Case report

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Intracardiac metastasis originated from chondrosarcoma

Nicola Maurea[a], Gianluca Ragone[b], Carmela Coppola[a], Antonietta Caronna[a], Carlo G. Tocchetti[a], Lucio Agozzino[c], Gaetano Apice[b] and Rosario V. Iaffaioli[b]

Primary cardiac tumors are extremely rare. By comparison, metastatic involvement of the heart is over 20 times more common and has been reported in autopsy series in up to one in five patients dying of cancer. Cardiac metastasis of chondrosarcoma is absolutely not frequent. In the recent literature, a cardiac metastasis from chondrosarcoma has never been described. We report the case of an 18-year-old man with a diagnosis of cardiac metastasis that originated from a left scapular chondrosarcoma. Chondrosarcoma is a skeletal tumor with various grades of malignancy, rapidly evolving, and with a strong tendency to metastasize, with low responsiveness to chemotherapy. The onset of characteristic systemic symptoms in the late stage of the disease led to the diagnosis of a mass localized in the right atrium. Management and differential diagnosis of infective heart lesions were also very complex in a rapidly evolving life-threatening condition. *J Cardiovasc Med* 12:000–000

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[a]Division of Cardiology, [b]Department of Colorectal Oncology, National Cancer Institute, G. Pascale Foundation and [c]Department of Public Health, Section of Pathology, Second University of Naples, Naples, Italy

Correspondence to Dr Nicola Maurea, Division of Cardiology, National Cancer Institute, G. Pascale Foundation, Via Mariano Semmola, 80131 Napoli, Italy
Tel: +39 081 5903519; fax: +39 081 5903829; e-mail: nicolamaurea@tiscali.it

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Introduction

Primary cardiac tumors are rare. As an example, in one series of over 12 000 autopsies, only seven were identified, for an incidence of less than 0.1% [1,2].

Cardiac tumors may be symptomatic or found incidentally during evaluation for a seemingly unrelated problem or physical finding. In symptomatic patients, a mass can virtually always be detected by echocardiography, magnetic resonance imaging (MRI), and/or computed tomography (CT). Because symptoms may mimic other cardiac conditions, the clinical challenge is to consider the possibility of a cardiac tumor so that the appropriate diagnostic tests can be conducted.

Mechanisms by which cardiac tumors may cause symptoms include the following:

(1) Obstruction of the circulation through the heart or heart valves, producing symptoms of heart failure;
(2) Interference with the heart valves, causing regurgitation;
(3) Direct invasion of the myocardium, resulting in impaired contractility, arrhythmias, heart block, or pericardial effusion with or without tamponade;
(4) Invasion of the adjacent lung may cause pulmonary symptoms and may mimic bronchogenic carcinoma;
(5) Embolization, which is usually systemic, but can be pulmonic;
(6) Constitutional or systemic symptoms.

In contrast to primary malignant cardiac tumors, metastatic involvement of the heart is relatively common. As an example, in one of the largest autopsy series of over 1900 patients dying of cancer, 8% had metastatic disease involving the heart [3]. Cardiac involvement may arise from hematogenous metastases, direct invasion from the mediastinum, or tumor growth into the vena cava and extension into the right atrium [4].

Malignant melanomas are particularly likely to metastasize to the heart [5–8]. Other solid tumors commonly associated with cardiac involvement include lung cancer, breast cancer, soft tissue sarcomas, renal carcinoma, esophageal cancer, hepatocellular carcinoma and thyroid cancer. There is also a high prevalence of secondary cardiac involvement with leukemia and lymphoma.

Indeed, the cardiac metastases of chondrosarcoma are extremely rare.

Cardiac or pericardial metastases should be considered whenever a patient with known malignancy develops cardiovascular symptoms, particularly if this occurs in conjunction with cardiomegaly, a new or changing heart murmur, electrocardiographic conduction delay, or arrhythmia. Emboli thought to originate in the heart should also raise the possibility of cardiac involvement with tumor. Rarely, cardiac metastases may be the first manifestation of malignant disease [9].

The specific symptoms will reflect the site of cardiac involvement, in a manner analogous to primary cardiac tumors. The diagnostic evaluation is the same as that for primary cardiac tumors and relies upon echocardiography, MRI and CT to ascertain the extent of cardiac involvement. In very carefully selected patients, resection of

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cardiac metastases has been used to provide symptom palliation and to prolong life [5,10,11].

