TITLE: DEMOGRAPHIC AND CLINICAL FEATURES OF LEFT ATRIAL TUMORS IN SOUTH INDIAN POPULATION: A CASE SERIES

Dr. H.S. Natraj Setty MD, DM, FACC; Dr. Raghavendra Murthy MS,MCH; Dr. Shivanand Patil MD, DM; Dr. Veeresh Patil MD,DM; Dr. Jayashree Kharge MD, DM; Dr. T.R. Raghu MD, DM; Dr. Rahul Patil MD, DM; Dr. Satvik Raj MD,DM; Dr. Babu Reddy MD,DNB; Dr. C.N. Manjunath MD, DM

Sri Jayadeva Institute of Cardiovascular Sciences and Research, Bengaluru, Karnataka, India

Corresponding author:

Dr. H.S. Natraj Setty MD, DM, FACC.

Assistant Professor of Cardiology,
Sri Jayadeva Institute of Cardiovascular Sciences and Research,
Bangalore, Karnataka, India

Address:

#493, 4th Cross, 7th Main,

J.P. Nagar 3rd Phase.

Bangalore – 69

Email: drnatrajsetty75@gmail.com

ABSTRACT:

Myxomas are the most common type of primary cardiac tumor. They cause a variety of clinical manifestations depending on size and anatomical location. Sometimes, manifestations are atypical challenging differential diagnosis and the therapeutic approach. Left atrial myxomas are commonly missed clinically and often lead to grave consequences. We present here a series of 6 cases of left atrial myxomas with demographic and clinical characterization of the patients that were managed successfully.

Keywords: Myxoma, 2D Echocardiography, Left atrium, Cardiac tumor

INTRODUCTION:

Primary tumors of the heart are a rare clinical entity. They occur with an annual incidence of 1 reported case per 100,000 inhabitants. Overall, 85% of them are benign tumors. And the most common type of benign tumor of the heart is myxoma. It comprises around 50 to 70% of all primary cardiac tumors. Most of the myxomas, around 75%, are located in the left atrium attached by a pedicle to the interatrial septum near the fossa ovalis. Around 18% of the remaining arises from the right atrium; 4% from the right ventricle, and 3% from the left ventricle respectively [1, 2]. Approximately 5% of myxoma patients show a familiar pattern of tumor development. They arise as a benign polypoid neoplasm. They usually originate from endocardial cells in the region of fossa ovalis and attach to the interatrial septum. Myxomas are pedunculated, friable and appear as a soft, gelatinous, mucoid, usually gray-white mass often with areas of hemorrhage or thrombi. They grow slowly and usually do not produce symptoms or signs until they enlarge. They are typically nonhomogeneous in texture with lucent centers or areas of calcification. They can be quite large, occupying most of the left atrium and resulting obstruction to left ventricular filling [3]. Once diagnosed, surgery provides cure. The patients require a close follow-up to prevent recurrence

SUMMARY OF THE CASES:

A total of 6 cases were recorded in our institution in the past 1 year. 4 of them were males and 2 females Average age of patients was 38.3 years. Two patients presented with complaints of dyspnea on exertion one of whom had progressed to the stage of generalized edema and anasarca, one patient presented with atypical chest pain, one with exertional palpitations. The fourth patient was totally asymptomatic and was diagnosed during the routine medical examination. The last patient was actually a previously diagnosed case of left atrial myxoma who had undergone surgical excision but had now presented with complaints of fatigue and during this evaluation was found to have a recurrence of left atrial myxoma. None of these patients had constitutional symptoms like chronic unexplained fever, weight loss. Most of these patients presented with symptoms of approximately 3 to 6 months duration. Routine tests like complete blood counts, chest x-ray, coagulation profile, and comprehensive metabolic panel were normal for all 6 patients. Echocardiography showed all 6 patients having mass in left atrium, 2 of which were arising from the interatrial septum, in all 6 cases, the mass was seen protruding in mitral orifice causing dynamic mitral valve obstruction during certain phases of the cardiac cycle.
All 6 patients underwent successful surgical excision of myxoma, Average size of myxoma mass was 4.85 x 3.2 cm. they were irregularly shaped sometimes giving the appearance of a cluster of grapes, with a smooth surface. They had a nonhomogenous texture with intermittent areas of calcification.

