Debulking of malignant cardiac tumour discovered at operation for presumed right atrial thrombus obstructing the tricuspid valve

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Introduction

Primary cardiac lymphomas (PCL) are rare neoplasms. They occur at any age and are rare in immunocompetent patients, accounting for 1.3% of all cardiac tumours and 0.5% of all extranodal lymphomas. PCL have been increasingly found in patients with acquired immune deficiency syndrome (AIDS).[1] PCL are difficult to diagnose, especially during the early stages of the disease when their manifestations are non-specific.[2]

Discovery of a malignant cardiac tumour at operation would usually be managed by biopsy and closure without resection, and inevitably would result in early mortality. We report a patient who is alive 12 months after resection of a primary cardiac non-Hodgkin's B cell lymphoma.

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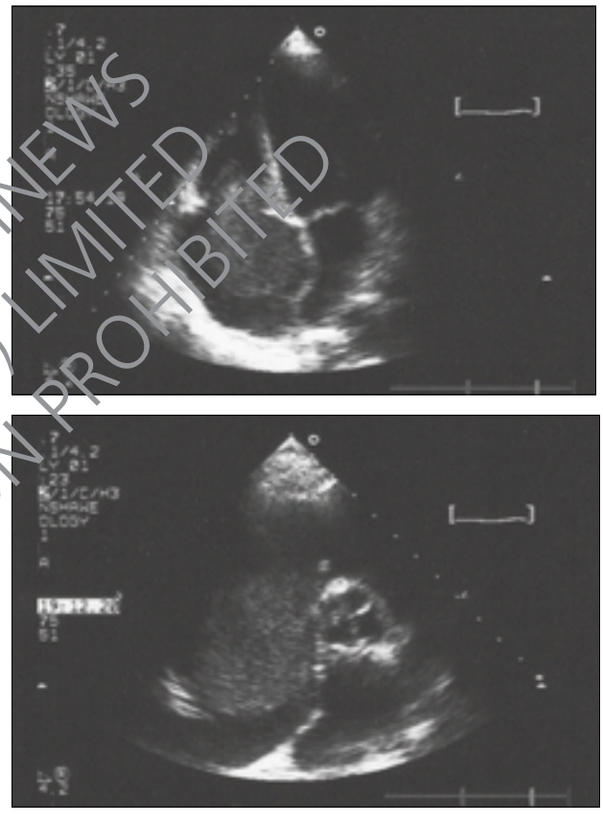
Case report

A 57-year-old woman presented in March 2001 with features of sick sinus syndrome. A permanent pacing system was inserted at her local hospital. Two weeks later, the pacemaker box site became infected and she was prescribed antibiotics. The pacing system was not removed and the infection resolved. A transthoracic echocardiogram (TTE) at that time was reported to be normal.

Four months later, the patient collapsed at home. For the two weeks before that she had had shortness of breath, tiredness, night sweats and abdominal pain. On examination, she was found to have hepatomegaly (5 cm), a new systolic murmur heard over the tricuspid area, and mild pyrexia (37.3°C). Blood tests were normal apart from microcytic anaemia and a raised C-reactive protein. TTE showed a large mass occupying the right atrial cavity and protruding through the tricuspid valve orifice. The working diagnosis was thought to be an infected intra-atrial thrombus related to the pacing wires, but a primary or secondary tumour could not be excluded.

Figure 1. Preoperative TTE images showing a right atrial mass protruding through the tricuspid valve



The patient was transferred to a cardiothoracic surgery centre for removal of the thrombus and restoration of function of the tricuspid valve. At this stage, the patient developed features of right heart failure, and a repeat TTE showed tricuspid valve obstruction (figure 1). Surgery was expedited and transoesophageal echocardiography (TOE) was deferred until after induction of anaesthesia.

