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CASE REPORT
Cardiovascular imaging

Case report of rare highly aggressive cardiac tumour: the intimal sarcoma

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Background Primary cardiac tumours are rare, and diagnosis may be difficult, as symptoms and cardiac imaging may mimic other cardiac diseases. The intimal sarcoma is the least commonly reported cardiac tumour with only few cases reported worldwide. In this case report, we present a case of an intimal sarcoma with a highly aggressive disease course.

Case summary A 60-year-old male with a history of prior aortoplasty due to congenital aortic stenosis, mechanical aortic valve replacement, and aortic stenting due to aortic dilatation presented with night sweats, malaise, and dyspnoea. Initial imaging (including transthoracic and transoesophageal echocardiography and emergency computed tomography) revealed masses suspected to be thrombi in the left atrium and ventricle. However, a positron emission tomography/computed tomography scan revealed that the masses were suspicious for malignancy. The patient underwent non-radical tumour resection and insertion of biological valve prostheses. Subsequent tissue analysis and pathology assessment revealed an intimal sarcoma. There were no curative treatment options, and the patient succumbed to his illness <3 months after surgery.

Discussion This case report presents a case of a highly aggressive intimal sarcoma. As complete tumour resection is of great importance when it comes to life expectancy in cardiac sarcomas, early diagnosis using non-invasive and invasive imaging modalities is essential to start early treatment and to improve outcomes in this patient group.

Keywords Intimal sarcoma • Cardiac tumour • Cardiac imaging • Case report • Rare • Aggressive

ESC Curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 2.4 Cardiac computed tomography • 6.8 Cardiac tumours • 7.5 Cardiac surgery

Learning points

- To be able to make an early diagnosis of cardiac tumours using non-invasive and invasive multimodality imaging.
- To recognize how aggressive the disease course of rare cardiac tumours can potentially be.

Introduction

Primary cardiac tumours are rare,[1] and diagnosis may be difficult, as symptoms and cardiac imaging may mimic other cardiac diseases. Complete tumour resection is important with regards to life expectancy, and it is therefore essential that physicians can make an early diagnosis using multimodality imaging and mass resection with subsequent pathology assessment. Here, we present a challenging case of the least commonly reported primary cardiac tumour, the intimal sarcoma,[1] with a highly aggressive disease course.

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Timeline

Time	Event
Initial presentation	Night sweats and malaise throughout a month, increasing exertional dyspnoea throughout a week, and fluctuating International Normalized Ratio levels for the past 5 weeks.
Day 1	Admitted to Department of Cardiology for physical examination, laboratory analyses, electrocardiogram, chest X-ray, transthoracic echocardiogram (TTE), transoesophageal echocardiogram, and emergency computed tomography (CT). A mass was found in the left atrium and left ventricle, but the nature of the mass was still unclear. The radiodensity of the mass was 50 Hounsfield units, indicating a thrombus.
Day 2	Positron emission tomography (PET)/CT revealed masses suspicious for malignancy (due to strong tissue metabolic activity) in the left atrium, left ventricle, and septum.
Day 3	Surgical treatment with non-radical tumour resection and patch implantation in the left atrium and left ventricle, bioprosthetic mitral valve replacement, and a freestyle bioprosthesis for the aortic root. The removed tumour mass was sent for pathology assessment. The surgery was complicated by pulmonary oedema, and the patient was put on veno-venous extracorporeal membrane oxygenation (ECMO).
Day 5	The patient came off ECMO.
Day 7	The patient was extubated.
Day 9	The patient was moved out of the intensive care unit.
Day 12	Postoperative transthoracic echocardiography showed well-functioning bioprostheses, no visible residual tumour masses, and a normal left ventricular ejection fraction.
Day 27	Final pathology assessment revealed an intimal sarcoma with non-radically resected tumour borders.
Day 37	It was concluded at a multidisciplinary conference that there were no further surgical or medical curative treatment options.
Afterwards	The patient was discharged and started palliative treatment. He developed dyspnoea, lost his appetite, experienced weight loss, and succumbed to his illness two and a half months after the surgery.

