# An unusual case of pericardial constriction

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## Introduction

**Pericardial constriction remains a rare condition. The precise aetiology is undefined in up to 50% of cases. We describe a case of rapidly progressive pericardial constriction and highlight how post-mortem examination remains useful in establishing unexpected diagnoses.**

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## Case report

A 60-year-old Afro-Caribbean woman presented with a short history of breathlessness and cough. Examination revealed a clear chest, tachycardia in the absence of fever, blood pressure of 110/60 mmHg (with a paradox of 25 mmHg) and no organomegaly, lymphadenopathy or skin lesions. Electrocardiography showed sinus tachycardia. Echocardiography revealed a moderate, circumferential pericardial effusion with evidence of right ventricular diastolic collapse. Uncomplicated pericardiocentesis via a subxiphoid approach yielded 700 mls of heavily blood-stained fluid.

Subsequent immunological screen and viral serology was negative. There was no evidence of an underlying bleeding diathesis and renal function was normal. Pericardial fluid examination revealed no malignant cells and no evidence of infection (including acid, alcohol fast- bacillus). Computerised tomography of the thorax revealed a markedly thickened pericardium and no parenchymal lung lesions. In the absence of reaccumulation of pericardial fluid over a 10-day period, despite negative viral serology, a diagnosis of viral pericarditis with haemorrhagic pericardial effusion was made. She was discharged home but represented 10 days later with chest pain. Examination revealed peripheral oedema, persistent tachycardia and raised venous pressure.

Repeat echocardiography showed no reaccumulation of pericardial fluid but the pericardium was noted to be thickened. The right ventricle was small and left ventricular function was good. Constrictive physiology was suggested by findings at cardiac catheterisation with typical left and right ventricular 'dip and plateau' patterns and equalisation of ventricular end-diastolic pressures at 28 mmHg.

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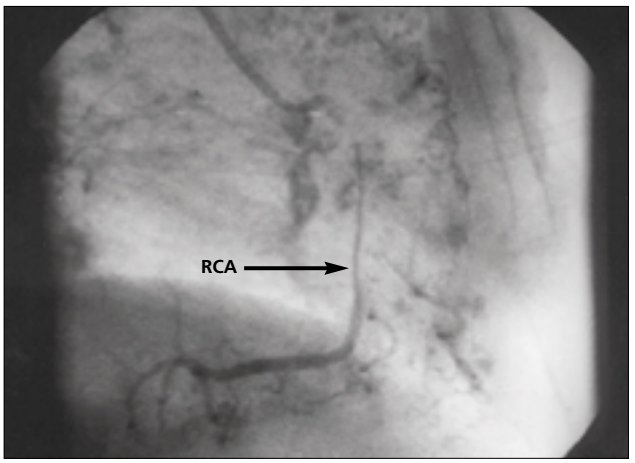
**Figure 1.** Right coronary angiogram (lateral projection) demonstrating fistulous connections from right coronary artery to pericardial space forming a 'vascular blush' effect




Coronary angiography demonstrated multiple coronary artery/myopericardial fistulous connections causing a 'vascular blush' effect (figure 1).

Whilst awaiting pericardiectomy she sustained a cardiac arrest. Resuscitation was unsuccessful.

Gross pathological examination at autopsy revealed thickened visceral and parietal pericardium. The pericardial cavity contained organised thrombus encasing the heart (figure 2). The cause of death was initially felt to be pericardial constriction due to pericardial haemorrhage in the setting of pericarditis, or potentially secondary to traumatic pericardiocentesis. Nevertheless, subsequent histological examination of the pericardium revealed a pleomorphic sarcoma forming slit-like and cavernous vascular spaces, staining positive with the endothelial marker CD 34 (figure 3).

