Primary right ventricular osteosarcoma

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L Zhang, J Ellis, D Kumar, T Deng. Primary right ventricular osteosarcoma. *Can J Cardiol* 2008;24(3):225-226.

A 41-year-old man suffering from dyspnea on exertion was found to have a mass located on the right ventricle. Resection of the tumour required reconstruction of the right ventricular outflow tract and replacement of the pulmonary valve. Histopathology confirmed the diagnosis of high-grade osteosarcoma.

Key Words: Cardiac tumour; Surgery

Primary cardiac osteosarcomas are an uncommon malignancy, accounting for approximately 3% to 9% of all cardiac sarcomas, and the majority arise from the left atrium (1). Primary cardiac osteosarcomas arising from the right ventricle have scarcely been reported in the past two decades (2). The present report describes the clinical and pathological features of a patient with a primary osteosarcoma in the right ventricle.

CASE PRESENTATION

A 41-year-old man without a significant medical history presented with progressive dyspnea on exertion associated with chest tightness for the previous seven months. Despite an initially negative echocardiogram, he was found to have a systolic murmur at the left sternal border one week before admission. There was no jugular venous distension, and no calf or ankle edema. His lungs were clear and an abdominal examination was normal. There were no abnormal findings in the musculoskeletal system. An electrocardiogram showed sinus bradycardia with unspecific ST segment elevation. Cardiac computed tomography showed a 2.3 cm mass at the right ventricular outflow tract (RVOT); tissue abnormalities of the mass were consistent with those of a sarcoma (Figure 1). The mass caused significant obstruction of the RVOT, with a gradient of 64 mmHg, as measured by echocardiography. A positron emission tomography scan revealed abnormal hypermetabolic fluorodeoxyglucose uptake in the RVOT, along with an active neoplastic mass in the RVOT below the pulmonary valve, involving the valve leaflets and wall of the RVOT. A systemic evaluation did not reveal any extracardiac origins of the tumour. Because of the patient's dyspnea and RVOT obstruction, a decision was made to perform surgery for definitive diagnosis and resection for possible cure and relief of symptoms.

The patient underwent resection of the mass and reconstruction of the RVOT using a cardiopulmonary bypass. The tumour measured 2 cm × 3 cm, was attached to the free wall of the RVOT and infiltrated the pulmonary valve. The tumour, along with a part of the free wall of the RVOT, was resected with grossly clear margins. The defect was repaired with a Gore-Tex patch (WL Gore & Associates, USA). The pulmonary valve also had to be resected, and a Medtronic Mosaic bioprosthetic valve (Medtronic Inc, USA) was implanted. The patient

Un ostéosarcome ventriculaire droit primaire

On a découvert une masse sur le ventricule droit d'un homme de 41 ans souffrant de dyspnée à l'effort. La résection de la tumeur a exigé la reconstruction de la chambre de chasse du ventricule gauche et le remplacement de la valvule pulmonaire. L'histopathologie a confirmé le diagnostic d'ostéosarcome de haut grade.

Figure 1) Contrast-enhanced computed tomographic scan revealing an intraluminal filling defect located at the right ventricular outflow tract, extending into the main pulmonary artery

did well postoperatively and had subsequently started chemotherapy as an outpatient. He was alive one year after surgery (at the time of the report).

Histologically, the tumour was consistent with a high-grade sarcoma; there was an appearance of numerous pleomorphic malignant cells and some spindle cells, with multiple areas of osteoid and chondroid formation (Figure 2). There were obvious, large, atypical malignant cells within an osteoid matrix. Extensive geographic necrosis and many mitotic figures were noted. Immunohistochemistry studies showed tumour cells positive for alpha-smooth muscle actin (Figure 3).

DISCUSSION

Primary cardiac tumours are rare. Among the primary cardiac sarcomas, angiosarcomas are the most common, followed by undifferentiated sarcomas, osteosarcomas, fibrosarcomas and malignant fibrous histiocytomas. Angiosarcomas usually originate from the right atrium or ventricle, while osteosarcomas are usually left-sided.

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Received for publication May 16, 2006. Accepted June 4, 2006

Figure 2) Hematoxylin and eosin staining revealing osteoid formation and calcification in a background of pleomorphic malignant cells (original magnification $\times 100$)

The largest series of primary cardiac osteosarcomas was reported by Burke and Virmani (1), who described nine cases. Patient age ranged from 24 to 67 years, and the symptoms mimicked those of a myxoma; all tumours were located in the left atrium or on the mitral valve. Complete excisions were achieved in eight patients. Except for one patient who was lost to follow-up, all postoperative survivors died from disease or had metastasis. Therefore, the behaviour of cardiac osteosarcomas is similar to that of osteosarcomas originating from other organs, and the prognosis is likely worse in patients older than 40 years of age (3).

Of note, cardiac metastasis of osteosarcomas is often seen on the right side of the heart. Therefore, the diagnosis of a primary cardiac osteosarcoma should be established with extra caution. In the present case, however, all the clinical evidence indicated that the malignancy originated from the right side of the heart. Furthermore, in a patient whose symptoms lasted for longer than six months, the primary origin of the osteosarcoma should be readily confirmed due to its aggressive nature.

The diagnosis of a cardiac osteosarcoma is established on histological and immunohistochemical examinations. The tumour cells are positive for alpha-smooth muscle actin, as observed in cases of non-cardiac osteosarcomas (4). Also, chondroid differentiation is a valuable clue for differential diagnosis.

Figure 3) Immunohistochemical staining for alpha-smooth muscle actin revealing strong positivity of malignant cells (original magnification $\times 200$)

Surgery is certainly the first-line treatment for cardiac malignancies. A reconstruction of the valves and chambers may be necessary to ensure a radial resection of the tumour. Heart transplantation is an alternative approach to treat an unresectable tumour, given that no evidence of remote metastasis exists (5). Although the prognosis of primary cardiac osteosarcoma is considered to be poor, surgery can prolong survival if performed early (1).

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