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FIBROSARCOMA OF THE HEART: CASE REPORT

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Introduction

CARDIAC tumours have aroused interest among clinicians and with advances in cardiology there is a greater awareness of their varied presentations. Because of their diagnostic interest and the fact that their infrequency precludes familiarity, a case of fibrosarcoma of the heart is reported.

Case Report

The patient was a nineteen year old soldier who was first seen in St. Bricin's military hospital in July 1969. He complained of praecordial pain of two weeks duration, with occasional radiation to the left shoulder, periodic dyspnoea at rest and nocturnal dyspnoea.

Past history was of abscesses in the neck at the age of two years, which were surgically drained. There was no history of rheumatic fever.

Physical examination then showed evidence of cardiac enlargement and a pericardial friction rub was heard all over the praecordium. He was pyrexial (99-102°F.). Chest X-ray (Plate 1) showed a marked generalised increase in cardiac size and E.C.G. tracing showed flattening of T waves in leads II, V_2-V_6 and inversion of T waves in leads III and A.V.F. Laboratory investigations: E.S.R. 42 mm/hour (Westergren); anti-streptolysin O titre-12 todd units; haemoglobin 12g./100 ml. A tuberculin test (1st strength PPD) was reported as weakly positive.

He was treated with penicillin and aspirin and improved generally for three weeks. His E.S.R. and temperature returned to normal and the friction rub disappeared with some reduction in cardiac size on X-ray. He was then discharged from hospital but readmitted after a few days with the same pain. This time it was worse on deep inspiration and he also complained of nausea and general malaise. On examination, then, his pulse was 120/min; jugular venous pressure markedly raised; temperature 101.4°F; blood pressure 110/70; the pericardial friction rub had returned and a chest X-ray showed an increase in the cardiac outline. His E.S.R. was 25 mm/hour. At this stage a tentative diagnosis of tuberculous pericarditis was made, he was started on anti-tuberculous chemotherapy and was transferred to the Mater Hospital on 21 September 1969.

On examination, he was not distressed but nervous. He was mildly pyrexial. Pulse 100 beats per minute, regular, paradoxical. Blood pressure

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90/60. Jugular venous pressure raised 5-6 cm increasing with inspiration. There was no abnormal chest wall movement and the heart did not appear to be enlarged. Heart sounds were diminished in intensity but normal. A pericardial rub was heard maximal in intensity at the left sternal border. The liver was not palpable and there was no peripheral oedema. Scars were noted at the right side of the neck. Chest X-ray showed a general increase in the cardiac size and on X-ray screening there was diminished cardiac movement. No calcification was noted. E.C.G. tracing was low voltage and showed T wave inversion in leads II, III, AVF and V_1-V_6.

