CASE REPORT

Multifocal cardiac leiomyosarcoma. Diagnosis and surveillance by transoesophageal echocardiography and contrast enhanced cardiovascular magnetic resonance

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Postgrad Med J 2002;**78**:492–493

Primary cardiac sarcomas are rare and typically undergo aggressive local spread. There is no reliable definitive treatment, although radical surgical resection can provide palliation in the medium term. A case of a pleomorphic leiomyosarcoma with dramatic images is presented. The relative usefulness of transoesophageal echocardiography and cardiovascular magnetic resonance imaging to define the extent of tumour involvement, allowing planning of treatment, is demonstrated.

A 45 year old man presented with a two month history of progressive breathlessness, exertional chest pains, and night sweats. Examination revealed jaundice, marked ascites, and peripheral oedema, sinus tachycardia of 100 beats/min, blood pressure 110/70 mm Hg with no paradox, engorged neck veins, right ventricular third heart sound, and a tricuspid regurgitant murmur. The lung fields were clear. Transthoracic echocardiography showed a large lobulated mobile mass obstructing much of the right atrium and right ventricle. Transoesophageal echocardiography (TOE) confirmed these appearances and revealed additional tumour forming an irregular layer in the left atrium and right sided pulmonary veins (fig 1). Coronary angiography demonstrated normal coronary arteries and a leash of collaterals supplying the tumour.

At surgery a superoseptal approach to the atria enabled removal of all the right heart tumour which was attached to the right atrial appendage via a stalk and macroscopic removal of the left atrial tumour (fig 2). Immunohistochemical staining and electron microscopy revealed the tumour to be a pleomorphic leiomyosarcoma.

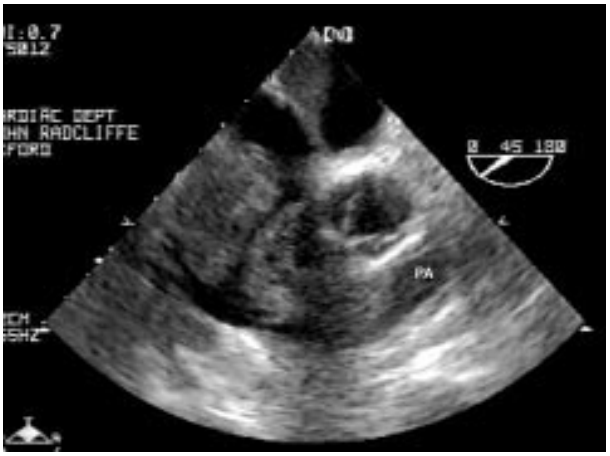

Figure 1 Transoesophageal echocardiogram showing the lobulated tumour in the right heart, to the left and below the centrally placed aortic valve. The tumour occupies much of the right atrium and prolapsed through the tricuspid valve during the cardiac cycle.

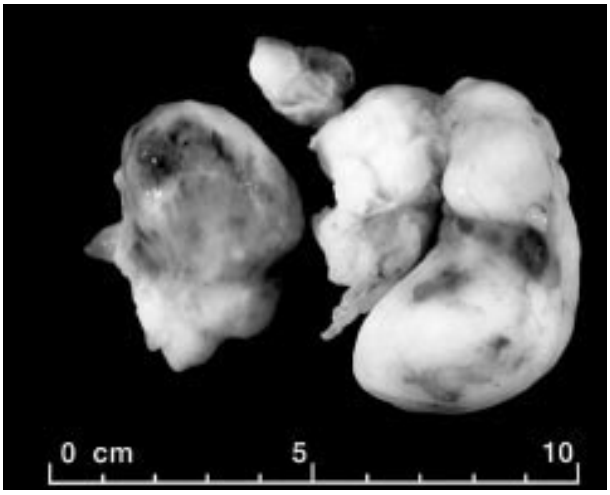

Figure 2 Tumour removed from the right heart.

One month after surgery he received four cycles of ifosfamide and doxorubicin and began surveillance TOE imaging. After 14 months there was evidence of tumour recurrence with a 1.5 cm round mass partially obstructing the right upper pulmonary vein and a 0.6 × 0.8 cm sessile mass adjacent to the left atrial appendage.

Cardiovascular magnetic resonance (CMR) was undertaken to assess the feasibility of further surgery. T1 weighted spin echo images showed a discrete pedunculated mass (1.5 × 1.5 cm) within the mouth of the right upper pulmonary vein, fig 3A (top left, long arrow), and a sessile lesion attached to the posteriolateral wall of the left atrium, extending between the insertion of the posterior mitral valve leaflet and the left lower pulmonary vein (short arrow). These masses enhanced after the intravenous administration of gadolinium diethylenetriaminepenta-acetic acid (DPTA), fig 3A (bottom left). Contrast enhanced magnetic resonance angiography confirmed the position and size of these masses but crucially that they were discrete without evidence of more distal involvement of the pulmonary veins (fig 3B).

