Acute Heart Failure due to a giant left atrial myxoma: A Case Report

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Abstract

Background
Cardiac myxomas are the most common primary benign tumor of the heart. Most of them occur between the 4th and 6th decade of life, are most frequent in the woman, and most frequently localized in the left atrium.

Case summary
We present a case of a 41-year-old female who presented with a history of left-sided heart failure. A left atrial mass of 87 x 88 x 65 mm was documented by cardiac magnetic resonance. She was taken to surgical resection of the mass. Histopathologic findings were diagnostic of cardiac myxoma. Generally, myxomas that are bigger than 6 cm are associated with the worst prognosis.

Discussion
Primary cardiac tumors are mostly benign, being in 50% of the cases a cardiac myxoma. The rest of them correspond to papillary fibroelastoma (26%), fibromas (6%), lipomas (4%), and others including calcified tumors, hemangiomas, teratomas, cysts, and rhabdomyomas. Our clinical case illustrates an unusual and rare presentation of cardiac myxoma with a double mitral lesion.

Keywords: myxoma; cardiac tumors; cardiovascular imaging; cardiac surgery; histopathology; case report.
Funding

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Authors contributions:

Fuentes-Mendoza Juan Alan MD – original draft writing, acquisition of data, patient care;
Pimentel-Esparza Juan Andrés – patients care, contributed to the writing and editing of the manuscript; Cervantes-Nieto Jorge Antonio – patients care and follow-up.

Fuentes-Mendoza Juan Alan was involved in the care of the patient care; drafting and revision of the manuscript; Major role in acquisition of data.
Pimentel-Esparza Juan Andrés: was involved and contributed to the writing and editing of the manuscript. Also involved in the follow-up of the patient.
Cervantes-Nieto Jorge Antonio: was involved in the care of the patient care and follow-up; Major role in acquisition of data.

All authors have read and approved the final manuscript.
3. Learning points

Learning Points

1. Myxomas are the most common cardiac primary tumor, but the obstruction of the left ventricular inflow and/or outflow tract is not common and it could be dynamic.

2. It has rarely been described as a cause of acute heart failure.

3. The bigger the size might predict worst prognosis and outcome.

4. Multi-image approach is very important for establishing the differential diagnosis of intracardiac masses and evaluate the hemodynamic changes caused by the obstruction.
5. Surgery is the definitive treatment and periodic follow-up with echocardiogram should be implemented at least every year to detect recurrence.

4. Acknowledgments

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5. Organised Timeline for case report

Timeline

<table>
<thead>
<tr>
<th>Time</th>
<th>Events</th>
</tr>
</thead>
<tbody>
<tr>
<td>Day 0</td>
<td>First hospitalization due to history of one-year exertional dyspnea and atypical chest pain.</td>
</tr>
<tr>
<td>Day 1</td>
<td>Transthoracic echocardiography revealed a heterogeneous tumor of 7.5 x 8.3 cm in the left atrium.</td>
</tr>
<tr>
<td>Day 2</td>
<td>Cardiac MRI was performed and confirmed the left atrial mass was adhering to the interatrial septum of 87 x 88 x 65 mm.</td>
</tr>
<tr>
<td>Day 4</td>
<td>The patient underwent successful surgical intervention for resection of the mass by transseptal approach and conservative management of the mitral valve with annuloplasty due to severe mitral annulus dilatation.</td>
</tr>
<tr>
<td>Day 5</td>
<td>The patient continued post-surgical recovery at the Coronary Care Unit for cardiac monitoring.</td>
</tr>
<tr>
<td>Day 30</td>
<td>After symptoms improvement, the patient is discharged and ambulatory follow-up planned.</td>
</tr>
</tbody>
</table>
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Introduction

Cardiac myxomas account for nearly half of the primary cardiac tumors and are mostly located in the left atrium. Their size ranges from 1 to 15 cm and depending on the size, they can manifest with obstructive, embolic, or constitutional symptoms (1-2). We present a, relatively, rare case of a patient with an obstructive giant left atrial myxoma, who present one-year exertional dyspnea.

