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CASE REPORT
Cardiovascular surgery

Heart team rescues a bleeding heart: a case report of cardiac angiosarcoma causing life-threatening tamponade

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Background There are many causes of pericardial effusion and if the accumulating fluid results in cardiac tamponade, it may lead to life-threatening haemodynamic collapse. Therefore, rapid diagnosis and treatment of cardiac tamponade is critical.

Case summary A 53-year-old woman presented to the emergency department with chest pain and undifferentiated shock. A computer tomography (CT) aortogram showed a pericardial effusion, active contrast extravasation (possibly arising from the right coronary artery), but no aortic dissection. Echocardiography confirmed a large pericardial effusion with tamponade physiology. The consensus between cardiology and cardiothoracic surgery was for urgent coronary angiography to identify the source of bleeding. This showed a network of vessels from the right coronary artery that appeared to supply a mass. Re-review of the CT scan and repeat targeted echocardiography showed the silhouette of a mass adjacent to the right atrium. The patient was taken immediately for cardiac surgery. A cardiac tumour extending through the right atrial wall was identified, resected, and subsequently was diagnosed histologically as a cardiac angiosarcoma.

Discussion Malignancy is responsible for only a small proportion of pericardial effusions and metastatic disease is overwhelmingly more common than primary cardiac neoplasms. This case highlights the use of multi-modality cardiac imaging to guide diagnosis and treatment, and the need to consider the rarer causes of haemopericardium in cases where the more common causes have been excluded.

Keywords Case report • Tumour • Tamponade • Angiosarcoma

ESC curriculum 2.1 Imaging modalities • 2.4 Cardiac computed tomography • 3.4 Coronary angiography • 6.6 Pericardial disease • 6.8 Cardiac tumours

Learning Points

- Bleeding primary cardiac neoplasms are a rare cause of haemopericardium.
- Multi-modality cardiac imaging provides complimentary information that can guide diagnosis and treatment decisions.

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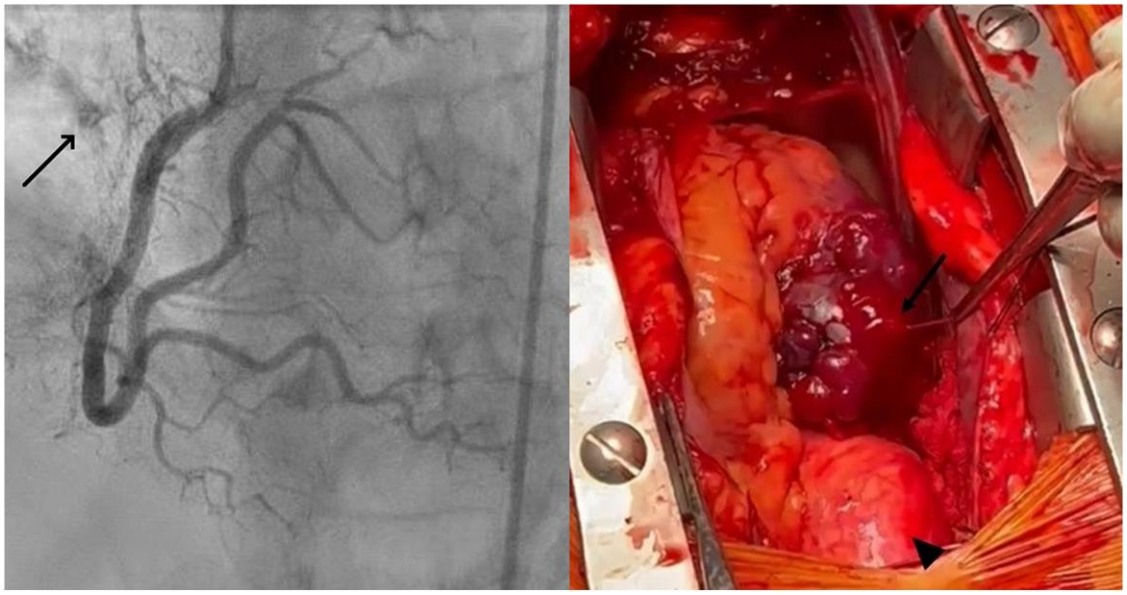
Introduction

Cardiac tamponade is medical emergency in which fluid accumulation within the pericardial space leads to increased intra-pericardial pressures, reduction of cardiac filling, decreased cardiac output, and shock. There are many causes of pericardial effusion and accurate diagnosis is required to guide therapy. Malignancy is responsible for only a small proportion of pericardial effusions[1] and metastatic disease is overwhelmingly more common than primary cardiac neoplasms.[2]

We present the case of a 53-year-old woman who presented with cardiac tamponade due to haemorrhage from a cardiac angiosarcoma.

