbohydrate macromolecules present in the myxoidstroma. Neutral mucopolysaccharides are stained "purplemagenta" by Schiff's reagent, acid mucopolysaccharides are stained "blue" by alcian blue, and the complex of the two is stained "purplish red" when detected by ABPAS staining. An antibody panel was utilized for the purpose of immunohistochemical staining. Positive staining was only assigned to the brown particles that were clearly visible through low power objectives.

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Microsoft Windows SPSS software was used to analyze the data. Student t-tests were used to compare the mean values between the two groups. For every comparison, the criteria level of significance was set at $P<0.05$.

RESULTS

Out of 11 cases of cardiac myxomas 9 were females (81%) (Figure 1) .Median age is 58 yrs (Range 34-68yrs). Two cases were less than 40yrs. Most common site was left atrium (63%) (Figure 2).

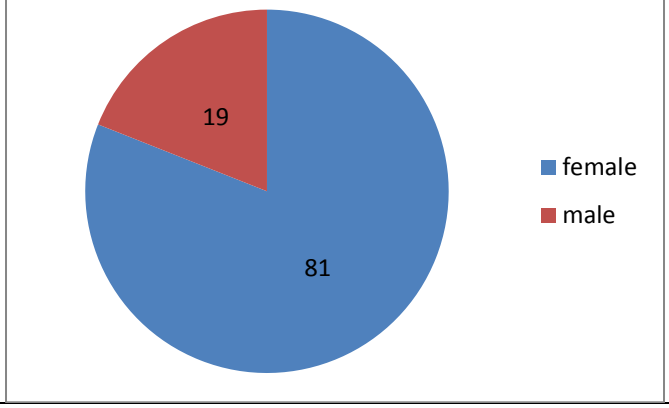

Figure 1: Distribution of age among cases

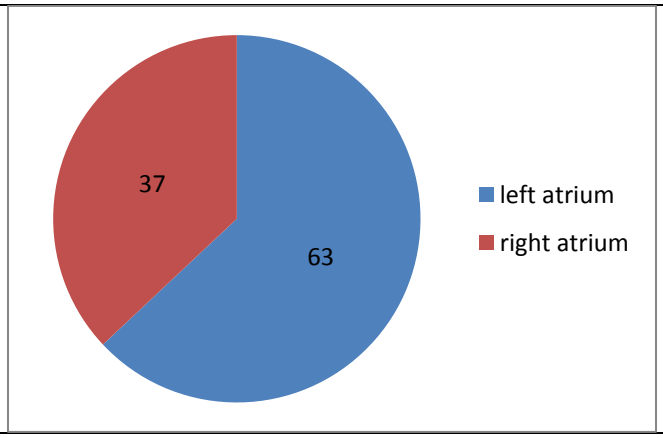

Figure 2. Common site of myxoma

Majority of patients presented with non specific cardiac symptoms such as breathlessness, unstable angina and generalized tiredness. Extra cardiac manifestations include stroke, fever and abdominal pain. Echocardiography could establish pre resection diagnosis in all cases. One patient had fever and positive blood culture for streptococcus viridians. Gross examination of specimen showed tumor diameter ranged from 2.5-7.5 cm. Surface of tumor was smooth in 8 cases (73%), and villous in 3 cases (27%)(Figure 3).Cut sections of all showed gelatinous and brownish areas (figure 4). (Table 1)

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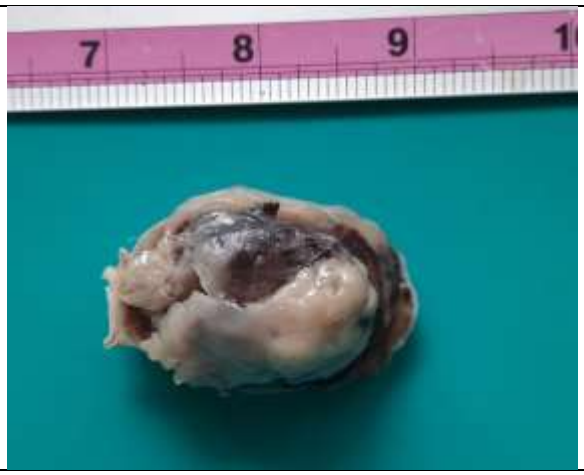

