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Case report

Management of a case of left atrial myxoma presenting with right ventricular dysfunction and pulmonary artery hypertension

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ABSTRACT

Left atrial myxomas are rare primary cardiac tumors, accounting for approximately 0.5% of all cardiac tumors per million individuals annually. Although histologically benign, their strategic location and potential for obstructive or embolic complications often render them clinically significant. These tumors predominantly arise from the interatrial septum and may present with a constellation of non-specific systemic and cardiorespiratory indications, mimicking various cardiac or pulmonary pathologies. A middle-aged subject with progressive dyspnea, fatigue, and signs of cardiorespiratory compromise was studied in this case study. Initial laboratory investigations revealed elevated renal parameters suggestive of end-organ involvement or hypoperfusion. Further evaluation with two-dimensional echocardiography (2D ECHO) revealed the presence of a large left atrial mass, consistent with a myxoma, prolapsing through the mitral valve during diastole. Associated findings included severe pulmonary artery hypertension (PAH) and right ventricular (RV) dysfunction. The left ventricular ejection fraction (EF) was preserved at 55%, indicating isolated right-sided compromise. The patient was hemodynamically stabilized using appropriate medical management prior to surgical intervention. Optimization involved correction of fluid balance, management of renal function, and supportive treatment for RV dysfunction and PAH. Once stable, the case experienced successful surgical excision of the tumor via median sternotomy and cardiopulmonary bypass. Histopathological examination established the identification of atrial myxoma. The postoperative recovery was predominantly unremarkable, with gradual improvement in cardiopulmonary function and normalization of renal parameters. This case highlights the importance of early recognition and comprehensive evaluation of cardiac tumors, particularly myxomas, which can have life-threatening consequences if not promptly diagnosed and managed.

Key words: atrial myxoma, right ventricular dysfunction, pulmonary artery hypertension, 2D ECHO

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INTRODUCTION

Primary cardiac tumors are exceedingly rare, with a prevalence of merely 0.03%, of which 75% are benign. Half of the benign instances are myxomas. The incidence rate of left atrial myxomas, which are comparatively benign primary heart tumor, is 0.5% per million annually [1, 2]. Females aged 50–60 years old had the highest incidence of cardiac myxomas [3]. Patients may have various symptoms linked to embolization, cardiac valve interference, circulation obstruction, and/or direct invasion of the heart or surrounding lung tissue [4, 5]. Cough, hemoptysis, peripheral edema, fatigue, pulmonary edema, orthopnea, dyspnea, and paroxysmal nocturnal dyspnea are the most common non-specific general and cardiorespiratory symptoms of atrial myxomas [1, 6].

Large tumors, in particular, can result in a mass effect on the heart chamber, significant hemodynamic abnormalities, comprising secondary pulmonary hypertension, and making the surgical management is more challenging [7]. There have been numerous documented occurrences of pulmonary hypertension (PH) associated with massive left atrial myxoma [8–11].

We describe a notable case of a cardiac myxoma accompanied by severe pulmonary hypertension and right ventricular dysfunction (RVD) requiring careful hemodynamic optimization before surgical removal.

CASE PRESENTATION

62-year-old male subjected arrived at the emergency department with various complaints, including chest pain, a sense of chest heaviness, generalized body swelling, bilateral pedal edema, abdominal discomfort, reduced appetite, breathlessness, and oxygen desaturation. With the history of systemic hypertension and on regular antihypertensive medication. Upon admission, the individual was alert, conscious, and appropriately oriented to time, place, and identity. His vital parameters were stable with a blood pressure of 118/88 mmHg and a heart rate of 100 bpm. Oxygen saturation (SpO_2) improved to 99% upon administration of oxygen via nasal cannula. On systemic examination, cardiovascular auscultation revealed soft first (S1) and second (S2) heart sounds without any additional murmurs or gallops. Respiratory examination showed clear breath sounds with bilateral basal crepitations, suggestive of early pulmonary congestion. Abdominal examination revealed a soft but distended abdomen, without guarding or rigidity. These clinical findings warranted further investigation for potential cardiac decompensation and systemic involvement.

