Asymptomatic metastatic osteosarcoma to the right ventricle: Case report and review of the literature

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Metastatic cardiac tumors are far more common than primary tumors, and benign primary cardiac tumors are more common than malignant tumors. We report a 22-year-old Saudi woman with right femur osteosarcoma who was found to have a large right ventricular mass by transthoracic and transesophageal echocardiography. Diagnosis was highly suggestive by cardiac magnetic resonance imaging (MRI) and fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) scan. We performed a review of the literature for metastatic osteosarcoma of the right ventricle.

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Abbreviations: RV, right ventricle, TTE, transthoracic echocardiography, TEE, and transesophageal echocardiography, MRI, magnetic resonance imaging, FDG, fluorodeoxyglucose, PET, positron emission tomography, CT, computed tomography.

Keywords: Cardiac tumors, Intracardiac metastasis, Cardiac osteosarcoma

Introduction

The heart may rarely be affected by primary or secondary tumors. Secondaries may occur by local extension or haematogenous spread [8,6]. Osteosarcoma very rarely metastasizes to the heart. About 27 cases in the last 55 years have been reported in the literature so far. However, the authors were unable to find any published description of a secondary intracardiac osteogenic sarcoma exactly like the one reported in this report, although, there have been rare lesions with somewhat similar features in the literature [1]. Very rarely, cardiac tumors are incidentally discovered in asymptomatic patients [3]. We report a rare case of asymptomatic metastatic osteosarcoma to the right ventricle (RV) with a review of the literature.

Case report

A 22-year-old Saudi woman was diagnosed with high grade right femur osteosarcoma. She had
limb-sparing surgery shortly after diagnosis followed by adjuvant chemotherapy. She did not have any cardiac complaints. During her elective admission under oncology, she had transthoracic echocardiography for pre-chemotherapy assessment and a well-defined RV mass was found (Fig. 1A and B). Differential diagnosis included tumor or thrombus.

Trans-esophageal echocardiography confirmed that the mass was attached to the RV free wall with a broad base. The mass was homogenous with multiple lobules and measured 4.5 × 3.2 cm (Fig. 2A and B).

Cardiac MRI with and without contrast was done. It showed a large mass in the RV. It was arising from the RV free wall and was occupying almost half of the RV (mid and apical cavity). It had irregular edges with intermediate enhancement on T1 images and is hyperintense on T2 stir images. There was some evidence of contrast uptake on T1 weighted contrast images. It did not seem to have a significant fatty component on T1 weighted images with fat saturation. The magnetic resonance imaging (MRI) features were consistent with tumorous involvement of the RV (Fig. 3A and B).

A transthoracic echocardiography performed 4 months before the current admission was normal with no masses detected in the RV. This ruled out the possibility of a primary tumor from the heart and also indicates rapidly progressing mass in the RV.

Right ventricular biopsy through a femoral approach was performed.

Five biopsies were taken; the histopathology revealed thrombus and myocardium with no
evidence of tumor which was attributed most likely to failure to pick up samples from the tumor mass. To improve diagnostic yield of the biopsy, echocardiography guidance can be used with 3-D transesophageal echocardiography (TEE) or intracardiac echocardiography.

Whole body fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) scans were obtained. It revealed that the RV mass showed intense increased FDG activity. The mass was highly suspicious for metastatic deposits. This lesion was along with other metastasis involving inguinal, deep femoral, popliteal and right hilar lymph nodes (Fig. 4A and B).

The patient had no cardiac symptoms, and was being considered for second line chemotherapy. This patient had a poor overall prognosis, therefore, the decision was not to perform cardiac surgery and closely follow the patient clinically and with serial echocardiography. If obstruction symptoms manifested, then surgery would be strongly considered.

Discussion

Osteosarcoma is a malignant tumor that most frequently involves the appendicular long bones of young adults within the second and 3rd decades of life [9]. The common demographic features for metastatic osteosarcoma includes female sex, second and third decade of age at the time of diagnosis and longer interval to onset of secondary disease [9]. The clinical characteristics involve hemodynamic compromise or precordial abnormality, with magnetic resonance imaging emerging as the gold standard for diagnosis [9]. The first report of MRI findings of cardiac osteosarcomas was presented in 2001 by Yamagishi et al. [11] in which the tumour 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 which are similar to some extent to our case. Since then, a few cases have been reported with MRI images, however, with no detailed description of MRI findings and no contrast enhancement study included [7,5].

In our case FDG-PET/CT scan was a powerful diagnostic tool for detecting tumour mass in the RV along with other metastases and enhances the level of confidence in diagnosis of tumour spread. Also, 3-D echo has improved the detection, and characterization of cardiac tumors. Contrast echo is useful in differentiating thrombus from tumor, and highly vascular tumors like angiosarcomas from other less vascular tumors.

Osteosarcoma very rarely metastasizes to the heart. Very few have been reported to occur in the right ventricle, all of them in the RV outflow tract. To our knowledge, no cases have been reported to occur in the RV apex and free wall and to be discovered before being symptomatic exactly like our case scenario. Platonov et al., has reported one case of metastatic osteosarcoma with the tumor mass attached to the tricuspid valve [9]. The size of metastatic osteosarcoma varies at presentation from small accidentally discovered nodules to huge mass filling the entire right ventricle and extending into the pulmonary valve and across the tricuspid valve and into the right atrial cavity [10]. Diagnosis in some of these cases was made during investigation for severe cardiac failure and in most of them at autopsy [10]. Very few cases diagnosed incidentally and were asymptomatic like our case [3].

Although surgery is generally contraindicated in the presence of metastatic disease, the role of surgical palliation of intracardiac tumor to relieve obstruction and to prevent embolization is nevertheless valid [10]. However, even with successful surgical resection with or without chemotherapy, the prognosis of patients with osteosarcoma remains poor [10]. Cardiac involvement is a strong
predictor of disease elsewhere and mandates careful surveillance, with surgical management likely providing the best outcome. No reports for surgical interventions for metastatic asymptomatic RV masses as in our case. Iyigun et al., reported 17-year-old female with recurrent metastatic osteosarcoma into the left ventricle 5 months after surgical removal of the first LV metastasis. Based on the collaborative decision, chemotherapy was initiated and in 2 months the size of the recurrent tumors had diminished [4].

The diagnosis of cardiac tumors with endomyocardial biopsy has been rarely described. There are reports of failure to get enough tissues to diagnose tumor masses. The use of two-dimensional echocardiography simultaneously with fluoroscopy has been reported as an aid to positioning of biopsy bioptome [2].

The unique features in our case was the location of the mass in the distal to mid part of the RV and despite being large mass, the patient is still completely asymptomatic from cardiac point of view. Anticoagulation has to be initiated in such patients with proven thrombus on top of the tumor by biopsy. There is also debate about the best time to intervene surgically in this clinical scenario.

Disclosures

None.

References