Angiosarcoma of the heart

MARGARET SHACKELL, ANNA MITKO, P. L. WILLIAMS, AND G. C. SUTTON

From the Cardiac Departments of Hillingdon and Harefield Hospitals and the Pathology Department of Hillingdon Hospital, Middlesex

SUMMARY Two cases of angiosarcoma of the heart are described. In one the tumour, which arose from the right atrium, was demonstrated during life by angiography. In the other, diagnosed only at necropsy, the tumour arose from the right ventricle. Both cases illustrate many of the typical features of this rare tumour and the difficulties of ante-mortem diagnosis.

Primary tumours of the heart are rare. Straus and Merliss (1945), in a review of 480 331 necropsies, found the incidence of primary cardiac tumour to be 0.0017 per cent. Angiosarcoma is among the least common. It usually arises from the right atrium and only rarely from the pericardium, right ventricle, or left atrium.

Case reports

Case 1

A 42-year-old man was admitted in July 1976 complaining of severe lumbar and epigastric pain. He had first developed lumbar pain after a fall 2 years previously, but it had become more severe in the 6 months before admission. In March 1976 he had been admitted to the orthopaedic department of another hospital complaining of lumbar and epigastric pain, malaise, and weight loss. During that admission he was found to be anaemic, but no definite diagnosis was made and he was discharged on oral iron and indomethacin. There was no history of contact with vinyl chloride.

On examination he had a low grade pyrexia, intermittent mental confusion, and mild tachypnoea. The jugular venous pressure was raised 3 cm and he was in sinus rhythm at 70 to 110/min. The blood pressure was 90/50 mmHg without paradox. A pericardial friction rub was audible throughout the precordium. The electrocardiogram was normal and the chest x-ray film showed globular cardiac enlargement with normal lung fields. The haemoglobin was 12.1 g/dl with normal indices, the white cell count $13.3 \times 10^9$/l with 70 per cent neutrophils, and the erythrocyte sedimentation rate was 77 mm/hour. A Mantoux test and investigations for collagen disease were negative. X-ray film of the lumbar spine showed mild degenerative changes only. M-mode echocardiography confirmed the clinical impression of a pericardial effusion.

Six days after admission cardiac tamponade developed. Twenty-five ml blood-stained pericardial fluid were aspirated, with relief of the tamponade. Malignant cells were not seen on cytological examination of the aspirate. One month after admission open pericardial biopsy was carried out through a left thoracotomy. No tumour was seen. The biopsy showed a grossly thickened pericardium with extensive fibroblastic proliferation and focal aggregates of lymphocytes, but no evidence of malignancy.

The jugular venous pressure returned to normal after pericardial biopsy, and remained so during the rest of his illness. However, the patient continued to deteriorate. In October 1976 a bone scan showed patchy loss of uptake in the lumbar region and malignant cells were found on bone marrow aspiration. The patient developed a sinus tachycardia with a gallop at this time, and the electrocardiogram showed nonspecific ST and T wave abnormalities, though the chest x-ray film showed a normal cardiac silhouette. His condition deteriorated further and he died in December 1976.

At necropsy the main tumour was in the anterior part of the right ventricle, measuring $5 \times 4 \times 4$ cm and infiltrating the full thickness of the myocardium and endocardium. The pericardium was firmly adherent to the heart and contained numerous haemorrhagic metastases on the outer surface. There was confluent subpericardial tumour, approximately 0.5 cm thick, surrounding the whole heart. There was also a large number of haemorrhagic metastases in the lungs, liver, spleen, and lumbar

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Angiosarcoma of the heart

Fig. 1  Photomicrograph of a solid part of the tumour from the right ventricle showing a pleomorphic, spindle-cell pattern, with numerous capillary sized vascular spaces. (H and E. × 150.)

Fig. 2  Vertebral metastasis showing the vasoproliferative nature of the tumour and outlining the vascular spaces. (Reticulin. × 150.)
vertebrae. The histological appearance of the tumour in the pericardial space was that of a solid spindle-cell sarcoma, but in other sites, such as the liver and right ventricular wall, there were large vascular spaces in the tumour itself. In the vertebrae the tumour had definite vasoformative properties.

The overall appearances were those of angio-
sarcoma (Fig. 1 and 2).

