Calcified Left Ventricular Mass: Unusual Clinical, Echocardiographic, and Computed Tomographic Findings of Primary Cardiac Osteosarcoma

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Primary cardiac osteosarcomas are rare and usually originate in the left atrium. In contrast, osteosarcomas metastatic to the heart most commonly involve the right cardiac chambers. This case report describes an unusual primary cardiac osteosarcoma, initially observed as a slowly growing, densely calcified mass of the left ventricle with subsequent secondary pulmonary metastasis. Although cardiac tumors may be asymptomatic, this patient had recurrent bouts of ventricular tachyarrhythmia. We describe the clinical, echocardiographic, and radiological observations spanning 6 years and the gross and microscopic features at autopsy.

The origin of the mass and its relation to the adjacent papillary muscle were uncertain but were thought possibly to represent calcification of the posteromedial papillary muscle. The patient was reluctant at that time to pursue any surgical option. Follow-up CT of the chest 3 months later showed no change in the size of the mass.

In August 1994, the patient presented elsewhere with unstable angina and subsequently underwent cardiac catheterization, which showed severe multivessel coronary artery disease. Uncomplicated triple coronary artery bypass grafting was performed elsewhere. Intraoperatively, the surgeon described the presence of a large posterior myocardial and endocardial calcification. No biopsy specimens were obtained. The patient's immediate postoperative course was uneventful. However, 1 month after the cardiac operation, the patient was readmitted to the hospital because of lower extremity deep venous thrombosis and a pulmonary embolism, which were treated with anticoagulation and an inferior venocaval filter.

The patient remained asymptomatic until October 1995, when he presented elsewhere with recurrent monomorphic ventricular tachycardia, up to 3 times daily. There was no evidence of acute myocardial infarction by cardiac enzymes. The arrhythmia was initially treated with sotalol, then lidocaine, and later oral amiodarone and tocainide. The patient underwent an electrophysiologic study while he was taking antiarrhythmic medications. No ventricular arrhythmias could be induced with up to triple ventricular extrastimuli from 2 right ventricular sites. Coronary angiography showed that the saphenous venous graft to the first obtuse marginal artery was occluded. Left ventriculography demonstrated a mildly dilated left ventricle with a large area of posterior calcification but normal left ven-
tricular function. The patient was dismissed home and was taking amiodarone and tocainide.

On a follow-up visit to our institution in January 1996, the patient denied having recurrence of symptomatic ventricular tachycardia or symptoms of angina or heart failure. An electrophysiology consultation recommended that tocainide use be discontinued. Transesophageal echocardiography showed that the calcified left ventricular mass was 6.5 cm in length (Figure 1, right). Repeated contrast-enhanced CT of the chest confirmed that the mass was now larger, 6.5 x 2.2 cm (Figure 3, left), and showed a new 1.5-cm, pleural-based, noncalcified nodule with insignificant nodule enhancement in the left lower lung (Figure 3, right).

The patient underwent a left posterolateral thoracotomy, and excision of the lung nodule showed metastatic sarcoma. With immunoperoxidase studies performed on paraffin-embedded lung tissue, neoplastic cells reacted with antibodies to vimentin but did not react with antibodies to keratin (AE1/AE3), actin, desmin, CD34, or S-100 protein. The results supported the diagnosis of osteogenic sarcoma. Work-up for primary bone cancer was negative. The patient was considered to have a primary cardiac osteosarcoma with pulmonary metastasis, and therefore systemic chemotherapy was initiated. He received 2 cycles of ifosfamide, etoposide, and mesna and 1 cycle of mitomycin, doxorubicin, and cisplatin. Repeated echocardiography showed no change in the size of the calcified left ventricular mass.

In late December 1996, the patient had an out-of-hospital cardiac arrest. On arrival at the emergency department,
he was in full cardiopulmonary arrest with a pulse rate of 20 to 30/min. He was resuscitated with atropine and epi­
nephrine. Electrocardiography demonstrated acute inferior wall ST elevation, suggestive of acute myocardial infarc­
tion. The patient’s rhythm remained stable thereafter, but he remained comatose. He also developed coagulase­
negative Staphylococcus bacteremia; respiratory, renal, and liver failure; and anoxic encephalopathy, from which he never recovered. The patient died a few days later from multiorgan failure.

Autopsy was limited to the heart, and the intact specimen was referred to one of us (W.D.E.) for evaluation. The large left ventricular osteosarcoma, 6.8 × 6.0 × 5.3 cm, involved the inferoseptal wall of the left ventricle and the posteromedial papillary muscle (Figure 4). Microscopic findings were typical for osteosarcoma (Figure 5). The right coronary artery, which supplied the tumor, was dilated and was involved with only mild atherosclerosis. The saphenous vein graft to the first obtuse margin branch showed old thrombotic occlusion, whereas the other 2 grafts were widely patent. Despite the presence of cardiomegaly (heart, 725 g), there were no old or recent myocardial infarcts.