Other causes of cardiac symptoms must also be considered. In particular, metastatic disease must be distinguished from the cardiotoxicity that may be associated with chemotherapeutic agents, particularly anthracyclines and target therapy.

Chondrosarcoma is the second most frequent primary bone malignant tumor, representing approximately 25% of all primary osseous neoplasms. Chondrosarcomas are a group of tumors with highly diverse features and behavior patterns, ranging from slow-growing, nonmetastasizing lesions to highly aggressive metastasizing sarcomas [12–14].

This type of cancer rarely affects individuals under age 20 and is most common between the ages of 50 and 70. The goal for treatment of chondrosarcoma is to remove the mass and reduce the likelihood of recurrence.

Treatment may include surgery, to remove the tumor, and chemotherapy, although it is not the primary treatment and has a limited role, which may be required if the cancer has spread to other body districts.

Case report

In June 2002, an 18-year-old man was admitted to our observation after the appearance of a small mass in his left scapular region, which increased in volume rapidly.

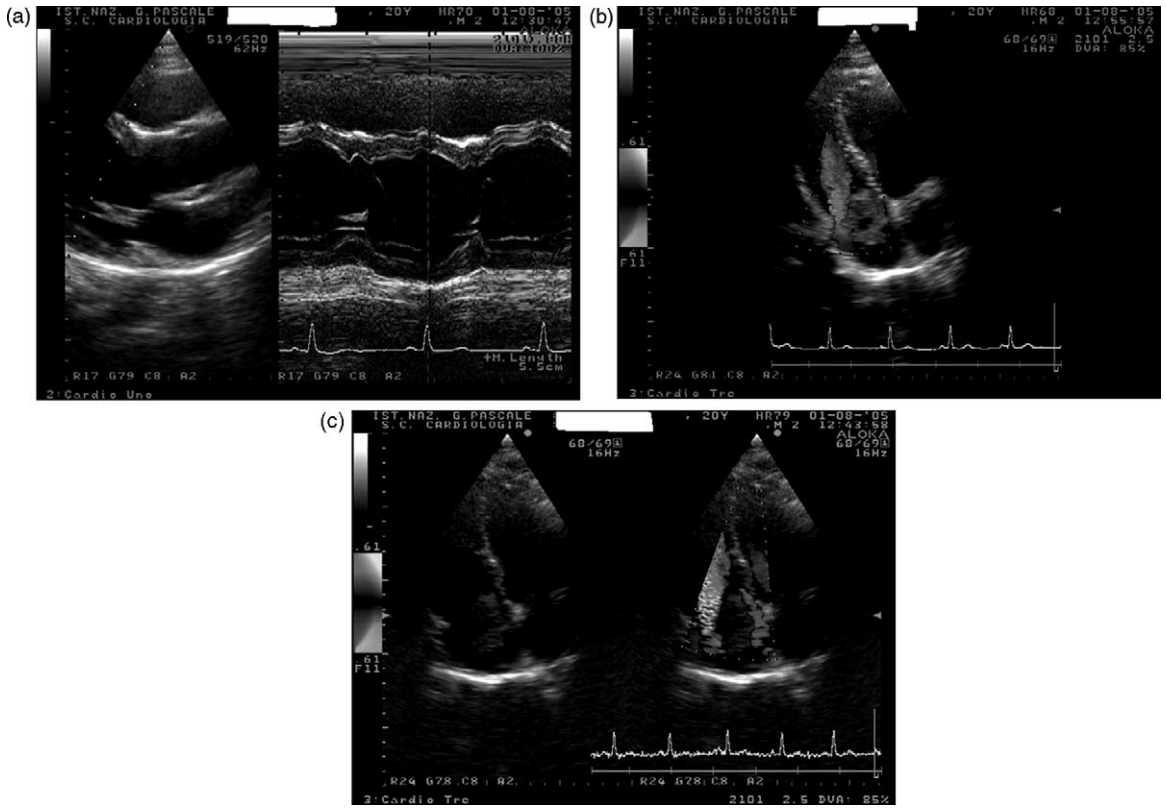
In July 2002, the patient underwent excision of the body of the left scapula for a lesion of 4 cm, whitish and spongy-crisp in consistence.

