Case Reports in Oncology

Case Rep Oncol 2013;6:224–228

DOI: 10.1159/000351123
Published online: April 19, 2013

1662–6575/13/0061–0224$38.00/0
www.karger.com/cro

Primary Osteosarcoma of the Heart: Experience of an Unusual Case

Düriye Sıla Karagöz Özen[a] Mehmet Akif Öztürk[b] Fatih Selcukbiricik[b]
Sinem Nihal Esatoglu[a] Zeynep Hande Turna[b] Pınar Beyaz[c]
Sergülen Dervisoglu[d] Mustafa Özgüroglu[b]

Departments of [a]Internal Medicine, [b]Medical Oncology, [c]Radiology, and [d]Pathology, Cerrahpasa Faculty of Medicine, Istanbul University, Istanbul, Turkey

Key Words
Cardiac tumor · Extraskeletal osteosarcoma · Mitral stenosis

Abstract
Primary cardiac osteosarcomas are uncommon tumors. They have an aggressive biology and hence poor prognosis. This report describes a 23-year-old male patient who was referred to our hospital with chest pain. Echocardiography showed a left atrial mass, and tumor excision revealed a cardiac osteosarcoma. Adjuvant cisplatin plus ifosfamide combination chemotherapy provided a disease-free survival of 9 months; unfortunately the patient died of metastatic disease thereafter.

Introduction

Metastatic tumors of the heart are more common than primary tumors. The frequency of primary cardiac tumors is 0.001–0.03% in autopsy series [1]. About 75% of these primary cardiac tumors are benign tumors such as myxoma, lipoma, papillary fibroelastoma and rhabdomyoma [2]. The most frequently reported malignant primary cardiac tumors are sarcomas. Angiosarcomas and myxofibrosarcomas are the most common sarcomas of the heart [3]. A cardiac osteosarcoma, however, is extremely rare.

In this paper, we would like to report a young male patient showing symptoms of mitral stenosis who was later diagnosed with a primary cardiac osteosarcoma

Düriye Sıla Karagöz Özen, MD
Department of Internal Medicine
Cerrahpasa Faculty of Medicine, Istanbul University
TR–34098 Istanbul (Turkey)
E-Mail silakaragoz@yahoo.com

Case Presentation

A 23-year-old man was referred to our clinic because of chest pain, cough and hemoptysis. His physical examination and chest X-ray revealed left pleural effusion. Thoracentesis was performed and exudative aspiration fluid cytology did not show atypical or malignant cells. Routine laboratory tests were normal except a mild increase in lactate dehydrogenase. Echocardiography (echo) revealed a left atrial mass measuring 3.8 × 3.8 cm in diameter, filling the left atrial cavity and extending into the mitral valve during diastole, causing severe mitral stenosis symptomatology (fig. 1). The presumptive diagnosis was a cardiac myxoma; because of the symptoms it had caused, tumor excision with mitral valve replacement was performed. Pathological examination revealed a high-grade conventional osteosarcoma arising from the cardiac wall. The tumor cells were mostly osteoblastic in nature. The lace-like pattern of the osteoid matrix was seen between these atypical osteoblasts. S-100 immunostain was negative and vimentin was positive. S-100 negativity excluded the possibility of a malignant nerve sheath tumor with extensive tumoral bone production (fig. 2, fig. 3).

Due to the possibility of a further primary tumor or distant metastasis, a bone scan and computed tomography (CT) were performed; the bone scan was normal, and the thoracic-abdominal CT scan did not show any signs of distant metastasis or another primary tumor. Adjuvant combination chemotherapy with cisplatin and ifosfamide (plus mesna) was done for six cycles every 3 weeks, owing to the histology and grade of the tumor. At the end of the therapy, physical examination, thoracic-abdominal CT and echo were completely normal, without a clue for recurrent disease.

9 months after the last cycle of chemotherapy, the patient presented to our clinic with left hemiparesis and abnormal speech. A CT scan of the cranium showed a central pontine mass (3.3 × 3 × 2.5 cm) extending to the right (fig. 4). The radiologists diagnosed the mass as a metastatic lesion. Neurosurgery consultation did not recommend surgery because of the close proximity to vital nervous structures. The patient was given radiotherapy with a dose of 37.8 Gy. However, during the course of hospitalization, his clinical status worsened, loss of consciousness occurred and he eventually died because the tumor compressed the vital structures at the brain stem.