At repeat surgery the tumour masses were excised including a full thickness portion of the left posterior atrial wall, which was repaired with a portion of aortic valve homograft. Unfortunately at four months after operation and 21 months

Abbreviations: CMR, cardiovascular magnetic resonance; DPTA, diethylenetriaminepenta-acetic acid; TOE, transoesophageal echocardiography

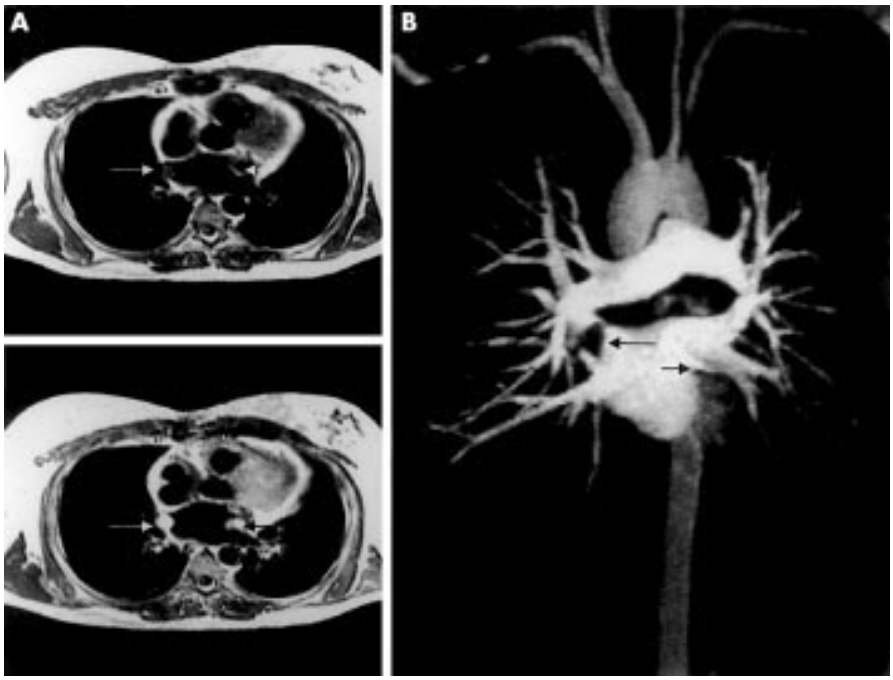

Figure 3 (A) T1 weighted spin echo images (upper left) and enhancement with gadolinium (lower left). The long arrows mark tumour at the mouth of the right upper pulmonary vein and the small arrows tumour adjacent to the left atrial appendage. (B) Contrast enhanced magnetic resonance angiography. Arrows show black signal defects due to tumour.

from the original operation he developed limiting exertional breathlessness and had signs of right heart failure with marked peripheral oedema. TOE demonstrated multifocal recurrence of tumour with a dumbell shaped mass traversing the interatrial septum, irregular thickening of right and left atrial walls, and separate masses of tumour arising from the tricuspid valve annulus and left atrial appendage. Atrial systolic function was absent and this was confirmed by the lack of an A wave on mitral and tricuspid valve inflow Doppler. The combination of severe tricuspid regurgitation and atrial electromechanical dissociation explains the symptoms of cardiac failure. Three months later, 24 months from the original operation, he died with congestive cardiac failure.

DISCUSSION

Primary cardiac tumours tend to present relatively late as a result of symptoms arising from obliteration of cardiac chambers and obstruction of great vessels or cardiac valves. Transthoracic echocardiography can identify intracardiac tumours with a sensitivity of 91% (myxomas excepted) but TOE and particularly CMR are required to delineate the degree of tumour extension and infiltration of paracardiac structures.[1] Enhancement with intravenous gadolinium DPTA is a typical feature of malignant cardiac tumours, although it can occur in well vascularised benign tumours such as haemangioma.[2]

Primary cardiac sarcomas have a poor prognosis with a median survival of approximately 12–16 months.[3 4] Complete surgical resection increases median survival to approximately 24 months, while adjuvant chemotherapy based on doxorubicin after surgical resection does not appear to affect the natural history.[4 5] One reported case described survival in the absence of local recurrence 20 months after heart transplantation performed for recurrence following initial radical resection of a left atrial leiomyosarcoma.[6] Transplantation is unlikely to be a realistic option for most cases because of the tendency of leiomyosarcomas to undergo aggressive local invasion with involvement of the pulmonary veins and the short time window for intervention.

In summary early radical surgery provides the best chance for palliation of cardiac leiomyosarcoma in the medium term. The feasibility of surgery requires a detailed anatomical assessment and is therefore best guided by the complementary imaging modalities of TOE and CMR.

Learning points

- Transthoracic echocardiography is mandatory in heart failure to exclude unusual treatable causes—for example, intracardiac tumour, pericardial effusion.
- Cardiovascular magnetic resonance imaging compliments TOE and is superior for defining the extent of extracardiac tumour involvement of vascular structures.
- Early detection and radical resection of cardiac sarcomas provides the only realistic chance of cure.

ACKNOWLEDGEMENTS

No financial support. Clinical case managed at the John Radcliffe, Oxford and contrast enhanced cardiovascular magnetic resonance was performed at the Royal Brompton Hospital.

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Submitted 20 September 2001
Accepted 4 April 2002

REFERENCES

1 **Engberding R**, Daniel WG, Erbel R, *et al*. Diagnosis of heart tumours by transoesophageal echocardiography:a multicentre study in 154 patients. European cooperative study group. *Eur Heart J* 1993;**14**:1223–8.
2 **Hoffman U**, Globits S, Frank H. Cardiac and paracardiac masses. Current opinion on diagnostic evaluation by magnetic resonance imaging. *Eur Heart J* 1998;**19**:553–63.
3 **Donsbeck AV**, Ranchere D, Coindre JM, *et al*. Primary cardiac sarcomas:an immunohistochemical and grading study with long-term follow-up of 24 cases. *Histopathology* 1999;**34**:295–304.
4 **Putnam JB**, Sweeney MS, Colon R, *et al*. Primary cardiac sarcomas. *Ann Thorac Surg* 1991;**51**:906–10.
5 **Llombart-Cussac A**, Pivot X, Contesso G, *et al*. Adjuvant chemotherapy for primary cardiac sarcomas: the IGR experience. *Br J Cancer* 1998;**78**:1624–8.
6 **Babatasi G**, Massetti M, Agostini D, *et al*. Recurrent left sided heart leiomyosarcoma: should heart transplantation be legitimate? *J Heart Lung Transplant* 1998;**17**:1133–8.