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GRAND ROUND
Heart failure

When cardiac surgery comes to its limits: a case report of pericardial mesothelioma invading the myocardium

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Background Primary pericardial mesothelioma (PPM) is a rare form of highly aggressive cancer. Many patients are diagnosed only at an advanced stage. Therefore, the overall survival rate is poor with a median survival of 3 months. In some rare cases, the PPM infiltrates the myocardium causing lethal myocardial dysfunction.

Case summary A 66-year-old patient was transferred to our centre with the provisional diagnose of pericarditis of unknown origin. Using extensive cardiac imaging [echocardiography, computed tomography (CT), positron emission tomography–CT, cardiac magnetic resonance imaging, left and right heart catheterization, coronary angiography], PPM was finally diagnosed. After consultation with the oncologists, the heart team decided to resect the tumour first due to impaired haemodynamics and then initiate adjuvant chemotherapy. Intraoperatively, myocardial infiltration of the tumour became apparent, which was not detected preoperatively despite intensive imaging. Complete resection of the PPM was not possible and effective decompression of the ventricle could not be achieved. The patient died on the first postoperative day.

Discussion Surgical therapy is indicated in many forms of cardiac tumours. However, when a tumour invades the myocardium, surgery often comes to its limits. In this case, myocardial invasion of PPM could not be detected despite extensive imaging. We therefore suggest that possible myocardial infiltration by PPM, and thus potential limitations of cardiac surgery, should be considered independently of imaging results when therapeutic options are discussed.

Keywords Case report • Myocardial invasion • Primary pericardial mesothelioma • Cardiac imaging • Cardiac tumour surgery

Learning points

- Primary pericardial mesothelioma is a rare and lethal form of cancer and represents a potential cause of pericardial effusion.
- Myocardial tumour infiltration must be considered even in the absence of evidence by cardiac imaging.
- Possible therapeutic options must be discussed and decided in a multidisciplinary team.

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Introduction

Malignant mesothelioma is a rare and highly aggressive tumour. Around 80% of all mesotheliomas originate from mesothelial cells of the pleura.[1] Only a few cases of primary pericardial mesothelioma (PPM) are reported in the literature.[2] In PPM, malignant cells originating from the pericardium can infiltrate into the myocardium causing functional impairment. PPM is found in middle-aged patients and is commonly associated with asbestos exposure.[3] Diagnosis of PPM is difficult especially in early stages of the disease.[4] Accordingly, many patients are diagnosed at an advanced stage of the disease or post-mortem.[5] Thereby the overall survival of patients suffering from PPM is poor, with a median survival of 3 months without surgical intervention.[6] Therapeutic options for PPM include resecting surgery, radiation, and chemotherapy with only limited data on treatment efficacy.[4,6] In patients with acute symptoms due to haemodynamic compromise, tumour resection is the only approach that can restore cardiac function.[7]

The following case report presents a patient with PPM in whom a surgical approach for ventricular decompression was taken after multidisciplinary team discussion.

Timeline

Case presentation

A 66-year-old Caucasian male patient, suffering from right heart failure due to pericardial effusion, was transferred to our centre. The patient's symptoms started 4 months earlier with shortness of breath and substantial weight loss. In the 2 weeks prior to hospitalization, the patient suffered from episodes of tachycardia, mild fever, and dizziness. The patient had an ongoing history of diabetes mellitus. In particular, there was no previous asbestos exposure. A pericardiocentesis performed at a distant hospital delivered 500 mL of serous exudate, but was incidentally inconspicuous with no malignant cells in cytology. Breathlessness remained unchanged even after successful drainage. The patient was transferred to our centre with the provisional diagnosis of pericarditis of unknown origin.

Transthoracic echocardiography (ECHO) showed dilatation of both atria and impaired left ventricular (LV) function (LV ejection fraction 42%). Predominantly the right ventricular was compressed apically by an 18 mm × 32 mm tumourous formation surrounded by a localized pericardial effusion (*Figure 1A*). Cardiac magnetic resonance imaging (MRI) and computed tomography (CT) scan showed a contrast-enriched neoplasm, originating from the pericardium (*Figure 1B*). Positron emission tomography (PET)–CT scan revealed a highly increased glucose metabolism in the neoplasia (*Figure 1C*). Therefore, a primary pericardial malignoma was diagnosed. There was no evidence for infiltration of the myocardium or metastasis to adjacent organs or lymph nodes. Based on cardiac imaging we hypothesized a constrictive effect of the tumour on the myocardium. This was supported by haemodynamic findings showing an increase and equalization of diastolic pressures in the left and right ventricle as well as a typical square root sign in the pressure curves. Cardiac output was markedly decreased at 1.8 L/min (*Figure 1D*).