At operation the external appearances were not consistent

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Table 1. Literature review of published case reports of cardiac lymphomas in the last 15 years (see appendix for full reference data)

Author	Time of diagnosis	Location	HIV status	Symptoms	Diagnosis	Treatment	Outcome
Jurkovich *et al*. 2000	AM	RA	-	Haemoptysis	TOE	Chemotherapy	Alive at 32 months
Skarin *et al*. 2000	AM	RA	-	SOB	TOE	Surgery, chemotherapy	Died at two months
Miyashita *et al*. 2000	AM	RV	-	Arrhythmia	Echo	Chemotherapy, XRT	Alive at 30 months
Kuo *et al*. 2000	AM	RA	-	SOB	Echo	Chemotherapy	Died at seven months
Mejhert *et al*. 2000	AM	RA, LA	-	SOB	Echo	Chemotherapy	CR at two months
Dennis *et al*. 2000	AM	RA	-	Syncope	Echo	Chemotherapy	-
Cordel *et al*. 2000	AM	-	-	SOB	-	Chemotherapy	Alive at 12 months
Montalbetti *et al*. 1999	AM	Pericardium	-	SOB	Echo	Chemotherapy	Died at nine months
Daus *et al*. 1998	AM	Pericardium	-	SOB	Echo	Chemotherapy	Alive at 12 months
Fava *et al*. 1998	PM	RA, RV	-	Cardiogenic shock	-	-	-
Makishima *et al*. 1998	PM	RA	-	SOB	Echo	Radiation therapy	Died within hours
Toko *et al*. 1998	PM	RV, LV	-	CCF	-	-	Died at seven days
Nakayama *et al*. 1997	AM	RA, pericardium	-	SOB, arrhythmia	Echo	Chemotherapy	CR at 12 months
David *et al*. 1996	AM	RA	-	Heart failure	Echo	Surgery, chemotherapy	Died at one month
Lynch *et al*. 1996	AM	Pericardium	-	SOB	Echo	Chemotherapy	CR at six months
Chao *et al*. 1995	AM	RA, AV groove	Negative	SOB	Echo	Chemotherapy	Alive at six months
Aboulafia *et al*. 1994	AM	Pericardium	Positive	SOB	Echo	Chemotherapy	-
Pavlidis *et al*. 1994	AM	Pericardium	-	SOB	Echo	Chemotherapy	Alive at 40 months
Yuh *et al*. 1994	AM	LV	-	CCF	-	Surgery, heart transplant	Alive? At 15 months
Dorsay *et al*. 1993	AM	RV	-	Chest pain	Echo	Chemotherapy	Alive at seven months
Medolago *et al*. 1992	AM	RV	-	CVA	Echo	Chemotherapy, XRT	Alive at 12 months
Chen *et al.* 1992	AM	RA	-	-	-	Surgery, chemotherapy	-
	AM	RA	-	-	-	Surgery, chemotherapy	-
	AM	LA	-	-	-	Surgery, chemotherapy	-
Holladay *et al*. 1992	PM	Pericardium	Positive	SOB, fever	-	-	-
Takagi *et al*. 1992	AM	RA	-	SOB	Echo	Surgery, chemotherapy	Died at seven months
Kasai *et al*. 1992	PM	-	-	Abdominal pain	-	-	-
Nand *et al*. 1991	AM	AV groove	Negative	Arrhythmia	Echo	Chemotherapy	Alive at 18 months
Zahari *et al*. 1991	AM	RA	-	SOB	Echo	Chemotherapy	Alive at 21 months
Horowitz *et al*. 1991	AM	RV	-	SOB	Echo	Surgery	Alive at eight months
Nagamine *et al*. 1990	AM	Pericardium	-	CCF	Echo	Chemotherapy	Alive at 11 months
Leventhal *et al*. 1990	PM	RA, RV, LA, LV	-	SOB	Echo	-	-
Castellie *et al*. 1989	AM	RA	-	Arrhythmia	Echo	Chemotherapy	-
Proctor *et al*. 1989	AM	RA	-	Weakness	Echo	Surgery	Hours
Guarner *et al*. 1987	PM	LV	Positive	SOB	-	-	-

Key: - = Information not available; AM = ante-mortem; PM = postmortem; HIV = human immunodeficiency virus; RA = right atrium; LA = left atrium; RV = right ventricle; LV = left ventricle; CCF = congestive cardiac failure; Echo = echocardiography; SOB = shortness of breath; CR = complete remission.

with intra-atrial thrombus. A hard nodular mass (5 cm x 9 cm) occupied most of the right atrial cavity and extended into the superior vena cava (SVC). TOE showed a right atrial mass obstructing the tricuspid valve orifice. The mass was not mobile and appeared to extend across the inter-atrial septum into the roof of the left atrium, and around the aortic root. A frozen section biopsy of this mass suggested a malignant tumour, possibly a non-small cell carcinoma. Bi-caval and aortic cannulation were undertaken to establish cardiopulmonary bypass because of the haemodynamic instability resulting from the obstruction of flow across the tricuspid valve. There was no possibility of the patient surviving unless the obstruction was relieved and the frozen section diagnosis could not be considered definitive. Therefore, de-bulking of the right atrial mass was attempted. The tumour was adherent to the tricuspid valve leaflets and invaded the right atrial free wall and the terminal SVC.