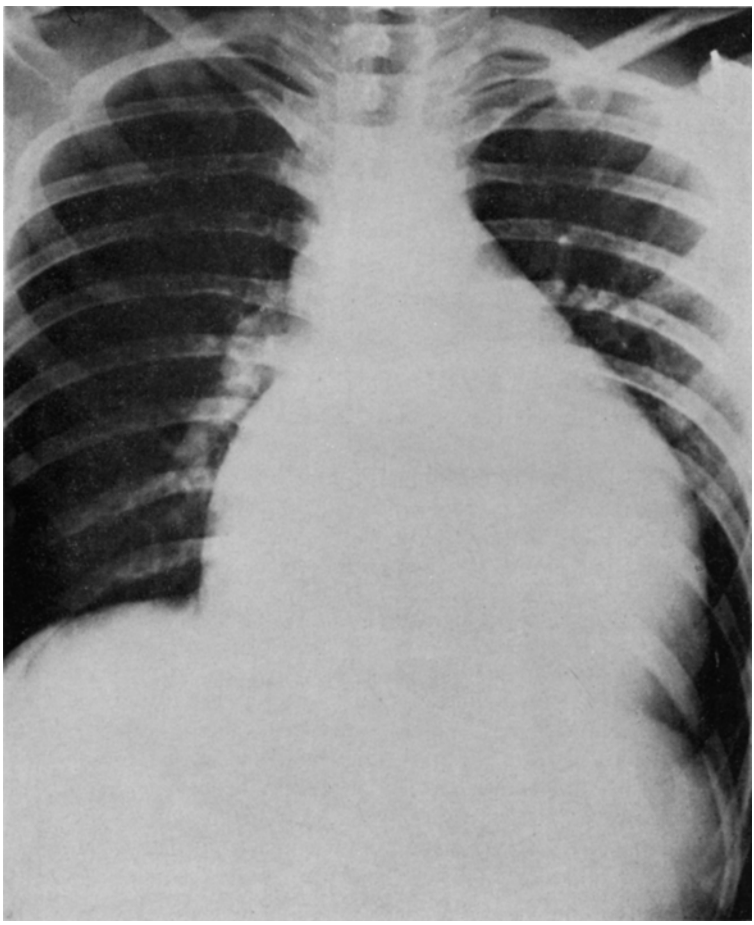

Plate 1—Chest X-ray (portable). Marked generalised cardiac enlargement.

Laboratory findings: E.S.R. 5-8 mm/hour (Westergren) on several readings; haemoglobin 14.7 g./100 ml; PCV 47 per cent; MCHC 31 per cent; white cell count 18,900/cu.mm; neutrophils 60 per cent; lymphocytes 34 per cent; monocytes 5 per cent; eosinophils 1 per cent. Urinalysis, blood urea, serum electrolytes, serum proteins were normal. Antistreptolysin-0 titre 125 Todd units; LE test (latex) negative. A tuberculin test (1st strength PPD) was now read as negative.

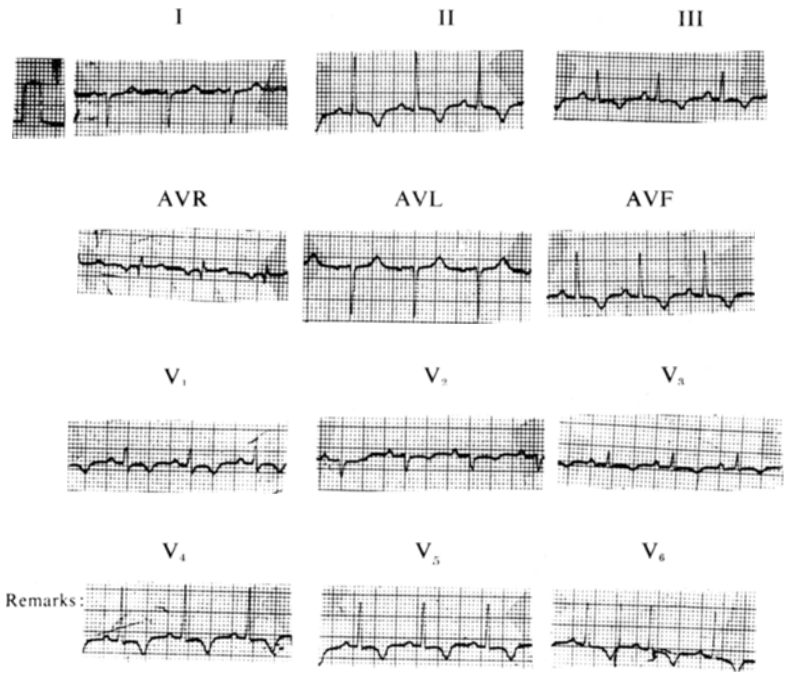

Plate 2—Electrocardiograph.

Pericardial paracentesis was attempted and a few cc of bloodstained fluid were removed with difficulty. The impression was got of a pericardium of great thickness. The fluid obtained was negative for organisms by Gram and Ziehl-Nielsen stains and culture was sterile.

