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Angiosarcoma of the heart
Unusual presentation and survival after treatment

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SUMMARY A 49 year old man presented with severe cyanosis and dyspnoea on exercise. Clinical examination together with echocardiography, cardiac catheterisation, and angiography showed a balloting tumour in the right atrium, intermittently occluding the tricuspid ostium, and an atrial right to left shunt. At operation a pedunculated vascular tumour was found with a broad base which was embedded in the atrial wall and continued into the interventricular septum. Histological examination showed angiosarcomatous features and signs of a less than radical excision. The patient, who made an uneventful recovery, was given postoperative radiotherapy. After 36 months there are no signs of recurrence or metastasis.

Primary tumours of the heart are rare, and in review of 480 331 necropsies Straus and Merliss[1] found an incidence of only 0·17 per 1000. Angiosarcoma is among the least common of such tumours,[2] and the signs and symptoms with which it presents are well documented.[3 4] The most common symptoms are malaise, chest pain, fever and haemoptysis, while the physical signs include raised central venous pressure and a precordial friction rub. The tumour usually arises from the right atrium[3 5] and only rarely from other parts of the heart.[6 7] The diagnosis in reported cases has usually been made at necropsy or immediately before death.[3] When operation has been attempted signs of inoperability have usually been encountered, and only a few patients have survived for more than six months.[8]

Case report

In June 1980 a 49 year old male truck driver was admitted to hospital. During the previous two years, he had become increasingly dyspnoeic during exercise and he had noted bluish discoloration of the skin during physical activity. On examination he was strikingly cyanotic, and even on minor exercise the cyanosis increased and he became breathless. The patient was in sinus rhythm at a heart rate of 84 beats/min and his blood pressure was 120/100 mm Hg; slight clubbing of the fingers was present. A tall a wave in the jugular venous pulse was noted. On auscultation a diastolic murmur with a distinct presystolic component was audible in the fourth intercostal space by the left parasternal border and an early ejection systolic murmur was weakly audible in the same area. Neither hepatomegaly nor dependent oedema was found.

The chest x ray film showed a normal heart configuration and normal lung fields. Pulmonary spirometry and pulmonary scintigram were normal. Blood haemoglobin concentration was 20·4 g/100 ml, the packed cell volume 67%, and the erythrocyte sedimentation rate 1 mm in the first hour. Pa_{O_2} was 41·3 mm Hg (5·5 kPa) and Pa_{CO_2} was 25·2 mm Hg (3·35 kPa). The electrocardiogram showed P waves with amplitude exceeding 0·25 mV in limb leads II and III and precordial lead V1 together with depression of the PR segment. The graded bicycle ergometer test had to be discontinued after 3 minutes at 48 W because the patient became breathless and increasingly cyanotic. Heart rate rose to 118 beats/min during exercise.

The echocardiogram showed a space occupying structure in the right atrium moving into the tricuspid orifice during diastole (Fig. 1). Cardiac catheterisation showed tricuspid stenosis with a pressure difference between the right atrium and right ventricle of 8 mm Hg, increasing to 20 mm Hg during atrial systole. A defect in the atrial septum and a pressure gradient of about 7 mm Hg from right to left atrium was also found. By the hydrogen method no left to right shunt could be detected. The fraction of total venous return shunted from right to left atrium was 43% (cal-