The right atrial free wall and superior vena cava junction were resected, and the tumour was stripped off the tricuspid valve leaflets. The pacing wires were removed, and the right atrium and superior vena cava were reconstructed with bovine pericardium. The patient was weaned from cardiopulmonary bypass easily, but the roof of the left atrium then disintegrated. Cardiopulmonary bypass was re-established, the patient was cooled to 18ºC, and the roof of the left atrium was reconstructed with pericardium. She made a good recovery.

Histopathological and immunological examination of the tumour confirmed the diagnosis of a diffuse large B cell lymphoma (which was CD79a and CD20 positive). Subsequent bone marrow biopsy and computerised tomograms of the thorax, abdomen, and pelvis showed no extra-cardiac involvement, and she was HIV-negative. Three weeks after surgery, the patient was transferred to an oncology centre and treated with

chemotherapy and radiotherapy. The patient received three cycles of the chemotherapy regime VAPEC-B (vincristine, doxorubicin, prednisolone, etoposide, cyclophosphamide and bleomycin) and methotrexate. Subsequent trans-oesophageal echocardiography showed no evidence of residual tumour. At 12 months follow-up, the patient is well, fully active, and apparently free of tumour.

Discussion

The definition of primary cardiac lymphoma (PCL) is an extranodal malignant lymphoma of any cell type involving only the heart and the pericardium, or both. More than 80% of PCLs are reported to arise from the right heart chambers.[3] The initial presentation in patients with PCL varies according to the site, size and the extent of tumour involvement. Presentations include congestive heart failure, pericardial effusion, superior vena cava syndrome, supraventricular arrhythmias, atrioventricular block and sudden death.[4] An intracardiac mass is diagnosed by echocardiography and histology obtained by percutaneous transvenous or open biopsy. The diagnosis of intracardiac masses has increased due to the wider availability of echocardiography. TOE is superior to TTE in the diagnosis of intracardiac lesions.[5]

PCL is an infrequent diagnosis:[6] however, in 28 of the 35 cases reported since the 1980s the diagnosis was made antemortem (table 1). Reported tumour sites included the pericardium (23%), left atrium (9%) and right atrium (42%), left ventricle (4%) and right ventricle (13%), and atrioventricular groove (9%). Reported treatment modalities in the 35 cases reviewed included chemotherapy alone in 17 cases (49%), radiotherapy alone in one case (3%), and combined chemotherapy and radiotherapy in two cases (6%). Surgery has been reported in nine cases (26%), of which seven patients had subsequent chemotherapy. In the remaining cases the diagnoses were made post-mortem.[7-9] Survival ranges from zero days to 40 months; the longest reported survivor had a pericardial tumour.[10] Of the surgical cases, four of the nine (44%) survived operation and proceeded to receive chemotherapy; survival of these patients was one, two, seven, and eight months.

The development of an early pacemaker infection understandably contributed to the working diagnosis prior to operation, but TOE was not undertaken prior to surgery because the patient had developed systemic hypotension. The decision to resect was taken in the context of an inevitable intra-operative death. Ideally, pre-operative work-up would have included a transoesophageal echocardiogram that would have delineated the extent of involvement of the tumour and a transvenous biopsy which might have yielded the diagnosis. With this knowledge, we might not have proceeded to surgery. Instead, chemotherapy and radiotherapy might have been undertaken. This treatment strategy could, however, have resulted in tumour necrosis, cardiac tamponade and death. In this case, the risk of cardiac rupture was reduced by surgical excision, reconstruction and post-operative adhesions.

Conclusions

Malignant cardiac tumours carry a grave prognosis. Primary cardiac lymphoma must be considered in the differential diagnosis of malignant cardiac tumours because it is responsive to non-surgical interventions. If a malignant cardiac tumour is discovered at operation, and salvage debulking is necessary, an open and close strategy would disenfranchise patients with lymphoma who might benefit from aggressive surgery.

Appendix

Listed below are the references from table 1:

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