An unusual cause of acute pericarditis: a case report

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Background
Inflammation of the pericardium, or pericarditis, is a frequent cause of acute chest pain in young patients. Pericarditis is typically associated with viral infections, but other potential causes may have distinct prognostic and therapeutic implications.

Case summary
A 26-year-old man presented with typical signs and symptoms of acute pericarditis. However, imaging disclosed an anterior mediastinal mass that compressed the right ventricular outflow tract. The coarse outflow murmur convincingly mimicked a pericardial friction rub on auscultation.

Conclusion
Clinicians should be aware of alternative aetiologies to pericarditis in patients who present with prolonged or refractory symptoms.

Keywords
Case report • Mediastinal mass • Pericarditis • Outflow tract obstruction • Right ventricular failure

Introduction
Inflammation of the pericardium, or pericarditis, is the most common pericardial disease and is a frequent cause of acute chest pain in young patients. Pericarditis is typically associated with viral infections, but may also be caused by rheumatic diseases, uraemia, and neoplasms. The typical signs and symptoms include a pericardial friction rub, concave ST-segment elevations, pleuritic chest pain, and a pericardial effusion. In most cases, treatment is primarily supportive, because pericarditis is often self-limited, and includes administration of non-steroidal anti-inflammatory drugs or colchicine. However, other potential causes of pericarditis may have distinct prognostic and therapeutic implications. Here we describe a patient with a mediastinal tumour who presented with the signs and symptoms of acute pericarditis.
Timeline

| Timing  | Event |
|---------|-------|
| +2–4 days | Transthoracic echocardiogram: Extrinsic compression of right ventricular outflow tract (RVOT) and main pulmonary artery by anterior mediastinal mass with shunting through restrictive ventricular septal defect. |
| +1 week | Transbronchial biopsy detected malignant cells |
| +1 month | Diagnosis of stage III mediastinal non-seminomatous germ cell tumour and initiation of chemotherapy |
| +4 months | Median sternotomy with resection of anterior mediastinal tumour |
| +6 months | Computed tomographic angiogram with contrast: resolution of RVOT compression |
| +8 months | Magnetic resonance imaging: new femoral bone metastases Initiation of salvage chemotherapy |

Case presentation

A previously healthy 26-year-old man without a significant past medical history was admitted to the emergency department with pleuritic substernal chest pain that had steadily increased in frequency and intensity for 2 weeks. The pain was sharp, localized to the left precordium and was exacerbated by deep respirations or changes in position. He also reported a dry cough and dyspnoea. In 2016, a workup for similar chest pain that included an electrocardiogram, transthoracic echocardiogram, and myocardial perfusion imaging study were reportedly unremarkable.

On physical examination, he appeared to be in moderate distress due to chest pain. He was tachycardic (pulse 100–110 b.p.m.), afebrile, and normotensive. A coarse systolic murmur was audible at the parasternal border and radiated throughout the precordium. S1 and S2 were distinct and there were no additional heart sounds. Electrocardiography disclosed diffuse ST-segment elevation and PR-segment depression (Figure 1). The patient was admitted to the hospital with an initial plan for medical therapy of presumptive acute pericarditis using non-steroidal anti-inflammatory drugs and colchicine.