Figure 3: Smooth surface of myxoma

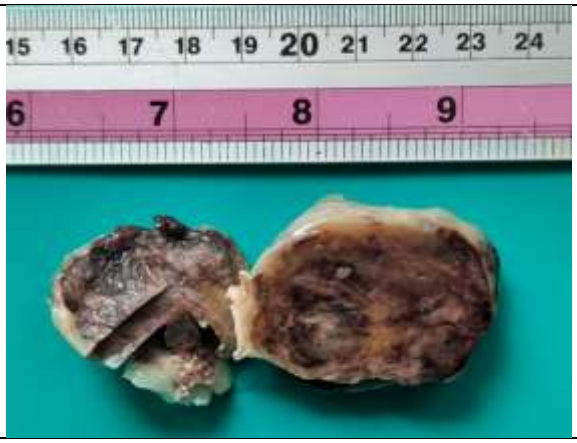

Figure 4: Brownish and myxoid cut surface of myxoma

Variable		Frequency (%)
Symptoms	Breathlessness	9 (81)
	Tiredness	8 (72.7)
	Unstable angina	7 (63.6)
Cardiac manifestations	Stroke	9 (81)
	Fever	10 (90.9)
	Abdominal pain	7 (63.6)
Surface of tumor	Smooth	8 (73)
	Villous	3 (27)

Table 1. Clinical features of cardiac myxoma

Histopathological diagnosis was based on characteristic spindle or stellate cells with eosinophilic cytoplasm and ovoid nucleus, embedded in amorphous myxoid matrix. All cases showed hemosiderin laden macrophages in background(figure 5).Gamnagandy bodies were noted in 2 cases (18%)and calcification was noted in one case(figure 6).

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Histopathological features	Frequency (%)
Hemosiderin macrophages	11 (100)
Gamnagandy bodies	2 (18)
Calcification	1 (9)

Table 2.Histopathological features of cardiac myxoma

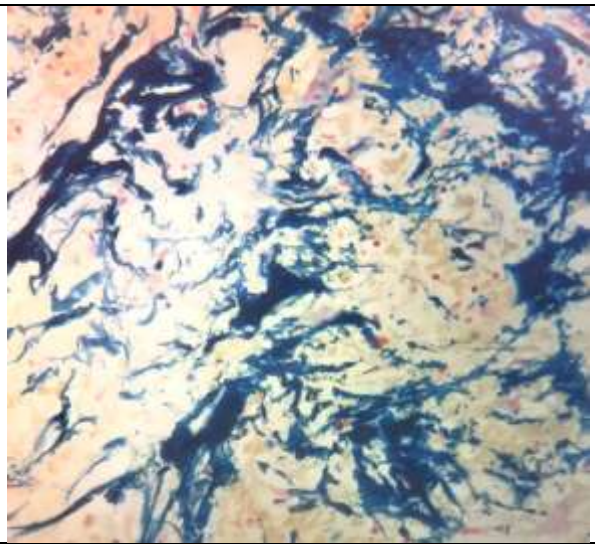

Fig 5: Perls stain-hemosiderin deposits 40X

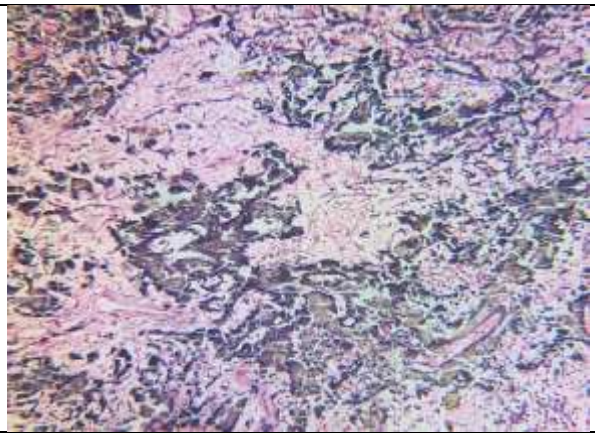

Fig 6: Gandy gamna bodies in myxoid background H&E 10X

DISCUSSION

The cardiac myxoma, which usually develops in the left atrium, is the most common primary tumor of the heart. Systemic embolization and left atrial blockage symptoms could be brought on by the tumor.[10]