Case presentation

A 60-year-old male, physically active and part of workforce, sought medical attention due to night sweats and malaise throughout a month as well as increasing exertional dyspnoea throughout a week. No weight loss was reported. The patient had a history of prior aortoplasty due to congenital aortic stenosis (Age 17), mechanical aortic valve replacement (Age 35), and aortic stenting due to aortic dilatation (Age 58). He was taking Losartan and Marevan, and his self-monitored International Normalized Ratio levels had been fluctuating for the past 5 weeks with several measurements below 2 and above 5. Physical examination revealed dyspnoea while talking, a blood pressure of 92/66 mmHg, a holosystolic murmur in the second intercostal space on the left side of the sternum, bibasal pulmonary crackles, and mild peripheral oedemas.

Laboratory results showed a haemoglobin of 9.2 g/dL (normal range 13.4–16.9), lactate dehydrogenase of 260 U/L (normal range 105–205), and C-reactive protein of 129 mg/L (normal range <10). The electrocardiogram (see Supplementary material online, *Figure S1*) showed normal sinus rhythm, left axis deviation, and poor R-wave progression. Chest X-ray (see Supplementary material online, *Figure S2*) showed pulmonary congestion, Kerley B lines, and bilateral pleural effusion. A TTE (*Figure 1A*) showed a functional mitral stenosis due to large masses in the left atrium with a mean gradient of 13 mmHg; masses protruding into the left ventricle creating an outflow gradient of 130 mmHg across the aortic valve; a tricuspid regurgitation gradient of 50 mmHg; and no pericardial effusion. A transoesophageal echocardiogram (TOE; *Figure 1B* and *C* and Supplementary material online, *Video S1*) showed massive masses in the left atrium and ventricle, seemingly perforating the mitral valve and extending into the left ventricular outflow tract (LVOT), resulting in functional LVOT obstruction and functional mitral stenosis. Emergency CT of the chest and abdomen revealed a mass in the left atrium and left ventricle, pulmonary congestion, one pathologically enlarged lymph node in the mediastinum, and several suspicious mediastinal lymph nodes. The radiodensity of the mass was 50 Hounsfield units, indicating a thrombus. As the nature of the detected mass was still unclear, a PET/CT scan (*Figure 2*) was performed, revealing masses suspicious for malignancy (due to strong tissue metabolic activity) in the left atrium, left ventricle, and septum. All lymph nodes in the mediastinum were without strong metabolic activity and thought to be benign.

After heart team discussion, it was decided to proceed with surgical treatment with subsequent tissue analysis and insertion of biological valve prostheses. Cardiac magnetic resonance imaging was not performed in this case as the final diagnosis had to be verified histologically, and surgery could not be postponed due to the physical state of the patient. In surgery, it was impossible to determine the origin of the tumour, as the tumour was firmly adherent to the left atrium, interatrial septum, mitral valve, ventricular septum, LVOT, and left ventricle. Apart from non-radical tumour resection (*Figure 3A*), the surgery included patch implantation in the left atrium and left ventricle (Supple Peri-Guard, model number 60PC-0404SN), bioprosthetic mitral valve replacement (Epic Mitral 31 mm FlexFit valve, model number E100-31M), and a Freestyle 27 bioprosthesis (model number FR995-27) for the aortic root. Non-radical tumour resection was chosen in order for functional cardiac anatomy to be preserved, realizing that a radical resection was not feasible in this case. The removed tumour mass was sent for pathology assessment. The surgery was complicated by pulmonary oedema, and the patient was put on veno-venous ECMO. The postoperative course was complicated by kidney failure (treated by continuous renal replacement therapy), shock liver, gastroparesis, and postoperative atrial fibrillation. The patient came off ECMO 2 days after the surgery with subsequent closure of the sternum, was extubated 4 days after the surgery, and was moved out of the intensive care unit 6 days after the surgery. Postoperative TTE (9 days after the surgery; see Supplementary material online, *Figure S3*) showed well-functioning bioprostheses, no visible residual tumour masses, and a normal left ventricular ejection fraction.

