Large left ventricular metastasis causing left ventricular outflow tract obstruction and haemolysis

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Introduction
Cardiac metastases occur frequently in patients with malignancies, particularly in the context of disseminated disease.1 They often remain clinically silent, or unidentified due to overshadowing by symptoms of the primary malignancy. However, secondary spread to the heart is important to recognize, as it occurs mainly in the context of disseminated disease and portends a poor prognosis,2 as illustrated by our case.

Case report
A 75-year-old man was admitted to hospital with 1 month of increasing exertional dyspnoea.

His background history included a soft-tissue fibrosarcoma of his left wrist, diagnosed 9 years ago. At that time, he had undergone a below elbow amputation, and the histopathology report was consistent with fibrosarcoma. There was no evidence of local or distant metastatic spread. He did not receive any adjuvant therapy, was monitored for 5 years, and then discharged from the care of his orthopaedic surgeon. Other history included symptomatic sinus node dysfunction, for which a dual-chamber pacemaker was inserted in 2002.

The patient had presented to his general practitioner 2 weeks prior to admission, due to worsening exertional dyspnoea and orthopnoea over the previous month. Clinical examination by his general practitioner revealed a regular pulse with heart rate of 72 bpm, blood pressure of 140/70 mmHg, and a soft pansystolic murmur audible at the apex. There were fine inspiratory crepitations in the lower thirds of the lung fields bilaterally. Radiographic examination showed a normal heart size and confirmed significant left ventricular (LV) failure with interstitial oedema and Kerley B lines. The patient was commenced on oral diuretic therapy, to which he responded well, with improvement in his symptoms and exercise tolerance.

A transthoracic echocardiogram was ordered to investigate for significant valvular pathology as a cause of LV failure. While awaiting this test, the patient’s symptoms progressed. He began to experience presyncope after standing from a supine position. Two weeks after his initial medical review, he presented to the emergency department following a syncopal episode at home. This occurred after several hours of standing upright, working in his garage, with initial presyncope symptoms followed by a loss of consciousness of 30 s.

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On presentation, clinical examination revealed an irregular pulse with heart rate of 82 bpm and blood pressure of 135/75 mmHg when supine. On standing, symptomatic hypotension occurred with a significant fall in systolic blood pressure to 70 mmHg. The patient’s symptoms and blood pressure improved rapidly once again lying supine. Jugular venous pressure measured 2 cm and the apex beat was not displaced. On auscultation, normal first and second heart sounds were heard, with a soft apical pan-systolic murmur as well as a fourth heart sound. Examination of the chest revealed no evidence of pulmonary oedema. There were no signs of tumour recurrence at the site of the previous forearm amputation and no significant lymphadenopathy on palpation of all lymph node groups.

Electrocardiogram revealed atrial fibrillation with a controlled ventricular rate at 84 bpm. An emergent transthoracic echocardiogram was performed.

The LV was of normal size with normal wall thickness and systolic function. There was a large homogenous tumour mass filling the majority of the LV cavity (maximum systolic length 10.6 cm, maximum diastolic length 8.3 cm, and maximum width 5.0 cm). It appeared to be attached to the distal infero-lateral wall and apex via a broad base. There was partial prolapse (3.6 cm × 2.3 cm) through the mitral valve in systole via a narrow isthmus of tumour (Figure 1). It also extended into the left ventricular outflow tract (LVOT) causing a dynamic outflow gradient. Diastolic parameters were indeterminate (due to the presence of atrial fibrillation and mass in the mitral value). The left atrium was moderately dilated (area 30 cm²). The mitral leaflets were structurally normal with leaflet opening and closure limited only by the LV tumour. There was mild complex regurgitation. There was no significant stenosis. The interatrial septum appeared intact. The estimated closure limited only by the LV tumour mass was estimated to be 1–3%.