Two days after paracentesis the patient's condition worsened and he suffered from increasing dyspnoea, chest pain and vomiting during which he became distressed and cyanotic. His venous pressure remained high, his liver was now palpable but there was still no peripheral oedema. He had several syncopal attacks, relieved by putting him in the supine position. At this stage, the validity of the diagnosis was in doubt and it was felt that something more than pericarditis was aetiological. His anti-tuberculous drug therapy was discontinued as he was unable to tolerate it and he was treated with digoxin, diuretics and venesection, 300 cc's of blood being taken off. His condition deteriorated and he abruptly went into severe congestive heart failure. Despite vigorous treatment there was no response and cardiac arrest was the terminal event. Resuscitation was unsuccessful.

At autopsy performed by Dr. W. Kealy, the heart weighed 1000 grams. The right atrium was distended by a large, yellow, smooth, multilobulated mass, 6 cms at its maximum diameter. It was attached to the posterior wall of the right atrium by a pedicle and extended through the tricuspid orifice into the right ventricle behind the chordae tendineae. It infiltrated through the posterior wall of the atrium and had expanded behind the heart into a large, soft, yellow, haemorrhagic mass, covering the right ventricle. The pericardium showed generalised fibrous adhesions and tumour tissue in-

vaded the posterior aspect. The lungs were moderately congested and oedematous. The liver was enlarged and showed signs of chronic venous congestion. Metastases were not found.

Microscopy showed a very cellular tumour composed of small spindle cells with hyperchromatic nuclei. Small amounts of intercellular collagen were present and infiltration of the myocardium was evident. Mitoses were seen but were infrequent. Staining for muscle striation and fat was negative.

Discussion

Primary tumours of the heart are rare. Strauss and Merliss in 1945 reported an incidence of 0.000017 per cent of 480,331 post mortems. Ben-

