

# An exceptionally giant left atrial myxoma: a case report and literature review

Bassil Khalil Al-Zamkan<sup>1</sup>, Anas Mohamad Hashem <sup>2\*</sup>, Samir Ahmed Alaaeldin<sup>1</sup>, and Mohamed Abdel Aziz<sup>1</sup>

<sup>1</sup>Cardiac Center Sharjah, Al-Qassimi Hospital, Ministry of Health and Prevention (MOHAP), Sharjah, United Arab Emirates; and <sup>2</sup>Department of Gastroenterology and Hepatology, Mayo Clinic, 200 First Street SW, Rochester, MN 55905, USA

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## Background

Cardiac myxomas are considered the most common benign heart tumours. The clinical manifestations mainly depend on the size of the tumour. They usually vary from asymptomatic, mild non-specific symptoms, to severe obstructive cardiac and systemic findings. We describe herein a significantly large left atrial myxoma in a patient misdiagnosed with respiratory asthma.

## Case summary

A 54-year-old lady, was diagnosed previously with asthma, presented with a history of dyspnoea on exertion, palpitations, and mild peripheral oedema. Chest X-ray suggested pulmonary congestion. Due to high suspicion of cardiac issues, transthoracic echocardiography was done revealing giant left atrial mass. Consequently, the mass was approached and excised surgically through the inverted T biatrial incision. Grossly, the mass measured 10 × 8 × 6 cm, and it had a smooth surface and was filled with gelatinous material. The histopathology confirmed benign myxoma without malignant features.

## Discussion

Our article mainly focuses on the diagnostic challenges of a patient with atrial myxoma. The major discrepancy between the tumour size and the severity of the patient's symptoms should draw physicians' attention to consider atrial myxoma over a long list of differentials, in order to take immediate action to reduce the mortality and improve the overall prognosis.

## Keywords

Atrial myxoma • Heart neoplasm • Giant • Left atrium • Fossa ovalis • Case report

## Learning points

- Atrial myxoma accounts for half of the primary cardiac tumours. It might present with vague obstructive cardiac symptoms and be misdiagnosed as other cardiopulmonary conditions.
- In few cases, including ours, there may be a discrepancy between tumour size and symptom burden. The symptoms were mild in spite of having a very large myxoma making the diagnosis challenging.
- Transoesophageal echocardiography is the imaging modality of choice for cardiac myxomas.
- Prompt surgical resection is required to provide definitive treatment and reduce risk of further complications such as embolic and systemic manifestations.

\* Corresponding author. Tel: +1 (507) 271 0663, Email: [dr.anashashem@outlook.com](mailto:dr.anashashem@outlook.com)

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## Introduction

While the incidence of cardiac myxomas is estimated to be 8–150 cases per million, it accounts for nearly half of the primary cardiac tumours (PCTs). The majority of the cardiac myxomas arise in the left atrium (LA); they are usually pedunculated, attached to the endocardium by a stalk. The clinical features of an atrial myxoma varies based on its size, ranging from 1 to 15 cm, and comprises a triad of obstructive cardiac symptoms, embolic signs, and constitutional manifestations.<sup>1</sup> Immediate diagnosis of myxoma is essential to provide urgent surgical treatment. We describe herein an extremely rare case of a patient with a giant left atrial myxoma, who was misdiagnosed with asthma and complained of mild obstructive cardiac symptoms only.

## Timeline

Time	Event
Day 1	Patient presented with a history of exertional dyspnoea, bilateral pedal oedema, and dizziness when leaning forward. One year ago, she was diagnosed with asthma. Chest X-ray showed findings suggestive of pulmonary congestion and the patient was managed as a case of heart failure.
Day 2	Transthoracic echocardiography (TTE) was performed which showed a huge intra-atrial mass suggestive of atrial myxoma. She was booked for a preoperative transoesophageal echocardiography (TOE) and open heart surgery.
Day 3	Based on TOE, the patient underwent open heart surgery. The left atrial myxoma was resected and the interatrial septum defect was repaired. The patient was monitored closely after the operation in the intensive care unit.
Day 8	The patient improved dramatically without any complications, hence she was discharged.
Day 15	The patient was followed-up in the out-patient clinic and TTE showed improvement of her heart function.

