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CASE REPORT
Coronary heart disease

Intraoperative embolism of a right atrial myxoma: a case report

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Background Atrial myxomas are the most common benign cardiac tumours. Clinical manifestations vary from constitutional symptoms, to valvular stenosis and embolic events, and surgical removal is the only suggested treatment.

Case summary A 50-year-old female patient was referred to our centre for surgical excision of a reported right atrial mass. A transoesophageal echocardiographic exam revealed two right atrial masses and the surgical plan was total removal of both masses. However, during the operation the surgeons were not able to locate the larger of the two masses and embolization to the pulmonary trunk was considered as the most likely explanation in this setting. A control suction of the right ventricle and the proximal part of the pulmonary arteries was performed to check if the mass had embolized distally but this did not yield any mass. The patient was transferred to the intensive care unit and remained stable for 2 h, until she developed an abrupt cardiogenic shock with signs of right heart failure. An emergent pulmonary computed tomography angiography demonstrated the embolized mass to the left pulmonary artery and the patient was retransferred to the operating room for emergent surgical removal of the mass. The patient showed immediate clinical and haemodynamic improvement after the removal of this mass and had an uneventful further hospitalization.

Discussion Multiple right atrial myxomas are rarely reported and surgical excision requires experience, as in case of embolization immediate removal must be obtained to prevent from right ventricle distress and cardiogenic shock.

Keywords Cardiac tumours • Right atrial myxoma • Pulmonary embolism • Case report

Learning points

- Cardiac surgeons should be alert for pulmonary embolism during a right atrial myxoma surgery. Careful and precise cannulation of the vena cavae and incision of the right atrium are the keys to success with minimal complication risk.
- Embolization of myxoma into the pulmonary artery system can result in acute right heart failure and cardiogenic shock. Multimodality imaging plays a facilitating role in the diagnosis and further surgical management of this potentially fatal complication.

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Introduction

Myxomas are the most common benign cardiac tumours, mainly situated in the left atrium.[1] Rarely, they can be found in the right atrium or in the ventricles and occasionally multiple myxomas may be present in cardiac chambers.[2] Clinical presentation varies from constitutional symptoms to valvular manifestations and embolic events.[3,4] Echocardiography is the first line method for the diagnosis,[5] and surgical removal is the solely suggested treatment. In this case report, we present a patient with two right atrial myxomas, one of which embolized to the pulmonary artery (PA) trunk intra-operatively, causing acute respiratory failure and cardiogenic shock.

Timeline

Day 0	
10:00	Admission to our clinic for evaluation and management of a right atrial mass
12:00	Baseline transoesophageal echocardiography (TOE) that confirms the presence of two right atrial, myxoma-like masses. Stable condition of the patient.
Day 1	
14:30	Patient transferred to the operation room for surgical removal of the masses.
15:39	Baseline intraoperative TOE that demonstrated two right atrial masses.
16:00	Cannulation and incision of the right atrium revealing the smaller mass only. Surgical removal of this mass.
16:28	Repeated TOE that confirmed the absence of the second larger atrial mass.
17:00	Suction of the right ventricle and the main pulmonary artery (PA).
19:00	Transfer of the patient to the intensive care unit (ICU) in a haemodynamically stable condition.
21:00	Clinical and haemodynamic deterioration, cardiogenic shock requiring massive doses of inotropes and high-flow oxygenation.
23:30	Emergent pulmonary computed tomography angiogram showing left PA occluded by the second mass.
Day 2	
01:00	Patient retransferred to the operation room. The second mass is removed after incision of the main PA.
02:00	Patient transferred back to the ICU in a stable clinical condition.
Day 3	
09:00	Stable condition of the patient.