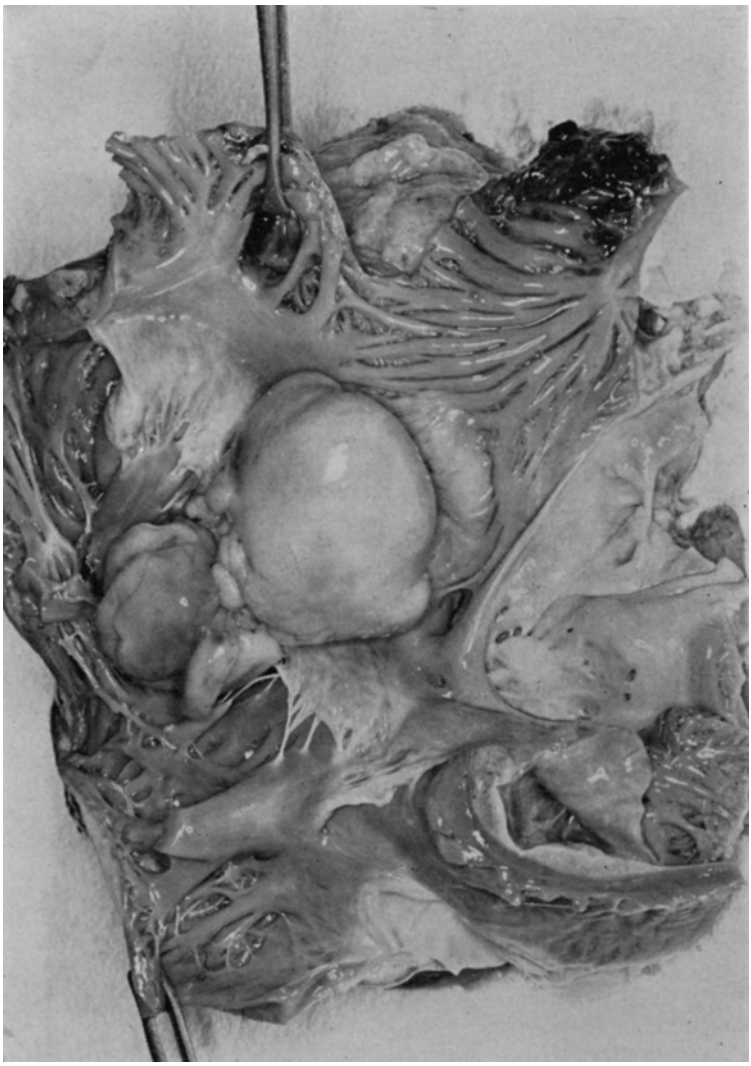

Plate 3—Multi lobulated tumour extending through the tricuspid orifice.

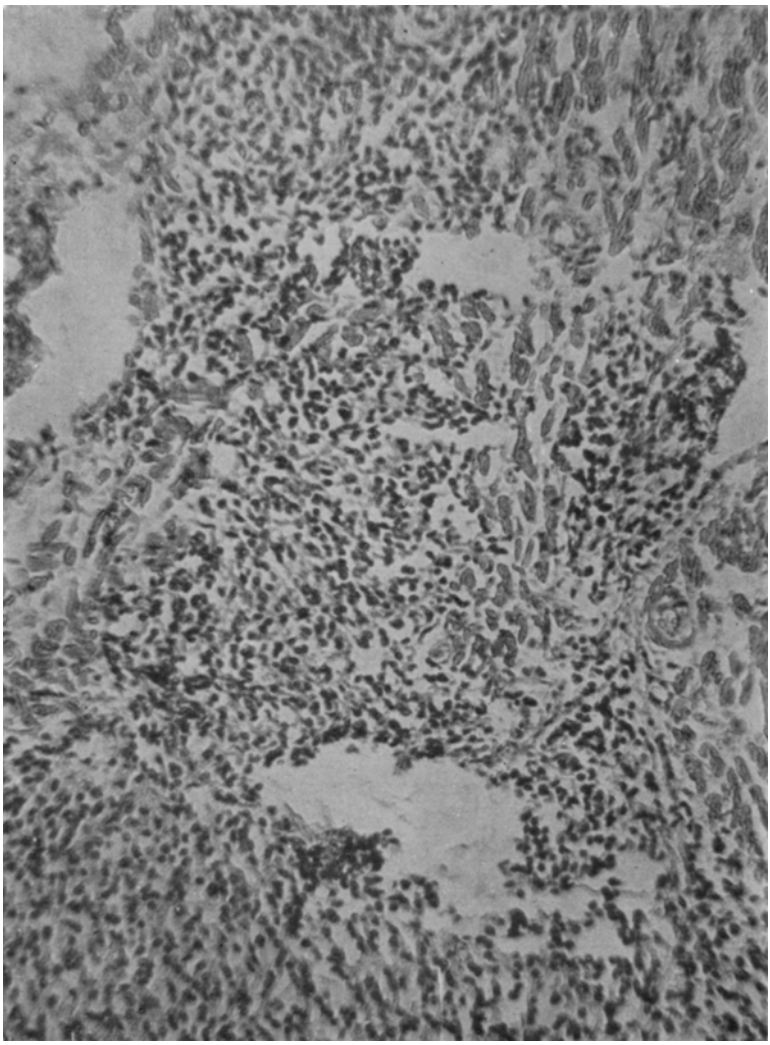

Plate 4—Microscopy. (H & E). Tumour showing infiltration of myocardium.

jamin and Lymburner reported in separate and much smaller surveys an incidence of 0.05 per cent and 0.03 per cent respectively. Secondary tumours are more common, 3.9 per cent of deaths due to cancer having cardiac metastases (Prichard 1951). All primaries are of mesoblastic origin and 25-30 per cent are malignant (Friedberg).

