Cardiac sarcoma presenting as heart failure and diagnosed as recurrent myxoma by echocardiogram

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We report a case of an extremely rare high-grade, undifferentiated cardiac sarcoma. The patient with left atrial myxoma resected 8 months ago initially presented to the emergency department with dyspnoea on exertion, productive cough, fever, and chills for a week. On examination, the patient was febrile and tachypneic. The neck veins were distended bilaterally and the carotid upstroke was mildly diminished. Lungs revealed diffuse, prominent rales. She was tachycardic with a regular rhythm, but a prominent diastolic low-pitched murmur consistent with mitral stenosis was present. The EKG was read as sinus rhythm with right bundle branch block and left posterior fascicular block and cardiac enzymes were negative. A chest radiograph (Figure 1) showed pulmonary oedema with bilateral pleural effusions. As patient did not respond the course of intravenous antibiotics, initial diagnosis of pneumonia subsequently changed to congestive heart failure, confirmed by a transthoracic echocardiogram (TTE). The TTE (Supplementary data, Movie 1) showed a large, mobile left atrial (LA) mass prolapsing through the mitral valve apparatus resulting into impaired left ventricular (LV) filling. A transoesophageal echocardiogram (TEE) (Figure 2) confirmed the presence a 4.9 x 2.4 cm mobile mass in the LA attached to the anterior wall leading to functional mitral stenosis with a mean gradient of 13 mmHg.

Cardio-thoracic surgery was consulted and surgical resection of the mass was recommended. A large mass almost completely filling the LA and prolapsing through the mitral valve was removed without complications. The surgical pathology (Figure 3) of the LA mass revealed a high-grade, undifferentiated cardiac sarcoma (Grade 2 on a scale of 3). The post-operative course was uneventful and the surgery produced immediate and effective symptom relief. A follow-up TEE reported a normally functioning mitral valve with mild mitral regurgitation and chest X-ray (Figure 4) showed the marked improvement in the pulmonary oedema. The patient opted to continue treatment in Peru and was lost to follow-up.

Interestingly, the patient had a similar presentation of severe pneumonia and CHF when diagnosed with her initial LA mass, which was surgically removed in 8 months earlier in Peru.

Discussion

Primary cardiac tumours, either benign or malignant, are rare.1 The combined incidence of primary cardiac tumours is ~0.02% based on pooled autopsy series.2 A mobile mass found in the left atrium is generally assumed to be a benign myxoma. However, that is not always the case. In the present case, a primary cardiac sarcoma was preoperatively diagnosed as left atrial myxoma based on TEE appearance and history.

The presentation, diagnosis, and treatment options of cardiac tumours are often not well known. The incidence of these tumours varies from 0.0017 to 0.019%,3,4 most of which are benign. The benign tumours account for ~63% of primary

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tumours and include myxoma, the most common, and followed in decreasing frequency by papillary fibroelastoma, rhabdomyosarcoma, fibroma, haemangioma, and various other rare tumours. The remaining 37% are malignant, predominantly consist of sarcomas. A large series reported, angiosarcoma, to be the most common primary malignant tumour followed by malignant fibrous histiocytoma, leiomyosarcoma, rhabdomyosarcoma, unclassified, and others.\textsuperscript{5,6}

Cardiac tumours may be either symptomatic or found incidentally during evaluation for unrelated problems or symptoms. When symptomatic, the clinical presentation of cardiac tumours depends on their location. Left atrial tumours can cause obstruction or regurgitation of the mitral valve presenting with signs and symptoms of mitral stenosis like dyspnoea, orthopnoea, paroxysmal nocturnal dyspnoea, pulmonary oedema, cough, haemoptysis, oedema, and fatigue from resultant congestive cardiac failure and pulmonary hypertension. Positional changes, such as lying down or lifting the legs that increase venous return, may worsen symptoms. Tumour fragment may embolise leading to systemic metastasis. On physical examination, a characteristic ‘tumour plop’ may be heard early in diastole. There has been one case of left atrial sarcoma reported where the patient presented with brain metastasis.\textsuperscript{7,8}

While myxoma is the most common left atrial tumour, this case shows that sarcoma can also arise from the left atrium. It may be difficult to differentiate benign myxoma and sarcoma solely on the basis of anatomical features and the clinical nature of the tumour.

Tumours can occur in all four cardiac chambers. Tumours in the right atrium produce haemodynamic changes similar to those seen with tricuspid stenosis presenting with right heart failure sign and symptoms such as fatigue, peripheral oedema, hepatomegaly, ascites and prominent ‘a waves’ in the jugular veins. A diastolic murmur similar to the ‘tumour plop’ heard with left atrial myxoma can be ascultated. The patient may present with pulmonary embolism as a result of tumour fragments released into the pulmonary circulation.\textsuperscript{8} The common tumours of the right atrium are myxoma and angiosarcoma.
Arrhythmias and conduction defects are common with intramural left ventricle tumours. Intracavitary tumours often present with systemic embolization or outflow obstruction leading to LV failure. Syncope is a common symptom of intracavitary tumours that obstruct the outflow tract.

Tumours arising in the right ventricle can often be misdiagnosed as pulmonary stenosis, restrictive cardiomyopathy, or tricuspid regurgitation. Right ventricular tumours present with signs and symptoms such as shortness of breath, syncope, peripheral oedema, hepatomegaly, ascites, and sudden death. All these result from interference with filling and/or outflow from the right ventricle, leading to right-sided heart failure.

Diagnostic approaches for cardiac tumours include echocardiography. In particular, TEE has proved an effective and relatively non-invasive tool. Other imaging modalities such as MRI and CT scan can also be used. The combination of echocardiography as well as cardiac MRI/CT may be useful in differentiating thrombus from tumour in lesions appearing to arise on a heart valve. The role of transvenous biopsy has been limited due to the risk of embolization. Treatment is immediate surgical resection. The neoadjuvant or chemotherapeutic agents have not been well studied.

Supplementary data

Supplementary data are available at European Journal of Echocardiography online.

References