CASE 2
A 48-year-old woman underwent subtotal thyroid-
ectomy for a multinodular goitre with substernal
extension in January 1977. The histology was of a
benign nodular colloid goitre. Her only symptom
after operation was nonspecific chest pain. But
whereas before operation the chest x-ray film had
been normal, afterwards it showed gross cardiac
enlargement. There were no abnormal physical
signs but the electrocardiogram showed generalised flatta-
ing of the T waves and frequent ventricular
ectopic beats. M-mode echocardiography showed
a large pericardial effusion. The haemoglobin was
10.3 g/dl with a normal blood film and the erythro-
cyte sedimentation rate was 50 mm/hour. Anti-
nuclear factor, autoantibodies, and Mantoux test
were negative, and a diagnosis of viral or idiopathic
pericarditis was made.

One week later a pericardial friction rub was
heard. The effusion cleared spontaneously, con-

dirmed by echocardiography, with conservative manage-

ment only and the patient was discharged 3
weeks after admission for subtotal thyroidectomy.

Chest pain and malaise continued after discharge
and treatment with tetracosactrin zinc 0.5 mg twice
weekly was started 2 months later. This was changed
to prednisolone 40 mg daily after one week, when
she was readmitted. The chest x-ray film now
showed a bulge on the right border of the heart
(Fig. 3). Right atrial angiography showed an
apparent outpouching of the right atrium (Fig. 4).
She was discharged again after 3 weeks, on pred-
nisolone, but continued to complain of nonspec-
ific pain in the chest and arms, together with
weight gain and shortness of breath. She was
admitted on 2 further occasions with an increase
in the jugular venous pressure and sinus tachy-
cardia, but at no stage was there clinical or echo-
cardiographic evidence of reaccumulation of peri-
cardial fluid.

At the end of June 1977, because of her un-
explained deterioration, cardiac catheterisation was
repeated. Right atrial angiography now showed a
mass about 2 cm in diameter, projecting into the
right atrium. A left ventricular injection showed an
abnormally contracting left ventricle, its anterior
surface, from the aortic root to the apex, being
almost completely akinetic. Injection into the left
coronary artery was followed by a blush over the
whole of the base of the heart, and injection into the
right coronary artery showed the proximal part of the
vessel to be displaced around a non-opacified area.
After arterial clearing there was again a blush in the

Fig. 3 Chest x-ray film (case 2) showing a bulge on the
right border of the heart.
region between the right coronary artery and the right border of the heart. The appearances were those of a tumour of the right atrium with involvement of the base of the heart.

Subsequent chest x-ray films suggested secondary deposits in the lungs. Needle biopsy of the lung was attempted in order to make a histological diagnosis, but no tumour was obtained. The patient died the next day.

At necropsy the heart weighed 1300 g. There were
multiple haemorrhagic tumour nodules, from 1 to 5 cm in diameter, scattered over the entire epicardial surface. The tumour appeared to be originating from the right atrium where there was an intracavitary extension, approximately 4 cm in diameter, reducing the cavity (Fig. 5). There were multiple pericardial adhesions to the surface over epicardial nodules. Metastases were present in brain, lungs, liver, and ovaries, mainly 0·5 to 2 cm in diameter. The histology was that of angiosarcoma (Fig. 6).

Discussion

The constancy of symptoms and physical signs in patients with angiosarcoma of the heart has been stressed by many authors (McNalley et al., 1963; Glancy et al., 1968; Hollingsworth and Sturgill, 1969). The most frequent symptoms are general malaise, chest pain, fever, and haemoptysis, and the physical signs include a pericardial friction rub and raised jugular venous pressure. The chest x-ray film may show cardiomegaly, sometimes with a bulge on the right border of the heart. The electrocardiogram may be normal at the time of presentation or show nonspecific ST and T wave abnormalities. Echocardiography may confirm a pericardial effusion, which typically is blood-stained on aspiration. Ports et al. in 1977 described 4 cases of right ventricular tumour, 3 myxomas, and 1 metastatic melanoma, in which the tumour was seen on M-mode echocardiography. In 2 cases the site of attachment of the tumour was shown by twodimensional echocardiography.