The majority of sarcomas arise from the atria, predominantly the right (Yater 1931). They include the fibrosarcoma, myxosarcoma, leimyosarcoma, rhabdomyosarcoma, angiosarcoma, reticulum cell sarcoma, lymphosarcoma, liposarcoma and neurosarcoma. Secondary spread occurs in about 40 per cent, mainly to the lungs and thoracic lymph glands (Wharton 1951). The fibrosarcoma occurs at any age, but is more frequent in children. It frequently spreads to the vertebral column or parenchymal organs (Heath

1968). It is made up of soft white tissue showing areas of haemorrhage and necrosis. As shown by this case it infiltrates the tissues of the heart and may extend from one atrium to the other. Symptoms are due to interference with passage of blood through the heart, impaired conduction, pericardial reaction, emboli, or, more rarely, extensive myocardial damage.

These may present in several forms, depending whether the tumour is endocardial, myocardial, pericardial or mixed.

1. Congestive heart failure unresponsive to therapy.
2. Acute circulatory failure leading to syncopal attacks or sudden death.
3. Chronic tamponade.
4. Acute pericarditis or pericardial effusion.
5. Arrhythmias or conduction defects.
6. Signs simulating valvular disease.
7. Emboli from blood clot attached to tumour or tumour itself. Notable in absence of atrial fibrillation.
8. Constitutional disturbance of a non-specific nature, e.g. chest pain, pyrexia, anaemia, leucocytosis, raised ESR, clubbing, haemoptysis; characteristic of myxoma (Goodwin *et al.* 1962) but reported by Sterns *et al.* in a primary sarcoma in 1966.

Sarcomas are usually mural but presenting features are frequently due to intra-cavitary extension (Goldberg *et al.* 1955). The chest X-ray may be helpful in that a bizarre cardiac outline, pericardial effusion or lung metastases may be shown (Steiner 1968). The ECG is non-specific (Friedberg 1966), though a pericarditis pattern may be shown where there is pericardial involvement (Harvey 1968).

Diagnosis is by suspicion, knowledge of clinical signs and diagnostic procedures, angiography being the best. Cleland *et al.*, 1969, point out the hazard of producing tumour or thrombus embolism to the lungs with the cardiac catheter.

Pericardial aspiration may show a blood-stained effusion, though this is not invariable and frequently tumour cells may be found in the fluid.

Our case presented as chronic tamponade. Hurst *et al.* pointed out that the occurrence of tamponade, in the absence of TB or other obvious cause, was strongly suggestive of neoplastic disease. The past history of abscesses in the neck directed the thinking towards a diagnosis of tuberculous constrictive pericarditis and, for a time, the patient was on anti-tuberculous chemotherapy, without notable effect, until unable to tolerate it.

In retrospect, the passage of the needle, during pericardial aspiration, through what was interpreted as thick pericardium was, in fact, through the tumour itself. Emmanuel and Lloyd 1962 reported a case of a right atrial myxoma presenting as a constrictive pericarditis attributed to TB as there was evidence of TB elsewhere in the body. They emphasised the importance of angiography in the diagnosis as cardiac catheterization failed to demonstrate the tumour. Due to the rapid advance of the disease and the

intolerance of the patient of diagnostic procedures, angiography was not possible in this present case.

It is suggested that the syncopal attacks suffered by this patient were due to transient tricuspid valve obstruction by the tumour. Such a presentation has been quoted by Harvey, 1968, as seen at a teaching symposium in Georgetown, 1966. Without angiographic evidence one cannot be certain of this and as has been noted the patient was of an intensely nervous disposition. Syncopal episodes are more common with tumours obstructing the mitral valve.

Surgery of malignant tumours of the heart is not often feasible. Castaneda *et al.*, 1968, scarcely mention it in a recent review of surgical considerations of heart tumours. Friedberg, 1966, however, points out that sarcomata arising in the right atrium have been removed successfully with survival for several years. They are radio-insensitive (Walter and Israel).

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