The mass was sent for pathological examination. The pedunculated portion of the mass (on left of photograph) which had been prolapsing across the mitral valve appeared pinkish and firm. There was a larger component of the mass, which had filled the LV and appeared soft, lobulated, and yellow. Histological features (spindle cells) suggested a leiomyosarcoma or fibrosarcoma. However, immunohistochemical testing and comparison with the original tumour specimen removed 9 years earlier, confirmed a recurrence of fibrosarcoma.

The patient recovered well post-operatively and returned home, but was re-admitted 6 weeks later with symptomatic conscious ventricular tachycardia, which reverted spontaneously to sinus rhythm. Treatment with oral amiodarone prevented further recurrences of ventricular arrhythmias during admission.

Repeat transthoracic echocardiogram showed rapid recurrence of the tumour, with a large 5.9 × 2.4 cm mass arising from the inferior and posterior walls, and extending almost to the mitral valve leaflet tips in diastole. The posterior mitral leaflet was restricted in its mobility, resulting in decreased coaptation. There was severe regurgitation with systolic blunting of the pulmonary vein inflow, and moderate left atrial dilatation (area 32 cm²).

The patient was discharged home after 3 days of observation. One month after discharge, he died suddenly, from presumed ventricular arrhythmia.

**DISCUSSION**

Metastases to the heart occur commonly in the setting of malignancy but often remain unrecognized due to the predominance of other symptoms related to the primary tumour. The most common tumours associated with cardiac metastases are malignant melanoma and lung, breast, and oesophageal carcinomas. Soft tissue sarcomas are also known to metastasise to the heart, often presenting with congestive cardiac failure from extensive myocardial involvement or restriction due to pericardial disease.

Pericardial metastases (often by regional lymphatic spread) are more common than myocardial or endocardial metastases. Myocardial metastases occur as a result of regional lymphatic spread, and endocardial metastases due to haematogenous spread. The manifestations of metastatic cardiac deposits include arrhythmias and congestive cardiac failure, as in our patient’s case, or conduction disturbances.
or thromboembolic complications. Clinically recognized metastases to the LV are rare, although isolated case reports do exist. A change in the clinical state of a patient with known malignancy, associated with a new or changing heart murmur may suggest the need for further evaluation. Echocardiographic imaging is easily available, can be used for serial testing, and may be performed at the bedside of an unwell patient. However, imaging quality may vary depending on body habitus and the operator’s experience. The differential diagnosis of an intra-cavitary mass identified on echocardiography includes thrombus and benign cardiac tumours such as myxoma. A heterogenous appearance and absence of underlying ventricular dysfunction may be of aid in identifying malignancy. Magnetic resonance imaging may be of use in further delineating the extent of myocardial invasion. There are, however, no pathognomic features of metastases, and ultimately a tissue biopsy is required for a definitive diagnosis.

Cardiac involvement by malignancy generally reflects widespread disseminated disease and, as this case illustrates, portends a very poor prognosis. Surgical resection offers the best chance for cure in carefully selected patients with solitary metastases. However, post-operative mortality is high and resection may be limited by the proximity of vital cardiac structures. The decision to treat with chemotherapy or radiotherapy is made on a case-by-case basis.

Surgical resection was offered in our patient’s case to relieve symptoms, which were likely to worsen significantly as the mass enlarged. His good pre-morbid functional state and absence of any other metastatic disease were also considerations in the decision to operate. Although endomyocardial biopsy would be an alternative method of confirming a malignant process, it would not allow any further delineation of the extent of myocardial infiltration or resection, or allow relief of LVOT obstruction.

In the emerging era of biological therapies for malignant disease, prolongation of survival and augmentation of the natural history of these diseases may mean that metastases that were once considered rare become identified more commonly. Importantly, the diagnosis of a cardiac metastasis may influence the approach to management, causing a shift from more aggressive treatments with curative intent, to a focus upon palliation.

Supplementary data
Supplementary data are available at European Journal of Echocardiography online.

Conflict of interest: none declared.

References