Acute heart failure due to a giant left atrial myxoma: a case report

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Background
Cardiac myxomas are the most common primary benign tumour 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 × 88 × 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 tumours 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 tumours, haemangiomas, teratomas, cysts, and rhabdomyomas. Our clinical case illustrates an unusual and rare presentation of cardiac myxoma with a double mitral lesion.

Keywords
Myxoma • Cardiac tumours • Cardiovascular imaging • Cardiac surgery • Histopathology • Case report

ESC Curriculum
6.4 Acute heart failure • 6.8 Cardiac tumours • 7.5 Cardiac surgery • 2.1 Imaging modalities

Learning points
• Myxomas are the most common cardiac primary tumour, but the obstruction of the left ventricular inflow and/or outflow tract is not common and it could be dynamic.
• It has rarely been described as a cause of acute heart failure.
• The bigger the size might predict worst prognosis and outcome.
• Multi-image approach is very important for establishing the differential diagnosis of intracardiac masses and evaluating the haemodynamic changes caused by the obstruction.
• Surgery is the definitive treatment and periodic follow-up with echocardiogram should be implemented at least every year to detect recurrence.

Introduction
Cardiac myxomas account for nearly half of the primary cardiac tumours and are mostly located in the left atrium (LA). 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 1-year exertional dyspnoea.
Timeline

| Time  | Events |
|-------|--------|
| Day 0 | First hospitalization due to history of 1-year exertional dyspnoea and atypical chest pain |
| Day 1 | Transthoracic echocardiography revealed a heterogeneous tumour of 7.5 × 8.3 cm in the LA |
| Day 2 | Cardiac MRI was performed and confirmed the left atrial mass was adhering to the interatrial septum of 87 × 88 × 65 mm |
| Day 4 | 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 |
| Day 5 | The patient continued post-surgical recovery at the Coronary Care Unit for cardiac monitoring |
| Day 30 | After symptoms improvement, the patient is discharged and ambulatory follow-up planned |

Case presentation

A 41-year-old female was admitted to the emergency department due to 2 weeks of dyspnoea at rest with a history of 1 year of exertional dyspnoea and atypical chest pain. She denied syncpe, palpitations, ankle oedema, 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 at 6th 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 (Figure 1A) was in sinus rhythm with left atrial enlargement pattern, and chest radiography demonstrated pulmonary congestion, Grade III cardiomegaly, and double contour image suggestive of LA dilatation (Figure 1B). Laboratory results showed mild microcytic hypochromic anaemia (Hb 11.6 g/dL), elevated C-reactive protein, and 2236 pg/dL NT-proBNP, rest of the blood tests were unremarkable. Transthoracic echocardiography (Figure 1C1–C3) revealed a left ventricular ejection fraction of 48%, estimated systolic pulmonary pressure of 66 mmHg, and a heterogeneous mass of 7.5 × 8.3 cm, which occupies the entire LA (LA volume 176.2 mL/m²), 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 7 cm, and dynamic pseudo-mitral stenosis. Right ventricle was not dilated, and function was preserved (TAPSE 21 mm and tricuspid annular systolic velocity 10.3 cm/s). Cardiac magnetic resonance imaging confirmed a giant left atrial mass adhered to the interatrial septum (87 × 88 × 65 mm), obstructing the mitral annulus and severe mitral

Figure 1  Electrocardiogram, chest X-ray, and transthoracic echocardiogram. (A) Electrocardiogram showing sinus rhythm with P wave negative in V1 suggestive of left atrium Dilatation (black arrows). (B) Chest X-ray showing Grade III cardiomegaly, and double contour image (arrow) suggestive of left atrium dilatation. (C) Transthoracic echocardiogram. C1: long parasternal axis view showing a giant mass (circle) in the whole left atrium. C2 and C3: apical four-chamber view showing a giant mass obstructing all the left ventricle inflow tract and causing a pseudo-mitral stenosis.
regurgitation with mild pericardial effusion (Figure 2). 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, polyhedral 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 1 month of admission, with a New York Heart Association functional Class II, and scheduled outpatient follow-up post-hospitalization (Figure 3).

Discussion

Primary cardiac tumours are mostly benign tumours, 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 tumours,
haemangiomas, teratomas, cysts, and rhabdomyomas. Histopathologic findings of cardiac myxomas are stellar, ovoid of speculated cells with a myxoid content, and vascular stroma. The size varies from 1 to 15 cm. About 70% of all myxomas originate in the LA, 20% in the right atrium, and 5% can be present in both.

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, a ratio of 0.75:1 for right-sided myxomas.

About 7% of all cardiac myxomas occur secondary to a dominant autosomal manifestation associated with skin lesions and endocrine tumours, 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 tumours 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%).

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 dyspnoea, palpitations, syncope, ankle oedema, and chest pain; acute heart failure in patients with atrial myxoma has rarely been described. A 30% of tumours in this location manifest some embolic event.

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 × 5.5 × 3.7 cm. Parameswaran et al. published a case with a myxoma of 11 cm weighing 105 g. Most myxomas range from 1 to 6 cm. Dang et al. described a mass of 15 × 6.6 × 3 mm. In our case, we report a total size of 16 cm in length, with a total weight of 125 g. There is no threshold for the definition of giant myxoma.

Cardiac magnetic resonance imaging 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 with normal myocardium and hypointense compared with the blood pool, as we documented in our patient. 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 haemodynamic repercussion of the masses, as in our patient, where associated haemodynamic 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 favour intervention over no intervention. 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. 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.

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.

Supplementary material

Supplementary material is available at European Heart Journal—Case Reports online.

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