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Left atrial rhabdomyosarcoma and the use of digital gated computed tomography in its diagnosis

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SUMMARY A case of left atrial rhabdomyosarcoma presenting in a patient with known mitral stenosis is described. Computed tomography of the left atrium, gated to the electrocardiogram cycle, was used to visualise the site and extension of the tumour.

With the exception of myxomata primary intracardiac tumours are rare. The presence or later appearance of an intracardiac tumour may be overlooked in a patient with rheumatic mitral valve disease.

We report a case of rhabdomyosarcoma of the left atrium in a patient with coincident mitral valve disease. We believe that this is the first such atrial tumour to be diagnosed with the help of gated computed tomographic scanning techniques.

Case report

A 56 year old woman presented to hospital in 1977 complaining of dyspnoea of seven days' duration. She had previously been well and in particular there was no history of rheumatic fever, ischaemic heart disease, or pulmonary disease. There was no antecedent chest pain. On examination she was found to be in left heart failure with rapid atrial fibrillation. An apical systolic murmur was heard. She responded to treatment with digoxin and a diuretic. During convalescence an M mode echocardiogram showed evidence of mild mitral stenosis. Direct current conversion was performed and she was discharged with a diagnosis of mild mitral valve disease and decompensation precipitated by the onset of atrial fibrillation. She was kept under regular review and in 1978 she was found to be in atrial fibrillation again.

In 1979 a second echocardiogram was performed and again showed evidence of mild mitral stenosis. In September 1983 the patient complained of increasing dyspnoea and vague upper chest discomfort. A chest radiograph showed elevation of the right hemidiaphragm, with possible overlying pulmonary collapse and consolidation, and a right pleural effusion. There was also increased opacity in the lower part of the right hilum. She was admitted to hospital for investigation.

She was noted to be anaemic (haemoglobin 9·6 g/dl). The cardiac rhythm was atrial fibrillation, and blood pressure was 130/80 mm Hg. The venous pressure was not raised. The only remarkable findings on auscultation were an opening snap and an apical systolic murmur of moderate intensity. Physical signs in the chest were in keeping with the radiological findings. The liver was not enlarged. The erythrocyte sedimentation rate was consistently higher than 100 mm in one hour. Plasma protein electrophoresis and serum immunoglobulins were normal, as was plasma biochemistry. An electrocardiogram showed controlled atrial fibrillation with a QRS axis of 90°. Tomography demonstrated that the shadowing at the right hilum was the result of left atrial enlargement. At this stage an echocardiogram showed a large mass occupying the left atrium which appeared to be fixed posteriorly and was not attached to the interatrial septum (Fig. 1).

A digital computed tomographic scan of the thorax, gated to the R wave of the electrocardiogram, was performed at the level of the left atrium. The scan confirmed the presence of the filling defect, and showed a rim of contrast material separating the lesion from the interatrial septum (Fig. 2). Bilateral pleural effusions were seen on other views.

Subsequent right heart catheterisation demonstrated a pulmonary artery pressure of 74/30 mm Hg and a mean pulmonary capillary wedge pressure of 42 mm Hg on the right and 28 mm Hg on the left. Pulmonary angiography showed normal pulmonary

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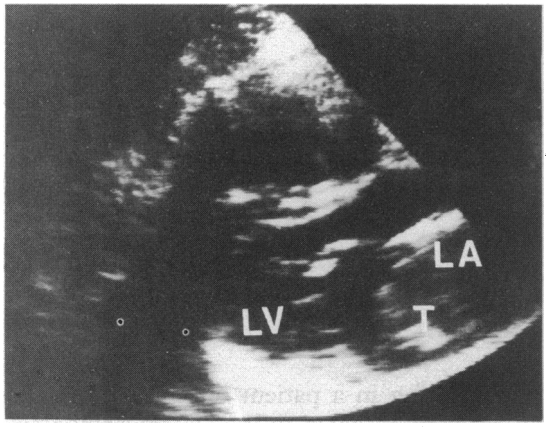

Fig. 1 *Preoperative cross sectional echocardiogram (long axis parasternal view) showing tumour (T) in the left atrium (LA). LV, left ventricle.*

arteries. Only the left pulmonary veins could be identified. The filling defect was seen to occupy most of the left atrial cavity. The mitral valve cusps were thickened. The appearance of the left ventricle and aorta was unremarkable.

