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CASE REPORT
Cardiovascular imaging

Left ventricular intimal sarcoma: a case report

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Background Secondary cardiac tumours are much more common compared with primary (100–1000 times). The majority of the primary cardiac tumours are benign; however, almost a quarter are malignant, and 95% of these are sarcomas. The rarest type of primary malignant cardiac sarcoma is intimal (spindle cell) sarcoma.

Case summary A 37-year-old woman presented with episodes of breathlessness. Initially treated for a chest infection, however, the patient continued to deteriorate and presented to the emergency department. A large pericardial effusion was discovered and drained, with samples sent for analysis. A repeat interval echo confirmed the resolution of the pericardial effusion with preserved left ventricular (LV) systolic function. The computed tomography (CT) of the thorax showed suspicious lesions in the heart and lung while the repeat echo raised suspicion of an infiltrative disease. A cardiac magnetic resonance imaging scan was performed, which suggested evidence of an undifferentiated sarcoma involving the posterior wall of the LV and an overlying thrombus. Computed tomography of the abdomen and pelvis did not show any evidence of abdominal metastasis. A CT-guided lung biopsy was arranged. On histological analysis, the report was overall strongly supportive of a diagnosis of intimal sarcoma. She underwent chemotherapy until recently.

Discussion Cardiac intimal sarcomas are the least reported type of primary malignant tumours of the heart. They are encountered more commonly in the large arterial blood vessels, including the pulmonary artery and aorta, and are extremely rare in the heart. A prompt diagnosis is essential as they are considered extremely aggressive.

Keywords Case report • Primary cardiac intimal sarcoma • Pericardial effusion

ESC curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 2.3 Cardiac magnetic resonance • 6.8 Cardiac tumours • 6.6 Pericardial disease

Learning points

- Majority of the primary cardiac tumours are benign. Almost a quarter of primary cardiac tumours are malignant, and 95% of these are sarcomas; the remaining 5% are lymphomas. Angiosarcomas are the most common type of primary cardiac sarcoma (about 37%) followed by undifferentiated sarcoma (24%), malignant fibrous histiocytoma (11–24%), leiomyosarcoma (8–9%), and osteosarcoma (3–9%).
- The rarest type of primary malignant cardiac sarcoma is intimal (spindle cell) sarcoma.
- Pericardial effusion often emerges as the primary presenting sign and should consistently arouse suspicion.

Introduction

Secondary cardiac tumours are much more common compared with primary cardiac tumours (100–1000 times).[1] Majority of the primary cardiac tumours are benign (~75%), and nearly 50% of these are atrial myxomas.[1] Almost a quarter of primary cardiac tumours are malignant,[1] and 95% of these are sarcomas; the remaining 5% are lymphomas. Angiosarcomas are the most common type of primary cardiac sarcoma (about 37%) followed by undifferentiated sarcoma (24%), malignant fibrous histiocytoma (11–24%), leiomyosarcoma (8–9%), and osteosarcoma (3–9%).[1] Less