Case presentation

A 50-year-old female patient was referred to our hospital for further evaluation and management of an incidentally reported right atrial mass. The patient had a history of ankylosing spondylitis under treatment with methylprednisolone and methotrexate, and factor V-Leiden mutation that had lead to several peripheral venous and arterial thrombosis during the last 5 years. The patient also experienced multiple episodes of pulmonary embolism that had been treated with anticoagulants. Although under warfarin treatment with a good time in therapeutic range of international normalized ratio, the patient developed recurrent pulmonary embolism which was later attributed to the presence of a newly diagnosed right atrial mass. The patient was suffering from heart failure symptoms with New York Heart Association class III–IV status due to chronic thromboembolism resulting in significant pulmonary hypertension (systolic pulmonary pressure = 55 mmHg). A new transoesophageal examination was performed at admission and it revealed two pedicled, mobile, 'myxoma-like' masses in the right atrium, one larger (21 × 20 mm) attached to the free wall and one smaller (12 × 4 mm) attached to the mid portion of interatrial septum (*Figure 1A*, *B*, *D*). These masses did not cause any extra symptoms and despite partial occlusion of the tricuspid valve by the larger mass, no functional stenosis was observed with a mean gradient of less than 1 mmHg on the valve. In order to avoid any further mass-originated embolic events and any further mass-related complications, surgical removal was suggested and the operation was scheduled within the same admission.

At her admission, the physical examination revealed normal first (S1) and second heart sound (S2), with no murmur; normal findings from the auscultation of the lungs and bilateral pedal oedema.

The operation was performed under general anaesthesia with a median sternotomy and cannulation of the ascending aorta, inferior and superior vena cava. The right atrium was vertically opened and surprisingly revealed the presence of only the smaller mass attached to the interatrial septum, which was removed along with its pedicle. Despite careful and meticulous visual and manual search of the right atrium, right ventricle (RV) and main PA, the larger mass could not be identified. A repeated intraoperative transoesophageal echocardiography (TOE) examination confirmed the absence of the second larger mass and as it was considered to have embolized into the pulmonary arteries. Persistent suction was applied into the right ventricle between the trabeculae and into the main PA in an attempt to capture and remove the mass. Lastly, the right atrium was closed and cardiopulmonary bypass was discontinued for clinical and hemodynamic evaluation of the patient. Since the patient was stable with satisfied oxygenation without any need of inotropic support, she was transferred to the intensive care unit (ICU). Unfortunately, two hours later she developed cardiogenic shock, with severe hypoxaemia and a bedside transthoracic echocardiography demonstrated signs of acute right ventricular failure and increased systolic PA pressure. As these findings were in keeping with acute pulmonary embolism, an emergent pulmonary computed tomography (CT) angiogram was performed and it revealed a large mass occluding the left PA (*Figure 1C*). The patient was retransferred to the operation room under massive inotropic support for emergent surgical embolectomy. After CBP, the main PA was incised longitudinally across its entire length and a 21 × 20 mm mass was removed from the initial part of the left PA using LeGardin graspers and suctioning. Remarkably, the RV showed a significant, dramatic recovery and the patient demonstrated immediate hemodynamic improvement allowing the surgeons to transfer her back to the ICU. After this intervention, the patient