At operation in November 1983 the right atrium and ventricle, and the left atrium were enlarged. Left atriotomy showed a friable, necrotic tumour with areas of fibrosis that almost completely filled the atrium. The tumour was removed without difficulty from the region surrounding the orifice of the left

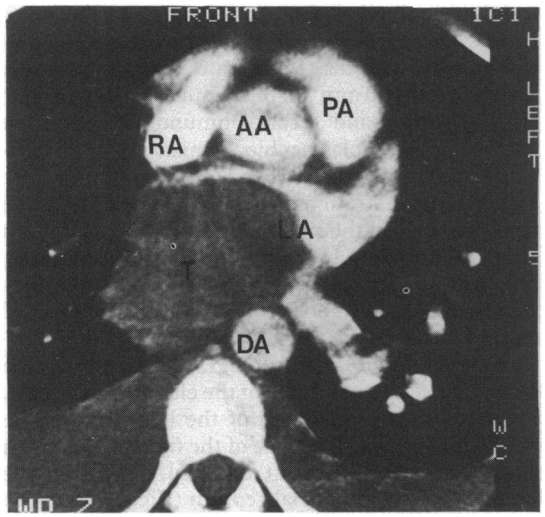

Fig. 2 *Preoperative digital gated computed tomogram of the thorax at the level of the left atrium. Tumour (T) is seen filling the left atrium (LA). The horn-shaped left atrial appendage is free of tumour. RA, right atrium; AA, ascending aorta; PA, pulmonary artery; DA, descending aorta.*

pulmonary veins but it was densely adherent to the orifice of the right inferior pulmonary vein, from which it appeared to arise. There was no septal or valvar involvement.

After removal of the tumour, the mitral valve was found to be stenosed, with 5 mm fusion of each commissure. Complete splitting of the commissures was obtained.

Postoperative convalescence was uneventful. Sinus rhythm was restored by direct current conversion. Histological examination of the tumour showed clusters of cells with strongly eosinophilic cytoplasm and striation. Mitotic activity was conspicuous. These appearances are typical of rhabdomyosarcoma. A postoperative echocardiogram showed no evidence of residual tumour. She was given a course of radiotherapy and remains symptom free.

Discussion

Primary cardiac sarcomas are very rare tumours. Angiosarcomas are the most common form and rhabdomyosarcomas the second most common form.[1 2] Rhabdomyosarcomas have been reported in patients ranging in age from 3 months to 80 years but, like other primary sarcomas, they usually occur in adults.[2] A slight male preponderance has been reported.[1] In a recent review of published reports, Schwartz *et al* found that 26 out of 40 reported cases of primary cardiac rhabdomyosarcoma occurred in males.[3] Unlike angiosarcomas, rhabdomyosarcomas do not appear to have a predilection for any particular cardiac chamber, and they occur with equal frequency on the right and left sides of the heart. In approximately 60% of cases coming to necropsy the tumour affects multiple cardiac sites.[2]

Cardiac tumours often remain undetected during life and the diagnosis is made at necropsy. Echocardiography greatly increased the potential for prompt and correct diagnosis.[4] Though echocardiography demonstrated the presence of a fixed left atrial mass in this patient, we obtained much higher resolution with the computed tomographic scan. When computed tomographic scans of pulsating organs such as the heart or aorta are taken conventionally the images are blurred and indistinct. Multiple tomographic scans all taken at one particular phase of the electrocardiogram cycle, and selected by computer can be reconstructed into a full computed tomographic image of the transverse section of the thorax at that phase of the electrocardiogram cycle. The resulting scans are much more distinct and details of intracavitary and intramural abnormalities of the heart can be seen. In our patient the computed tomographic scan was gated to the R

wave. The gated scan distinctly showed the tumour and the rim of contrast separating it from the interatrial septum. Since this non-invasive technique is safer than cardiac catheterisation, in which there is a risk of embolism by tumour fragments, we recommend its use when the echocardiogram is suggestive of tumour and more precise definition is required.

The need to maintain a high level of suspicion for unusual abnormalities is well illustrated by this case in which symptomatic deterioration was at first considered to have resulted from mitral valve disease.

The demonstration of a right ventricular rhabdomyosarcoma by means of computed tomographic scanning has previously been reported.[5] We believe, however, that this is the first report of a left atrial sarcoma defined by this method.

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