After consultation with oncologists, the heart team decided to first resect the tumour due to impaired haemodynamics and then initiate adjuvant chemotherapy.

March 2020: patient suffers first symptoms	**Patient presents with shortness of breath and substantial weight loss**
5 July 2020: initial examination at a distant hospital	Provisional diagnosis of pericarditis of unknown origin was made
14 July 2020: presentation at our centre	• Extensive imaging such as echocardiography, computed tomography (CT), cardiac magnetic resonance imaging, positron emission tomography–CT, left and right heart catheterization, and coronary angiography was performed • A primary pericardial mesothelioma was diagnosed • The case interdisciplinary consultation between cardiologists, cardiac surgeons, and oncologists was performed • Decision to resect the tumour due to impaired haemodynamics
3 August 2020: surgery	• Intraoperative diagnosis of tumour penetration into the myocardium • A complete tumour resection and thus haemodynamic stabilization was not possible • ECMO implantation was rejected due to lack of weaning option given the extent of myocardial tumour involvement
4 August 2020: 1st postoperative day	The patient died on the first postoperative day from multiorgan failure due to haemodynamic instability caused by persistent tumour compression

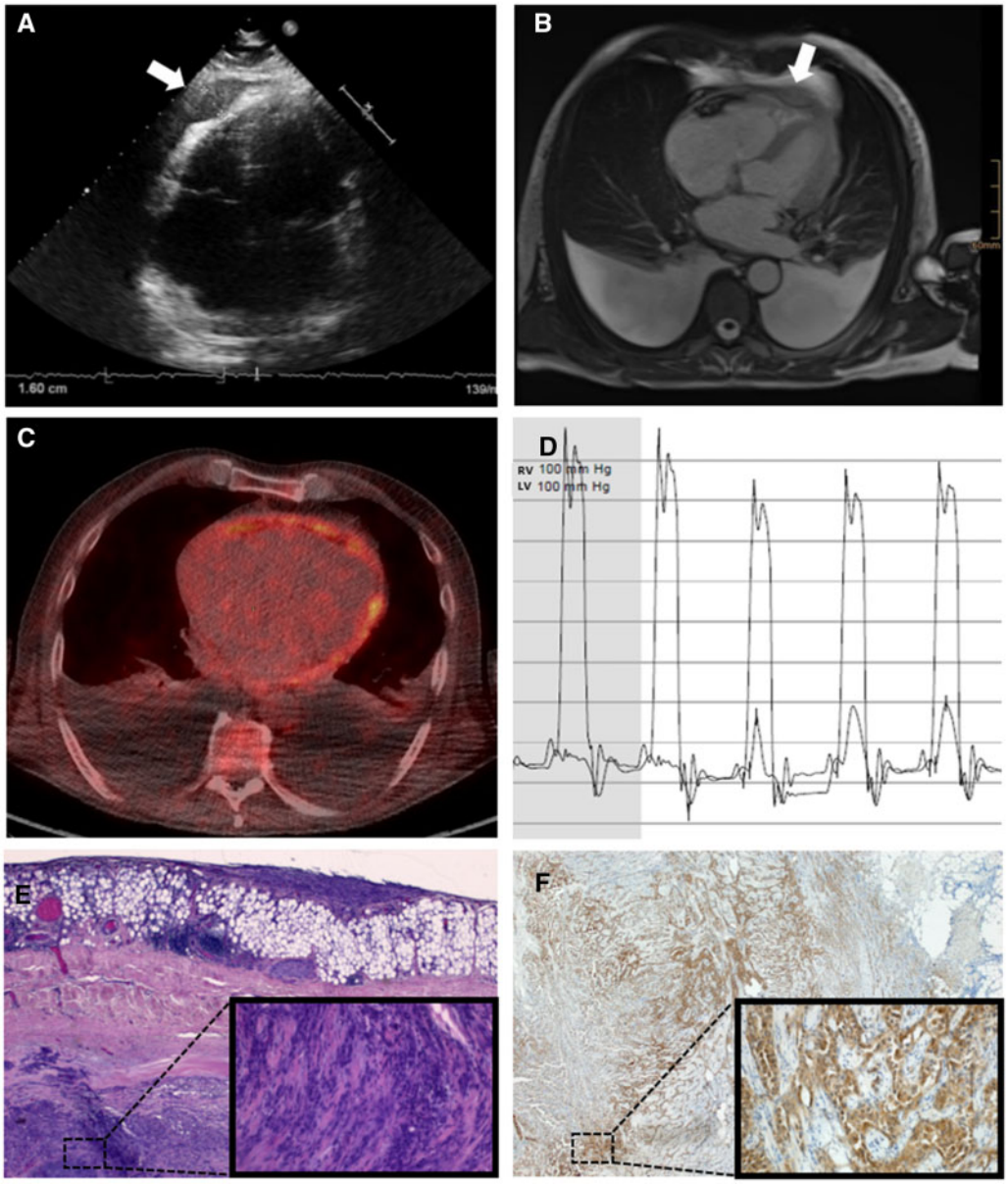

Figure 1 (*A*) Transthoracic echocardiography: 18 mm × 32 mm formation compressing the right ventricular apically. (*B*) Cardiac magnetic resonance imaging: contrast-enriched neoplasm, originating from the pericardium. (*C*) Positron emission tomography–computed tomography scan: highly increased glucose metabolism in the neoplasia in the right and left ventricular wall. (*D*) Simultaneous recording of haemodynamic pressure curves in the left and right ventricle: increase and equalization of diastolic pressures and a typical square root sign in both ventricles. Cardiac output was significantly decreased at 1.8 L/min. (*E*) Histology: spindle-shaped, partly tubular malignant tumour. (*F*) Immunohistochemistry: calretinin positivity in the glandular area of the tumour.

With the onset of anaesthesia, high doses of vasopressors and inotropes were required to stabilize cardiac output and blood pressure. Full sternotomy was performed to ensure sufficient access to the tumour. Rapid histological examination revealed a spindle-shaped, partly tubular malignant tumour (*Figure 1E and F*). After resection of the pericardial neoplasia, tumourous penetration into the myocardium