## Case presentation

A 54-year-old woman presented to the emergency department with abdominal pain and bilateral lower limb swelling. She had a 1-year history of recurrent episodes of dyspnoea with dry cough and was managed as bronchial asthma. Despite appropriate treatment, her symptoms persisted. She also complained of dysphagia, palpitations without noticeable chest pain and feeling dizzy when leaning forward. On admission, she was haemodynamically stable, with normal blood pressure (120/78 mmHg) and pulse (72 beats/min). Examination revealed normal heart sounds, no murmurs, bilateral peripheral pitting oedema, and no other remarkable findings.

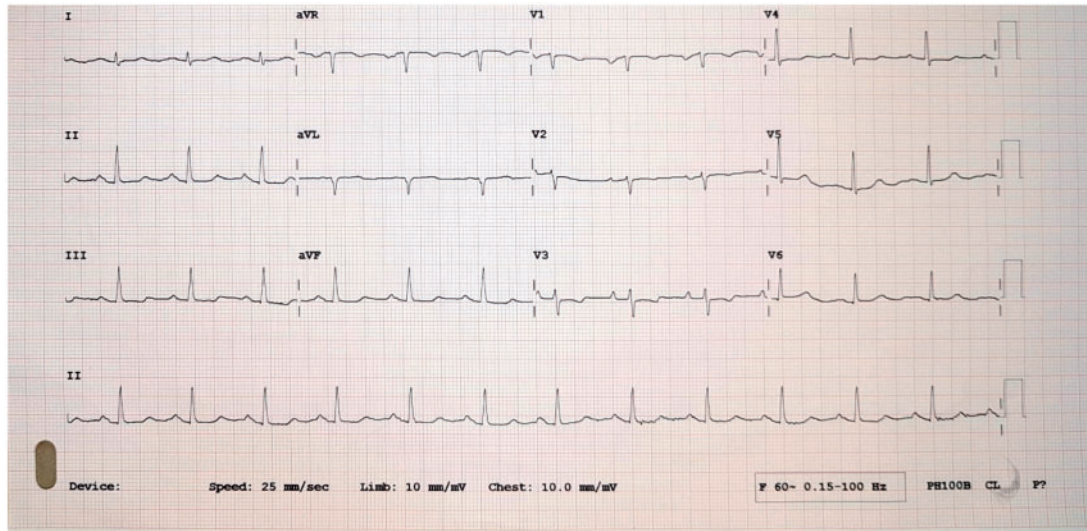
Electrocardiogram showed normal regular sinus rhythm, bifid P waves, and no ischaemic changes (Figure 1). Laboratory findings, including blood culture, were normal. Chest X-ray showed accentuated vascular markings bilaterally with diffuse reticular peripheral and basal lines, suggestive of pulmonary congestion (Figure 2). Consequently, a transthoracic echocardiography (TTE) was done which revealed a Grade III diastolic dysfunction with normal left ventricular systolic function. No regional wall motion abnormality was noted; however, moderate mitral and tricuspid regurgitations were observed on Doppler ultrasound. Additionally, it showed a large pedunculated mass (Video 1), approximately 9 × 7 cm attached to the upper third of the atrial septum, as well as severe pulmonary arterial hypertension (PAH) (84 mmHg) with a competent pulmonary valve.