In the present case review we studied the clinical and histopathological features of 11 cases related to cardiac myxoma observed during the study period of 2017 to 2023 in our hospital. Our findings are consistent with earlier research, which has demonstrated that tumors are the most prevalent primary tumor and that about 90% of myxoma are found in the atria, with

ISSN:0975 -3583,0976-2833 VOL14, ISSUE 12, 2023

the left atrium housing 63% of those tumors.[11] In the right side, only 37% of myxomas are found.[12]

As stated in previous studies myxomas occur most often in patients aged 30 to 70, with female predominance, and in families with a tendency to develop myxomas.[13] In our study also average age of patients was 58 years and 81% patients were female and rest were male.

Myxomas can present in a variety of clinical ways, depending on the location, size, and motility of the tumor.[14-17]A prior study found that dyspnea (54%), followed by palpitation (35%) is the most prevalent symptom.[18]Lower limb edema and dyspnea are believed to be the result of an atrioventricular valve blockage. However, the narrowing of the outflow tract and atrial fibrillation caused by the intracardiac blockage may further exacerbate dyspnea and palpitations. It is believed that coughing causes frank pulmonary edema and pulmonary venous hypertension. An inadequate blood flow could be the cause of angina. Another common sign of myxomas, which have been linked to the papillary surface, is embolism. Certain constitutional disorders may not yet have a clear cause. According to certain research, the cytokine interleukin-6 (IL-6) might be in charge of such. There is ongoing debate on the connection between IL-6 and constitutional disorders. Sometimes tumor fragment embolism from right atrial myxomas can cause pulmonary hypertension. Syncope can be caused by right ventricular myxomas that mimic pulmonary valve stenosis. In the present case series majority of patients presented with non specific cardiac symptoms such as breathlessness, unstable angina and generalized tiredness. Extra cardiac manifestations include stroke, fever and abdominal pain.

Echocardiography is a widely accepted sensitive preoperative diagnostic technique, even when certain occupied lesions seem to elude its detection. In certain instances, thrombus may be mistaken as myxoma. However, it is also shown that echocardiography is the most precise and dependable preoperative technique for predicting the shape, motility, attachment, and diameter of cardiac myxomas. In our series echocardiography could establish pre resection diagnosis in all cases.

On gross examination specimen of our cases showed tumor diameter ranged from2.5-7.5 cm. Surface of tumor was smooth in maximum cases and villous in some cases. Cut sections of all showed gelatinous and brownish areas. In previous studies the tumors exhibit a wide base, with the majority exhibiting pedicles and a soft, gelatinous, and highly friable surface when sliced. They can have several papillary, villous, finger-like projections or they can be smooth and shiny."Petrified" myxomas are myxomas that have become extremely calcified.[19]

During examination of histopathological features our case series showed calcification and gamnabodies, and hemosiderin deposits. In the previous literature it has been established that usually, necrosis, polymorphism, and mitotic figures are absent. It is usual to see Hemosiderin-Laden macrophages. Gamma Gandy bodies and hemosiderin pigment deposition were visible in. Mucin positivity can be established by the Alcian blue PAS stain.[20] According to reports, the myxoma cells exhibit reactivity towards S-100 protein, calretinin, vimentin, desmin, smooth muscle myosin, CD56, α1 antitrypsin, and α 1 antichymotrypsin. Myxoma cells' calretinin reactivity can be very helpful in distinguishing the disease from other conditions, especially fibroblastic lesions.[21]

CONCLUSION

We analysed 11 cases of cardiac myxomas diagnosed during 2017 to 2023. Most were females in the age range of 34-68 yrs. Cardiac symptoms such as breathlessness, angina and tiredness were

ISSN:0975 -3583,0976-2833 VOL14, ISSUE 12, 2023

the most common symptoms. Most common site was left atrium near fossa ovalis. Tumor size ranged from 2.5 cm to 7.5 cm in greatest diameter.70% of tumor showed smooth outer surface. Histopathological features include stellate cells, myxoid matrix, and ring like vascular structures. Calcification, gandygamna bodies were noted in a few cases. Inflammatory cells and hemosiderin deposits were common findings. Longterm follow up is excellent with no surgical recurrences noted in 11 cases.

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