Subsequently, plain chest films revealed bilateral nodular opacities and a widened mediastinum. Transthoracic echocardiography (TTE) identified an anterior mediastinal mass (Figure 2). The mediastinal mass and surrounding oedema compressed the main pulmonary artery to a minimum diameter of 7 mm and generated a 48-mmHg gradient across the RVOT. Correspondingly, the right ventricle was moderately dilated (57 mm) with reduced systolic function. Left ventricular systolic function was preserved (ejection fraction 65–69%). There was no evidence of direct pericardial involvement or invasion by the mass. Colour Doppler also detected a small, transient right to left shunt through a restrictive ventricular septal defect (VSD) proximal to the site of maximal RVOT compression (Video 1). Computed tomographic angiography identified multiple enhancing pulmonary nodules and demarcated the complete extent of the mass (70 mm × 76 mm × 92 mm) in the anterior mediastinum (Figure 3). Initial laboratory values were unremarkable. The red blood cell count was 4.78 M/µL (reference values: 4.60–6.20 M/µL) and the white blood cell count was 13 K/µL (reference values: 4.5–12 K/µL). Serum tumour necrosis factor, AFP, and β-hCG were undetectable. Transbronchial biopsy and subsequent pathologic analysis confirmed that the mediastinal mass was a metastatic stage III mediastinal non-seminomatous germ cell tumour. Over the next four months, the patient received VIP chemotherapy using cisplatin, etoposide, and ifosfamide with an initially favourable response. After multiple rounds of chemotherapy, the patient underwent a median sternotomy with successful resection of the anterior mediastinal mass. Treatment resulted in decompression of the RVOT, restoration of RV systolic function, and resolution of the patient’s chest pain (Figure 4). However, at 8 months, imaging detected new disease progression with enlarging pulmonary parenchymal lesions and probable bony metastases. This occurred in concurrence with an up-trending hCG serum level after the initial rounds of chemotherapy (2 mIU/mL) until detection of the lesions and metastases (4630 mIU/mL). hCG serum levels increased one month afterward to 9570 mIU/mL. The patient underwent additional rounds of salvage chemotherapy.

Discussion

The clinical hallmarks of acute pericarditis are pleuritic chest pain, a pericardial friction rub, suggestive changes on electrocardiography, and pericardial thickening or pericardial effusion. Most cases of pericarditis are caused by a viral infection, such as Coxsackie A9, Mumps, and Epstein–Barr Virus, and respond to anti-inflammatory medications. In this case, the age of onset, clinical presentation, ECG findings, and apparent friction rub were so consistent with pericarditis that plans for empiric therapy were initiated.

A pericardial friction rub is pathognomonic for pericarditis and is best heard at the end of an expiration with a characteristic coarse triple cadence. In contrast, pulmonic or right ventricular outflow tract murmurs are associated with a delayed second heart sound and a systolic ejection murmur that varies with inspiration. In this case, the presence of a small VSD may have contributed to the illusion of a friction rub. VSD murmurs are characterized by a
The combination of the two outflow tract murmurs convincingly mimicked a pericardial friction rub on auscultation. Transthoracic echocardiography revealed the source of the pulmonic murmur: a mass compressing the right ventricular outflow tract and main pulmonary artery.

In adults, anterior mediastinal masses are most frequently caused by thymomas, lymphomas, and germ cell tumours. Thymomas rarely metastasize and account for <1% of adult malignancies. In contrast, lymphomas and germ cell tumours are more common and frequently metastasize to the liver, bone marrow, or lungs. Common symptoms include dyspnoea, chest pain, cough, and fever. Mediastinal tumours may cause nonspecific symptoms related to impingement or invasion of adjacent mediastinal structures, including the right ventricular outflow tract and main pulmonary artery.
Neoplastic pericarditis is relatively rare but may account for as many as 7% of acute pericarditis cases and is most frequently associated with adenocarcinomas, lymphomas, and leukemias. Cancer cells most frequently spread to the pericardium by direct extension, lymphatic invasion, or haematogenous metastasis, but pericarditis may also occur as part of the paraneoplastic syndrome. Pericarditis may be the first symptom of metastatic tumours and is associated with increased mortality. Malignancies should be suspected in younger patients with prolonged or refractory pericarditis and weight loss or signs of systemic illness.

Conclusion

We describe an unusual presentation of acute pericarditis due to a mediastinal tumour. Clinicians should be aware of such alternative aetiologies when patients present with prolonged or refractory symptoms.

Lead author biography

Dr. Prakash is an adult congenital cardiologist and genetics researcher with a special interest in left ventricular outflow tract defects. He completed an MD and PhD at Baylor College of Medicine and teaches at The University of Texas Health Science Center at Houston.

Supplementary material

Supplementary material is available at European Heart Journal - Case Reports online.
Slide sets: A fully edited slide set detailing these cases 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 in line with COPE guidelines.

Conflict of interest: none declared.

Funding: none declared.

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