The pathology assessment revealed an intimal sarcoma with non-radically resected tumour borders (*Figure 3B–E*). Immunohistochemistry showed that the tumour was positive for MDM2, FLI1, and INI1, and

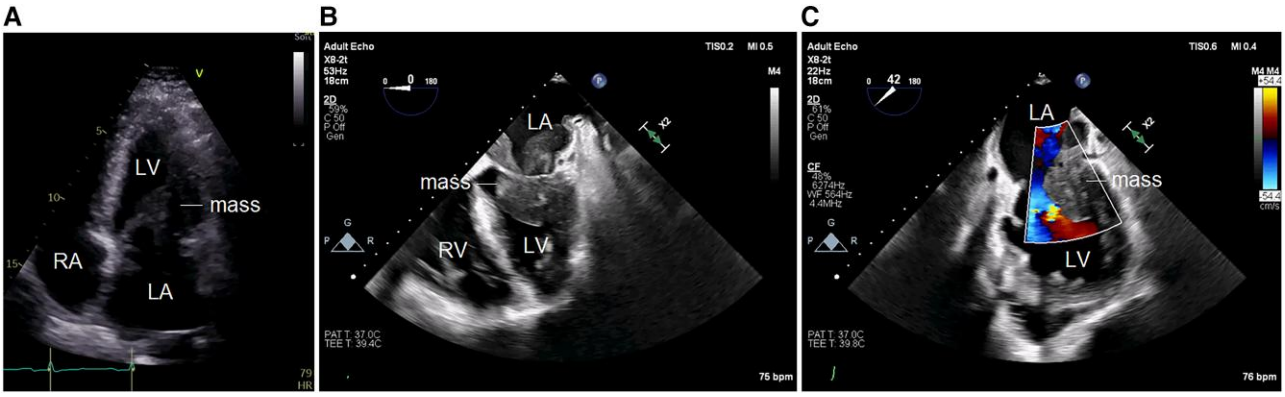

Figure 1 Preoperative transthoracic and transoesophageal echocardiogram. (*A*) Transthoracic echocardiogram, apical four-chamber view. Transthoracic echocardiogram showed a functional mitral stenosis due to large masses in the left atrium with a mean gradient of 13 mmHg; masses protruding into the left ventricle creating an outflow gradient of 130 mmHg across the aortic valve; a tricuspid regurgitation gradient of 50 mmHg; and no pericardial effusion. (*B* and *C*) Transoesophageal echocardiogram, four-chamber view, 0° rotation (*B*) and two-chamber view, 42° rotation (*C*). Transoesophageal echocardiogram showed massive masses in the left atrium and ventricle, seemingly perforating the mitral valve and extending into the left ventricular outflow tract, resulting in functional left ventricular outflow tract obstruction and functional mitral stenosis. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

