

Original Research Article

Systemic manifestations of atrial myxoma

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ABSTRACT

Background: Approximately 70% of the cardiac tumors are benign, atrial myxoma is the most common, and is seen more in females. Presentation is variable, and patients can present with cardiac symptoms or systemic manifestations. Prognosis may be grave if the presentation is late or if the diagnosis and surgery is delayed. Accurate diagnosis, early surgical intervention gives excellent results.

Methods: The study was conducted in the department of cardiovascular and thoracic surgery, all the patients who had diagnosis of cardiac myxoma irrespective of age, sex and associated diseases were included in the study. Transthoracic echocardiography was the initial investigation of choice, in some vascular Doppler studies, and CT scan was also done. All were operated under cardiopulmonary bypass, myxoma was approached by left atriotomy, tumor along with its base was excised, and the defect so created was closed directly or a patch repair was done.

Results: Majority were in 5th decade of life, females were more. Presentation included breathlessness, edema, hemiparesis, monoparesis, aphasia. Clinical examination revealed anemia, dyspnea, edema, audible murmur, weakness of limb/limbs and aphasia. Echocardiography established diagnosis in all. Left atrium was the common chamber involved. Interatrial septum on left atrial side was the common site of myxoma origin. All had uneventful post-operative period, and there was no mortality.

Conclusions: Atrial myxoma, though very rare, can present with a grim phenomenon, early and accurate diagnosis, followed by excision under cardiopulmonary bypass gives excellent results.

Keywords: Embolism, Excision, Left atrium, Myxoma

INTRODUCTION

The first report of cardiac tumor appeared in 1559, the prevalence of primary tumors of the heart in autopsy series has been reported between 0.007, 0.35 to 0.3 percent, In 1931 the first extensive review of the literature was made, and a useful classification of the system was designed.¹⁻⁵ Myxomas are globular in shape, have a soft gelatinous consistency, exhibit a yellow brown or greenish hue and often contain areas of hemorrhage and necrosis. They usually have short broad-based attachment but can be sessile. Papillary forms may have a frond-like surface that is friable making emboli more likely. Myxomas are exclusively intracavitary and

are rarely seen deeper than endocardium on histological sections.⁴ Cardiac myxomas are found most commonly in adult females with a median age at presentation of 49 years, but patients below 20 and above 90 years have also been diagnosed with these tumors. Left atrium is the most common site in about 86% of cardiac myxoma, and 90 percent of myxomas are solitary. These tumors usually occur without any predisposing genetic disorder, biatrial and multiple cardiac myxomas are also reported.

Symptomatology is related to hemodynamic effects of the tumor, embolism, or constitutional symptoms and may include, dyspnea on exertion, palpitations, congestive heart failure, syncope, hemoptysis, embolic events, atypical chest pain, paroxysmal nocturnal dyspnea, fever,

fatigue, weight loss, night sweats, severe dizziness, seizures, peripheral edema, or sudden death. Dyspnea occurs when pulmonary venous blood entering the left heart is obstructed by the left atrial tumor, the obstruction is progressive, but when obstruction is intermittent, syncope, often related to postural change or sudden death may occur.⁶ Impairment of valve closure, either by obstruction or leaflet damage, may cause regurgitation.⁷ The symptoms of pulmonary hypertension result from chronic pulmonary artery occlusion by right heart myxomatous emboli and mitral valve obstruction by left atrial cardiac myxomas.

The constitutional symptoms are produced by the hemorrhage and degeneration within the tumor, production of active substances by the neoplasm, micro embolism of small tumor fragments, or a systemic autoimmune reaction to shed myxoma cells. The embolization of myxoma may present as acute arterial occlusion or may be fatal, embolization is common because of the friability of the tumor and intracavitary location. Finally, the patient may be symptomatic from cardiac arrhythmias which improve after removal of the tumor. Clinical examination in a lean thin anemic patient may include, fever, mitral diastolic/systolic murmur, right heart failure, tumor plop, clubbing, fibrillation, monoparesis, hemiparesis, monoplegia, hemiplegia, peripheral or pulmonary edema.

The laboratory investigations may reveal a raised erythrocyte sedimentation rate, pulmonary emboli, elevated globulins, abnormal chest skiagram, polycythemia, anemia, pulmonary artery hypertension, mitral/tricuspid valve disease, in addition to an atrial mass. The investigation of choice is echocardiography (transthoracic {TTE}/transesophageal {TEE}), radionuclide imaging, computed tomography (CT), magnetic resonance imaging (MRI), and rarely angiocardiology may be needed in selected patients. Majority of the patients present with complications, because of the rarity of the disease, lack of knowledge, varied presentation, delayed diagnosis, and lack of diagnostic facilities at peripheral centers. Early suspicion, specific referral to cardiac centers, thorough evaluation can prevent complications, even with complications surgical excision gives excellent results.