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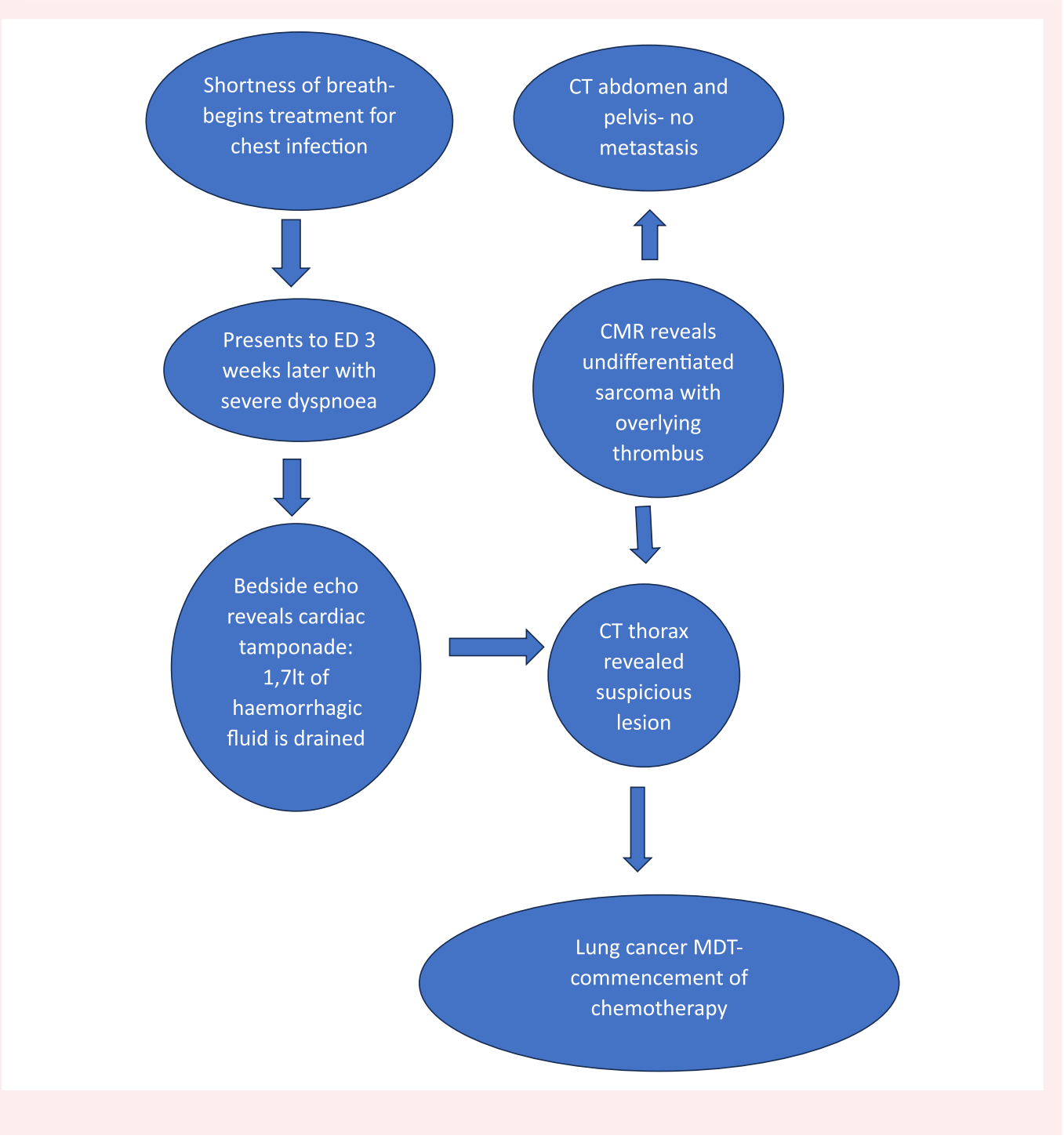
common types of primary cardiac sarcomas are rhabdomyosarcoma, liposarcoma, fibrosarcoma, synovial sarcoma, and haemangiopericytoma.[1] The rarest type of primary malignant cardiac sarcoma is intimal (spindle cell) sarcoma, and to our knowledge, there are only about 13 cases reported so far, all of which except one are of atrial origin.[1–15] Pericardial effusion is often the presenting sign in such cases.

We present a case of primary cardiac intimal sarcoma involving the left ventricle (LV). There is only one case found on literature review involving the LV.[2]

Summary figure

Case summary

A 37-year-old woman, who was normally fit and well without any medical background history, presented to her general practitioner with episodes of breathlessness. She was initially treated for chest infection with antibiotics and inhalers while her initial blood workup revealed increased inflammatory markers. The patient, however, continued to deteriorate and presented to the emergency department with 3-week history of worsening breathlessness. Cardiology advice was sought, and she was noted to have a global heart enlargement on chest X-ray (*Figure 1*) and small QRS complexes on electrocardiogram (ECG; *Figure 2*). Urgent bedside echocardiography (echo) revealed a



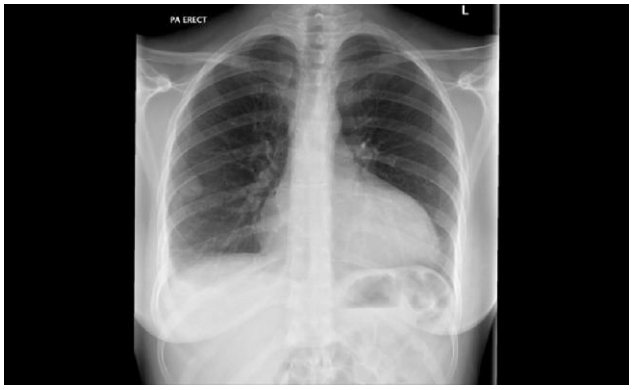


Figure 1 Chest X-ray showing global heart enlargement.