DISCUSSION:

Primary cardiac tumors are rare, with an incidence of 0.0017 to 0.19 in autopsy series, in which 60% of them are benign. Myxomas are the most common heart tumors, with the majority located in the left atrium. 80% of them originate in the interatrial septum [4].In our cases also, all 6 patients had left atrial myxoma. , They are slowly growing and usually do not produce symptoms or signs until they enlarge [5]. About 75% are pedunculated and may prolapse through the mitral valve and obstruct ventricular filling during diastole [6]. Myxomas are thought to originate in undifferentiated and totipotent mesenchymal stem cells. They produce vascular endothelial growth factor, which probably contributes to the induction of angiogenesis and the early stages of tumor growth [7]. Myxoma shape and texture can be quite varied. Myxomas may be smooth surfaced but are more often irregularly shaped or have the appearance of a “cluster of grapes”. They are typically nonhomogeneous in texture with lucent centers or areas of calcification. Myxomas can be quite large, occupying most of the left atrium and resulting obstruction to left ventricular filling. Most cardiac myxoma cases during life are diagnosed by echocardiography. Thus, allowing for subsequent potentially curative surgical removal [8]. With the development of thoracic computed tomography and magnetic resonance imaging of cardiac structure, these techniques have been applied to the detection of cardiac tumors, although echocardiography has remained the primary method of diagnosis of cardiac myxoma [9]. The pathogenesis of cardiac myxoma is poorly understood, especially for those that are sporadic. Carney syndrome accounts for the majority of familial cases of cardiac myxomas. It is an autosomal dominant syndrome characterized by myxoma formation in cardiac and several extra-cardiac locations [10]. Surgical excision is the treatment of choice for most benign cardiac tumors. Recurrence is 1% to 5% after resection. We Report a case series of 6 patients aged 30-45 years, who has a large mass in the left atrium underwent successful resection of the tumor. The patients had a prompt recovery on follow up for 2years.

CONCLUSION:

Cardiac myxomas are the most frequent finding among primary cardiac tumors. Cardiac myxomas are commonly found in the left atrium, near the fossa ovalis and other locations, such as the right atrium or left ventricle. Clinical manifestations depend on the involvement in valvular obstruction, distant arterial embolisms, or nonspecific, constitutional symptoms. Myxomas may remain asymptomatic may be occasionally found on routine clinical examination. Early diagnosis can be challenging, because the nonspecific cluster of symptoms may be misleading. Transoesophageal echocardiography is the cornerstone for diagnosis of atrial myxoma. Cardiac myxomas should be managed with surgical resection.

CONFLICT OF INTEREST: NONE
ETHICAL COMMITTEE: APPROVED
FUNDING: NONE

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Table 1:

Patient No	Age	Sex	Presenting	Size in cm	Myxoma

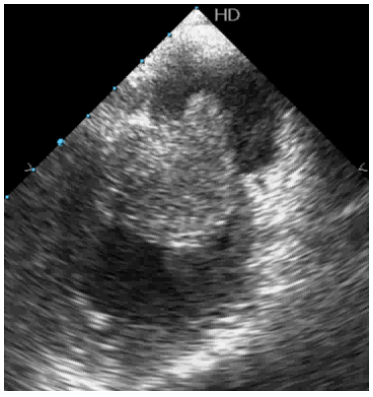
			complaints		Location
I	38	M	Palpitation	5.0X3.5	Left atrium
II	42	M	Dyspnea	5.3X2.6	Left atrium
III	31	M	Asymptomatic	5.2X3.1	Left atrium
IV	36	M	Fatigue	4.0X3.5	Left atrium
V	38	F	Chest pain	3.5X2.6	Left atrium
VI	45	F	Dyspnea	6.1X4.1	Left atrium

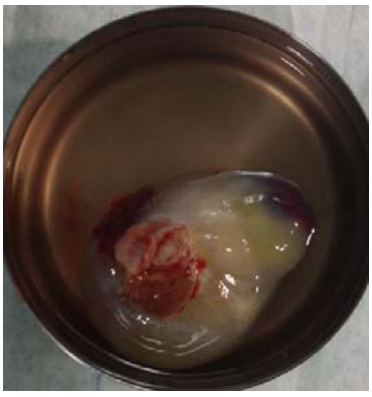
Figures:

Figure 1A:2D Echocardography parasternal long axis view showing large left atrial mass.