Preoperative transoesophageal echocardiography (TOE) (Figure 3A, Video 2) was performed and confirmed the presence of a huge mobile mass filling the LA. It had a heterogenic core of multiple lucent areas and a smooth outer surface. On Doppler scan, a moderate degree of mitral and tricuspid regurgitation was evident by the presence of mosaic blood flow. Subsequently, a surgical excision was performed via conventional median sternotomy. In order to surgically approach such a huge tumour, it was decided to use the inverted T-shaped bi-atrial technique. A 3 cm incision was made in the LA parallel to the interatrial groove and anterior to the right superior pulmonary vein. Then, a second incision perpendicular to the first was made in the right atrium about 4 cm posterior to the interatrial groove towards the sulcus terminalis. The second incision met the first one near to its middle, forming an inverted T-shaped opening. At this point, the interatrial septum was divided towards the fossa ovalis. This revealed a pedunculated mass with a short stalk (about 1 cm in length), arising from the interatrial septum (IAS), at the fossa ovalis. The tumour, its stalk and part of the IAS were resected, followed by closure of the defect using a suture line without a pericardial patch. Due to presence of mitral regurgitation on TOE, two sutures were used opposite to the middle of the posterior mitral leaflet to reduce the size of the mitral ring, minimizing the leak (Figure 3B, Video 3). Since the tricuspid valve showed mild regurgitation and intact anatomy, no intervention was required.

Macroscopically, the specimen measured 10 × 8 × 6 cm (Figure 4A) and weighed 119 g (Figure 4B). The outer surface was smooth with punctate haemorrhage, whereas a cut surface showed a yellow-to-brown gelatinous material (Figure 4D). Microscopically, a low cellular myxoid growth with numerous cords, nests, and glandular pattern of stellate to spindle cells surrounded by eosinophilic to bluish matrix were observed. No mitotic figures or malignant features were seen. Additionally, there were cystic formations and vascular differentiation with occasional calcifications (Figure 5). Five days after the operation, the patient's condition improved uneventfully with complete resolution of her presenting symptoms. One week later, a follow-up echocardiography showed improvement of her heart function and normalization of PAH.

## Discussion

Primary cardiac tumours are very rare with an incidence of 0.0017–0.03%, whereas secondary cardiac tumours are 30 times more

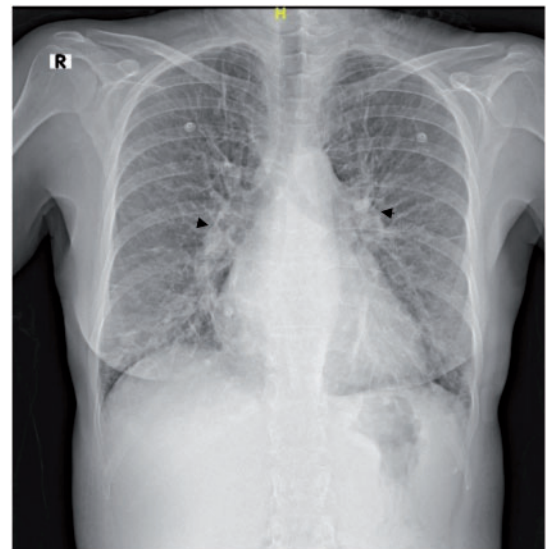


**Figure 1** Electrocardiogram, on admission, showing normal regular sinus rhythm, bifid p waves (P mitrale), and no ischaemic changes.

common when compared with PCT. Seventy-five percent of PCTs are benign, 50% of which are myxomas. The peak incidence of myxoma occurs amongst 50- to 60-year-old females. When compared with males, females are 2.05 times more likely to develop this condition.<sup>1-3</sup> The majority of cardiac myxomas (85%) are pedunculated and incorporated into the endocardium by a short stalk (<1 cm), while 15% are sessile.<sup>3</sup> Burke *et al.*,<sup>4</sup> who studied 107 cardiac myxoma cases found that 75% arise in the LA, 18% in the right atrium, 3% in either or both ventricles, and 1% on the valves.<sup>5</sup> When originating in the LA, 64.7% arise from the fossa ovalis border of IAS, 11.9% from the mitral annulus, 9.1% from the back wall of LA, and 8.1% at top of LA.<sup>6</sup>