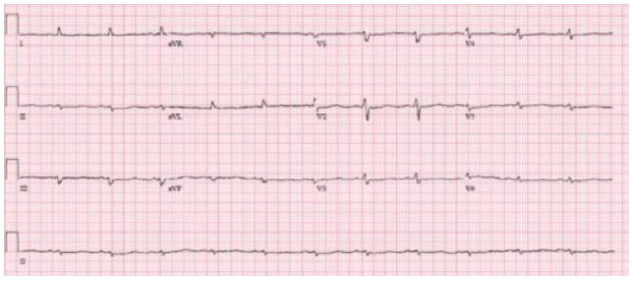


Figure 2 Patient's electrocardiogram before pericardiocentesis.

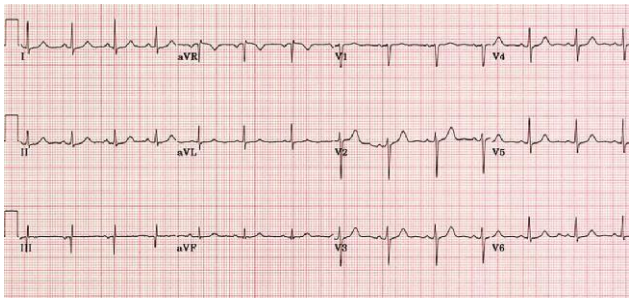


Figure 3 Patient's electrocardiogram after pericardiocentesis.

large pericardial effusion with the appearance of a 'swinging heart' with evidence of tamponade. Due to haemodynamic instability, emergency pericardiocentesis was performed leading to the drainage of 1.7 L of haemorrhagic fluid. Following this, instant relief with improvement of haemodynamics was achieved while a new ECG revealed enlargement of QRS complexes (*Figure 3*). Samples were sent to microbiology, cytology, and biochemistry. A repeat echo showed a non-mobile mass attached to the posterior wall of the LV, which raised suspicion for an infiltrative disease, and work-up for immunology, connective tissue disease, complement, bleeding diathesis, and viral screen was ordered. Pericardial effusion was sanguineous with lymphocytosis and mononuclear cell predominance. The cytologic microscopic examination showed that malignant cells were present. Histologically, the tumour cells in this case were composed of hypercellular malignant spindle cell with fascicular growth pattern.

Importantly also, the computed tomography (CT) thorax (*Figure 4*) showed suspicious lesions in the heart and lung. A cardiac magnetic resonance imaging (MRI) scan (*Figure 5A–C*) with characters of heterogeneous and significant patchy enhancement showed evidence of an undifferentiated sarcoma involving the posterior wall of the LV and

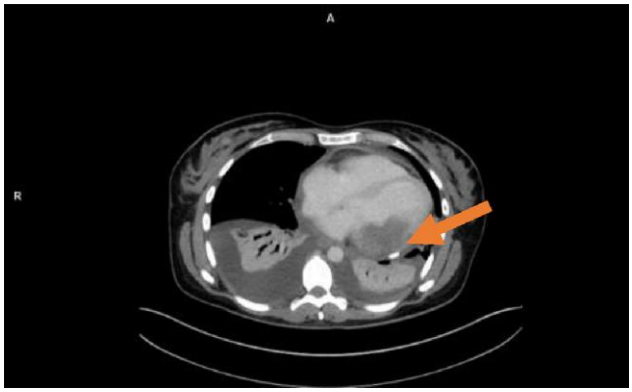


Figure 4 Computed tomography thorax showing suspicious lung and heart lesions.