Summary figure



Left: Coronary angiography (left anterior oblique projection) showing a network of vessels arising from the right coronary artery with extravasation of contrast into the pericardial space (arrow). Right: Clinical photograph taken during cardiac surgery. The heart is exposed via a median sternotomy approach. A cardiac tumour is visible (arrow) with active haemorrhage. The ascending aorta is visible at the lower end of the image (arrowhead).

Case presentation

A 53-year-old previously well schoolteacher was brought by ambulance to the emergency department of a tertiary hospital at ~4 a.m. in early June 2024. She woke with chest pain and dyspnoea and, upon arrival of paramedic services, was drowsy, pale, and hypotensive. Left and right blood pressure (BP) readings were unequal with a systolic BP of 100 mmHg in the left arm and unrecordable BP in the right arm. 800 mL of intravenous (IV) fluid and 100 µg of IV adrenaline was administered before arrival to hospital. Electrocardiogram showed sinus tachycardia with PR depression and no ST or T wave changes. In the emergency department, there was ongoing hypotension, and BP remained unequal. Resuscitation continued with further IV fluid boluses, placement of an arterial BP line, and commencement of a noradrenaline infusion. The BP normalized, and the patient's level of consciousness improved. A computer tomography (CT) aortogram was prioritized and this showed no evidence of aortic dissection, but a large pericardial effusion was noted (*Figure 1*; Supplementary material online, *Video S1*). Bedside echocardiography confirmed a large pericardial effusion with signs of tamponade. An urgent cardiology consult was requested, and preparations were made for pericardiocentesis in the emergency department in case the patient's haemodynamic status were to worsen. However, the patient remained stable and pericardiocentesis was not performed. Subsequently, the emergency physician was notified that the CT aortogram showed evidence of haemopericardium with active extravasation adjacent to the right coronary artery; however, the source of bleeding was unable to be identified. Urgent cardiothoracic surgery consultation was sought.

Further history revealed minimal past medical history with a distant left hemithyroidectomy, depression, and family history of premature coronary artery disease. Her only regular medication was venlafaxine. She had a viral upper respiratory tract infection 1 week prior to presentation.

Consensus between cardiology and cardiothoracic surgery was to proceed to an invasive coronary angiogram to identify the source of bleeding and consider deployment of a covered stent if feasible.

Coronary angiography revealed a fine network of vessels arising from the right coronary artery. These vessels appeared to supply a mass with extravasation of contrast from this mass into the pericardium (*Figure 2*; Supplementary material online, *Video S2*). The coronary arteries were otherwise normal. Further scrutiny of the CT scan, as well as targeted repeat echocardiography in the catheterization lab, was suggestive of a mass adjacent to the right atrium (*Figure 3*) with differentials including a paraganglioma or lymphoma. After heart team discussion, the decision was made to proceed immediately for surgical exploration and resection of the tumour.

The patient underwent a median sternotomy, and the pericardium was opened. There was clot present within the pericardial space (see Supplementary material online, *Video S3*) and the BP doubled once the pericardium was released. There was a tumour over the right atrium and anterior atrioventricular groove (*Figure 4*; Supplementary material online, *Video S4*). The decision was between conservative management, a wedge biopsy, or resection. Due to ongoing bleeding from the tumour, the decision was made to explore the atrial involvement of the tumour and consider resection. There was no palpable aortic disease. Cardiopulmonary bypass was commenced. The right atrium was

