

## An Unusually Heavy Heart

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### Key Words

Myxoma · Cardiac tumour · Echocardiogram · Neoplasm · Intra-atrial masses

### Abstract

A 47-year-old female presented with worsening insidious symptoms of breathlessness and leg swelling over the last 12 months. Whilst her ECG and chest X-ray were relatively unremarkable on admission, an echocardiogram demonstrated a large-sized left atrial myxoma. This tumour was resected by the cardiac surgeons the next day, with very good postoperative results. The ensuing discussion details the anatomy and physiology of myxomas, their epidemiology, clinical features, diagnostic investigations and management.

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### Introduction

Our 47-year-old patient presented to a hospital in Mauritius with breathlessness, intermittent palpitations and ankle swelling, which had been ongoing for the last 12 months. Her previous medical history revealed that she was previously fit and well. She took no medication and had no known drug allergy of note. There was no obvious family history and no relevant smoking, drug or alcohol history.

On examination, she had unremarkable vital signs, but suffered from right-sided heart failure with bilateral pitting oedema and a raised jugular venous pressure. Haematological and biochemical tests were as follows ([table 1](#)). A 12-lead resting ECG demonstrated T-wave inversion anterolaterally from V2 to V5, with the absence of chest pain ([fig. 1](#)). Chest X-ray (CXR) revealed an enlarged cardiac silhouette as shown below ([fig. 2](#)). In view of her deteriorating symptoms and above findings, she subsequently went on to have a transthoracic echocardiogram, which showed a large atrial myxoma in the left atrium ([fig. 3, fig. 4](#)).

Therefore, an urgent surgical resection of this intra-atrial mass was performed, prior to which our patient underwent a diagnostic coronary angiogram showing unobstructed coronaries (fig. 5, fig. 6).

The resection, occurring the following day, was uneventful with good postoperative results (fig. 7, fig. 8, fig. 9).

## Case Report

### *Anatomy*

A little more than half of the benign tumours resected in adults are found to be myxomas on tissue biopsy. A myxoma is an extremely rare intracavitary mass typically measuring 3–8 cm in diameter [1]. In the majority of cases, myxomas arise on the septal wall of the left atrium surrounding the foramen ovale. Less commonly, it is found in the right atrium or even in the ventricles. Multiple myxomas are familial and have a tendency to occur at a younger age than single myxomas; familial multiple myxomas are linked with a variety of skin and endocrine tumours. Familial myxomas are usually transmitted in an autosomal dominant fashion [1].

### *Pathology*

There are numerous types of myxomas, namely sessile or pedunculated, spherical or polypoid. They often have a jelly-like shiny surface with areas of haemorrhage or thrombosis [1]. The mobility of the tumour is associated with the degree of attachment to the interatrial septum and the dimension of the stalk [2]. Inflammatory debris, iron pigments, fibrotic tissue, calcified matter and bone material are frequently present. Although some hypothesize myxoma to be an organized thrombus, new molecular techniques tend to point towards a neoplastic process (table 2) [1, 2].

### *Epidemiology*

The incidence of primary cardiac tumours ranges from 0.002% in district general hospitals and up to 0.1% in larger tertiary teaching hospitals. Of these tumours, myxomas represent half [1].

### *Gender*

The female-to-male ratio of myxomas detected in the population is roughly estimated as 3:1, but female predominance is less strong in familial atrial myxomas [3].

### *Age*

Myxomas have been described in individuals ranging from 3 to 83 years. The average age for these predominantly atrial tumours is 56 years, whereas the mean age for familial cases is 25 years [2].

### *Clinical Presentation*

The clinical presentation can be acute, subacute or insidious over a few years and may initially suggest any type of cardiac, extracardiac or multisystemic involvement.

### *Cardiac Symptoms*

Intra-atrial myxomas can sometimes imitate valvular conditions. In the left atrium, myxomas that end up prolapsing across the mitral valve may suggest mitral disease and can

result in a multitude of symptoms including dyspnoea, cough, chest tightness and haemoptysis [1]. Febrile symptoms as well as lethargy may also be associated and may indicate an underlying endocarditis.

Right intra-atrial and intra-ventricular myxomas result in right-sided heart failure manifesting with hepatomegaly, peripheral oedema and ascites, and signs of tricuspid regurgitation, revealed by a new systolic murmur [1].