Discussion

Metastatic tumors of the heart are more common than primary tumors. The frequency of primary cardiac tumors is 0.001–0.030% in autopsy series [1]. About 75% of these primary cardiac tumors are benign tumors such as myxoma, lipoma, papillary fibroelastoma and rhabdomyoma [2]. The most frequently reported malignant primary cardiac tumors are sarcomas, angiosarcomas and myxofibrosarcomas being the most common sarcomas of the heart [3].

Osteosarcoma is one of the most prevalent primary skeletal tumors. Extraskeletal osteosarcoma is a rare condition, mostly seen in the soft tissue of the lower limbs. Cardiac osteosarcomas are extremely rare; they account for only 3-9% of all cardiac sarcomas [4].

A malignant cardiac tumor mass can cause valve dysfunction, arrhythmias and heart failure. Embolization of tumor pieces and local invasion to vital structures can also cause symptoms. Mass effect, obstruction or functional effects cause a wide spectrum of signs and symptoms in malignant cardiac tumors. These include neurological deficits, palpitations, cough, hemoptysis, pleural effusion, chest pain, syncope, ankle edema, elevated liver

enzymes (in the presence of right heart failure) and other symptoms of heart failure, valve dysfunction and thrombus/tumor embolization.

Primary cardiac osteosarcomas are seen in a broad range of ages and do not show any gender predominance [5]. Compared with extracardiac sarcomas, prognosis is very poor for patients with cardiac sarcomas. Based on this, it is important to distinguish the benign tumors of the heart from sarcomas. Imaging techniques such as echo, CT and MRI help in some way to explain the nature of the tumor, at least in the presence of distant metastasis. Bone scans are also important for patient follow-up [6]. However, tumor excision and pathological confirmation are still the only possibilities to prove the diagnosis [3]. Tumor excision is the most important part of the treatment of cardiac osteosarcomas. However, the difficulties of complete tumor resection and proximity to vital structures limit the benefits of surgery. Thirty-three percent of the patients with high-grade tumors die within 5 days of the initial surgery [3]. Local recurrence rates are high in the absence of adjuvant therapy.

In our case, a mass that almost filled the left atrium and caused a mitral valve stenosis-like clinical picture was treated with surgery and valve replacement, which ultimately improved the symptoms. As the tumor was a high-grade osteosarcoma, chemotherapy was combined with surgery. It seems reasonable to administer adjuvant cisplatin plus ifosfamide combination chemotherapy to such a patient by extrapolating the data from skeletal osteosarcomas. At the end of the treatment, the patient was completely asymptomatic, followed-up with a bone scan, echo and CT, which did not show any metastasis and/or local recurrence for 9 months. Left hemiparesis was thought to be related to the metastatic disease or a pontine glioma. Because of the close proximity to the vital structures, surgery or a biopsy could not be performed. However, radiological examination led to the diagnosis of metastases of a primary osteosarcoma. In conclusion, this paper reported the use of adjuvant cisplatin plus ifosfamide in a patient with curatively resected high-grade cardiac osteosarcoma that yielded a disease-free survival for about 9 months.

Disclosure Statement

The authors have no conflicts of interest to disclose.

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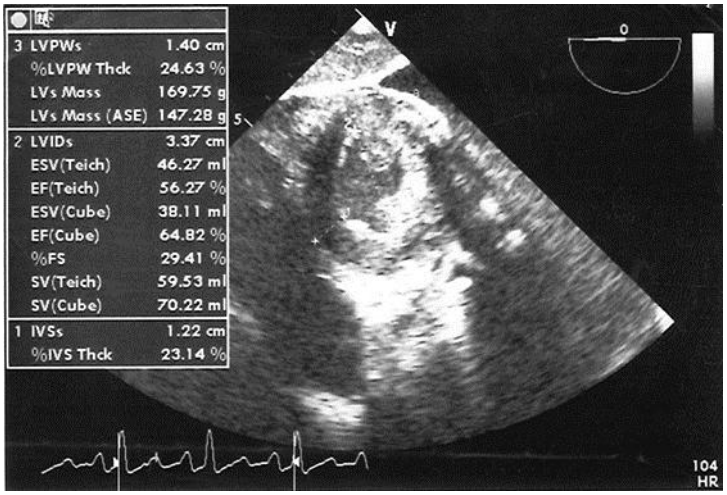

Fig. 1. Echo of the left atrial mass: heterogeneous mass filling the left atrial cavity and causing mitral stenosis.

