



Unusual Cardiac Metastasis of Nonvisceral Soft Tissue Leiomyosarcoma in the Right Ventricle: A Case Report and Literature Review

우심실로 전이된 내장 외 연부조직 평활근육종:
증례 보고 및 문헌 고찰

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Leiomyosarcoma (LMS) is a soft tissue sarcoma that originates from smooth muscle cells. It is commonly found in the uterus but can occur throughout the body, including the retroperitoneal space, abdominal cavity, and any vascular structure. Although there are many case reports of uterine or vascular LMS metastasizing to the heart, cardiac metastasis from nonvisceral lesions has only been reported in two cases. Herein we report a rare case of a patient presenting metastatic LMS from the left flank in the right ventricle observed with echocardiography and enhanced computed tomography.

Index terms Neoplasm Metastasis; Heart Neoplasms; Leiomyosarcoma; Echocardiography; Multidetector Computed Tomography

INTRODUCTION

Leiomyosarcoma (LMS) is a soft tissue sarcoma that originates from smooth muscle cells. It is commonly found in the uterus but it can occur throughout the body including the retroperitoneal space, abdominal cavity, and any vascular structure. In previous reports, metastatic cardiac tumors were up to 20 times more common than primary tumors and the incidence of cardiac metastatic LMS ranges from 6% to 34% (1). However, these case reports are about the LMS cardiac metastasis from the uterus and vasculature. We

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found only two cases that reported cardiac metastasis from a nonvisceral lesion (2, 3). In this report, we describe a rare case regarding a patient with nonvisceral soft tissue LMS metastasizing to the heart. We also review the literature, concentrating on radiologic imaging findings.

CASE REPORT

A 71-year-old male presented at the cardiothoracic surgery department with dyspnea and chest discomfort. The symptoms started 20 days ago. His D-dimer and N-terminal pro-brain BNP (NT-proBNP) levels were elevated to 3.60 ug/mL and 8090 pg/mL, respectively. The patient had a history of a mass excision 12 years prior for LMS in the left flank, followed by a wedge resection, radiotherapy, and chemotherapy due to metastasis in the upper lobe of the left lung.

Chest radiography showed a mild rightward shift of the trachea, enlarged mediastinal structures and cardiomegaly (Fig. 1A).

Transthoracic echocardiography revealed a huge inhomogeneous echogenic mass in the right ventricle (RV) that appeared lobulated contour with internal hyperechoic foci. Echocardiography also showed tumor invasion to the RV wall, tricuspid valve, and tricuspid chordae (Fig. 1B).

Multidetector CT scan was performed on a 64-slice CT system to obtain pre-contrast and post-contrast CT scans with iodinated non-ionic contrast. In the contrast-enhanced chest CT, a localized lobulated mass was found in the RV of the heart. The mass was measured to be approximately 9.0 cm × 3.4 cm × 6.4 cm in size. There were no calcification or hemorrhage or fat component throughout the entire mass on the pre-contrast images (Fig. 1C). The irregular shaped mass with a lobulated margin showed peripheral enhancement and filled the entire RV. The mass invaded the interventricular septum with irregular thickening and had a small polypoid extrusion into the right ventricular outflow tract on the contrast-enhanced images (Fig. 1D). The CT scan did not find an enlarged lymph node nor a pericardial effusion in the chest.

¹⁸F-fluorodeoxyglucose (¹⁸F-FDG) PET/CT was performed for further work up. ¹⁸F-FDG PET/CT showed an uneven, irregular hypermetabolic huge mass involving the entire RV (Fig. 1E). The maximum standardized uptake value (maxSUV) was 8.4 g/mL.

Emergency mass excision was performed to treat the right sided heart failure. Pathologically, the mass proved to be a LMS (Fig. 1F) and the results of the specimen were reviewed and compared with those taken 12 years ago from his flank. Both showed a high grade sarcoma compatible with LMS, suggesting that it was a metastatic rather than a primary cardiac tumor. In immunohistochemical staining, the tumor cells showed a positive reaction on smooth muscle actin. Tumor cells showed a negative reaction on Desmin, S-100 protein, and CD34 which was pathologically consistent with a LMS. The patient expired 10 days after the surgery.

