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Pericardial obliteration by angiosarcoma

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SUMMARY Primary cardiac angiosarcoma caused pericardial obliteration and death from tamponade in a 66 year old man.

Angiosarcoma of the heart is an uncommon malignancy with a very poor prognosis. It most commonly presents as a bloodstained pericardial effusion, and fever, pericardial pain, weight loss, and night sweats are typical. Few patients survive more than a few months, partly because of delayed recognition, with tuberculosis being the commonest misdiagnosis. Echocardiography may be helpful in diagnosis, and there are a few reports of early operation and extended life span[1]; however, many cases are diagnosed at necropsy. The mode of death is obscure in most cases, though in some necropsy series the pericardial space was obliterated by the tumour, and death was caused by tamponade. We report such a case.

Case report

A 66 year old white man presented with a three month history of progressive exertional dyspnoea, dry cough, and anorexia. For two weeks he had experienced profuse night sweats and aching central chest pain made somewhat worse by deep breathing but not by any postural change; he had continued to work. There was no relevant past medical or family history, and no exposure to any industrial chemicals.

On examination he was feverish (37·8°C) but looked well. He had a resting sinus tachycardia, an impalpable apex beat, and very quiet heart sounds. Jugular venous pressure extended above the angle of the jaw. Arterial blood pressure was 95 mm Hg systolic in expiration with a 20 mm Hg paradoxical wave. A posterior-anterior chest *x* ray showed massive globular cardiomegaly, and a cross sectional echocardiogram confirmed a very large pericardial effusion, though the heart appeared to be normal.

The effusion was tapped and 3 l of bloodstained pericardial fluid was obtained: there was an immediate improvement in haemodynamic function. No malignant cells were obtained from the fluid, although numerous lymphocytes and some debris were seen. No acid fast bacilli were seen on smear or cultures. A Mantoux skin test was strongly positive at 1/1000. A presumptive diagnosis of tuberculosis was made and treatment was started with rifampicin, isoniazid, ethambutol, pyrizinamide, and prednisone.

He continued to lose weight and two weeks later was readmitted with lethargy and malaise. Findings were essentially unchanged except that he had lost 6 kg in weight, his jugular venous pressure was elevated by 10 cm, and the arterial paradox was 10 mm Hg. A repeat echocardiogram showed a small amount of residual effusion and material was seen to be floating in the fluid. This was thought to be fibrin and old blood. A repeat pericardiocentesis obtained only 300 ml of bloodstained fluid. No malignant cells or acid fast bacilli were seen in the fluid. There was no detectable arterial paradox after the procedure.

A computed tomogram of the thorax and upper abdomen confirmed a pericardial effusion with a normal heart (fig 1). No solid tumour was seen in the thorax. Multiple metastases were seen within the liver, however. That night the patient was found to

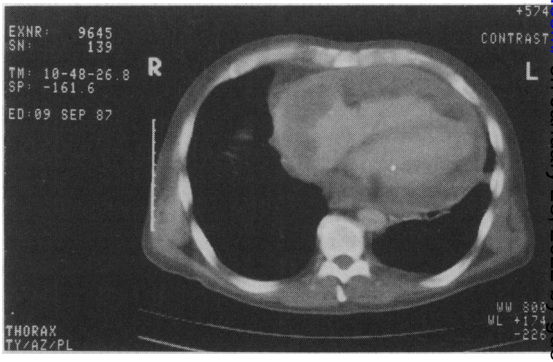

Fig 1 *Computed tomogram of the thorax at mid-thoracic level showing the heart surrounded by pericardial tumour.*

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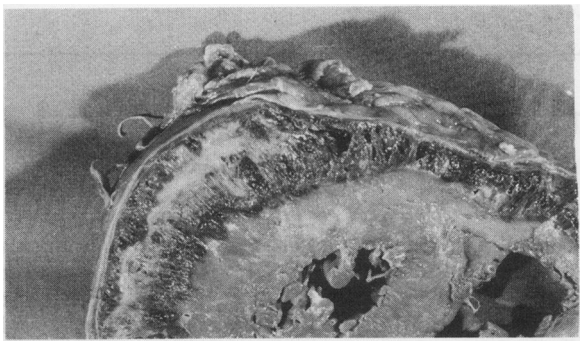

Fig 2 *Pathological specimen showing the heart encircled by necrotic tumour.*

be asystolic, and in view of the radiographic findings resuscitation was not attempted.

At necropsy important findings were confined to the heart and liver. The pericardial cavity was obliterated by a purple, haemorrhagic tumour infiltrating into the pericardial surface of the heart, with no clear plane of cleavage between the tumour and the heart (fig 2). The wall of the right atrium that was lateral and inferior to the opening of the superior vena cava was almost completely replaced by tumour. Multiple necrotic secondary deposits were found within the liver; no metastases were found in any other organs. Histology of the tumour showed it to be an angiosarcoma with extensive haemorrhage and necrosis.

Discussion

Primary angiosarcoma of the heart typically originates in or around the right atrium: dyspnoea may result from obstruction of the caval veins or of the right ventricular inflow and from pericardial tamponade. The fever, sweats, and high erythrocyte sedimentation rate are presumably a reaction to necrotic tumour. In contrast to hepatic angiosarcoma, there is usually no industrial exposure to environmental carcinogens. Although echocardiography may show a polypoid mass within the right atrium or pericardium, findings are not always diagnostic.

Glancy *et al* reviewed 41 cases of cardiac angiosarcoma at necropsy and found that in 13 the pericardial space was obliterated by tumour.[2] In the reviews by Janigan *et al* a similar proportion (20 of 43 cases) had pericardial obliteration by tumour,[3] and in both reviews a majority presented with haemopericardium. Many patients were previously fit and presented abruptly with large effusions in pericardial tamponade. Lin *et al* also described a case like ours in which an encasing pericardial angiosarcoma gave the appearance of a pericardial effusion.[4] Although echocardiography usually readily shows polypoid lesions within the pericardial space, we found great difficulty in differentiating between the fibrin and thrombus commonly seen in proteinaceous effusions and concentric sessile tumours. The computed tomograms in this case showed metastases but did not differentiate between pericardial fluid and tumour. Shin *et al* reported a cardiac angiosarcoma diagnosed by computed tomography, but the tumour was polypoid and clearly arose from the right atrium.[5]

Complex diagnostic aids have important limitations in the diagnosis of primary angiosarcoma of the heart if the tumour is confined in the thorax to the pericardial space and is not polypoid. Early open biopsy in persistent unexplained bloody pericardial effusion, as suggested by Strohl,[6] may allow early diagnosis.

References

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