METHODS

The study was conducted in the department of Cardiovascular and thoracic surgery, all most all the patients were under treatment/evaluation of quacks, paramedics, professionals, other specialties for weeks to months. A thorough history, detailed general and systemic examination was contemplated in all. Besides routine investigations, TTE was done in all, CT scan, vascular doppler studies and MRI were performed in selected patients. All the patients with diagnosis of atrial myxoma, irrespective of age, sex and associated diseases, were included in the study. Jugular venous cannulation

was avoided. Heart was approached through median sternotomy, all the patients were operated under cardiopulmonary bypass, cannulation was done gently, aorta cross clamped before giving cardioplegia, and the myxoma was approached through left atriotomy. Excision of the tumor along with its base was done, the defect so created was repaired directly or a patch closure was done. Thorough saline irrigation/suction of atria and ventricle removes all the tumor emboli. The intra / peri and post-operative period was monitored for any morbidity and mortality. Patient with PTFE patch were put on antiplatelet drugs for six weeks. All the patients were followed in outpatient department. Follow up TTE was done at 18 and 36 months after operation.

RESULTS

Of the seven patients 71.42% were female, 51.14% were in 5th, 28.57% in 4th and 14.28% in the 3rd decade of life. Breathlessness was the common symptom, Table 1, anemia, and audible murmur the common signs, Table 2.

Table 1: Symptomatology in patients with atrial myxoma.

| Symptoms | Number of patients (%age) |
|----------------|---------------------------|
| Breathlessness | 71.42 |
| Edema | 57.14 |
| Limb Pain | 57.14 |
| Hemiparesis | 42.85 |
| Monoparesis | 28.57 |
| Aphasia | 28.57 |

More than one symptom were present in a patient.

Table 2: Clinical findings in patients with atrial myxoma.

| Clinical Findings | Number of patients (%age) |
|-----------------------------------|---------------------------|
| Anemia | 57.14 |
| Dyspnea | 42.85 |
| Audible murmur | 42.85 |
| Weakness of Limbs | 42.85 |
| Aphasia | 28.57 |
| Vascular insufficiency (in limbs) | 28.57 |
| Atrial fibrillation | 28.57 |

Patients were very sick and 57.14% of the patients were in NYHA functional class IV, and 42.85% in NYHA functional class III. TTE evidence of atrial myxoma was evident in all. Vascular doppler done in 85.71% and CT head in 71.42% of the patients, showed features of peripheral vascular embolization in 57.14% and brain infarct in 42.85% of the patients respectively. Besides decompensating left ventricular symptomatology, embolization and the neurological complications were the major problems. Heart was approached by median sternotomy, and myxoma by bi-atrial approach in majority. 85.71% patients had myxoma on interatrial

septum left atrial side, and 14.28% had myxoma on interatrial septum right atrial side. Complete excision of the tumor with its base and adjacent septal wall was done in all, Figure 1 and 2, the defect so created was closed directly in 42.85% and in others patch closure of the defect was done. There were no intraoperative complications, and post-operative period was uneventful. Histopathology confirmed the diagnosis of myxoma in all. At a follow-up of two month none of the patient had features of peripheral emboli clinically or on doppler study, however, 14.28% had residual neurological deficit even at a follow-up of two year. All the patients had improved NYHA functional class, and by six months post operatively 85.71% were in functional class II. There was no recurrence, and no mortality.



Figure 1: Myxoma being scooped out of atrium.

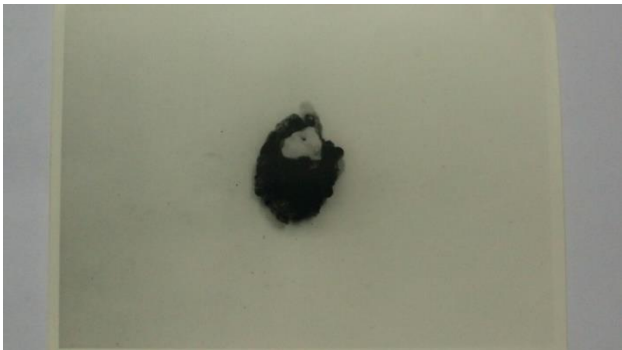


Figure 2: Excised myxoma along with its base.