#### Thromboembolic Events

In about two thirds of intracavitary masses, a thromboembolic event is one of the presenting features. Systemic emboli stem from left-sided tumours. Cerebral emboli are relatively common, with seizure activity or cerebrovascular accidents manifesting. Emboli have also been reported to arise in renal, femoral and coronary arteries [1]. Finding neoplastic tissue consistent with a cardiac source in biopsy samples or resected thrombotic material is occasionally the first clue to the diagnosis of a myxoma. Right-sided myxomas can embolize to the pulmonary vasculature, causing infarction, pulmonary hypertension and cor pulmonale [1].

#### *Features and Physical Examination Findings: Myxoma*

Signs can include: general fatigue, arthralgia, raised jugular venous pressure with prominent 'a' wave, a loud first heart sound, early diastolic 'plop' and a pansystolic murmur arising from mitral regurgitation if the valve is damaged by tumour; in Carney complex, myxomas can also be associated with skin hyperpigmentation and endocrine hyperactivity [5].

#### *Relevant Investigations*

##### CXR

Although poorly specific, CXR may sometimes show a calcification which can be seen with myxomas.

##### Echocardiography

In the last decades, echocardiography had revolutionized the way that myxomas are diagnosed. It makes it possible to visualize the tumour and exclude differentials [1]. In particular, the M-mode in echocardiography is known to be reliable in left atrial myxomas. Both transthoracic and transoesophageal echocardiography can reveal even small tumours and provide information as to their size, attachment, location as well as whether these myxomas are mobile or not [1]. Furthermore, Doppler techniques enable the operator to assess valvular haemodynamics.

The transoesophageal method is better in recognizing and assessing right atrial myxomas as well as evaluating their mode of attachment. It also offers superior images of the great vessels; however, it is less useful in the case of intramural or ventricular tumours [1].

##### CT

CT scanning on the other hand is superb in assessing intramural attachment and for delineating margins and demonstrating invasion into extracardiac surroundings [1].

##### Cardiac MRI

MRI of the heart gives an accurate 3D resolution, enabling the precise localization of the myxoma with respect to the endocardium, epicardium and myocardium, and as an added advantage, it can also distinguish fat from thrombus or tumour [1].

### Management of Myxomas

Myxomas remain the most commonly resected cardiac tumour. Surgery involves the removal of a small layer of atrium surrounding the myxoma stalk, and this effectively prevents a recurrence in 95% of situations [1]. Other operative techniques used to reduce recurrence include laser coagulation at the base of the myxoma and a total resection of the fossa ovalis region, ensued by repair (frequently with a pericardial patch).

### References

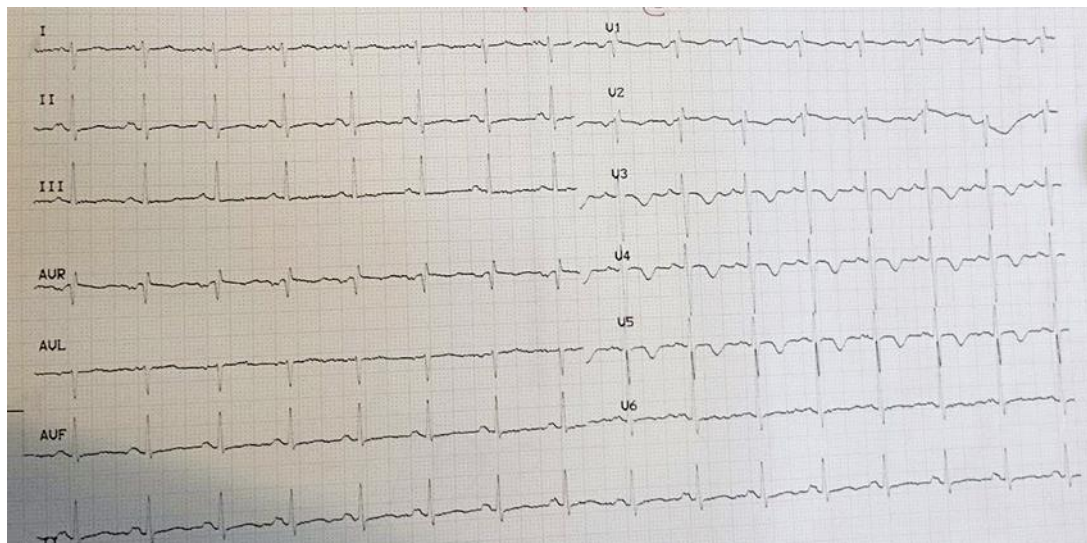
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**Table 1.** Haematological and biochemical serum results

