An unusual cause of pericarditic chest pain

Introduction

Pericarditis is a relatively common cause of hospital admission in young to middle-aged men. The aetiology is usually infective, inflammatory or autoimmune, although it is rarely caused by underlying primary cardiac malignancy.

Primary tumours of the heart are rare, with a reported autopsy prevalence of 0.001–0.28% (Reynen, 1996). Approximately 75% are benign and 50% of these are myxomas. The most frequent malignant tumours (35–40%) are angiosarcomas (Menq et al, 2002), which also predominantly affect middle-aged men.

Discussion

The location, size, friability and histological type of primary cardiac tumours determine the spectrum of symptoms and clinical findings. Because of the rarity of these tumours, consideration of cardiac sarcoma in a patient with non-specific cardiopulmonary symptoms and signs may be easy to overlook. The clinical manifestations may be divided into four general mechanistic categories: systemic (produced by tumour-secreting products or tumour necrosis), embolic, intrinsically cardiac and phenomena secondary to metastatic disease.

In this case, the electrocardiogram showed ST segment changes, but arrhythmias and atrioventricular block also occur. Angiosarcomas are often first detected by two-dimensional echocardiography. Inexplicably, around 80% of these are located in the right atrium (Menq et al, 2002). Pericardial spread can result in symptoms suggestive of pericarditis.

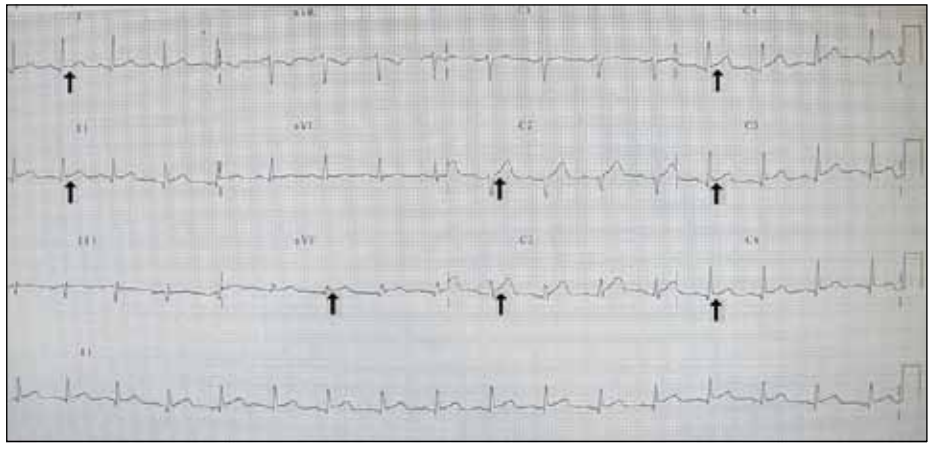
Detailed anatomy is best provided by axial computed tomography and magnetic resonance scanning. Positron emission tomographic imaging provides additional information on the functional (metabolic) activity of the tumour and its distribution. Preoperative angiography allows evaluation of involvement of the great vessels, coronary circulation and peri-tumour neovascularization (Sinatra et al, 2003).

Histologically, cardiac angiosarcomas usually exhibit evidence of endothelial differentiation, with formation of vascular channels and/or papillary structures. They grow rapidly and are characterized by friability, which may cause embolic phenomena, and a tendency towards bleeding. Cytogenetic analysis of these tumours may show numerical and structural chromosomal changes. Immunohistochemical study has shown a high expression of mutated p53 gene products (Zu et al, 2001). This can be used to further support evidence of endothelial differentiation by demonstrating CD31, CD34 and vWF immunophenotype in the tumour cells.

Dr Simon W Dubrey *is Consultant Cardiologist,* **Dr Richard Grocott-Mason** *is Consultant Cardiologist,* **Dr Jacqueline Doyle** *is Specialist Registrar in Cardiology, Hillingdon Hospital, Uxbridge, Middlesex UB8 3NN,* **Dr Tarun Mittal** *is Consultant Radiologist and* **Dr Alexandra Rice** *is Consultant Histopathologist at Harefield Hospital, Harefield, Middlesex*

Correspondence to: Dr SW Dubrey

Figure 1. Resting electrocardiogram showing widespread ST elevation (black arrows) in the anterior and inferolateral leads.



Case Report

A 56-year-old man was admitted describing a 1-day history of pericarditic type chest discomfort. On examination, the pulse was regular at 90 beats/min, blood pressure was 102/65 mmHg and heart sounds were normal. The jugular venous pressure was not elevated. The chest X-ray showed a normal cardiac silhouette with clear lung fields. An electrocardiogram was highly suggestive of pericarditis (*Figure 1*). Initial blood tests, including a troponin I of 0.04 μg/litre, were all normal. Transthoracic echocardiography showed a large mass within the right atrium (*Figure 2*). A contrast-enhanced computed tomographic scan of the thorax, abdomen and pelvis suggested direct extension of the atrial mass into a 2.5 cm thick soft tissue mass in the mediastinum, but no systemic spread (*Figure 3*).