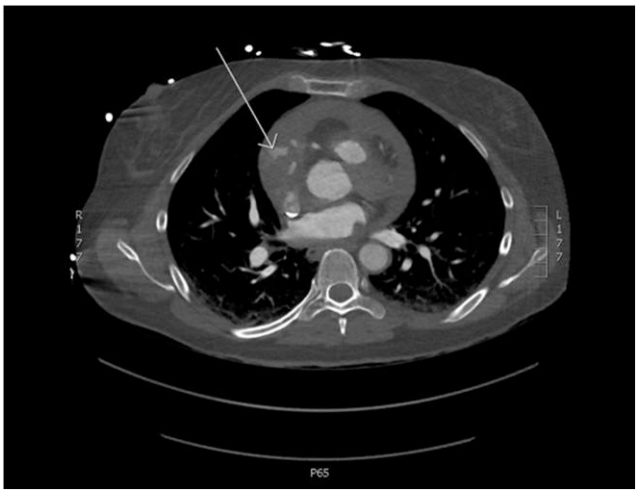


Figure 1 CT scan: CT aortogram showing haemopericardium with active contrast extravasation (arrow). CT, computer tomography.

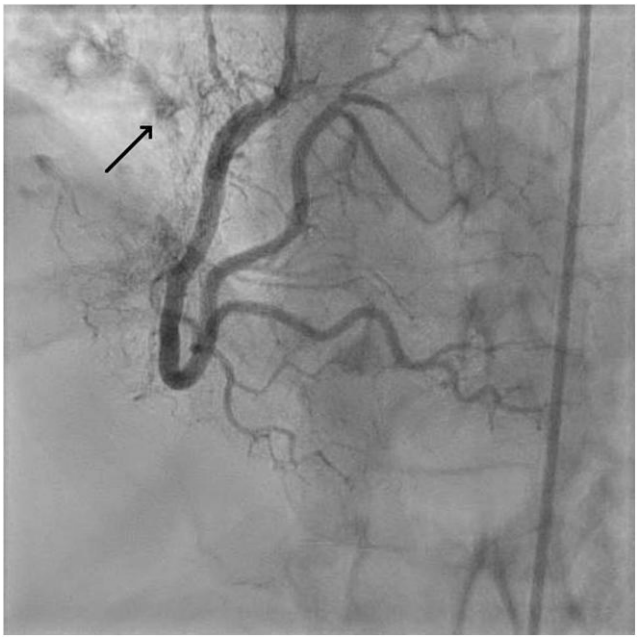


Figure 2 Coronary angiography: coronary angiography (left anterior oblique projection) showing a network of vessels arising from the right coronary artery with extravasation of contrast into the pericardial space (arrow).

opened parallel to the septum. The mass extended through the right atrial wall immediately adjacent to the right coronary artery and was carved off this with sharp dissection. This left a 3 mm cuff of atrium anterior to the tricuspid valve. The superior margin was divided inferior to the sinoatrial node. The defect in the right atrial wall was reconstructed with bovine pericardial patch. Epicardial pacing wires were placed for safety. The patient was weaned from cardiopulmonary bypass without inotropic support and haemostasis was secured. The chest was closed, and the patient was transferred to the intensive care unit (ICU) in a stable manner.

The patient was extubated, weaned from vasopressor support and discharged to the cardiothoracic ward from ICU the following day.

A repeat transthoracic echocardiogram on day 5 showed normal left ventricular function, low normal right ventricular function, no residual pericardial effusion, and prominent septal bounce thought to reflect some degree of persistent ventricular interdependence. The patient was discharged on postoperative day 6.

Histological assessment identified the tumour as an angiosarcoma with evidence of fat invasion, focal venous invasion, and extending

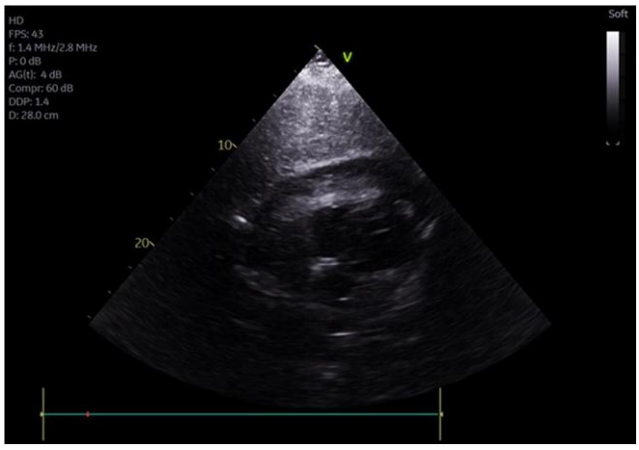


Figure 3 Echocardiography: subcostal 4-chamber view showing pericardial effusion with an echodense structure overlying the right atrium.