Case Presentation

A 41-year-old female was admitted to the emergency department due to two weeks of dyspnea at rest with a history of one year of exertional dyspnea and atypical chest pain. She denied syncope, palpitations, ankle edema, fever, cough, weight loss, malaise, fatigue, or other constitutional symptoms. She had a history of diabetes mellitus. Her baseline medications included metformin 850 mg daily. She denied other diseases or familiar history of heart disease or oncologic diseases.

At initial physical examination, she was found with hypotension, sinus tachycardia, jugular venous hypertension, apex displaced at 6th intercostals space, cardiac auscultation revealed intense second heart sound with an apical mild mid-diastolic murmur, intensified in left lateral decubitus position and irradiated to the anterior left axillary line, and a systolic mitral murmur at the supine position. No irradiation of the murmur was heard at another heart auscultation point.

Electrocardiogram (panel A) was in sinus rhythm with left atrial enlargement pattern, and chest radiography demonstrated pulmonary congestion, grade III cardiomegaly, and double contour image suggestive of left atrium dilatation (panel B). Laboratory results showed mild microcytic hypochromic anemia (Hb 11.6 g/dL), elevated CPR (C-reactive protein), and 2236 pg/dL NT-proBNP, rest of the blood tests were unremarkable. Transthoracic echocardiography (panel C1-C3) revealed a left ventricular ejection fraction of 48%, estimated systolic pulmonary pressure of 66 mm Hg, and a heterogeneous mass of 7.5 x 8.3 cm, which occupies the entire left atrium (LA
volume $176.2 \text{ ml/m}^3$) which was displacing interatrial septum, caused severe eccentric mitral secondary regurgitation due to annular dilatation and lack of leaflets coaptation with a Vena Contracta width of 7cm, and dynamic pseudo mitral stenosis. Right ventricle was not dilated, and function was preserved (TAPSE 21mm and Tricuspid annular systolic Velocity 10.3 cm/s). Cardiac magnetic resonance imaging (CMR) confirmed a giant left atrial mass adhered to the interatrial septum (87 x 88 x 65 mm), obstructing the mitral annulus and severe mitral regurgitation with mild pericardial effusion. She was taken to surgery for resection of the mass by transseptal approach and conservative management of mitral valve with annuloplasty due to mitral valve annulus dilatation, as planned previously in the Heart Team Session, and the intraoperative course was uneventful. Histopathologic findings showed lepidic cells without atypia, polyhedric cells, and pseudo-glandular structures surrounded by a myxoid extracellular matrix compatible with atrial myxoma. The patient continued post-surgical recovery at the Coronary Care Unit, where she evolved to recovery. The patient was discharged with symptomatic improvement after one month of admission, with a New York Heart Association (NYHA) functional class I, and scheduled outpatient follow-up post-hospitalization.

Discussion

Primary cardiac tumors are mostly benign tumors, in 50% of the cases a cardiac myxoma. The rest of them correspond to papillary fibroelastoma (26%), fibromas (6%), lipomas (4%), and others, including calcified tumors, hemangiomas, teratomas, cysts, and rhabdomyomas (3,4). Histopathologic findings of cardiac myxomas are stellar, ovoid of speculated cells with a myxoid content and vascular stroma. The size varies from 1 to 15cm. About 70% of all myxomas originate in the left atrium, 20% in the right atrium and 5% can be present in both (5).

Myxomas are more frequent in women between the 4th and 6th decade, with a ratio of woman/man of 2.0:1 respectively for left-sided myxomas and (6), a ratio of 0.75:1 for right-sided myxomas (6–8). About 7% of all cardiac myxomas occur secondary to a dominant autosomal manifestation associated with skin lesions and endocrine tumors, also known as the Carney complex. This syndrome is caused by a mutation in the regulatory subunit of an alpha kinase protein type 1 dependent on cAMP. This complex is usually present in younger patients; they are associated with multiple myxomas in atypical locations and risk recurrence after surgical treatment.
Benign tumors locally invasive can provoke alterations in cardiac contractility and valvular dysfunction, therefore, showing clinical findings of heart failure, fatal arrhythmias, and pericardial effusion. Pinede et al. described a lethal triad of complications: obstructive (67%), embolic (29%), and constitutional (34%) (9).

According to its location, left atrial myxomas are more frequent, situated classically on the border of the fossa ovalis in the interatrial septum or the mitral annulus. About 80% of patients manifest dyspnea, palpitations, syncope, ankle edema, and chest pain; acute heart failure in patients with atrial myxoma has rarely been described (10). A 30% of tumors in this location manifest some embolic event (6, 11).

