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Morphology of Cardiac Myxoma: A Case Series with Review of Literature

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Authors' contributions

This work was carried out in collaboration between both authors. Author AD performed surgeries, wrote the protocol, managed literature searches and performed analysis of study. Author DK processed and reported the surgical specimen, designed the study, wrote the first draft of manuscript and managed literature searches. Both the authors read and approved the final manuscript.

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Case Series

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ABSTRACT

Although rare, atrial myxomas constitute the most common benign cardiac tumors. Atrial myxoma presents a fascinating problem because of its rarity, manifestations and curability. Symptoms may be related to the hemodynamic effects of obstruction, to embolic phenomena or to systemic manifestations which may mimic collagen disease. Echocardiography is currently the most important noninvasive diagnostic modality available for imaging cardiac tumors. They have an excellent prognosis following surgical excision with occasional recurrence. We report eleven cases of atrial myxoma in seven females and four males of 14 - 66 year age group, with nine in left atrium and two in right atrium. Two-dimensional Color Doppler Echocardiography revealed pedunculated masses protruding into atrial chambers. All patients underwent successful surgical excision of the tumor and are asymptomatic since then. None of the patients had recurrence.

Keywords: Myxoma; benign; right and left atrium.

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1. INTRODUCTION

Cardiac myxoma is a primary benign neoplasm and accounts for 75% of surgically excised cardiac masses [1]. Autopsy series have shown that primary cardiac tumors constitute 0.001–0.19% [2]. The mean age of presentation is 50 years and has no sex predilection [3]. Familial syndromes associated with myxomas have activating mutations in the GNAS1 gene, encoding a subunit of G protein (in McCune-Albright syndrome) or encoding subunit of cyclic AMP dependent protein kinase (Carney complex) [4]. Myxomas commonly present as solitary mass arising from endocardial surface mainly in the left atrium (75%) followed by right atrium (15-20%) i.e. approximately 4:1 ratio [3-6]. Cardiac myxomas have varying clinical presentation and uncertain histogenesis [4]. Although most cases occur sporadically, familial lesions and lesions associated with a clinical complex have also been reported [5]. Biatrial (2%) are being reported increasingly while ventricular myxomas are distinctly rare [6]. Their cell of origin is thought to be primitive multipotent mesenchymal cell in subendocardial location [4]. On gross examination myxomas are gelatinous masses and have smooth lobulated or papillary surface. [7] Microscopically, it is characterized by an abundant mucopolysacchride matrix. The tumor cells may show arrangement in single cell pattern, trabecular pattern or in a vasoformative pattern and are subdivided further depending on predominant cells [6]. The degenerative changes like cystic spaces, hemorrhages and fibrin exudation are usually seen. Occasionally they may show calcification, bone formation with hematopoietic components and hemosiderin laden macrophages. These tumors are a major cause of patient morbidity and mortality [3]. Symptoms depend on location, size and mobility varying from no clinical signs or symptoms to various manifestations of chronic or acute congestive heart failure, syncope and arrhythmias with or without systemic findings, hence the suspicion of the diagnosis on the basis of clinical symptoms is very rarely achieved [2]. Our patients presented with vague complaints with constitutional symptoms and were diagnosed on 2 D Echocardiography, CT and MRI scans. Surgery for atrial myxoma can be undertaken solely on the basis of echocardiographic findings. The CT and MRI scans show the lesion to be endocardium based without myocardial extension. As the clinical presentation is diverse and nonspecific, imaging plays a key role in the diagnosis [8]. Surgical excision is curative for atrial myxomas and most of the patients can expect an excellent outcome. All of our patients underwent surgical excision of lesion. Their postoperative recovery was uneventful and all are asymptomatic on follow up visits. All the surgically resected specimen were sent for histopathology examination and all of them were reported to be myxoma (benign).

2. GROSS APPEARANCE

Received surgically resected gelatinous masses of 5 to 8 x 3 to 5 x 1 to 2 cm with lobulated surfaces, gelatinous and frondlike (Fig. 1) and a pedicle (Ball valve type) (Fig. 2). Cut surface was gelatinous soft (Fig. 3). No hemorrhage or necrosis were seen.

