MR Imaging Findings of a Primary Cardiac Osteosarcoma and Its Bone Metastasis with Histopathologic Correlation

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An osteosarcoma of cardiac origin is extremely rare, and a comprehensive description of MR imaging (MRI) findings of cardiac osteosarcoma and its metastasis in the femur have not been reported in the literature. We present a case of cardiac osteosarcoma in a 47-year-old woman and its metastasis to the femur, focusing on the description of MRI findings of the cardiac and metastatic bony osteosarcoma with a histopathologic correlation.

Index terms: Bone neoplasms; Bone neoplasms, diagnosis; Bone neoplasms, metastasis; Bone neoplasms, MR; Neoplasms, heart

INTRODUCTION

Primary cardiac tumors have a reported incidence of 0.002–0.3% in autopsy series. Cardiac osteosarcomas are very rare, accounting for less than 10% of primary malignant cardiac tumors (1, 2). An osteosarcoma is a malignant osteoid or bone producing tumor. Most reports in the radiology literature describe the characteristic CT imaging findings of osteosarcomas as mineralized lesions (3). However, its MRI findings have not been described in detail. Herein, we present a case of a 47-year-old woman with primary cardiac osteosarcoma which metastasized to the femur, 18 months after tumor resection and we describe

MRI findings of the primary cardiac tumor and its bone metastasis with histopathologic correlation.

CASE REPORT

A 47-year-old woman was admitted to the hospital with gradually worsening dyspnea, orthopnea, and nocturnal paroxysmal dyspnea. A non-contrast chest CT (Mx 8000 CT scanner, Philips) revealed bilateral pulmonary congestion and pleural effusion with left atrial enlargement and a low-attenuated mass in the left atrium (LA). Neither distinct calcification within the mass nor mediastinal lymph node enlargement was noted. MR images (Intera 1.5T, Philips) demonstrated a 46 × 41 mm, well defined mass with a broad base at the posterolateral wall of the LA. The mass showed mildly heterogeneous and high signal intensity (SI) on double inversion-recovery T1-weighted images (T1WI), with fat saturation (Fig. 1A). In addition, triple inversion recovery T2-weighted images (T2WI) with fat saturation and cine-cardiac images showed heterogeneously high SI (Fig. 1B, C). On delayed contrast-enhancement MR images, the mass was heterogeneously enhanced (Fig. 1D).

The patient underwent surgical removal of the mass,

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Fig. 1. MRI and histologic finding of cardiac osteosarcoma and its femoral metastasis.

A. Axial double inversion-recovery T1-weighted image with fat saturation (TR/TE, 960/8.6 msec) showed well-demarcated mass of mildly heterogeneous and high signal intensity without evidence of invasion into surrounding structures. **B, C.** Axial triple inversion-recovery T2-weighted image with fat saturation (TR/TE, 960/100 msec) and four chamber view of cine image (3211/1605 msec) showed heterogeneous signal intensity of mass. **D.** Axial image of delayed contrast-enhanced MRI (TR/TE, 4457/1427 msec) showed heterogeneous enhancement. **E.** Mass measured about $5.0 \times 3.7 \times 2.3$ cm and had focal hemorrhage and necrosis on gross specimen. **F.** Photomicrograph showed atypical spindle cells with abundant collagen material and displayed focal immature osteoid production (Hematoxylin & Eosin stain, $\times 40$).

Fig. 1. MRI and histologic finding of cardiac osteosarcoma and its femoral metastasis.

G. T1-weighted coronal image (TR/TE, 651/20 msec) showed mass that was isointense to muscle at right femoral neck. Peripheral rim with low signal intensity was seen. **H.** T2-weighted axial image (TR/TE, 4053/100 msec) showed heterogeneously high signal intensity lesion with peripheral hypointense rim at right femoral neck. **I.** Gadolinium-enhanced T1-weighted coronal image with fat suppression (TR/TE, 540/17 msec) was performed and lesion showed heterogeneous enhancement within central portion of non-enhancement. **J.** Atypical spindle cells with abundant collagen material were seen on photomicrograph (Hematoxylin & Eosin stain, $\times 40$).

which was found to be smooth, well-encapsulated with focal necrosis and hemorrhage (Fig. 1E). Histologically, the tumor was mainly composed of atypical spindle cells with high nuclear density and frequent mitosis in the background of abundant collagen and had foci of hemorrhage and coagulative necrosis. In addition, the presence of minimal osteoid deposition foci confirmed the tumor as a fibroblastic osteosarcoma (Fig. 1F). At that time, a bone scan was performed to identify any primary bone lesion or

bone metastasis. The result was negative and, therefore, osteosarcoma of bone-origin and the presence of bone metastasis were ruled out. The patient was further treated with adjuvant chemotherapy and radiotherapy. Periodic check-ups with chest CT revealed no evidence of regional recurrence.

Eighteen months after surgery, the patient complained of right hip pain and consequently underwent a hip MRI. The examination revealed a mass at the right femoral neck.

