Malignant Nature of Cardiac Liposarcoma Revealed by Fluorine-18 Fluorodeoxyglucose Positron Emission Tomographic Imaging

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Abstract

Primary cardiac liposarcoma is very rare and usually asymptomatic. It is often diagnosed at an advanced or incurable stage without being presented in surgical operation. We report a man in his early sixties with primary cardiac liposarcoma originating from the ventricular septum. The malignant nature of the tumor was suspected by positron emission tomography using fluorine-18 fluorodeoxyglucose as the tracer. The final diagnosis was made histopathologically following surgery. Treatment with carbon ion radiotherapy was applied, but failed to induce tumor regression.

Key words: cardiac neoplasm, fluorine-18 fluorodeoxyglucose, liposarcoma, positron emission tomography

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Introduction

Primary cardiac tumors are rare, with an autopsy incidence ranging from 0.001% to 0.030% (1). Among primary cardiac tumors, liposarcoma is extremely rare, representing only 1% of primary cardiac malignancies (2). In this report, we present a case of primary cardiac liposarcoma that was suspected based on multimodality imaging including positron emission tomography with fluorine-18 fluorodeoxyglucose as tracer and finally histopathologically diagnosed following surgery performed to prevent pulmonary tumor embolism.

Case Report

An asymptomatic man in his early sixties was referred to the cardiology department for assessment of a mass in the ventricular septum that was incidentally found on postoperatively scheduled computed tomography of the chest. He had undergone distal gastrectomy and cholecystectomy for gastric cancer seven years prior and thymoma resection four years prior. Both of his previous tumors showed no evidence of recurrence. He had no significant family history.

Physical examination revealed regular heart sounds at 60 beats/min and blood pressure of 100/60 mm Hg. Electrocardiogram showed normal sinus rhythm without an abnormal Q wave or ST-T change and chest radiograph was normal. Laboratory tests were within normal limits. Computed tomography showed a tumor in the ventricular septum measuring 47×25 mm that was partially enhanced with contrast medium. There was no evidence of pleural or pericardial effusion. On cardiac magnetic resonance imaging, the tumor showed low-signal intensity on T1-weighted image and high-signal intensity on T2-weighted image. On the delayed image of cardiac magnetic resonance imaging with gadodimide hydrate, the whole tumor showed delayed-enhancement (Fig. 1). Based on these imaging modalities, we could not characterize the tumor as benign or malignant. Fluorine-18 fluorodeoxyglucose positron emission tomography was performed to further investigate the nature of the tumor and to rule out extracardiac malignancy. Whole-body fluorine-18 fluorodeoxyglucose positron emission tomography confirmed a high-uptake tumor in the ventricular septum; however, there was no evidence of extracardiac fluorodeoxyglucose-positive lesions (Fig. 2). Thus, based on

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these findings, a primary malignant cardiac tumor was strongly suspected.

Transcatheter echocardiography-guided transvenous myocardial biopsy was performed twice for diagnostic purposes but we could not obtain tumor tissue. Coronary angiography demonstrated normal coronary arteries without any feeding artery to the tumor. Complete surgical resection was considered impossible because of the tumor size. Furthermore, chemotherapy or radiation could not be performed due to the lack of a definite histopathological diagnosis.

One year later, a floating daughter tumor extending from the grown primary ventricular septal tumor measuring 64×37 mm in the right ventricle was found on a regularly scheduled echocardiography. The patient remained asymptomatic. He was urgently referred to the cardiac surgery team to prevent pulmonary tumor embolism. At surgery, several yellow tumors could be seen through the tricuspid valve orifice in the right atrial approach with cardiopulmonary bypass. They were relatively fragile and appeared friable. The maximum daughter tumor was 20 mm in diameter (Fig. 3). The main tumor was large and extended near the coronary sinus in the inferior wall of right ventricle. Since complete resection was impossible, partial resection was performed only for the floating tumors. Histopathological examination
Liposarcoma is the second most common soft tissue sarcoma. It commonly occurs in the lower extremities and retroperitoneum; however it is rarely observed in the heart (2). The overall survival rate of patients with cardiac liposarcoma remains dismal, as with other cardiac sarcomas. The mean survival time of cardiac liposarcoma is 8.3 months (range 2-24 months) (3). The majority of cardiac liposarcomas originate from the right heart, particularly the right atrium (4); however, they are also reported in both ventricles and in the pericardium (4-6).

Liposarcoma has been classified into four pathogenic subtypes: well differentiated, myxoid, round cell, and pleomorphic (poorly differentiated) type. The prognosis of liposarcoma depends on the histological type. The well-differentiated forms present a low-grade malignancy, whereas the poorly differentiated ones are highly malignant, with a pronounced tendency to recur and metastasize after surgery (7). The present case was a histologically pleomorphic type.

Cardiac liposarcomas usually remain clinically silent and are often diagnosed at an advanced or incurable stage. According to previous reports, liposarcomas grow in a spherical and lobulated configuration, filling the intracardiac cavity (8). Affected patients present with symptoms of heart failure related to obstructed blood flow, arrhythmias, tumor embolism, or metastases. Some patients develop pericardial tamponade (4). Complete resection is considered essential for a positive outcome (9). The prognosis of cardiac sarcoma depends on its early diagnosis; however, accurate diagnosis is challenging and rarely possible antemortem.

A variety of diagnostic modalities including computed tomography and cardiac magnetic resonance imaging have been used; however, these modalities are not capable of differentiating between benign and malignant lesions (9). Although there are few data on cardiac tumors, fluorine-18 fluorodeoxyglucose positron emission tomography enabled us to determine that the tumor was malignant due to the finding of high fluorodeoxyglucose uptake (10). A localized high fluorodeoxyglucose uptake in the ventricular septum on whole body positron emission tomography indicated that the tumor was a primary malignant cardiac neoplasm. Of course, noninvasive imaging techniques including fluorine-18 fluorodeoxyglucose positron emission tomography did not allow a definitive diagnosis. We performed myocardial biopsy twice; however, unfortunately we could not obtain tumor tissue. Histological analysis of resected daughter tumors revealed a diagnosis of cardiac liposarcoma. Adjuvant chemotherapy or radiation may be beneficial in palliating...
patient’s symptoms and improving quality of life (11). Accordingly, we prescribed carbon iron radiotherapy that was the latest methodology of radiotherapy; however, no significant effect on tumor size was observed.

In conclusion, we report a very rare case of cardiac liposarcoma originating from the ventricular septum histopathologically diagnosed antemortem. When a patient presents with a cardiac tumor, fluorine-18 fluorodeoxyglucose positron emission tomography imaging could be helpful in obtaining crucial information to determine whether the tumor is benign or malignant.

The authors state that they have no Conflict of Interest (COI).

References