Figure 1B: Macroscopic appearance of the tumor is smooth surfaced irregular shaped typically nonhomogeneous in texture.

Figure 1C: Microscopy showing Composed of stellate or globular myxoma cells with abundant eosinophilic cytoplasm, indistinct cell borders, oval nucleus.





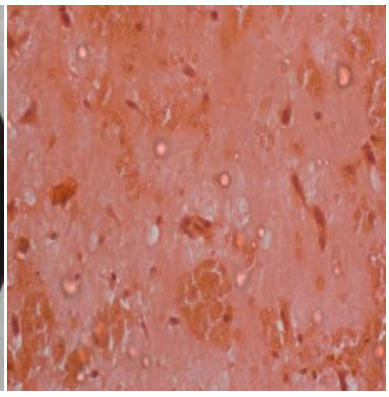


Fig 2B:2D Echocardography parasternal long axis view showing large left atrial mass.

Figure 2B: Macroscopic appearance of the tumor is smooth surfaced, irregularly shaped, the appearance of a cluster of

Fig 2C: Microscopy showing components of myxoma are spindle and stellate cells.

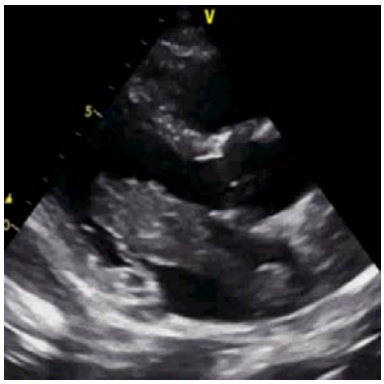


Figure 3A:2D Echocardography apical four chamber view showing large myxoma occupying the left atrium producing mitral valve obstruction.

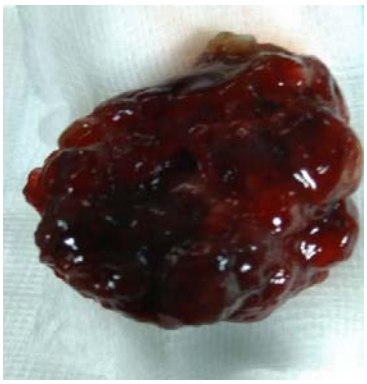


Figure 3B: Macroscopic appearance of the tumor is smooth surfaced, irregularly shaped, the appearance of a cluster of grapes.

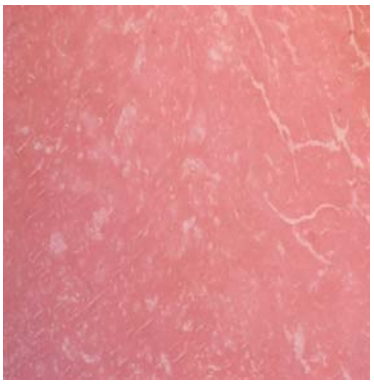


Figure 3C: Microscopy showing tumor displaying myxoid change and stellate cells having bland nuclei arranged in reticular meshwork.

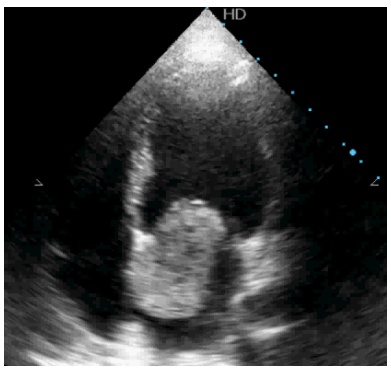


Figure 4A: 2D Echocardography apical four chamber view showing left atrial mass attached interatrial septum.

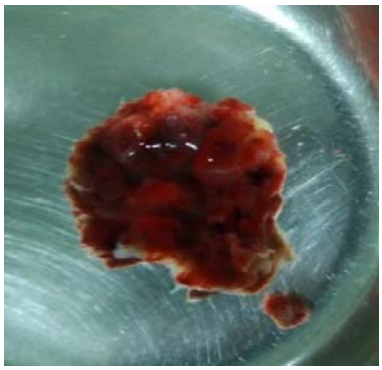


Figure 4B: Macroscopic appearance of the tumor is smooth surfaced, irregularly Oval shaped.