Marker	Result
Haemoglobin, g/dl	11.5
White cell count, 10 <sup>9</sup> /l	5.51
Platelets, 10 <sup>9</sup> /l	244
Sodium, mmol/l	125
Potassium, mmol/l	3.3
Urea, mmol/l	2.3
Creatinine, mmol/l	84

**Table 2.** Classification of myxoma [4]

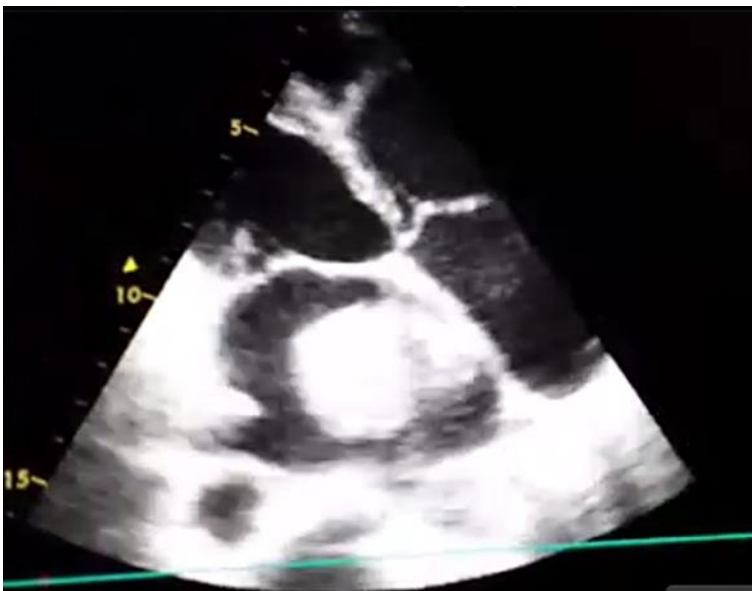
Myxoma	Margin	Vascular pattern	Cellularity	Stroma	Staining characteristics	Recurrence rate, %
Cutaneous	Multilobular, poorly/moderately defined	Thin-walled vessels	Spindle and stellate cells, some inflammatory cells	Profuse mucin with clefts	Vimentin	20–30
Intramuscular	Poorly defined; infiltrates muscle	Hypovascular/hypervascular variant	Spindle-shaped cells	Profuse mucin with cystic spaces	Vimentin	None
Juxta-articular	Poorly defined; infiltrates surrounding tissue	Focally vascular	Spindle cells with some atypical cells and mitoses	Profuse mucin with cystic spaces lined with fibrin or collagen	Vimentin	30–35
Aggressive angiomyxoma	Infiltrative	Evenly distributed blood vessels with frequently significant hyalinization	Round, spindle or stellate cells	Loose myxoid to collagenous	Vimentin	36–72
Superficial acral fibromyxoma	Infiltrative	Mild-to-moderate vasculature	Spindle and stellate cells, some mast cells	Myxoid to collagenous	CD34 EMA 20-30	Rare
Neurothekeoma	Well defined and multilobular	Hypovascular	Spindle-shaped cells	Bundles of cells interspersed with collagen	S-100, EMA	45–50