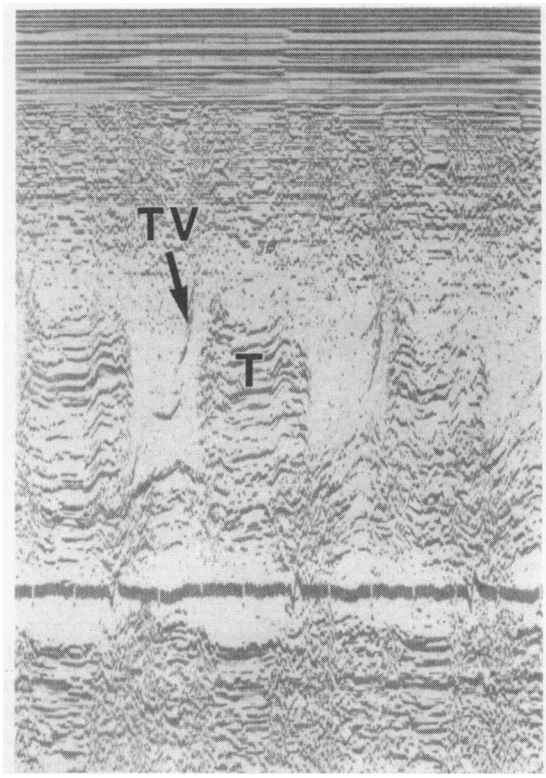

Fig. 1 *M mode echocardiogram of the tricuspid valve (TV). Tumour echoes (T) are seen behind the leaflets. The echo producing structure nearly fills the ostium during diastole.*

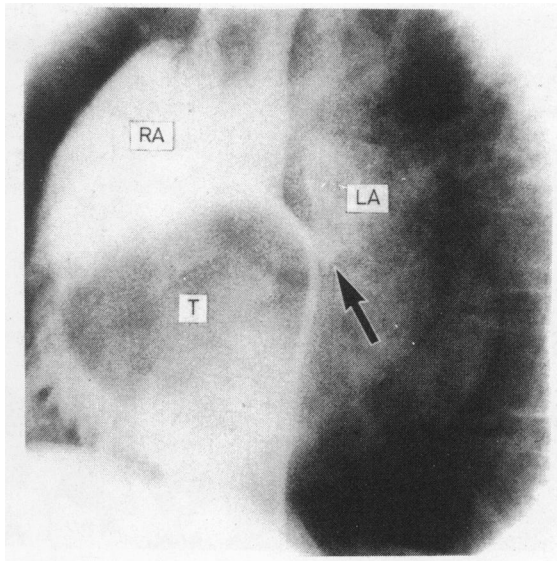

Fig. 2 *Angiocardiogram, right oblique view. Tumour (T), left atrium (LA) and right atrium (RA), and the atrial right to left shunt (arrow).*

culated from oxygen saturations in blood samples from the right atrium, the pulmonary vein, and the femoral artery). Angiocardiography also showed a space occupying structure in the right atrium moving partially into the right ventricle (Fig. 2) during diastole and a right to left shunt at the atrial level.

During operation on total cardiopulmonary bypass with cold cardiac arrest, a movable tumour filling almost the whole right atrium was found. The foramen ovale was wide open and the tricuspid orifice was widened. The tumour was round with a diameter of about 7 cm and was coloured blue grey with swollen veins. The oval base had a diameter of about 3 cm and the tumour continued below the fibrous trigone into the intraventricular septum, from which a rounded and rather well defined protuberance was enucleated. The cutting edges were apparently into normal myocardial tissue, and no obvious infiltrative growth was seen. Neither the mitral apparatus, the bundle of His, the coronary sinus, nor the circumflex branch of the left coronary artery was damaged. The foramen ovale and the inflicted atrial septal defect were closed with running monofilamental nylon 4–0. The patient made an uneventful recovery and experienced immediate subjective functional improvement. The excised tumour (Fig. 3) had a distinct external capsule and histological examination showed a well differentiated haemangiosarcoma, with vascular spaces lined with atypical endothelium and partly filled with erythrocytes (Fig. 4). A few mitoses and scattered

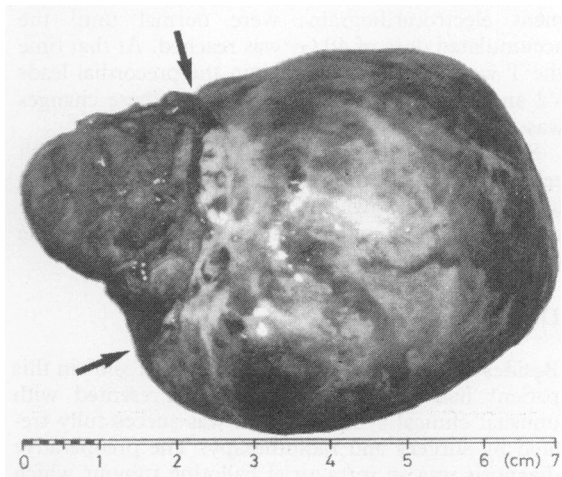

Fig. 3 *Photograph of resected tumour. Arrows show the borderline between the intramyocardial and the free pedunculated parts.*