**DISCUSSION**

Primary cardiac tumors are rare, with an incidence of 0.002% to 0.33% in reported autopsy series, and fewer than one third are malignant. Of all malignant tumors, 33% are angiosarcomas, 20% are rhabdomyosarcomas, 15% are mesotheliomas, and 10% are fibromas. Primary extraskeletal osteosarcomas are very rare, accounting for approximately 1.1% of all cardiac tumors and are mostly seen within the left atrium. Primary cardiac tumors can involve not only the heart but also the surrounding structures; thus, they can have overt manifestations and have been referred to as “great imitators” of cardiovascular disorders. They may cause cardiac obstruction, heart failure, chest pain, syncope, pulmonary hypertension, and cardiac arrhythmias with sudden death. The clinical manifestations are often related to the precise anatomical site of the cardiac tumor, rather than the histological types.

Our patient had documented spontaneous, monomorphic ventricular tachycardia in the absence of a prior history of myocardial infarction and in the presence of normal left ventricular function. This tachycardia was presumably related to the osteosarcoma, which involved the endomyocardium. Recurrent ventricular tachycardia has
Figure 5. Photomicrograph of primary cardiac osteosarcoma. Malignant spindle cells are associated with focal bone formation (hematoxylin-eosin; original magnification x300).

Cardiac osteosarcomas are heterogeneous, with large areas of fibrosarcoma or malignant fibrous histiocytoma and smaller regions of osteosarcoma and chondrosarcoma. Primary cardiac osteosarcomas most commonly involve the left atrium and the mitral valve and can metastasize, as in the current case. This case is unique because of the left ventricular origin of the osteosarcoma.

Definitive differential echocardiographic features of primary cardiac osteosarcoma have not been described. Echocardiography and CT provide important information on the size and shape, site of attachment, and pattern of movement of the tumor. Malignant tumors often involve the right side of the heart and invade the myocardium, with extension into the pulmonary veins or pericardial space with secondary hemorrhagic pericardial effusion. Because primary osteosarcomas are rare and most often arise in the left atrium,2 their appearance and location may simulate a myxoma. Despite individual variability, myxomas typically have a short broad-based attachment and are pedunculated, soft, and gelatinous, and they frequently contain areas of hemorrhage and necrosis. However, in up to 10% of myxomas, calcification may develop. Two additional features of osteosarcomas, their origin from nonseptal atrial walls and their tendency to extend into pulmonary veins, are useful for distinguishing osteosarcomas from myxomas and other left atrial tumors.10 The echocardiographic appearance of metastatic osteosarcoma is similar to the primary type except that metastatic tumors often involve multiple sites within the heart, tend to be larger, and are often intracavitary.10,11

Computed tomography and, more recently, magnetic resonance imaging have emerged as the preferred techniques for the evaluation of pericardial and paracardiac masses.12,13 Computed tomography not only can detect masses within the cardiac chambers but also can define fully their extent within the myocardial wall and outside the heart. The contrast resolution of CT may provide some insight into the composition of the mass, such as the presence of fat or calcium. A CT scan is sensitive for detecting even minute amounts of calcium. A review by Chaloupka et al18 described their experience with 2 cases of primary sarcoma of the heart and great vessels in which CT was useful for evaluating the full extent of tumor and suggesting a more definitive diagnosis. In one of the cases, calcification was present, suggesting that at least part of the tumor might contain osteosarcomatous elements, a finding later confirmed histologically.

Distinguishing tumor ossification from other causes of myocardial calcification may be difficult if there is also a known history of myocardial infarction, infection, or trauma. Myocardial calcification occurs most commonly in patients with left ventricular aneurysms who have had a myocardial infarction. However, the calcium deposits in these patients are usually curvilinear within the periphery of the infarct or aneurysm and occasionally may be homogeneous when an entire infarcted area calcifies.9 Tumor calcifications have been reported in several types of benign cardiac tumors, including rhabdomyomas, myxomas, fibromas, teratomas, hematomas, and angiomas.2,3,5 Calcification varies from a speckled pattern to a round clump of calcium mimicking mitral annular or valve calcification and may be visible on plain x-ray films or seen only with fluoroscopy or CT. The extent of calcification is generally minimal in benign tumors, unlike the tumor ossification in the current case. Dense calcification is unusual and should raise suspicion for an extraskeletal osteosarcoma.

For malignant cardiac tumors, a palliative surgical procedure can improve the patient’s quality of life, and chemotherapy and radiation therapy can sometimes extend survival from 1 to 3 years.6,7 The prognosis of this subset of patients after surgical resection is reported to be poor. More recently, some limited success in the treatment of cardiac sarcoma was achieved with orthotopic cardiac transplantation in combination with systemic chemotherapy.18

CONCLUSION

Malignant primary cardiac tumors occur infrequently but are potentially life threatening. This case report of an uncommon primary left ventricular cardiac osteosarcoma with pulmonary metastasis was well documented by serial echocardiography and CT. To our knowledge, this is the first report of a primary cardiac osteosarcoma arising from the left ventricle. Most primary cardiac osteosarcomas originate from the left atrium and therefore can be clinically mistaken for myxomas. In our patient, the large ossi-
fied mass may have contributed to the development of recurrent ventricular tachyarrhythmias.

REFERENCES