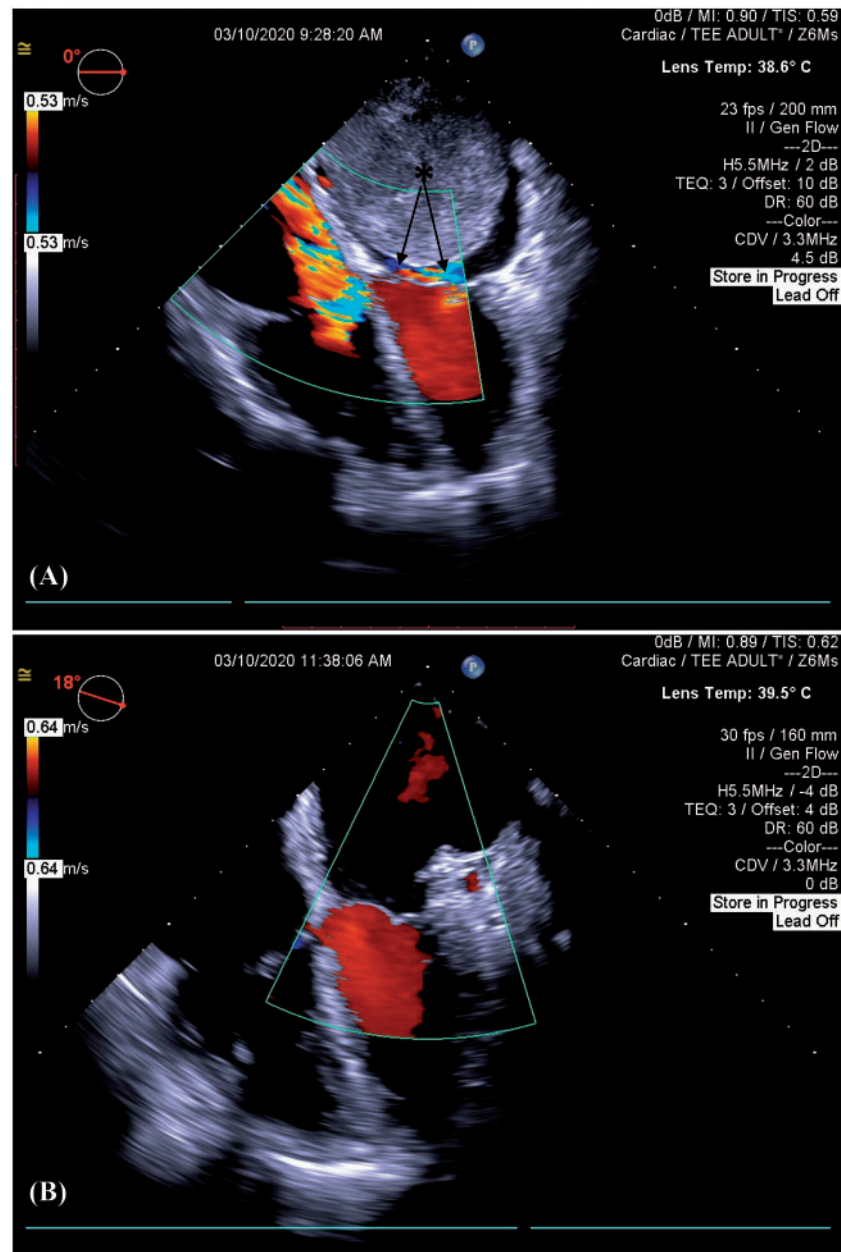
Clinically, atrial myxomas vary from asymptomatic, especially with small tumour size (<4 cm), to non-specific symptoms (exertional dyspnoea, lower limb and pulmonary oedema, angina, syncope, and palpitation), and constitutional symptoms (20–60%) such as fever, fatigue, and weight loss. In severe cases, symptoms might include systemic embolization (30–40%), obstructive symptoms (67%) and unexpected sudden cardiac death.<sup>7</sup> Pinede *et al.*<sup>1</sup> studied 112 cases of left atrial myxoma, examined the tumours macroscopically and correlated the size with the symptoms. The tumour diameter ranged from 1 to 15 cm, and weighed between 15 and 180 g, with a mean of 37 g. They discovered a statistically significant association ( $P=0.009$ ) between having a large atrial myxoma (>5 cm) and developing cardiac symptoms. On the other hand, no significant association was found between the size of myxoma and presence of embolic or systemic symptoms. Paradoxically, larger myxomas went undiagnosed for a longer period of time since the onset of symptoms ( $P=0.004$ ).