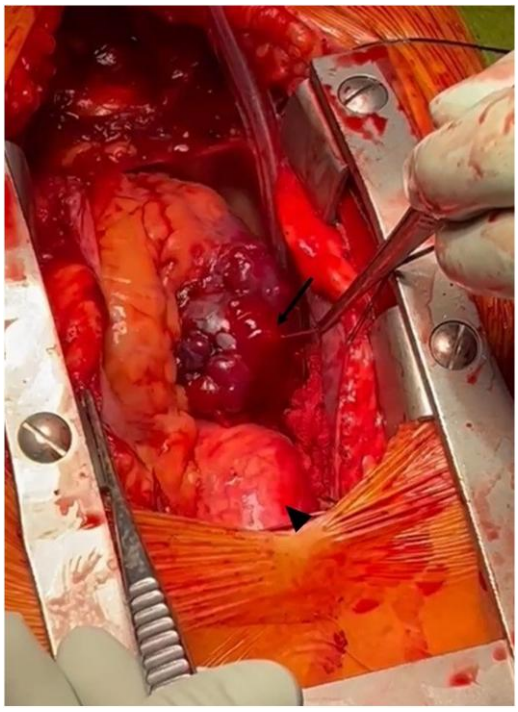


Figure 4 Cardiac surgery: clinical photograph taken during cardiac surgery. The heart is exposed via a median sternotomy approach. The cardiac angiosarcoma is visible (arrow) with active haemorrhage. The ascending aorta is visible at the lower end of the image (arrowhead).

to the surgical margins. The patient was discussed at the sarcoma multi-disciplinary meeting and planned for chemotherapy and radiotherapy. She completed 20 Gy of radiation in five fractions to the right atrium to prevent local recurrence and bleeding. A fluorodeoxyglucose positron emission tomography scan did not identify any nodal or distant metastases. She was offered chemotherapy with gemcitabine and docetaxel; anthracyclines were avoided due to concern of cardiotoxicity.

The outcome for patients with cardiac angiosarcomas is poor with median survival reported at 3.8 ± 2.5 months without surgical resection.[3] The rarity and variable presentation of this condition limit the ability to develop standardized treatment guidelines, but algorithms have been proposed.[4] Surgical resection may offer the best improvement in outcome, and chemotherapy and radiation are also commonly offered.[3,4] Median survival after surgical resection has been reported at 14 months[3] and there has been a reported case where the patient was still alive after 9.5 years.[5] Our patient's lack of metastatic disease may imply a better long-term prognosis.

Discussion

This case describes a rare cause of pericardial effusion and cardiac tamponade. Malignancy is responsible for ~6% of pericardial effusions,[1] increasing to 15%–20% of moderate to large effusions.[6] Metastatic disease involving the heart is overwhelmingly more common than primary cardiac neoplasms.[2] Primary cardiac tumours are exceedingly rare with an incidence of ~0.02%.[7]

This case highlights the complimentary nature of multi-modality imaging. The patient's diagnosis was investigated with the use of CT, echocardiography, and coronary angiography. The initial CT scan was able to identify the pericardial effusion, characterize it as a likely haemopericardium and noted the presence of active extravasation of contrast into the pericardial space. Echocardiography confirmed the presence of a large pericardial effusion with signs suggesting tamponade physiology. Coronary angiography identified a network of small capillaries that appeared to supply a mass adjacent to the right atrium. In the context of the patient presenting with obstructive shock, all these modalities were used to form the provisional diagnosis of cardiac tamponade secondary to an actively bleeding cardiac tumour, and to guide the decision to proceed immediately to cardiac surgery to control the bleeding, diagnose and resect the mass.

Lead author biography

Dr Theodore Sklavos completed medical school at the University of Queensland in 2017 and is now undergoing advanced training in cardiology at the Princess Alexandra Hospital in Brisbane, Australia. His interests include medical education and cardiac electrophysiology.

Supplementary material

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

Consent: The authors confirm that written consent for the publication of this case report, including images, videos, and text, has been obtained from the patient in accordance with Committee on Publication Ethics guidelines.

Conflict of interest. None declared.

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Data availability

Non-identifiable data underlying this article will be made available upon reasonable request to the corresponding author.

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