DISCUSSION

Cardiac myxomas are primary, intracavitary tumors, occurring within any of the cardiac chambers, but have a predilection for the atria, particularly the left. The disease is very rare, and even the best and busiest cardiac centers are doing at the most 1 to 2 cases in a year. Majority of the patients being females, though significant, cannot be taken as authentic because of less number of patients, however, myxomas being more in females is well known.⁸ Myxomas are seen more in 4th and 5th decade of life, but age is no bar and even children can be diagnosed to have such a disease.⁹ Symptomatology is related more to hemodynamic effect of tumor. Symptomatology in the present study is in accordance to the reported

observations of other studies, left atrial myxoma with systemic embolization usually a fatal complication has been reported to occur in 25 to 50 percent of patients preoperatively, similar observations have been made in other studies.¹⁰⁻¹³ Clinical signs may vary from patient to patient, but our observations are invariably in accordance to other studies.¹⁴⁻¹⁶ The patients in present study were very sick, majority had neurological/embolic events such as recurrent limb pain, change in temperature of the limb, paresis or plegia of a limb or side and even aphasia.

Majority of the patients with complications may be because of the fact, that the patients don't have access to quality health centers, initially get treatment from local quacks, paramedics and even professionals-who either don't have specific diagnostic facility, or whose advice / referral patients are not following. It is believed that these patients only come to a specialized center after a major complication event such as mono or hemiparesis, limb ischemia or cardiac failure. Since the first North American application of ultrasound to visualize cardiac neoplasms in 1968, echocardiography has become the most important non-invasive modality to diagnose myxomas with a sensitivity of 100%.¹⁷ Transthoracic echocardiography a non-invasive technique, widely available, is the initial investigation of choice, and in majority nothing further is required to confirm the diagnosis. It not only images the myocardium and cardiac chambers but also identifies the mass, provides information about any obstruction to the circulation and the likelihood that the tumor could be the source of embolization. TEE is more informative, the superior diagnostic utility of TEE is due to the proximity of esophagus to the heart, lack of intervening lung and bone, and the ability to use high- frequency imaging transducers that afford superior spatial resolution, and helps in planning surgical management.^{18,19}

CT scan is a useful diagnostic tool especially in patients where MRI is not available, but MRI is preferred which in addition to furnishing detailed anatomic images, the T1- and T2- weighted sequences reflects the chemical microenvironment within a tumor, thereby offering clues as to the type of tumor that is present, cardiac mural infiltration, pericardial involvement, and extracardiac tumor extension. Interatrial septum as the common site of myxoma origin is established in majority of the studies. Early surgical intervention for atrial myxomas which is done on urgent basis, mitigates morbidity and usually offers cure.

Our observations are at variance to the studies, where only right atriotomy approach has been used to excise the myxoma, but are in accordance to the technique, where a longitudinal incision in the left atrium, posterior to interatrial groove is first performed, and the tumor visualized, the exposure of the tumor is facilitated by a simultaneous right atriotomy, with excision of a full-thickness portion of the interatrial septum, including the fossa ovalis.²⁰⁻²³ The myxoma which is attached to the

fossa ovalis, is removed through right atrial incision if small or through left atrial incision if large, the bi-atrial approach is particularly useful if the base of the left atrial myxoma is sessile.²⁴

In patients with right atrial myxoma both cavae should be cannulated directly or venous drainage should be provided by superior vena cava and femoral vein cannulation. Our observations are at variance to the studies, as far as sporadic occurrence of myxoma in middle age women is concerned, but are almost in accordance with regard to isolated forms accounting for 95 percent, left atrium being the site in 76 to 88 percent, and without associations with other conditions.²⁵ Myxoma is suspected clinically, diagnosed on echocardiography/macrospectically, and confirmed by histopathology, one feature which distinguishes them from thrombi is that they are covered by endothelium and have endothelium lined crevices and clefts. None of our patients had any local recurrence, which is contrary to the studies where 4 to 5 percent recurrence has been reported, but in large series no recurrence has been reported.^{23,26} No deaths in the present study is at variance to studies, where less than five percent mortality is reported.²⁶⁻²⁸

CONCLUSION

In conclusion atrial myxoma though rare, can present with a grim phenomenon, clinical presentation is not specific, diagnosis is made by TTE, in case of doubt other modalities of investigation should be used, once a diagnosis of atrial myxoma is made, surgery should not be delayed, jugular cannulation should be avoided, tumor should be approached through left atriotomy, should be excised along with its base, thorough irrigation, suction of all the chambers should be done, the defect so created should be closed by direct or patch repair.

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