INVESTIGATION

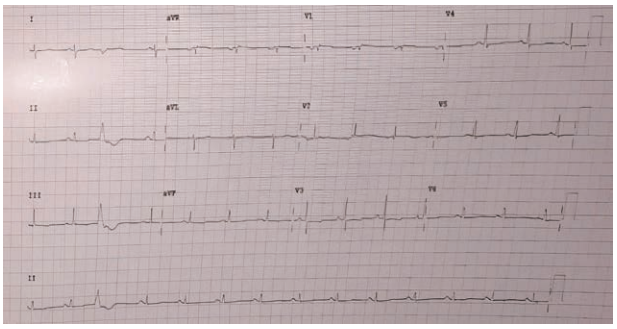
The patient's hematological profile was largely within normal limits, except for a mild reduction in red blood cell indices, suggestive of possible microcytic anemia. Serum electrolyte levels, including sodium, potassium, and chloride, were within normal reference ranges. Renal function tests revealed elevated serum urea and blood urea nitrogen, with a normal serum creatinine level. Serological tests for HIV, HCV, and sHBsAg were non-reactive (tab. 1).

Table 1. Laboratory findings of the patient.

Complete blood count	Observed value	Reference value
Hemoglobin	13.10 gm/dL	13–18 gm/dL
Total leucocyte count	10.39×10^3/µL	$4.5\text{-}11 \times 10^3$/µL
Total erythrocyte count	5.89×10^6/µL	$3.5–5.5 \times 10^6$/µL
Platelet count	251.00×10^3/µL	$150–450 \times 10^3$/µL
MPV	10.70 fl	
PCT	0.27%	
PDW	16.70%	
RBC indices		
HCT (PCV)	40.60 %	
MCV	69.00 fl	82–95 fl
MCH	22.30 pg	25–33 pg
MCHC	32.30 gm/dL	33–37 gm/dL
RDW CV	18.60%	11.0–16.0%
Differential WBC count		
Neutrophils	69%	40–70%
Lymphocytes	22.00%	20–45%
Eosinophils	0.1%	0–6%
Monocytes	0.8%	0–8%
Basophils	0.0%	0–1%
Serum electrolytes		
Serum sodium	134.10 mmol/L	130–148 mmol/L
Serum potassium	3.93 mmol/L	3.50–5.50 mmol/L
Serum chloride	99.10 mmol/L	85.0–108.0 mmol/L
Renal function tests		
Serum urea	65.99 mg/dL	15.0–45.0 mg/dL
Serum blood urea nitrogen	30.84 mg/dL	7–23 mg/dL
Serum creatinine	1.23 mg/dL	0.6–1.4 mg/dL
Anti-HCV	non-reactive	non-reactive
HBsAg	non-reactive	non-reactive
HIV I and HIV II	non-reactive	non-reactive

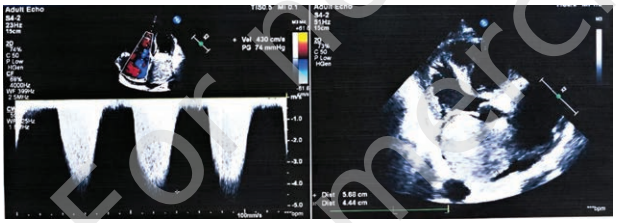
ECG showed sinus rhythm. The right ventricular strain was evident by right axis deviation and right ventricular hypertrophy. The ventricular premature contraction, and left atrial enlargement, and borderline T abnormalities were present (fig. 1).

Figure 1. Sinus rhythm. Ventricular premature complex, left atrial enlargement, Right ventricular hypertrophy, Borderline T abnormalities.



2D ECHO showed large left atrial myxoma, mild aortic regurgitation (AR), severe pulmonary artery hypertension (PAH), right ventricular (RV) dysfunction, ejection fraction (EF) was 55% with grade I left ventricular diastolic dysfunction (LVDD) (fig. 2).

Figure 2. 2D ECHO showing large left atrial myxoma.



Through the right radial approach, coronary angiography was done and diagnosed as ischemic heart disease with unstable angina. The left main coronary artery, left circumflex artery, and right circumflex artery were normal. The left anterior descending artery was type III LAD and was found to be normal. Considering the magnitude and manifestations of decompensated right ventricular failure accompanied by severe pulmonary hypertension, the patient underwent hemodynamic optimization prior to definitive surgical excision.