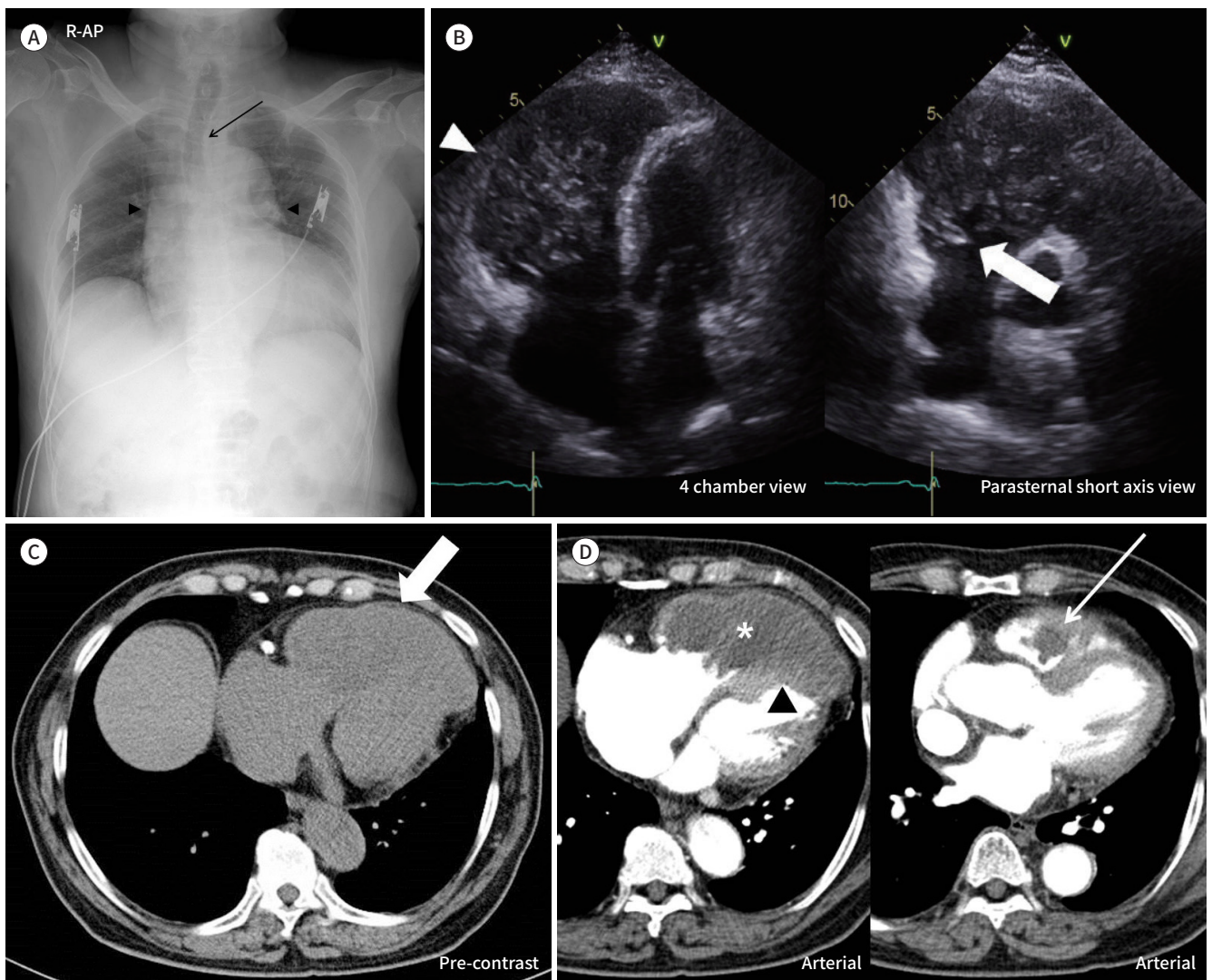
DISCUSSION

Although the incidence of cardiac metastasis occurs approximately 20 to 100 times more frequently than that of the primary tumor, metastasis from extracardiac sarcoma to the heart is uncommon (4). In previous reports, the cardiac metastases from other organs account for 2.3% to 18.3% (5). However, we found only two previous reports of cardiac metastasis from nonvisceral soft tissue LMS. According to the literature, the most common sources of extracardiac

metastases to the heart are the lung, followed by the breast, stomach, liver, lymphoma, leukemia, and melanoma. Among them, most cardiac metastatic LMS originate from the uterus. Nonvisceral LMS metastasizing to the heart is rare. Metastatic cardiac tumors have four spreading routes, including direct contiguous extension, hematogenous spread, retrograde lymphatic extension, and intracavitary diffusion through either the inferior vena cava or the pulmonary veins (6). The lymphatic spreading route is probably the most common route because the primary tumors with high prevalence, such as lung cancer and breast cancer frequently use this route (7). Bussani et al. (5) have suggested that the lymphatic system plays the most important role in metastatic heart disease and is associated with metastases to the myocardium or epi-

Fig. 1. A 72-year-old male with metastatic leiomyosarcoma from the left flank in the right ventricle.