Positron emission tomographic scanning revealed focal intense uptake of fluorine-18-fluorodeoxyglucose tracer within the right atrium and within pericardial fat between the heart and pericardium.

A tissue biopsy was obtained via anterior mediastinotomy. Histology showed infiltration of fat and fibrous tissue by a malignant spindle cell tumour, which appeared to be forming vascular spaces within it (*Figure 4*). On immunohistochemistry the tumour cells were diffusely strongly positive for endothelial marker CD31, confirming the diagnosis of angiosarcoma. Other tumour markers, including CD34, MNF116, calretinin, CK5/6, SMA, desmin and S100 were negative.

The tumour, a locally extensive angiosarcoma, was considered unsuitable for primary resection. The patient has been referred for chemotherapy with doxorubicin and potentially surgical resection if the tumour shrinks.

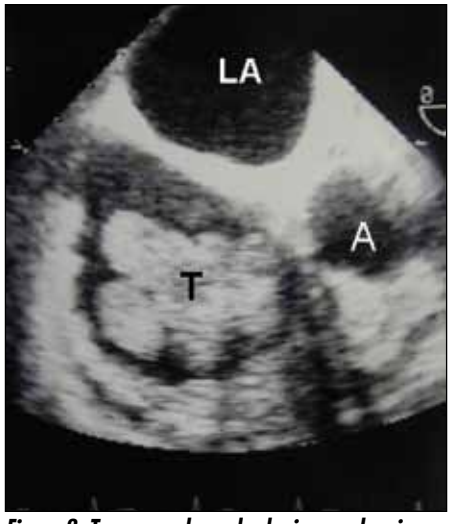


Figure 2. Transoesophageal echo image showing a lobulated tumour of dimensions 2.9 x 3.9 cm within the right atrium. A = aorta; LA = left atrium; T = tumour.

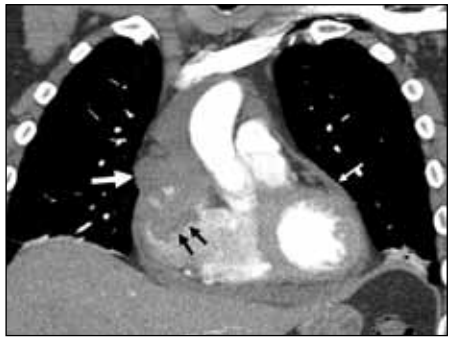


Figure 3. Computed tomographic scan (coronal plane), with intravenous contrast in the arterial phase, showing a mass in the right atrium (two black arrows) with two vascular spaces and extension through the pericardium (single large arrow). There is generalized pericardial thickening and an effusion (single small arrow).

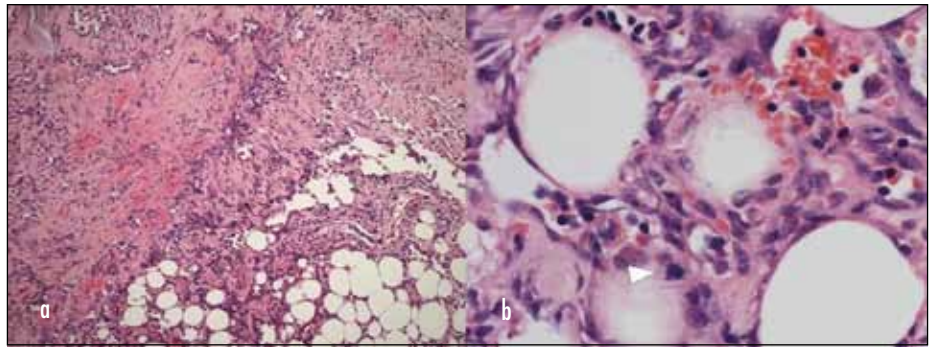


Figure 4. Histology of mediastinal biopsy showing (a) proliferation of irregular anastomosing vascular channels infiltrating fat and fibrous tissue (haematoxylin and eosin x100). b. A high-powered image shows hyperchromatic malignant cells with mitotic figures (arrow head) (haematoxylin and eosin x400).

Cardiac angiosarcomas are aggressive and associated with a poor prognosis. The mean survival is reported as 9–10 months (Kurian et al, 2006). Metastases are found in 66–89% of cases at the time of diagnosis, the pericardium, lungs, mediastinal nodes and vertebrae being frequent sites (Brandt et al, 2005).

As no randomized studies exist to guide treatment, management is individualized and multidisciplinary. Cardiac sarcomas are rarely cured, but prolonged survival is possible with surgical resection. The role of orthotopic heart transplantation for malignant cardiac tumours continues to be debated. Bench surgery (explantation and autotransplantation) is a relatively new approach and may help achieve more complete tumour resection (Reardon et al, 2006).

Post-surgical adjuvant radiation and chemotherapy have not proven consistently beneficial, but may ameliorate symptoms and improve quality of life.

Conclusions

This case illustrates the importance of performing an echocardiogram in patients with pericarditis and the role of subsequent imaging modalities. **BJHM**

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