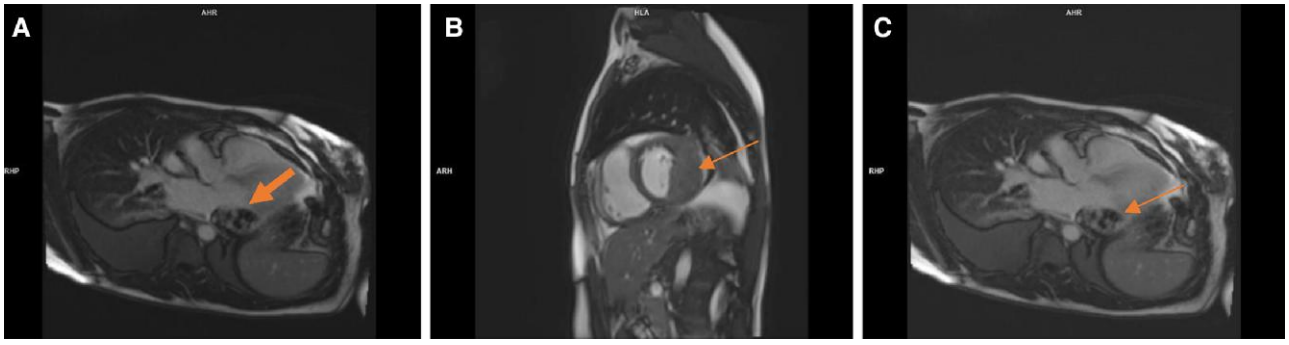


Figure 5 (*A*) Cardiac magnetic resonance showing undifferentiated sarcoma involving the posterior wall of left ventricle and an overlying thrombus. (*B*) Cardiac magnetic resonance showing undifferentiated sarcoma involving the posterior wall of left ventricle and an overlying thrombus. (*C*) Cardiac magnetic resonance showing undifferentiated sarcoma involving the posterior wall of LV and an overlying thrombus.

an overlying thrombus. Hence, the patient was commenced on anticoagulation. Computed tomography abdomen and pelvis did not show any evidence of abdominal metastasis, but the lung involvement earlier noted on non-contrast CT was confirmed. A CT-guided lung biopsy was arranged following consultation with the respiratory team. Ultrasound pelvis (both transabdominal and transvaginal) was negative for any masses.

She was referred for a multi-disciplinary team (MDT) discussion and discharged home with outpatient follow-up with the respiratory and oncology team. Lung cancer MDT advised that the tumour is not amenable to surgery or radiotherapy and advised to pursue with urgent oncologist referral. She was later reviewed in cardiology outpatients, and the MDT outcome was discussed with the patient while a referral for sarcoma MDT was anticipated. On histological analysis, low-level amplification for MDM2 was noted and the report was overall strongly supportive of a diagnosis of intimal sarcoma. Repeat interval echo confirmed the resolution of the pericardial effusion with preserved LV systolic function.

Following the diagnosis of metastatic cardiac intimal sarcoma, the patient commenced a chemotherapy regimen comprising doxorubicin and ifosfamide in mid-2021, which continued until autumn of 2021. After completing four cycles of chemotherapy, the patient experienced an episode of ventricular tachycardia, leading to the initiation of amiodarone and bisoprolol for cardiac management. Additionally, the patient's clinical course was complicated by the development of cancer-related venous thromboembolism (VTE), necessitating treatment with dalteparin. Subsequently, due to progressive disease, palliative therapy with docetaxel and gemcitabine was initiated on the first quarter of 2022 but discontinued after three cycles. A third-line chemotherapy regimen involving trabectedin was initiated on spring; however, disease progression was confirmed during subsequent assessments.

Discussion

In this report, we describe a case of an extremely rare malignant primary cardiac tumour. Cardiac intimal sarcomas are remarkably aggressive and least reported type of primary malignant tumours of the heart, and their clinical presentation is variable, ranging from common complains such as fatigue and dyspnoea to syncope. Patients often experience a rapid deterioration in health, and the prognosis is very poor with a mean survival of 3 months to 1 year. Pericardial effusion is often the presenting sign in such cases.[15]

Cardiac intimal sarcomas are encountered more commonly in the large arterial blood vessels, including the pulmonary artery and aorta, and are extremely rare in the heart.[15] We have managed to get the tissue diagnosis, but it highlighted the limited options for making a tissue diagnosis of the primary cardiac tumours involving the LV.

Lead author biography

Nikolaos Tsanaxidis is an interventional cardiologist at Papageorgiou General Hospital, Thessaloniki, Greece. He was trained at New Cross Hospital, Heart & Lung tertiary Centre, Royal Wolverhampton NHS Trust, UK. He is a board-certified cardiologist in the UK and Greece since 2018 and also holds a PhD from Larissa University Hospital, Greece. He is a professional member of the European Society of Cardiology (ESC) and EAPCI and a member of the British Cardiovascular Intervention Society (BCIS).

Consent: The authors confirm that written consent for submission and publication of this case report, including images and associated text, has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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Data availability

Data cannot be shared for ethical or privacy reasons. The data underlying this article cannot be shared publicly due to privacy reasons. The data will be shared upon reasonable request to the corresponding author.

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