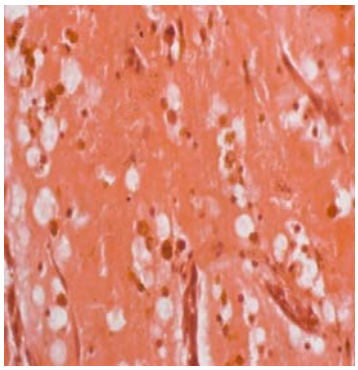


Figure 4C: Microscopy showing tumor displaying myxoid change and stellate cells having bland nuclei arranged in reticular meshwork.

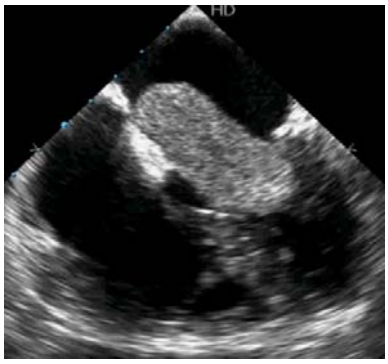


Figure 5A:2D Echocardography apical four chamber view showing left atrial mass attached interatrial septum.

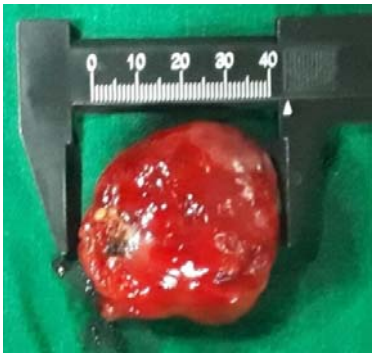


Figure 5B: Macroscopic appearance of the tumor is smooth surfaced, oval-shaped, gelatinous surface.

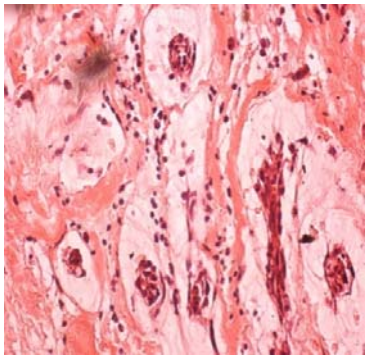


Figure 5C: Microscopic showing large areas of hemorrhage are noted, dark-colored pigmentation and a few pigment-laden macrophages are present.

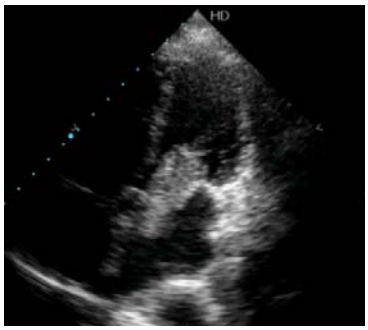


Figure 6A:2D Echocardography apical four chamber view showing left atrial mass attached interatrial septum

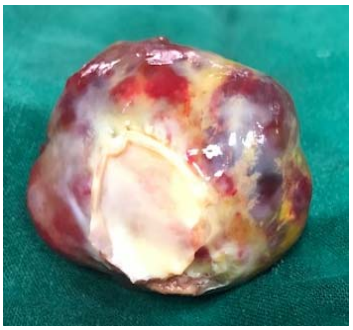


Figure 6B: Macroscopic appearance of the tumor is smooth surfaced, oval-shaped, gelatinous surface.

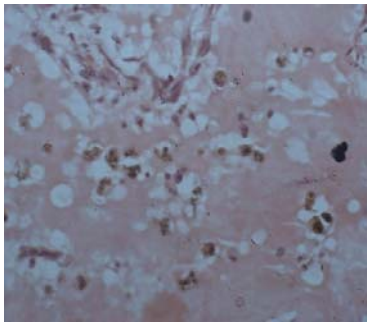
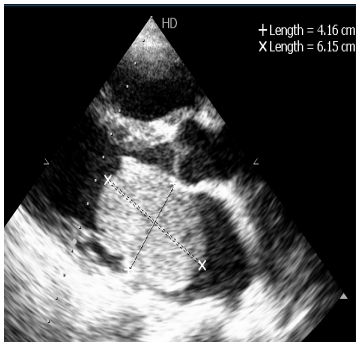


Figure 6C: Microscopic showing Large areas of hemorrhage are noted, dark-colored pigmentation and a few pigment-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+ Length = 4,16 cm
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