3. MICROSCOPIC FEATURES

Multiple sections from various areas showed tumor masses composed of uniform spindle shaped stellate cells with eosinophilic cytoplasm and rounded nuclei containing inconspicuous nucleoli (Fig. 4). The cellularity was variable. No nuclear atypia or abnormal mitoses was detected. At many places the stellate cells were seen aligned around blood vessels in ring like pattern associated with mononuclear inflammatory cell infiltrate (Fig. 5). The stroma was myxoid. No hemorrhages, necrosis or calcification were seen. The final diagnosis of atrial myxoma was made taking into account the echocardiography findings.

4. DISCUSSION

Left atrial myxoma was first described in 1845. However, the first antemortem diagnosis was made in 1951. Clarence Crafoord in 1954 performed the first surgical excision of a left atrial myxoma [9]. The antemortem diagnosis of primary cardiac tumors was difficult (is rarer still remove this)) but with the advent of angiocardiography and the adequate techniques of their surgical removal, the consideration of this diagnosis has become critical [2]. Grossly these are gelatinous tumours attached by a delicate or broad stalk protruding into cardiac chamber, ranging in size from 1-15 cm with smooth or lobulated surface [1]. Cardiac myxoma have been reported in the age group of newborn to 70 years, but most frequently manifests in the third to sixth decade and are often reported to be more common in females [1,2,3,9], which we have also observed. Approximately 50% of patients with myxomas may experience

symptoms due to central or peripheral embolism or intracardiac obstruction, but 10% of patients may be completely asymptomatic. Some patients may have murmur on auscultation. These murmurs can have similar findings like mitral and tricuspid stenosis for left and right atrial myxoma, respectively. It is typically mid-diastolic and occurs because myxoma causes atrioventricular valve obstruction [10]. Symptoms are characterized by a triad of hemodynamic obstruction, embolization and constitutional symptoms. Symptoms of mitral valve obstruction, the first arm of the classic triad of myxoma presentation, is present in majority of patients. The second frequent presentation in the classic triad is symptoms of embolism, essentially cerebral emboli with stroke. Males are statistically at greater risk than females of developing embolic complications. The third arm of the classic triad consists of constitutional symptoms with fever, malaise, weight loss, fatigue, myalgia and arthralgia are seen irrespective of location of myxomas [11]. Occasionally, constitutional symptoms are the sole manifestation of myxoma. which may be because of release of major acute phase chemical mediator cytokine interleukin-6 by tumor cells [4,10]. Younger and male patients have more neurologic and females have more systemic symptoms.