The lesion was isointense to muscle on T1WI (Fig. 1G) and showed a heterogeneously high SI on T2WI (Fig. 1H). The mass was well-demarcated, with a peripheral low SI rim on both T1WI and T2WI. Gadolinium-enhanced T1WI showed heterogeneous enhancement with central non-enhancing portions (Fig. 1I). A bone scan using technetium-99m (Vertex™ V60, Philips, Best, The Netherlands) was performed to evaluate the presence of other bone lesions, and showed an increased uptake at the known lesion in the right femur and no other pathologic uptake. A biopsy and following histological analysis confirmed the lesions as metastases from the previously resected cardiac tumor on the basis of histologic similarity between them (Fig. 1J).

DISCUSSION

Most cardiac tumors are metastatic tumors which are 20–40 times more common than primary tumors. About 75% of all primary cardiac tumors are benign and the remaining 25% being primary malignant tumors. The vast majority (95%) of primary malignant tumors are sarcomas and osteosarcomas, which are relatively quite rare, accounting for less than 10% primary cardiac malignant tumors (1, 2). Primary cardiac osteosarcomas exhibit a predilection for the LA, whereas the majority of malignant cardiac tumors, such as metastatic tumors, angiosarcomas or lymphomas, commonly arise in the right atrium (4). Due to its left atrial location, patients with cardiac osteosarcomas generally present as congestive heart failure with respiratory symptoms as in our case (5). Moreover, they tend to have a broad base of attachment, at a location away from the fossa ovalis and invading the surrounding structures, such as the mitral valve or pulmonary veins (2).

Osteosarcomas are a heterogeneous group of tumors containing malignant, bone-producing cells. Histologically, the tumor contains variable amounts of spindle-cells, osteoid, bone, or cartilage. Depending on the predominant component, osteosarcomas can be subgrouped as osteoblastic, chondroblastic, or fibroblastic (3). Macroscopically, the tumor may be well circumscribed and pseudoencapsulated, or infiltrate into the surrounding tissues and calcification, necrosis, or hemorrhage within the tumor can be seen (5, 6). The characteristic CT findings of cardiac osteosarcomas have been reported as a low attenuation mass with dense mineralization (3, 7). Moreover, osteosarcomas often have dense calcification to form hardy stone masses or may also have minimal

calcification in the early stage. However, a tumor lacking the identifiable calcification on CT presents as a nonspecific soft tissue mass as it did in our case, which made us miss the osteosarcoma as part of our differential diagnosis. As the microscopic examination showed, the tumor had too minimal a portion of osteoid deposition to reveal calcification on CT.

The first report of MRI findings of cardiac osteosarcomas was presented in 2001 by Yamagishi et al. (8) in which the tumor appeared as a huge mass of heterogeneous SI in the LA. The report focused on the role of MRI in differentiating malignant from benign tumors based on a broad-based attachment and invasive features. Since then, a few cases have been reported with MR images, however, with no detailed description of MRI findings and no contrast-enhancement study included. According to previous reports, cardiac osteosarcomas appear to be irregularly lobulated and may have variable heterogeneous SI on T1WI and heterogeneously high SI on T2WI (2, 4, 6, 7, 9). In our case, the tumor showed mildly heterogeneous and high SI on both T1WI and T2WI. In addition, the heterogeneity of the signal on T2WI was more definite on cine images with long echo time (Fig. 1C). This heterogeneity is attributed to the heterogeneous histologic composition since some portion of the tumor was composed of spindle cells with collagen material, while the other portion consisted of necrosis and hemorrhage. It is known that hypercellular areas are well-enhanced after gadolinium administration and show high SI on T2WI, whereas low SI on T2WI corresponds to hypocellular areas (10). Our case showed heterogeneous enhancement and the areas of enhancement are well-matched to the areas of high SI on T2WI, suggesting hypercellular areas. As discussed earlier, the tumor showed no calcification which would have shown low SI on all pulse sequences, due to minimal osteoid formation discernable only on histologic examination.

Primary cardiac osteosarcomas are very aggressive with a high frequency of recurrence and metastasis. Metastasis was observed at various sites including skin, lung, liver, bones, brain and adrenal glands (5). Our case also revealed metastasis at the femur, although the patient underwent chemotherapy and radiotherapy after surgical removal of the tumor. A comparison of the metastatic bone lesion with the cardiac lesion indicated less heterogeneity and denser enhancement with the lesser portion of non-enhancement which meant lesser extent of central necrosis in the metastatic bone lesion. The peripheral rim of low SI on

both T1WI and T2WI showed enhancement on gadolinium-enhanced images. This finding is considered to correspond to a fibrous pseudocapsule, which has been reported in some extraosseous osteosarcomas (10).

We present a case of primary cardiac osteosarcoma with a detailed description of MRI findings and histopathologic correlation of cardiac osteosarcoma and its bone metastasis.

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