became obvious. This was not to be expected based on previous imaging. Complete resection of the myocardial involvement by PPM was not possible due to massive infiltration. Therefore, an effective decompression of the ventricle could not be achieved. In the absence of treatment options, implantation of an Extracorporeal membrane oxygenatio (ECMO) for haemodynamic stabilization was rejected. Consequently, the patient died on the first postoperative day from multiorgan failure due to haemodynamic instability caused by persistent tumour compression.

Discussion

Surgical therapy is indicated in many forms of cardiac tumours, especially in benign myxomas.[8] In PPM, surgery is a possible curative

therapy if the tumour is confined to the pericardium. However, when a tumour invades the myocardium, surgery often comes to its limits. In our patient, a PPM invaded the myocardium, causing constriction as well as restriction of the myocardium. Tumourous infiltration of the myocardium was not suspected from cardiac imaging. Because of the life-threatening haemodynamic condition, urgent surgery was indicated. However, intraoperatively it became evident that the intramyocardial tumour could not be entirely resected.

Cardiac imaging revealed a primary pericardial tumour. However, myocardial invasion of the tumour could not be demonstrated despite extensive cardiac imaging (ECHO, CT, PET–CT, MRI). Similarly, dignity and malignancy of the tumour could not be accurately defined by imaging. Although a PPM was suspected preoperatively, it could only be identified on the basis of the intraoperative histological frozen section.

In a recently published retrospective study, myocardial infiltration was reported in no <45% of all patients undergoing cardiac surgery for PPM.[9] We therefore suggest that myocardial infiltration by PPM, and thus potential limitations to cardiac surgery, should be considered even independent of imaging results, when therapeutic options are discussed. Analysis of pericardial fluid is also of limited value in most cases.

Lead author biography

Professor Gerhard Pölzl is head of the heart failure unit at the University Hospital Innsbruck in Austria. He graduated from the University of Graz in Austria and completed his residency in training in Linz, Austria. He is an expert in heart failure and orphan diseases in cardiology.

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 authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient's next-of-kin in line with COPE guidance.

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