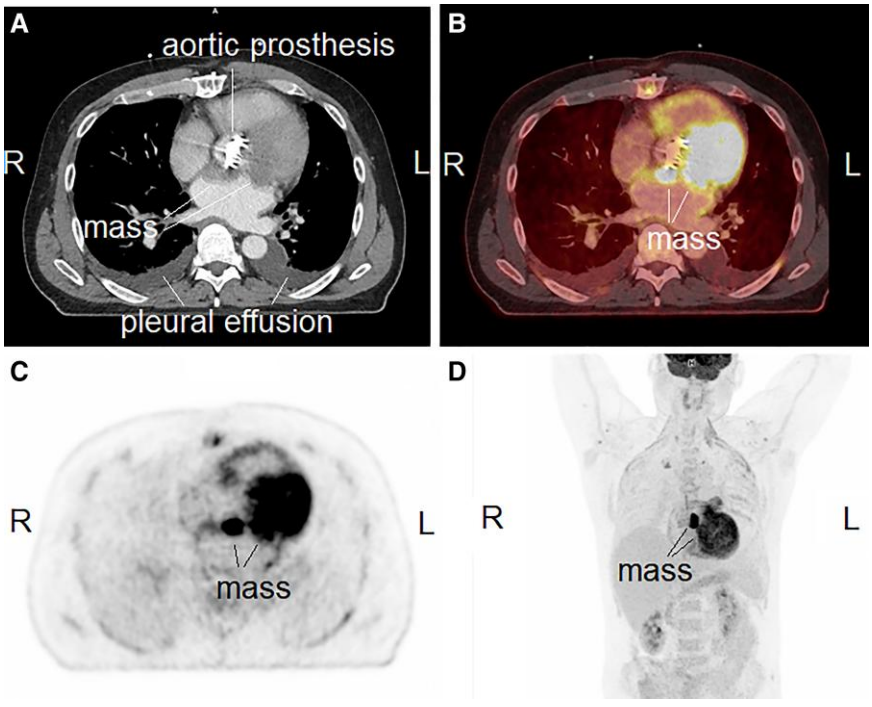

Figure 2 Positron emission tomography/computed tomography. (*A*) The Positron emission tomography/computed tomography scan revealed masses suspicious for malignancy (due to strong tissue metabolic activity) in the left atrium, left ventricle, and septum. All lymph nodes in the mediastinum were without strong metabolic activity and thought to be benign. The PET tracer used was ^{18}F-fluorodeoxyglucose. Panels *A*–*C* show axial view images, and Panel *D* shows the frontal view. Computed tomography is contrast-enhanced in the venous phase. L, left; R, right.

there was a heterogeneous expression of Calponine and Actine. There was no expression of CD34, ERG, CD31, S100, MYF4, Myoglobine, Desmine, Caldesmone, SMMS1, Beta-catenine, D2-40, or CK-AECAM.

The case was discussed at a multidisciplinary conference, and it was concluded that there were no further surgical or medical curative treatment options. The patient was discharged and started palliative treatment. He