Right atrial myxomas can be present in the cava vein and near the coronary sinus. They are mostly asymptomatic, but pulmonary embolism can be the initial presentation. Some authors like Medhat have reported myxomas of 7.7 x 5.5 x 3.7 cms (5). Parameswaran et al. (9) published a case with a myxoma of 11 cm weighing 105 grams. Most myxomas range from 1-6cm. Dang et al. described a mass of 15 x 6.6 x 3 mm (6). In our case, we report a total size of 16 cm in length, with a total weight of 125 grams. There is no threshold for the definition of giant myxoma.

CMR is the most reliable imaging technique for tissue characterization of cardiac masses. Myxomas usually appear as well-defined, smooth, oval, or lobular lesions that are commonly pedunculated and typically appear hyperintense compared to normal myocardium and hypointense compared with the blood pool, as we documented in our patient (12). However, the echocardiogram also continues to be an essential tool in the diagnosis of cardiac masses since, in addition to characterizing the image, it gives us information about the hemodynamic repercussion of the masses, as in our patient, where associated hemodynamic changes were secondary to the large size of the myxoma. Therefore, the multi-image approach in the characterization of cardiac masses has a fundamental role in diagnosing and treating this pathology.

The therapeutic management of cardiac myxoma, regardless of its size, is usually urgent surgical excision, typically curative. Performing an early resection at the diagnosis is often routine, funded by the known risks that favor intervention over no intervention (13). In the case of left atrial myxomas, be resected through the interatrial septum; however, in some cases, as it adheres to the interatrial septum, total resection of the fossa ovalis and reconstruction of the
septum must be performed (14). The surgical procedure for cardiac myxoma has a low rate of
postoperative complications, and the postoperative period is usually uneventful, presenting only
minor complications in most cases (15).

Conclusions
In conclusion, we describe a rare case of a giant left atrial myxoma that caused acute heart
failure due to its massive size, causing an obstruction that behaved as a severe mitral
regurgitation and pseudo-stenosis. That was successfully resolved with resection of the mass
and preservation of the mitral valve apparatus.

Disclosures
The author(s) declare(s) that there is no conflict of interest.

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Informed Consent
The author confirms that written consent for submission and publication of this case report
including images and associated text has been obtained from the patient in line with COPE
guidance.
References


Figure 1 Electrocardiogram, chest X-ray, and Transthoracic echocardiogram. Panel A: Electrocardiogram showing sinus rhythm with P wave negative in V1 suggestive of LA Dilatation (black arrows). Panel B: Chest X-ray showing grade III cardiomegaly, and double contour image (yellow arrow) suggestive of Left Atrium Dilatation. Panel C: Transthoracic echocardiogram. C1: long parasternal axis view showing a giant mass (yellow circle) in the whole left atrium. C2 and C3. Apical four-chamber view showing a giant mass obstructing all the LV inflow tract and causing a pseudo mitral stenosis.
Figure 2 Magnetic Resonance. T2 weighted localized (D1) Sagittal (D2) Coronal (D3) Axial images. In the left atrium a well-defined, smooth, oval lesion with a narrow pedicle attached to the interatrial septum of 87 x 88 x 65 mm. T2 weighted, (D4, D5) T2 weighted with fat saturation, (D6) first-pass perfusion, and (D7) late gadolinium enhancement (LGE) four-chamber sequences showing the typical appearance of a left atrial myxoma (LAM). The lesion shows limited enhancement on first-pass perfusion sequences and heterogeneous enhancement on LGE sequences which correspond with regions rich with myxomatous tissue and focal inflammation.
Figure 3 Surgical and histopathologic findings. Panel E: surgical resection. Panel F: macroscopic view of the mass, with a length of 16 cm x 7 cm, complete weight of 125 grams. Panel G: Hematoxylin-eosin 10X: G1: with lepidic cells without atypia, surrounded with a myxoid extracellular matrix. G2: Hematoxylin-eosin 40x: polyhedral cells forming perivascular hoses. G3: Hematoxylin-eosin 10x, pseudoglandular structures surrounded in a myxoid matrix with hemorrhage and hemosiderin