Diagnosis of atrial myxoma based on its presentation is challenging. The diagnostic modality of choice is echocardiography which aids in identifying any intra- or pericardial mass, its site, size and dimensions. Furthermore, it differentiates atrial myxoma from any thrombus or vegetation.<sup>8</sup> Transoesophageal echocardiography is considered to be superior to TTE. A study conducted by Shyu *et al.*<sup>9</sup> where 17 patients



**Figure 2** PA view chest X-ray showing bilaterally accentuated hilar vascular markings 'arrowhead', associated with diffuse reticular peripheral and basal lines (Kerley B lines), bilaterally.

were investigated for intracardiac tumour using both TTE and TOE, 14 of whom underwent cardiac surgery, found that TTE had 4 false-positives and 2 false-negatives when compared with TOE which had only one false-positive and no false-negatives. They emphasized the inadequacy of TTE for detecting small masses and the possible superiority of TOE including better visualization of the mass location, attachment, and dimensions. Other potentially useful diagnostic imaging modalities include computed tomography scan and magnetic resonance imaging. Their main advantage over echocardiography is their accuracy in visualizing soft tissue with all internal details of the



**Figure 3** (A) A midoesophageal four-chamber view of the transesophageal echocardiography showing a huge left echogenic mass filling the left atrium (asterisk) with a moderate degree of mitral and tricuspid regurgitation evident by the presence of mosaic blood flow (arrow). (B) Postoperative transesophageal echocardiography midoesophageal four-chambers view revealing minimal systolic mitral regurgitation. 'P' an indicator of the probe site.

myxoma, along with identifying the tumour attachment and the dimensions of its stalk.<sup>10</sup> Histopathological examination is considered the definitive confirmatory test for atrial myxomas.<sup>11</sup>

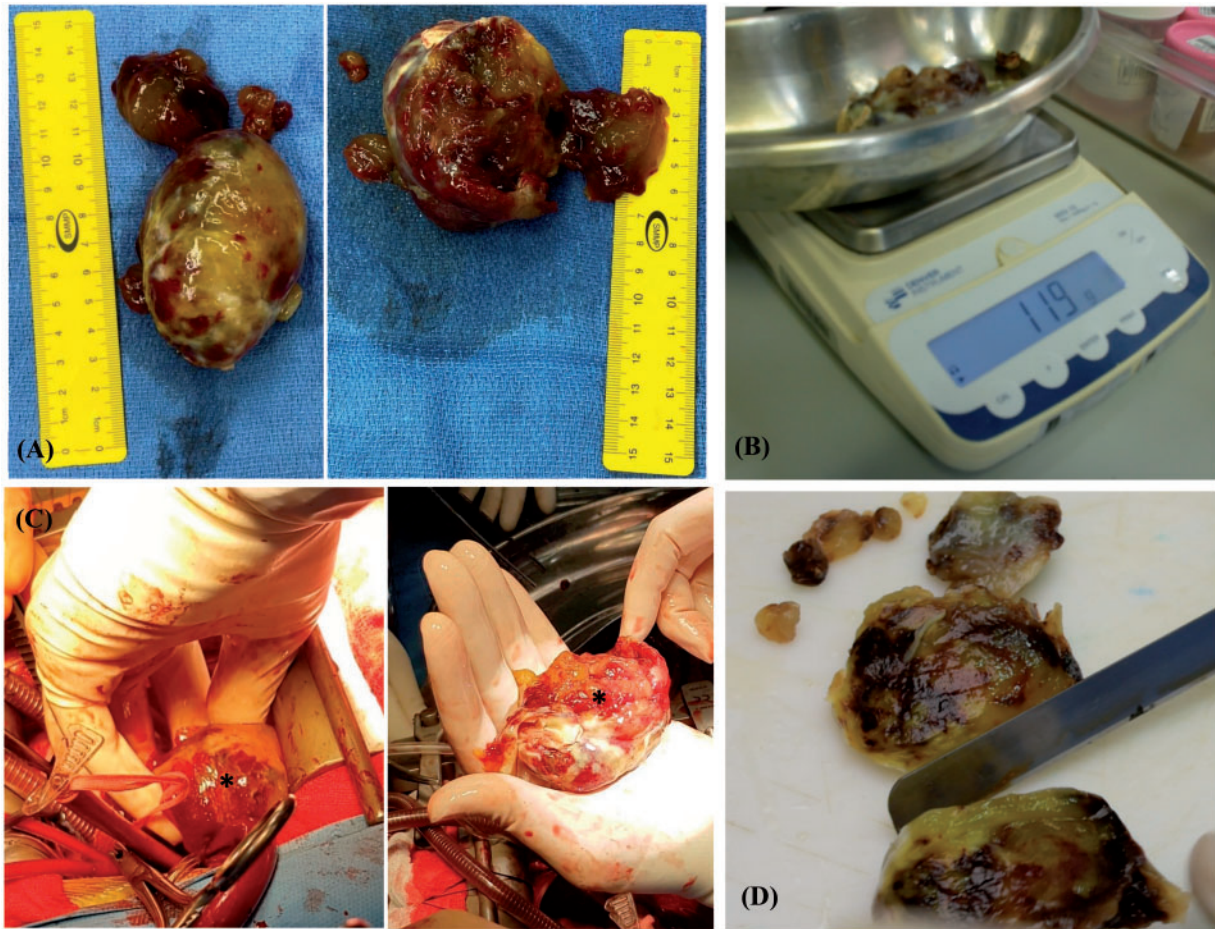
Following the diagnosis, immediate surgical intervention should be done promptly, to prevent any further complications.<sup>12</sup> Accessing LA alone is not sufficient to inspect all four chambers of the heart, especially in events of large myxomas, which limits adequate mass manipulation and radical resection.<sup>13</sup> The best technique to excise left atrial myxomas is known as inverted T-shaped biatrial incision, which provides an optimal exposure of all