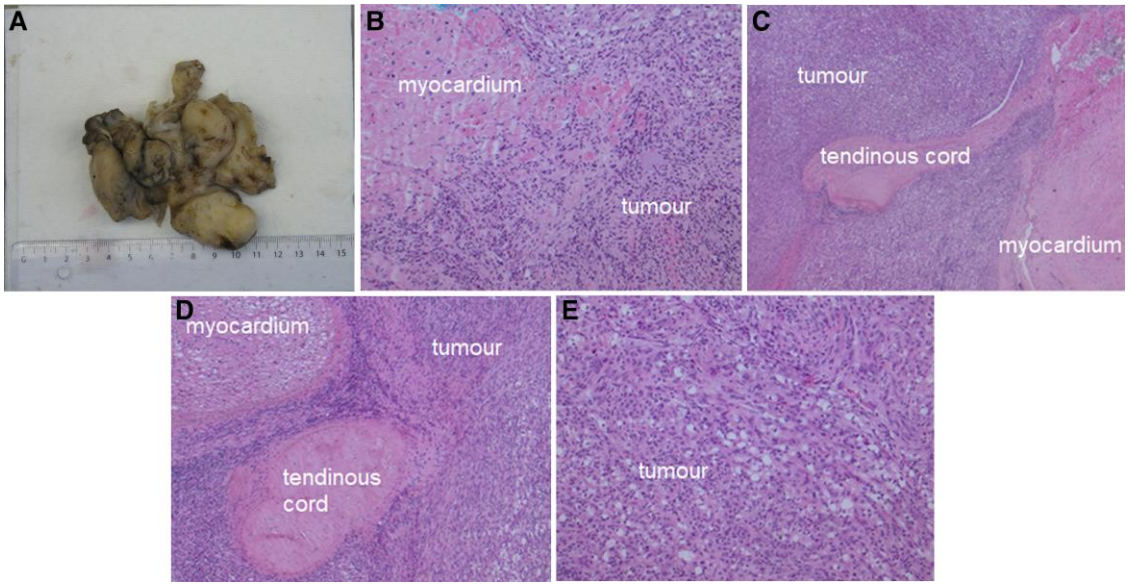

Figure 3 Gross pathology and histopathology. (*A*) Gross specimen after tumour resection. The resected tumour measured 85 × 80 × 30 mm. (*B–E*) Histopathology, haematoxylin, and eosin stain. The pathological assessment revealed an intimal sarcoma with non-radically resected tumour borders. (*B*) Myocardium (left) and infiltrating tumour cells, lymphocytes, and macrophages (right). Degenerated myocytes are seen in the transition zone. (*C*) Myocardium (right) and tendinous cord (middle). The tumour (left) fills out the whole ventricular cavity and has grown into the endocardium which has thickened as a reaction to the tumour. (*D*) Myocardium (upper left), tendinous cord (middle), and tumour. The myocytes have reversible changes (including fluid accumulation and degeneration of myofibrils), most likely as a reaction to the tumour. (*E*) Close-up of tumour with blood vessels.

developed dyspnoea, lost his appetite, experienced weight loss, and succumbed to his illness two and a half months after the surgery.

Discussion

Primary cardiac tumours are rare with an autopsy incidence of 0.001–0.030%. Of the 25% of primary cardiac tumours that are malignant, 95% are sarcomas, and 5% lymphomas.[1] The most common type of cardiac sarcoma is the angiosarcoma (37%); others include undifferentiated sarcoma, malignant fibrous histiocytoma, leiomyosarcoma, and osteosarcoma.[1] Further, several very rarely encountered primary cardiac tumours exist, with the least commonly reported being the intimal sarcoma, also called a spindle cell sarcoma.[1] Only few cases of cardiac intimal sarcomas have been reported,[2–7] with most cases being in males aged 45–50,[3,5–7] but there have also been reports on a 69-year-old male[2] and a 29-year-old pregnant female.[4] Intimal sarcomas are more commonly encountered in large arteries, and common sites of metastasis of intimal sarcomas in general include lungs, kidneys, lymph nodes, brain, and skin[2,8] as well as tumour emboli to bone, peritoneum, liver, and mesenteric lymph nodes.[2,9] Imaging morphology plays an important role in the preoperative differential diagnosis of cardiac tumours.[10] Features suggesting malignancy include broad-based lesions, infiltration of the myocardium/pericardium, lesions including the entire cardiac chamber, hilar lymphadenopathy, necrosis, or metastases.[3] Cardiac intimal sarcomas typically involve the left atrium, which is in contrast to angiosarcomas which typically involve the right atrium.[7] Patients with cardiac sarcomas live for a mean of 3 months to 1 year,[1] although survival up to 11 years has been reported.[5] In this case report, we present a case of an intimal sarcoma with a highly aggressive disease course where the patient succumbed to his illness just two and a half months after initial presentation to a physician, <3 months after onset of cardiac symptoms (i.e. dyspnoea), and three and a half months after onset of constitutional symptoms. Though the surgery was largely palliative in this case, early surgery is still valuable because of the improvement in haemodynamics and thereby dramatic palliation in symptoms. Further, it has been reported that life expectancy is almost twice as long for patients undergoing complete tumour resection compared with patients with incomplete excision.[11] Thus, early diagnosis using non-invasive and invasive imaging modalities is essential to start early treatment in order to improve outcomes in this patient group.

Lead author biography

Dr Adelina Yafasova, MD, graduated from University of Copenhagen in 2021 as a medical doctor. After completing her intern year in internal medicine, she is about to start her PhD studies at the Department of Cardiology, Copenhagen University Hospital, Denmark. She has previously done research in the field of cardiology, including on heart failure and autoimmune diseases.

Supplementary material

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

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The patient provided informed written consent for the data in this manuscript to be published in accordance with COPE guidelines.

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