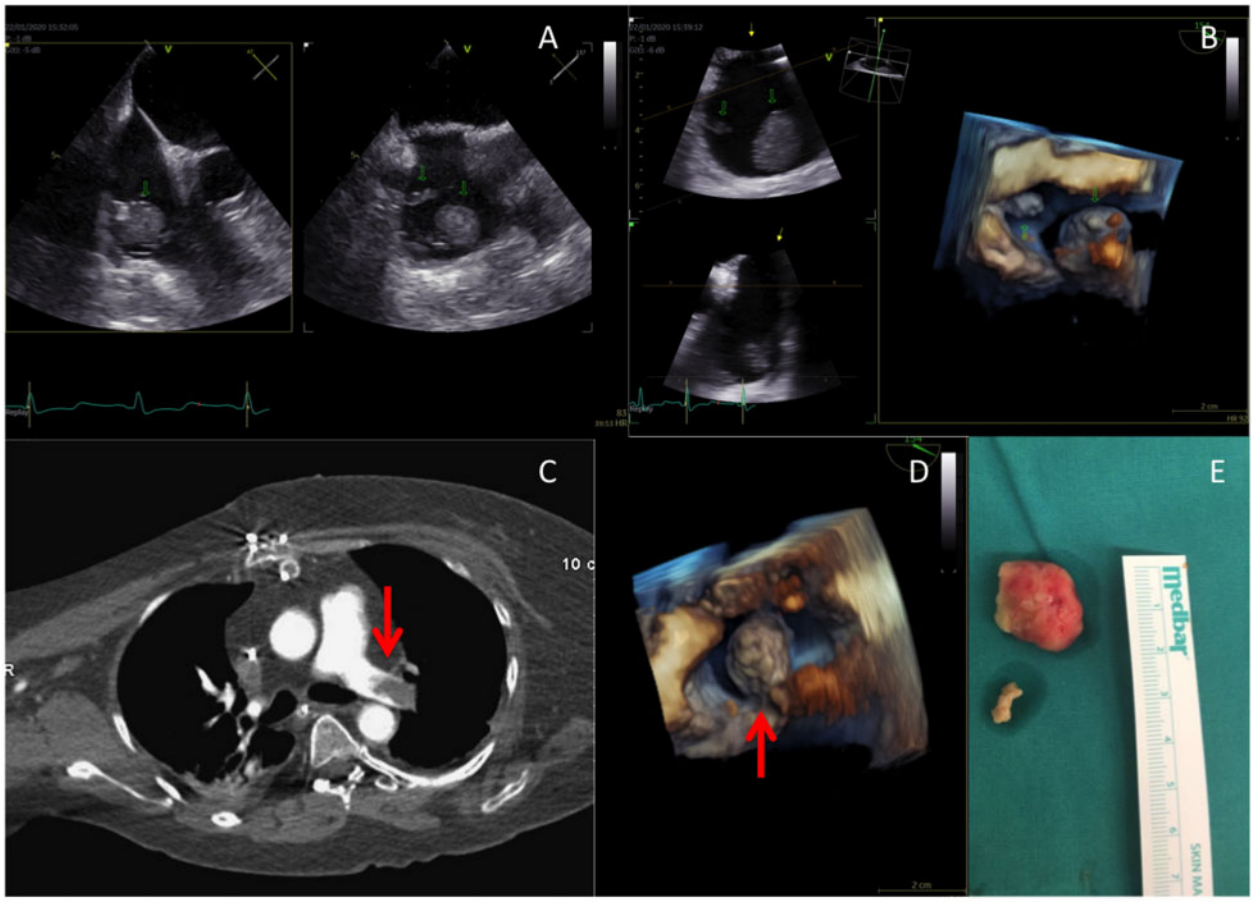


Figure 1 (*A*) Biplane views of both right atrial masses. (*B*) Four-dimensional views of both right atrial masses. (*C*) Contrast-enhanced computed tomography pulmonary angiography scan showing the embolized myxoma into the left pulmonary artery (arrow). (*D*) Four-dimensional views of the larger right atrial mass attached to the free wall. (*E*) Post-operative measurement of both right atrial masses.

had an uneventful further hospitalization period without extra oxygen and inotropic support need, improvement of her symptoms and the pedal oedema by the physical examination. Histopathologic examination of these two masses confirmed the diagnosis of multiple cardiac myxomas (*Figure 1E*).