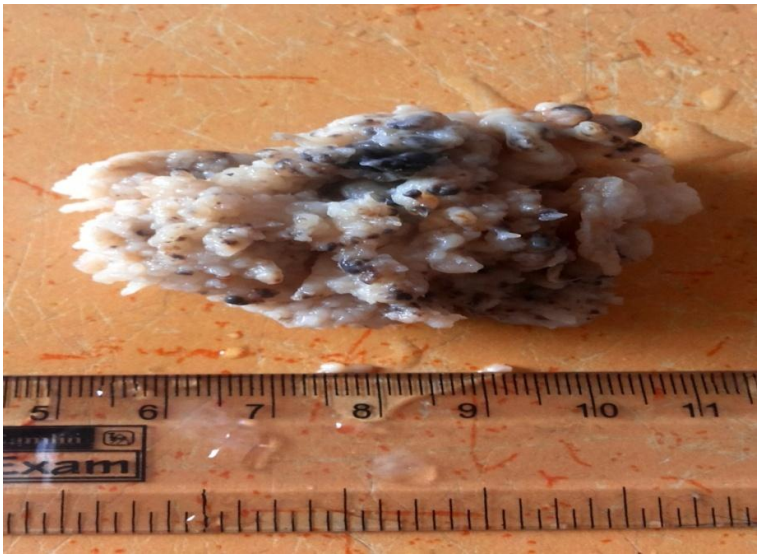

Fig. 1. Gross appearance of left atrial myxoma

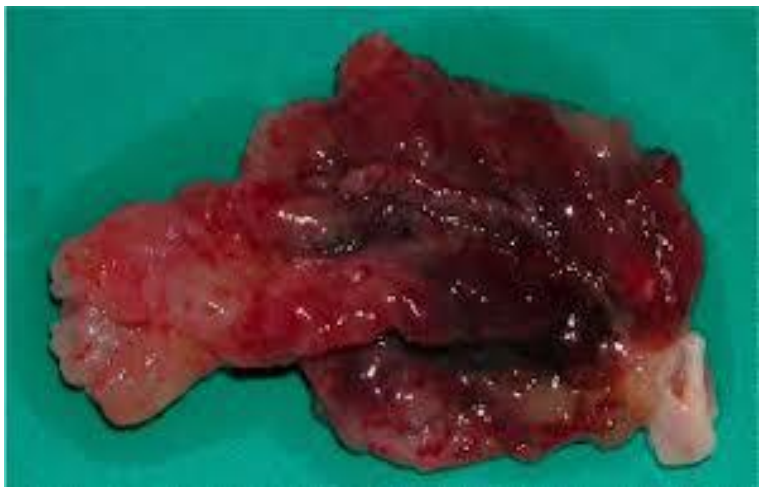

Fig. 2. Gross appearance of right atrial myxoma (Ball valve type)