**Fig. 1.** Twelve-lead ECG on admission.



**Fig. 2.** CXR showing a cardiomegaly.



**Fig. 3.** Large left atrial mass seen on transthoracic echocardiogram.

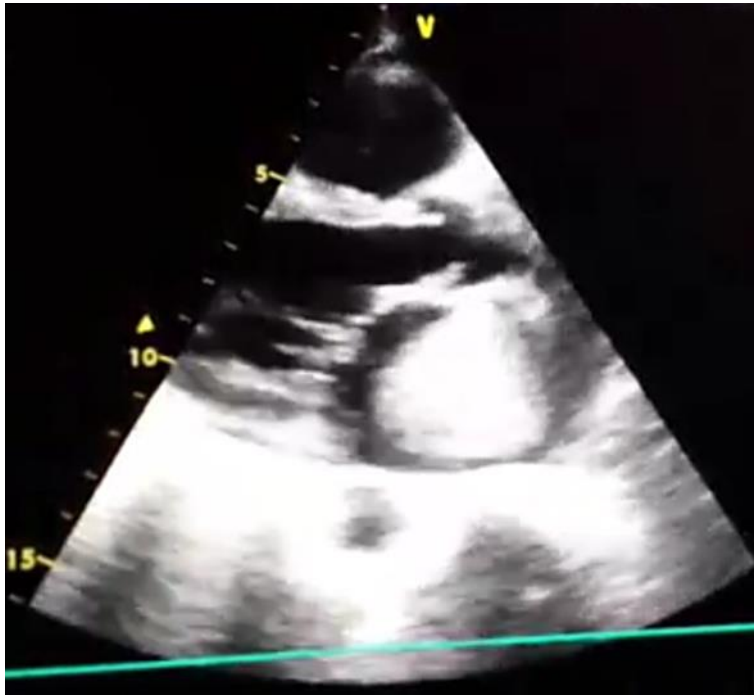


Fig. 4. Parasternal long axis view of the myxoma.

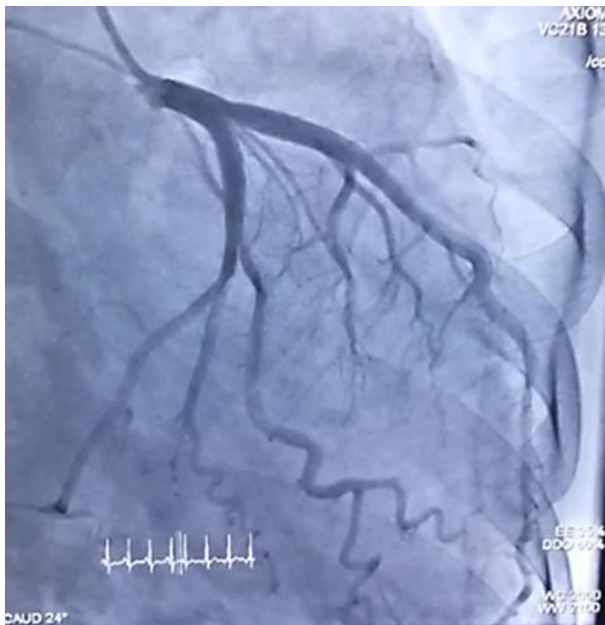


Fig. 5. Left coronary angiography demonstrating an unobstructed coronary tree.

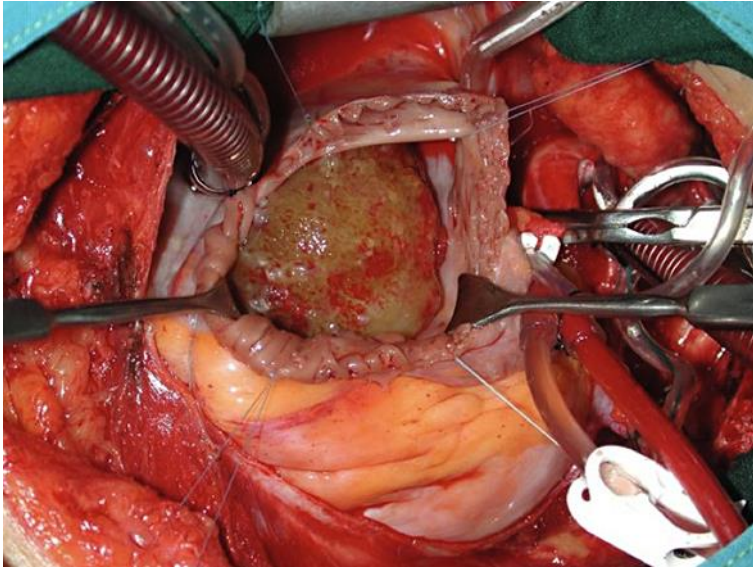


**Fig. 6.** Right coronary angiography demonstrating an unobstructed coronary tree.



**Fig. 7.** Intra-operative photo showing the resection of the tumour.





**Fig. 8.** Post-operative resected myxomatous tissue.



**Fig. 9.** Resected tumour with typical shiny gelatinous surface.