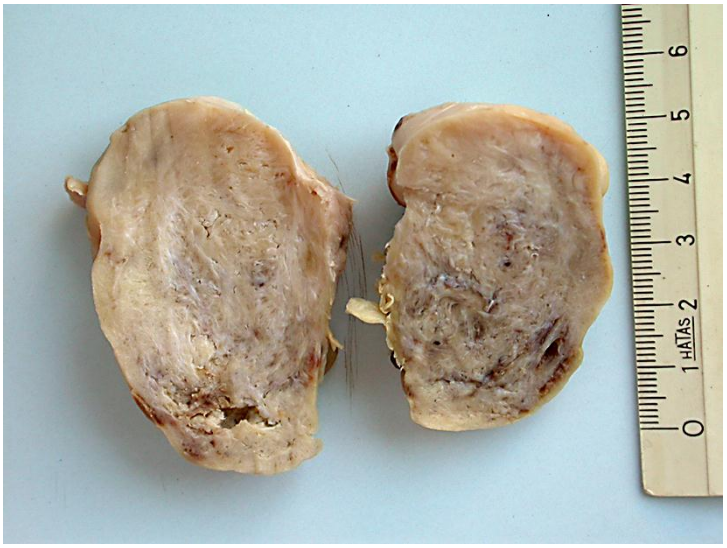

Fig. 2. Photomicrograph of the osteosarcoma: fleshy, white tumoral mass with irregular discrete margins in the atrial wall.

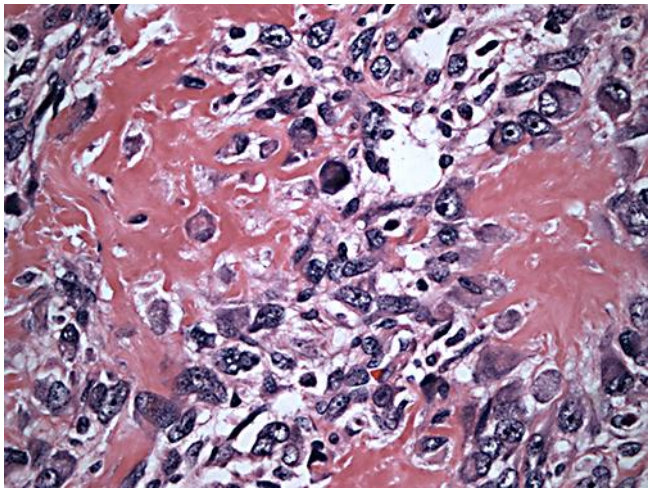

Fig. 3. Photomicrograph of the osteosarcoma: extensive osteoid production by tumoral osteoblastic cells.

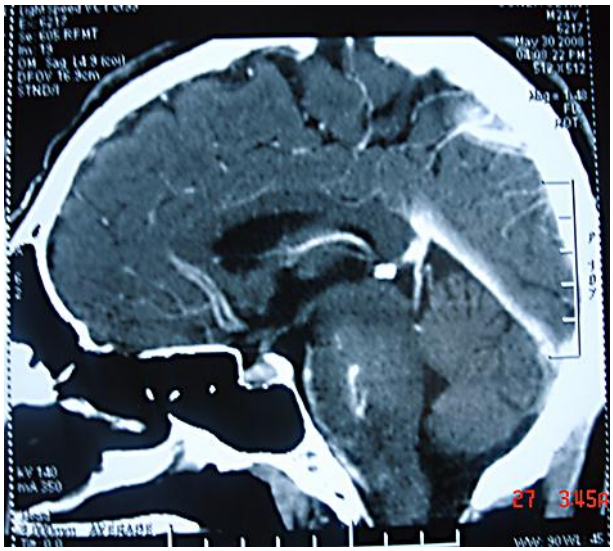

Fig. 4. CT scan of the central pontine mass: hypervascular pontine mass with calcification.