four chambers enabling radical excision and sufficient surgical correction of the mitral valve incompetency, if present.<sup>14</sup> The mortality rate from the surgical intervention is low, and the long-term prognosis is satisfactory.<sup>15</sup>

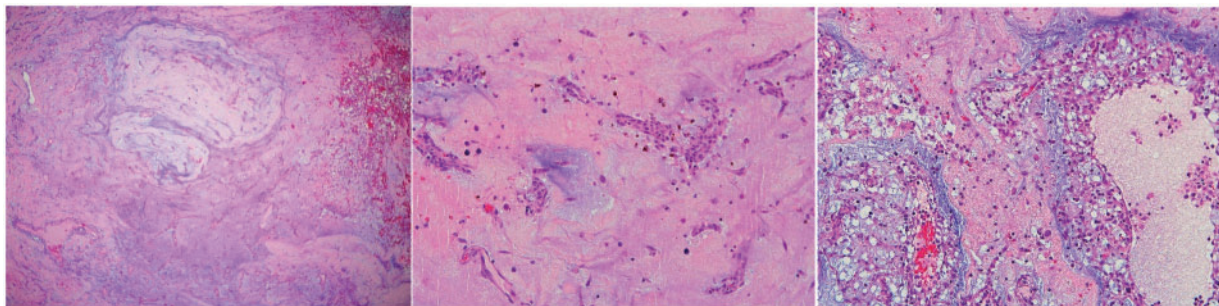
## Conclusion

To conclude, we described a case with an unusually giant atrial myxoma. It continues to be a significant challenge for emergency