MANAGEMENT

The patient was treated symptomatically with nasal O_2 support and ionotropic support with injection noradrenaline and injection dopamine along with injection heparin 5,000 IU t.d.s., injection furosemide 10 mg b.d., tablet carvedilol 3.125 mg b.d., tablet spironolactone 25 mg o.d., tablet ivabradine 5 mg b.d., injection cefpodoxime 1 g b.d., injection pantoprazole 40 mg b.d. and injection ondansetron 4 mg t.d.s.

Following a 48-hour period, the patient exhibited decongestion and symptomatic improvement. Hence, the patient was discharged but was readmitted with similar symptoms a few hours later. After the symptoms had been entirely resolved and renal parameters returned to normal levels, the patient was assessed for fitness for surgery. The patient was planned for intracardiac tumor excision. He was posted for surgery on the 7th day after initial admission.

Under general anesthesia, with the patient in the supine position, a midline sternotomy was done. Pericardial effusion was present. The pulmonary artery was tense, suggestive of severe PAH. The patient was heparinised. Auto bicaval cardiopulmonary bypass (CPB) was done. The aorta was cross clamped. The left atrial appendage (LAS/RA) was opened. A large 6 × 6 cm left atrial myxoma attached to the LAS/RA was seen. The mass was removed. A thorough wash was given to the LAS/RA, which was closed. The heart chamber was deaired, and a clot shot was given. The patient was declamped and weaned off from CPB. The patient was protaminized. After hemostasis was attained, the mediastinal drain was kept, and chest closure was done.

The patient was shifted to the ICU on ambu bag support on stable hemodynamics. The patient was put on synchronized intermittent mandatory ventilation (SIMV) ventilation mode. The patient was given inotrope support for blood pressure. Ventilatory support was gradually tapered. Extubation is done after 4 h as per standard protocol. Post extubation, the patient was given nasal O_2 support. Postoperative period was uneventful.

2D ECHO on the 4th post-operative day showed global LV hypokinesia, and EF was 35%. Right atrial (RA) and right ventricular (RV) dilatation were present, along with RV dysfunction and severe tricuspid regurgitation (TR). Bilateral mild pleural effusion was also seen. Right ventricular systolic pressure (RVSP) was 70 mmHg, and right atrial pressure (RAP) was 1 mmHg. The patient was discharged on the fifth day following the operation.

The excised specimen was given for pathological analysis. On gross examination, soft tissue bits aggregated to 6.5 × 4.3 × 3.9 cm, and the cut surface showed myxoid areas. Histology of the representative sections showed a tumor comprised of numerous lipidic cells on the background of myxoid area the myxoid regions with few dilated vessels and scattered inflammatory cells confirmed the diagnosis of left atrial myxoma.

DISCUSSION

After a left atrium enlargement was seen on angiography, a failed surgical procedure was used in 1951 to make the first known attempt to diagnose a left atrial tumor in a living individual [12]. The first clinical use of 2D echocardiography, which made it possible to see and describe myxomas, did not occur until the early 1970s [13]. As technology has advanced, so too has the safety and accuracy of diagnosing and removing such tumors.

Entrapment of the embryonic foregut within the heart tissue, which permits neuronal and epithelial differentiation, is one of the hypothesized causes of cardiac myxomas [13]. There are numerous ways in which myxomatous tumors of the left atrium (LA) can cause pathologic symptoms. LA myxomas can result in mitral regurgitation or heart failure by blocking blood flow through the heart's chambers and/or valves [6]. Local cardiac or pulmonary tissue invasion by myxomas may result in arrhythmia, pericardial effusion, cardiac tamponade, hypercontractility, or lung tissue invasion [6, 14].

Lastly, embolic diseases, the most severe of which are neurologic, can result from the tumor's ejection of fragments or thrombi [14, 15]. The tumors typically appear as masses that are 1–15 cm in size, weigh 15–180 g, and grow at a pace of 1.3–6.9 mm per month [14, 16]. Smaller tumors generally exhibit a friable or villous appearance, making them more susceptible to embolization, whereas larger tumors display a smoother surface [17]. Furthermore, ventricular arrhythmias and heart block are conduction abnormalities that may arise from direct pressure on the myocardium or conduction system [18].