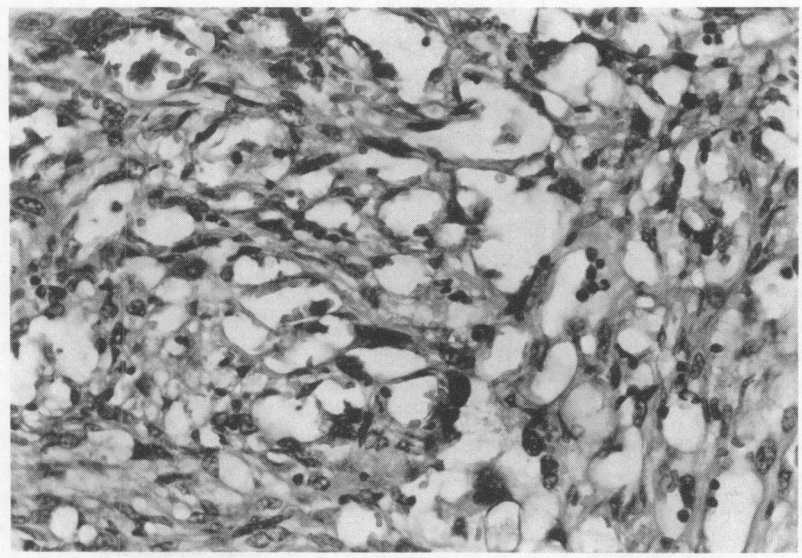

Fig. 4 *Light micrograph of paraffin embedded tumour tissue. Vascular cavities covered by spindle shaped cells with atypical nuclear pattern and pleomorphism are seen. Haematoxylin and eosin stain (original magnification × 350).*

large nuclei were seen, although in general only moderate nuclear pleomorphism was present. Tumour tissue had infiltrated into fat tissue and heart muscle and extended to the resection border. Because the resection was probably incomplete postoperative radiotherapy was given (Dr O Solheim). The irradiation was planned on the basis of computed tomography and given with photon beams by a 16 MV linear accelerator. Two oblique anterior fields with wedge filters included most of the heart. The total dose in the critical tissue was 50 Gy. During the radiation treatment electrocardiograms were normal until the accumulated dose of 40 Gy was reached. At that time the T waves became negative in the precordial leads V2 and V3, but as the importance of these changes was uncertain, the irradiation was continued.

Subsequent clinical examinations, together with echocardiography and computed tomography of the heart, have not shown any signs of tumour recurrence or metastasis 36 months after the operation. The patient is in good health.

Discussion

Besides being a rare tumour, the angiosarcoma in this patient had two rare features: it presented with unusual clinical symptoms and was successfully treated by surgery and radiotherapy. The preoperative diagnosis was an intra-atrial balloting tumour which intermittently obstructed the tricuspid ostium. The raised right atrial pressure had effected an atrial right to left shunt through the foramen ovale, explaining the considerable cyanosis on effort. A case of atrial myxoma with a similar clinical picture has been reported,[9] and this diagnosis was considered most likely. At operation, however, it was apparent that the tumour was not a myxoma but was probably malignant. A wide excision was not easily performed especially along the extension into the interventricular septum. The intracardiac defect was easily repaired, and apart from a temporary tricuspid insufficiency no postoperative sequelae have been detected.

The histological picture was typical of angiosarcoma or haemangioendotheliosarcoma,[3 4 10] although not highly undifferentiated. Pleomorphism and mitotic activity were moderate. Infiltrating growth along the line of resection indicated less than radical excision, and postoperative radiotherapy was considered appropriate.[11 12]

In an extensive study on previously reported cardiac angiosarcoma (56 cases) Gröntoft and Hellquist[3] state that no other type of sarcoma is so strictly located to the right atrium. In their study, the mean age of the patients at diagnosis was 41 years and the tumour was three times as common in men as in women. Only seven cases were diagnosed before death, with an average survival of six months, and 65% had distant metastases at necropsy. Pedunculated growth, without early infiltration and distant metastases, as described in this report, is obviously rare.

This case shows that cardiac angiosarcomata may present clinically like myxomata and that treatment can be successful.

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