Discussion

Primary cardiac tumours are rare, with an autopsy prevalence of 1 in 2000.[6] Benign cardiac tumours account for 75% of all, and myxomas are the most common type among them.[1] Myxomas consist of an acid-mucopolysaccharide stroma with mucoid extracellular matrix and they are named by this gelatinous, mucoid nature. Approximately 75% of them appear in the left atrium, attached to the fossa ovalis of the septum, whereas 20% are located in the right atrium and only 5% in ventricles or in multiple positions.[2] The clinical manifestations vary from general symptoms to valvular symptoms due to obstruction or embolic events. General symptoms include cachexia, dizziness, fatigue, fever, and joint-muscular pains,[3] valvular symptoms may mimic stenosis of the atrioventricular valves and embolic events may lead to stroke, myocardial infarction or peripheral embolization.

Right atrial myxomas may further present with right heart failure symptoms like pedal oedema, ascites and hepatomegaly due to functional tricuspid stenosis[7] or can cause pulmonary embolic events[4] as in our case. The first line imaging method of choice for diagnosis is echocardiography with sensitivity that can reach up to 100% if transoesophageal echo is used.[5]

The only treatment for right atrial myxomas is surgical removal with recurrence rate ranging from 2% to 3% in sporadic cases to 22% in patients with Carney complex.[8] The challenging aspect of this operation is the correct bicaval cannulation and the precise excision of the mass with a wide part of normal tissue in order to hinder embolization of the mass to the pulmonary trunk.[9] Myxomas attached to the free wall of the right atrium are more prone to embolize during the cannulation or at the time the surgeon opens the right atrium. When low or high-lying tumour pedicles preclude safe atrial cannulation, the distal part of the superior vena cava or the femoral or jugular vein can be used for cannulation. The right atrium should be opened widely for the resection and then be reconstructed again. In our case, both vena cavae were distally cannulated but when the free wall of

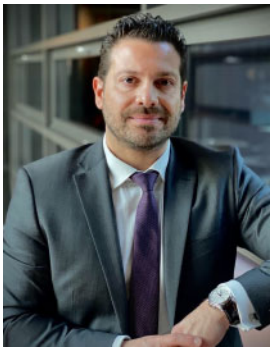
the right atrium was opened, no mass was detected. That way we assume that the pedicle of the larger myxoma was cut during the opening the right atrium, which caused the embolism towards the pulmonary trunk. Intraoperative transoesophageal echo plays a crucial role in the optimal planning of the operation and the continuous assessment during the operation in case of an unexpected haemodynamic deterioration.

Multiple right atrial myxomas have previously been reported in the literature[10] and pulmonary embolism seems to be a common manifestation accounting for 30–40% of all symptoms.[11–13] In patients where multiple myxomas are present, surgeons should be able to confirm the resection of all of them by direct observation of the right atrium comparing with the preoperative TOE images. Confirmation utilizing TOE at the end of the operation is of utmost importance. Combination of suction between the trabeculae of the right ventricle and the main pulmonary arteries and direct visualization and removal of the embolized mass is the only intraoperative therapeutic approach for the patient to prevent a bad outcome. In case of nonability to find the total number of demonstrated masses, pulmonary CT angiogram must be performed immediately in order to demonstrate the potentially embolized mass and plan a redo operation for a definitive, curative surgical removal with good long-term survival.

Conclusion

Right atrial myxomas may be complicated with pulmonary embolism even intraoperatively. Careful planning of these procedures as well as early diagnosis and surgical management of intraoperative complications such as embolization, are of critical importance for a satisfactory final result with low rates of mortality and morbidity.

Lead author biography



Dr Konstantinos Papadopoulos is a consultant cardiologist, head of Echocardiography lab, specialized in advanced cardiac echo techniques (TOE, 3D, Stress Echo, Speckle tracking). He is experienced in guiding structural heart diseases and transcatheter heart valves, especially MitraClip and PASCAL implantation. He is a fellow of the EACVI (FEACVI), member of the WG of valvulopathies and has an active role as a researcher in several trials (RESHAPE-HF2, EVOLUT-R FORWARD, INTREPID, POSE, SPACER Trial, CLASP, CLASP-P5). He has just completed his PhD on MitraClip and deformation imaging and has several pubmed publications.

Supplementary material

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

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.

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