The histological examination showed neoplastic tissue consisting mainly of chondroid tissue with foci of malignant osteoid tissue and metaplastic epithelium infiltrating the medullary canal, the cortical bone and adjacent soft tissues. Excision margins were tumor-free. The histological appearance, as well as the radiographic signs, and the young age led to the diagnosis of chondroblastic osteosarcoma, moderately differentiated (G2).

From November 2002 to April 2003, the patient underwent polychemotherapy with the alternation of cisplatin (120 mg/m^2), methotrexate (12 g/m^2), Adriamycin (70 mg/m^2) and iphosphamide (6 g/m^2) for eight cycles. The administration of these chemotherapies made necessary the application of a central venous catheter (CVC).

In May 2003, the patient underwent the excision of residual scapula for recurrence of disease.

Fig. 1



Transthoracic echocardiogram. Normal kinesis and biventricular dimensions, presence of an oval mass with mixed echogenicity occupying most of the right atrium; infiltration of tricuspid valve plan, obstruction of the right ventricular filling. Vena cava dilation with inspiratory collapse less than 50%.

In this case, nonresponsiveness to chemotherapy, local relapse in the absence of metastases, and the histological aspect led to a diagnosis of a highly malignant chondrosarcoma (G2, focally G3).

After surgery, radiotherapy was performed on the left scapular region.

Subsequently, the follow-up was negative until March 2005, when a CT scan showed multiple lung metastases (histologically confirmed).

In June-July 2005, two cycles of chemotherapy with Adriamycin and iphosphamide were administered (Adriamycin 60 mg/m^2 and iphosphamide 9 g/m^2).

In August 2005, the patient suddenly experienced dyspnea on slight effort [New York Heart Association (NYHA) class III]. ECG and blood tests were normal. The chest radiograph confirmed the presence of lung metastases. Transthoracic echocardiogram (Fig. 1) showed normal biventricular size and contractility with presence of an oval, voluminous mass with mixed echogenicity occupying most of the right atrium, partially adhesive to the interatrial septum. This mobile lesion infiltrated the tricuspid valve plan, with obstruction of right ventricular filling. The vena cava was slightly dilated with an inspiratory collapse less than 50%. CT of the chest confirmed the presence of a mass in the right atrium.

In our Cardiology Division, this mass was diagnosed as metastasis from chondrosarcoma. But during the consultation with the cardiac surgeons, doubts arose, and the possibility that this mass could be a big vegetation of the tip of the CVC was considered (in fact, the patient had fever with the characteristics of a septic hyperthermia). Blood cultures were done, but results were negative.

Anyway, as our patient was highly symptomatic for dyspnea, he underwent surgical excision of the cardiac mass.

The surgical approach, with excision of the mass and closure of a small foramen ovale, was as follows: median sternotomy; incision of the pericardium; separate cannulation of aorta and cava; excision of an epicardial mass that was sent to histopathology; cardiopulmonary bypass with closure of the pulmonary artery; right atriotomy; neoformation observed, adhesive to the interatrial septum, occupying the whole cavity, gelatinous in consistency (Fig. 2); excision of the mass and closure of a small foramen ovale; closure of the right atrium; hemostasis; drainage and closure in layers.

Sections from the heart mass showed histopathologic features of neoplastic tissue attributable to malignant mesenchymal neoplasm, with chondroid differentiation compatible with metastasis of the previous sarcoma; myocardial tissue with infiltration by sarcoma with chondroid differentiation.

Fig. 2

Neoformation excised from interatrial septum.

Unfortunately, the patient died 2 months later due to cancer progression.

Discussion

There is neither consensus nor guidelines for the optimal management of patients with cardiac metastasis of sarcomas. Surgical resection has been attempted in some cases [15,16], and this could be an option if the tumor is restricted in its growth pattern and if the patient is a candidate for surgery.

The present case was peculiar because of the rare type of metastasis, the young age of the patient for this type of cancer, and the fact that the patient underwent surgery, upon the recommendation of surgeons, also on the basis of the possibility of a diagnosis of venous catheter infection.

Anyway, surgery allowed the relief of symptomatology (dyspnea) of the patient.

Heart metastases are often unrecognized. In patients suffering from metastatic disease, with widespread lesions, cardiac metastasis should always be suspected.

In neoplastic patients with CVCs presenting with atrial masses, as in our case, the differential diagnosis among metastases, thrombosis or infection can be difficult.

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