Osteogenic Sarcoma Metastatic to the Heart via Infradiaphragmatic Venous Extension — A Case Report

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We report a rare case of osteogenic sarcoma of the right pelvis with intracavitary metastases to the right heart through tumor growth along and within the inferior vena cava. Excision of the cardiac metastases was done as a palliative procedure, although the patient died of disseminated disease 6 months later.

Key Words: Osteogenic sarcoma • Metastatic cardiac tumor

INTRODUCTION

Secondary intracardiac neoplasms arise most frequently from renal cell carcinoma, bronchogenic carcinoma, Wilms’ tumor, chondrosarcoma, and osteogenic sarcoma.1,2 However, the authors were unable to find any published description of a secondary intracardiac osteogenic sarcoma exactly like the 1 reported in this paper, although there have been rare lesions with somewhat similar features in the literature.2,4

CASE REPORT

A 16-year-old girl was admitted because of an acute onset of right thigh pain occurring over 1 week. The pain was non-radiating, dull and aching in character, did not incapacitate her, but was more intense on walking. A year prior to admission, she had noted occasional episodes of chest tightness and exertional dyspnea; the conditions were exacerbated on deep inspiration and on lying down.

She received medication for this with no improvement. Three days prior to entry, she had gone to another hospital where a grade 1/6 cardiac murmur was detected and echocardiographic study revealed a tumor in the right atrium; she was referred to this hospital for further evaluation. The patient had no experience of trauma, fever, cough or hemoptysis, and her general health had been good.

On admission, no definite cardiac murmur or tumor plop could be identified on cardiac auscultation. The positive physical findings were limited to the right lower limb. There was a tender, poorly defined enlargement of the upper portion of the right thigh (circumference: right thigh, 48 cm; left thigh, 43 cm). No peripheral cyanosis or edema was noted, and no inguinal lymph nodes were palpable. Electrocardiogram showed normal sinus rhythm and right axis deviation. Chest X-ray was normal and there was no evidence of pulmonary metastases or pleural effusion.

Echocardiography showed a highly mobile serpentine mass in the right atrium, extending from inferior vena cava. The mass lay within the right atrium in systole and passed through the tricuspid valve into the right ventricle during diastole (Figure 1A to C). Magnetic resonance imaging demonstrated that the primary tumor mass arose from the right pelvis.

Since she was otherwise free from disease, excision of the cardiac metastases was done as a palliative procedure. A worm-shaped, yellow-gray, firm, glistening, smooth-
surfaced tumor was found in the right atrium. Because there were no attachments or obvious invasion of tumor into the myocardium, it was excised at the junction of the inferior vena cava and the right atrium (Figure 1D). The pulmonary trunk was clean and free of intracavitary tumor growth or embolus. Microscopic examination showed that the intracardiac tumor consisted of distinctly osteoblast-like cells with small, round, deeply stained nuclei embedded in amorphous homogeneous eosinophilic material resembling osteoid, consistent with the diagnosis of metastatic osteogenic sarcoma (Figure 1E).

The patient was discharged 15 days later after an uneventful postoperative course. Unfortunately, she died of disseminated metastases 6 months later despite a variety of chemotherapeutic regimens.

**DISCUSSION**

Clinical reports of both primary and secondary cardiac involvement with osteogenic sarcoma have been reported previously. Although osteosarcoma metastases to the heart, in contrast to primary osteosarcomas of the heart, are often right-sided, suggesting intravascular and intracardiac seeding and/or spread, documented cases of...
the tumor reaching the right heart via infradiaphragmatic venous extension were sparse.\textsuperscript{2-4} Inasmuch as there was no demonstrable evidence of systemic, blood-borne, visceral metastases or true endocardial implants in our patient, it seems clear that the intracardiac neoplasm represented a free-growing tumor extending from the primary osteosarcoma in the pelvis.

Although certain clinical manifestations can be suggestive of a metastatic cardiac tumor, especially in those tumors invading the pericardium, no clinical finding or set of findings is pathognomonic.\textsuperscript{1,3} The reported symptoms and signs in patients with metastatic osteogenic sarcoma of the heart include dyspnea, syncope, peripheral cyanosis and subcutaneous edema secondary to massive tumor invasion, and compression or obstruction of the venous system.\textsuperscript{3} Electrocardiography may show nonspecific changes, bundle branch block, and cardiac arrhythmia.\textsuperscript{3} Chest radiography may show pulmonary metastases and/or bony structure metastases (radiographically visible) inside the heart, which is the most unique feature of osteogenic sarcoma involving the heart.\textsuperscript{3} In our case, the initial symptoms of chest tightness and exertional dyspnea were considered nonspecific. As there was no identifiable tumor plop or cardiac murmur, the results of cardiac examination did not suggest this diagnosis. Because there was no formation of bone in the intracardiac tumor, the chest X-ray was normal. The tumor was detected unexpectedly, as with the majority of reported patients, during echocardiography.

Transthoracic and transesophageal echocardiography remains the procedure of choice in screening and fully elucidating the position and size of cardiac tumors.\textsuperscript{1} It also provides real-time portrayal of those extremely mobile tumors;\textsuperscript{1,3,6} their easy prolapse through the atrio-ventricular valve may cause hemodynamic compromise, clearly illustrating the need for removal before attaining large size and causing obstruction or embolization, as in our case. Although surgery is contraindicated in the presence of metastatic disease, the role of surgical palliation of intracaval and intracardiac tumor to relieve obstruction and to prevent embolization is nevertheless valid.\textsuperscript{3,6} However, even with successful surgical resection with or without chemotherapy, the prognosis of patients with osteosarcoma remains poor.\textsuperscript{2,4,6}

REFERENCES