## Discussion

In up to 50% of cases the cause of pericardial constriction remains undetermined. Only once before has angiosarcoma been described as causing pericardial constriction.[1] Whilst primary malignant tumours of the heart or pericardium are rare, angiosarcomas are the most commonly reported histological

**Figure 2.** Gross pathological specimen of sectioned left ventricle revealing encasement of myocardium by what was initially presumed to be thrombus but latterly shown to be angiosarcoma

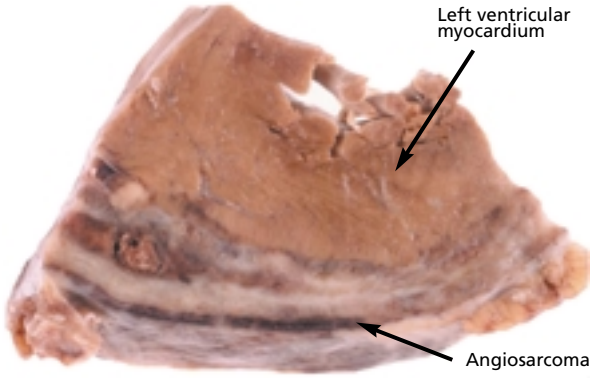
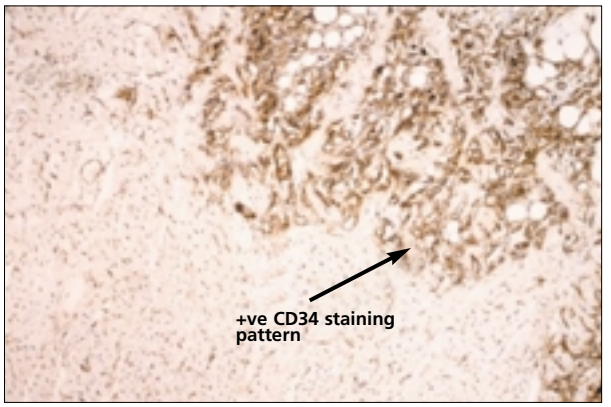



**Figure 3.** Histological specimen revealing a pleomorphic sarcoma forming slit-like and cavernous vascular spaces, staining positive with the endothelial marker CD 34




type.[2] As with angiosarcomas of other organs, those involving the heart display highly variable histological patterns, which often overlap those seen in Kaposi's sarcoma.[3] Cardiac angiosarcomas are most often located in the right atrial wall with frequent extension into the pericardium, vena cava or tricuspid valve. In Africans, Kaposi's sarcoma can occur in the heart in the absence of skin lesions, but lesions are usually small and localised with few clinical sequelae.[4] More recently, with the emergence of human immunodeficiency virus (HIV) infection and acquired immunodeficiency syndrome (AIDS), the incidence of Kaposi's sarcoma has risen.

Immunohistochemistry has been used to differentiate between angiosarcoma and Kaposi's sarcoma. There appears to be no significant difference in the positivity of endothelial markers Factor VIII, CD31 and CD34 in tumour cells but reactivity of CD34 does appear stronger in cases of Kaposi's sarcoma.[5]

In the above case, anatomical evidence would suggest that this tumour was an angiosarcoma, although immunohistochemical findings shared some features with Kaposi's sarcoma. In the described case, exact histology has no bearing on clinical outcome but clarification of the true nature of similar tumours removed at surgery may have relevance with regard to adjuvant chemotherapy. Kaposi's sarcoma in HIV disease is often disseminated and has been linked to human herpes virus (HHV) 8 infection. Serological markers for HIV or HHV-8 were not measured in this case but the patient had no risk factors and little clinical evidence to suggest underlying HIV infection and AIDS.

### Key messages

- Pericardial constriction can be caused – rarely – by angiosarcoma
- Angiosarcomas display highly variable histological patterns which can overlap those seen in Kaposi's sarcoma
- Angiosarcoma can be differentiated from Kaposi's sarcoma using immunochemistry; the reactivity of CD34 appears stronger in cases of Kaposi's sarcoma

## References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