The prognosis is poor, with few patients surviving more than a few months from presentation. At necropsy there is obliteration of the pericardial sac by tumour and often pulmonary and rib metastases. Though it is often difficult in the presence of extensive infiltration of surrounding structures by the tumour to determine the precise site or origin, it usually appears to arise from the right atrium. Other sites are much less common.

We have found 15 cases reported in which the probable or possible site of origin was the pericardium (Glancy et al., 1968; Hollingsworth and Sturgill, 1969; Hansson et al., 1970; Poole-Wilson et al., 1976), 6 in which it was the right ventricle (Lange and Christiansen, 1947; Amsterdam et al., 1949; Tatsumi et al., 1949; Glassy and Massey, 1950; Groom, 1956; Thompson et al., 1977), and 2 in which it was the left atrium (Gross and Englert, 1937; Hager et al., 1970).

Few cases of angiosarcoma of the heart have been diagnosed in life. In a review of the published reports in 1968, Glancy et al. found, out of 41 cases of angiosarcoma of the heart, 12 in which the diagnosis had been suspected clinically and only 7 in which it was confirmed before death. We have found a further 8 cases in which the diagnosis was made before death (Allaire et al., 1964; Hollingsworth and Sturgill, 1969; Hager et al., 1970; Laws et al., 1973; Poole-Wilson et al., 1976; Rossi et al., 1976; Thompson et al., 1977).

In the 7 cases cited by Glancy et al. the diagnosis was made by biopsy of the tumour or its secondary deposits in 4, by demonstrating tumour cells in the pericardial fluid in 2, and the diagnosis was suspected at right heart angiography in 1, though not confirmed histologically during life. In the 8 other cases we have found, the diagnosis was made at thoracotomy in 6 cases (Allaire et al., 1964; Hollingsworth and Sturgill, 1969; Hager et al., 1970; Laws et al., 1973; cases 1 and 4; Poole-Wilson et al., 1976) and by angiocardiography followed by thoracotomy in 2 (Rossi et al., 1976; Thompson et al., 1977).

Despite the fact that we suspected malignancy from an early stage in our first case, we were unable to confirm this until a bone-marrow aspiration 10 weeks after initial admission, and in spite of cytological examination of the pericardial fluid and open pericardial biopsy. It was particularly unusual to fail to make the diagnosis at open exploration of the pericardium, especially as this was carried out at a time when the disease must have been far advanced. The histological diagnosis of angiosarcoma of the heart was only made at necropsy. There are a number of other published cases in which cytological examination of the pericardial fluid or pericardial biopsy was unhelpful (Hollingsworth and Sturgill, 1969; Hager et al., 1970; Hansson et al., 1970; Patt et al., 1974; Poole-Wilson et al., 1976). Nevertheless, open pericardial biopsy or thoracotomy remain the most fruitful methods of diagnosis.

In our second case a cardiac tumour was diagnosed shortly before death by angiocardiography. In this case the unusual features were the spontaneous resolution of the pericardial effusion and the appearance of an outpouching of the right atrium at the first angiocardiogram. In retrospect the latter feature was probably produced by the tumour, though it was not suggestive of this at the time. Spontaneous resolution of pericardial effusion in this condition has not been described previously, though Patt et al. in 1974 described a case of resolution occurring after a single pericardiocentesis.

In neither of our cases did M-mode echocardiography show any evidence of a tumour. Echocardiography is an invaluable aid in the diagnosis of pericardial effusion, but the M-mode technique may not be useful in detecting pericardial or myocardial tumours. Two-dimensional echocardiography is

Margaret Shackell, Anna Mitko, P. L. Williams, and G. C. Sutton
Angiosarcoma of the heart

more likely to provide diagnostic information, as described by Ports et al. (1977).

We conclude nevertheless that the best methods of making the diagnosis of primary angiosarcoma of the heart are by open exploration of the heart and pericardium, and by angiocardiography. The former should enable a histological diagnosis to be made, though, as our first case illustrates, this is not invariably so. Angiocardiography should disclose a characteristic appearance of a tumour, though correct interpretation may be difficult, as our second case illustrates.

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Requests for reprints to Dr G. C. Sutton, Cardiac Department, Hillingdon Hospital, Uxbridge, Middlesex UB8 3NN.
Angiosarcoma of the heart.

M Shackell, A Mitko, P L Williams and G C Sutton

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