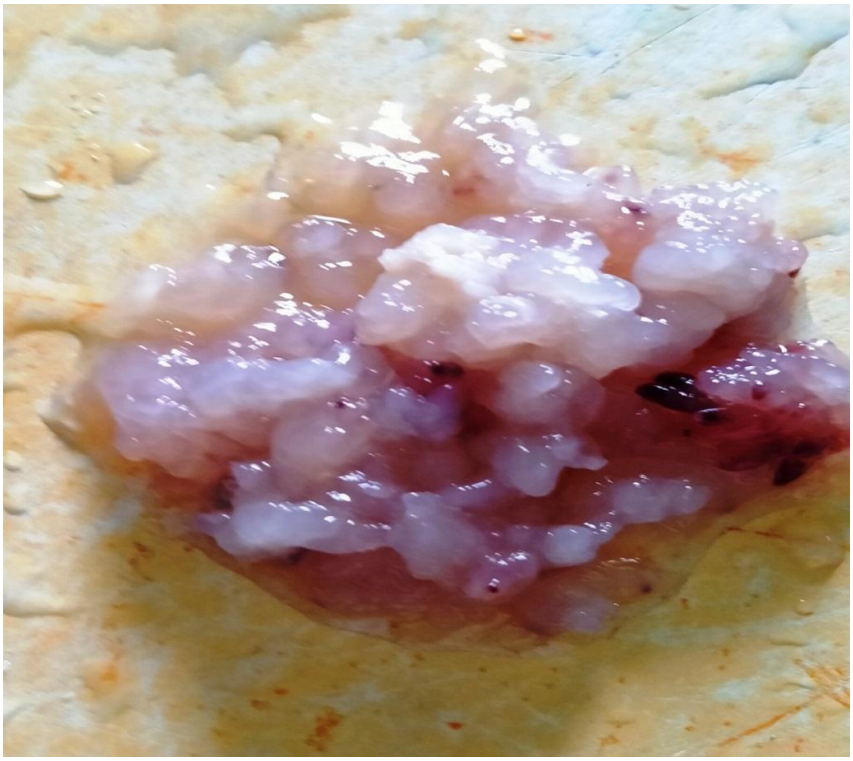

Fig. 3. Gelatinous appearance of myxoma

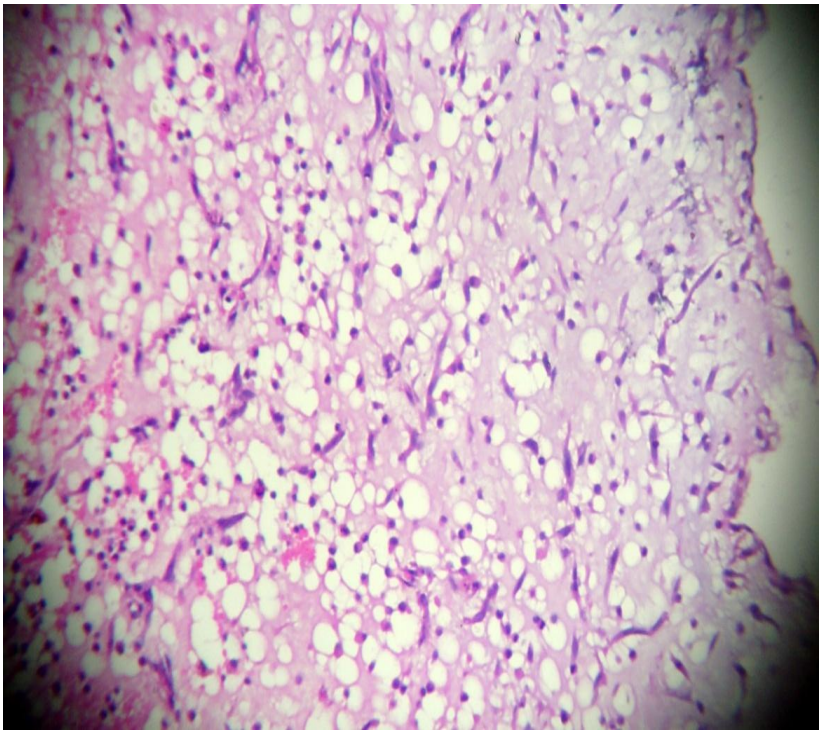

Fig. 4. Shows spindle shaped cells with elongated nuclei and myxoid background. No mitoses or necrosis seen. (H & E 10x X 40x)

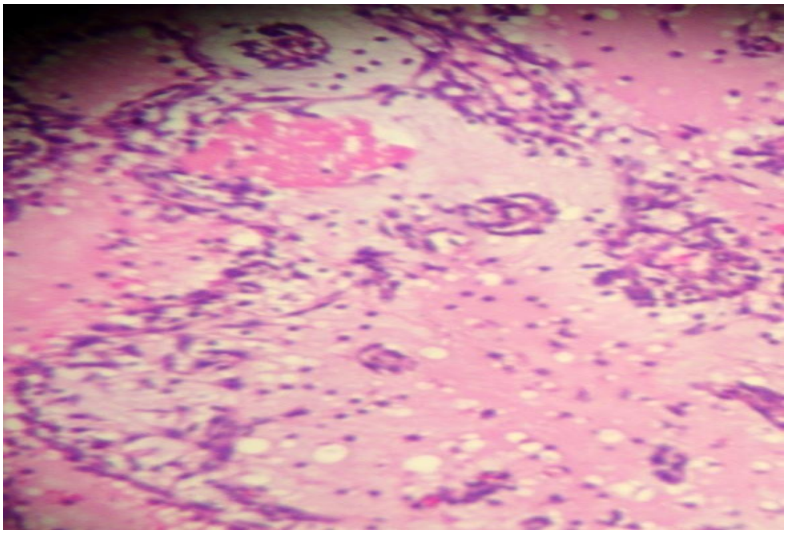

Fig. 5. Tumour cells arranged mainly around blood vessels in rings with mononuclear infiltrate (H & E 10x X 10x)

Screening for myxomas should involve a thorough history with physical examination and a 2D echocardiogram which provides an opportunity to identify these masses noninvasively [4]. Transthoracic echocardiography is approximately 95% sensitive and transesophageal echocardiography approaches 100% for the detection of cardiac myxomas [2]. A high index of suspicion is important for early diagnosis. Immediate surgical treatment is indicated because of the high risk of embolization or of sudden cardiac death. Complete excision of the tumour prolongs symptom free survival and is associated with a low recurrence rate [12]. Since late recurrence is known, years to months later probably because of incomplete excision, long-term clinical and echocardiographic follow-up is essential [2,3,9].

Surgery is a potential cure for myxomas. The rate of operative mortality ranges from 0–3%. In our study, the mortality was 0%.The major causes of peri-operative morbidity and death are embolism and myocardial infarction. Early post-operative dysrhythmias are transient and the long-term results are usually excellent. The tumor may recur if (a) it is incompletely excised, (b) if intraoperative fragmentation occurs, or (c) in familial myxoma [1].

5. CONCLUSION

Constitutional and obstructive symptoms are the common mode of presentation followed by embolic manifestation. Transthoracic 2D echocardiography (TTE), supplemented by CT and MRI scans and trans esophageal echocardiography (TEE), when necessary, forms the mainstay of diagnosis. Surgical excision of atrial myxoma is curative and gives excellent short term and long term results.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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