Fatigue, constitutional symptoms, orthopnea, and dyspnea are common presenting symptoms. For big masses that fully obliterate the left atrium (as in our instance), myxoma-induced circulation obstruction might result in acute pulmonary hypertension, RV failure, and heart failure symptoms [4, 7, 8, 18]. Our case had bilateral pedal edema, abdomen discomfort, chest pain, chest heaviness, body swelling, decreased appetite, dyspnea, and desaturation.

The chronic nature of this tumor and the ensuing cardiac symptoms from its hemodynamic consequences are indicated by the patient's high pulmonary arterial pressure and RV dysfunction. Patients with left atrial myxoma frequently experience pulmonary symptoms, which may be caused by restriction of mitral flow that mimics severe mitral stenosis. Nonetheless, tricuspid regurgitation, severe RV systolic dysfunction, and pulmonary hypertension are well-known side effects [2, 4, 7, 8, 19, 20]. Although the number of subjects with atrial myxomas with RV failure is unknown, there exist additional documented instances, which raises the possibility that it is an underappreciated side effect of left atrial myxomas [2, 4, 7, 8, 19, 21].

Since atrial myxomas are only clinically suspected in approximately 5% of myxoma patients, TTE is crucial in the diagnostic process [4, 7, 8, 20]. TTE also makes it possible to see hemodynamic compromise and late-stage cardiac alterations brought on by obstructive left atrial tumors, as was the situation with our patient. Early transthoracic echocardiography should be performed on anyone exhibiting heart failure symptoms. If a left atrial myxoma is found, it should be removed immediately to avoid difficulties in the later stages [4, 20].

An echocardiogram is a straightforward and non-invasive diagnostic method that graphically shows the tumor mass, circulation obstruction, and emboli's sources [1, 14, 22]. Because transesophageal echocardiograms have better spatial resolution sensors and less blocking tissue, they offer better imaging than transthoracic echocardiograms, which are less invasive [6, 14]. Additional diagnostic techniques include cardiac CT, cardiac MRI, and PET scans (distinguishing between myxoma and heart metastases) [6, 14]. Because of the possibility of embolism, transvenous biopsies are not advised [6, 14, 15].

As seen in our study, ECG may reveal findings like RVH, right axis deviation, right bundle branch block in case of pulmonary hypertension, ventricular premature contraction [7, 8], or QT prolongation in case of emboli [1].

However, 35% of participants have laboratory abnormalities, including anemia, elevated globulins, elevated erythrocyte sedimentation rate (ESR), and elevated C-reactive protein (CRP) [14–23]. Depending on the patient's posture, a "tumor-plop", or low-pitched diastolic sound immediately following S2, may be detected during a physical examination [14, 22]. In our investigation, renal parameters were raised. A mid-diastolic murmur, often referred to as a "tumor-plop", is a prevalent physical examination finding associated with large left-sided tumors, as the mass briefly impacts the mitral valve [7, 18].

Histologically, myxomas show dispersed, irregular cells within a myxoid stromal matrix composed of glycosaminoglycans and mucopolysaccharides [1, 14, 24]. Histological sections from our investigation revealed a tumor composed of many lipidic cells on a backdrop of myxoid regions with a few dilated arteries and sporadic inflammatory cells.

Prompt surgical resection is the mainstay of treatment for LA myxomas to reduce the risk of cardiac or embolic consequences [14, 15]. Although median sternotomies are the traditional resection method, there is a lower chance of infection or aesthetic concerns with more recent options, such as right anterolateral micro thoracotomies [15]. Recovery is quick, and the post-operative prognosis is generally favorable (less than 5% intraoperative death rate) [6, 14]. Arrhythmias, nodal conduction problems, infection, and tumor recurrence (5% of cases) are among the post-operative consequences [6, 14]. For recurring LA myxomas, a heart transplant is recommended [14]. Furthermore, in cases of acute pulmonary hypertension, hemodynamic stabilization may necessitate inotropic support. Due to their propensity to reoccur, serial imaging requires long-term follow-up [7, 18].

CONCLUSION

Left atrial myxomas, though benign, can lead to serious hemodynamic and embolic complications. Early diagnosis through echocardiography is essential, especially in patients with unexplained cardiac symptoms. Surgical resection remains the definitive treatment with excellent outcomes. Post-operative follow-up is vital due to recurrence risk. Awareness of RV dysfunction and systemic involvement is crucial in clinical evaluation.

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