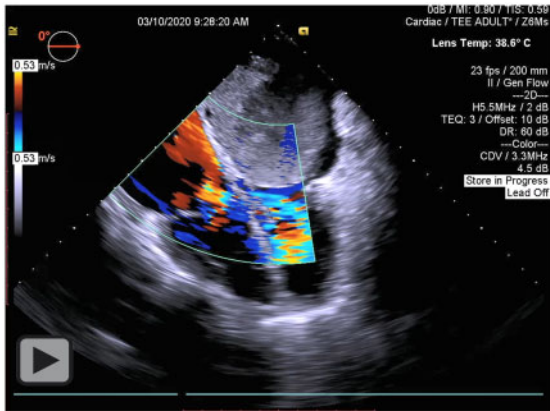




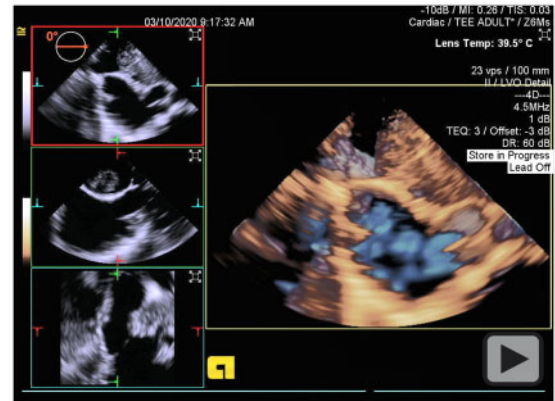
**Figure 4** (A) As shown in the picture, the resected specimen measuring  $10 \times 8 \times 6$  cm. The outer surface is smooth and covered with punctate haemorrhage. (B) On a digital weighing scale, the mass weighs 119 g. (C) Intraoperative photographs revealing a huge left atrial tumour mass (asterisk). (D) A cut section of the specimen showing a yellow–brown gelatinous and haemorrhagic areas.



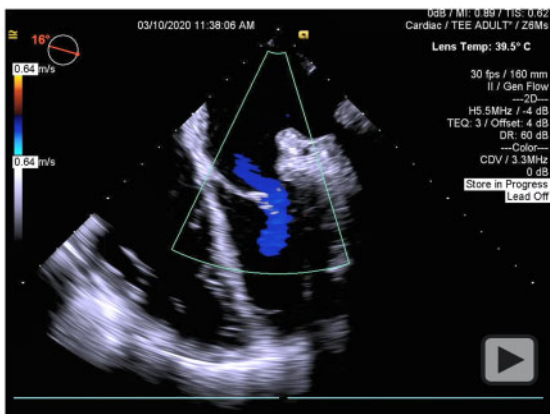
**Figure 5** A histopathologic view of haematoxylin and eosin stained sections confirms the presence of low cellular myxoid growth with numerous cords, nests, rings, and glandular pattern stellate to spindled cells that have a pink cytoplasm surrounding a small round benign nucleus. No nucleoli or mitotic figure are seen. Background rich in small-sized capillaries with occasional hemosiderin pigments suggests stromal haemorrhage. Multiple cystic formations with vascular differentiation along with calcifications are also seen.



**Video 2** 3D cine view of the preoperative TOE showing the atrial mass attached by a stalk to the interatrial septum.



**Video 1** Cine four-chamber view of the preoperative TOE showing a huge left echogenic mass filling the left atrium with a moderate degree of mitral and tricuspid regurgitation evident by the presence of mosaic pattern of blood flow during systole.



**Video 3** Cine four-chamber view of the postoperative TOE revealing minimal systolic mitral regurgitation

physicians due to its non-specific symptoms and overlap with other conditions. The discrepancy between mild symptoms and the serious consequences the condition carries, necessitates high index of suspicion and thorough investigation to timely diagnose it among a long list of differentials.

## Lead author biography



Dr Basil Khalil Al-Zamkan received the German Board as a Cardiac Surgeon in 1998, and Thoracic Surgeon in 2007, Heinrich Heine University, Düsseldorf, Germany. He worked as a Senior Consultant for Cardiovascular and Thoracic Surgery (1997–2009) at Clinic of the Klinikum Krefeld. He published articles in international journals and

has membership in the German Society for Cardiovascular and Thoracic Surgery, European Society of Thoracic Surgery, European Society of Cardiovascular and Thoracic Surgery, and Emirates Cardiac Society. Currently, he is working as a senior consultant of cardiovascular and thoracic surgery at Al-Qassimi Hospital, UAE.

## Supplementary material

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

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**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

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