- A.** Chest radiography shows mild, rightward shifting of the trachea (arrow), enlarged mediastinal structures (arrowheads), and cardiomegaly.
- B.** Echocardiography shows a huge hyperechoic mass involving the cavity of the right ventricle (arrowhead) and tricuspid valve (arrow).
- C.** Axial image from pre-contrast CT shows the irregularly shaped, hypodense mass with a lobulated margin (arrow) in the right ventricle.
- D.** Axial images from contrast-enhanced CT show peripheral enhancement of the tumor (asterisk), invasion of the interventricular septum with irregular thickening (arrowhead), and a small polypoid extrusion into the right ventricular outflow tract (arrow).



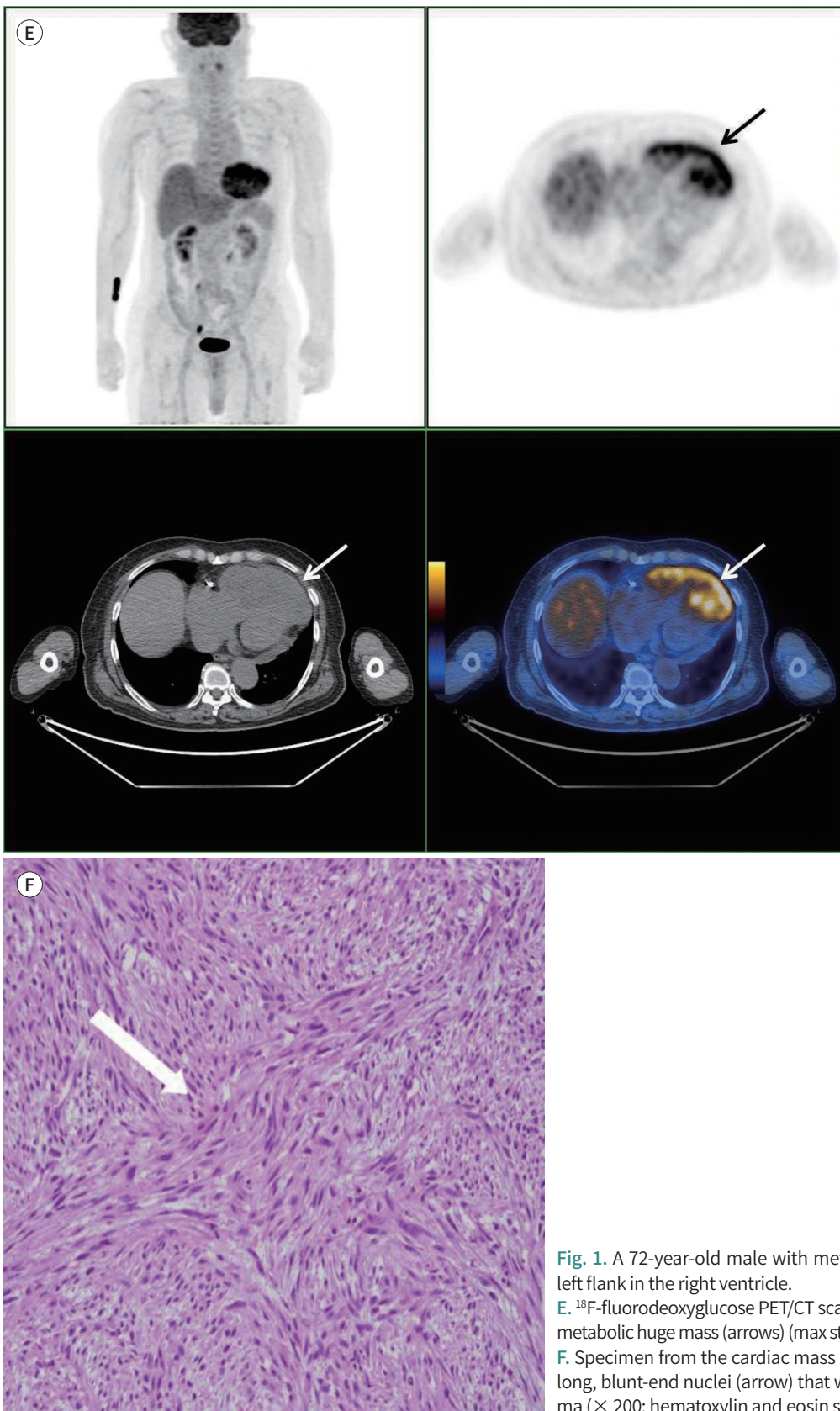


Fig. 1. A 72-year-old male with metastatic leiomyosarcoma from the left flank in the right ventricle.

E. ^{18}F -fluorodeoxyglucose PET/CT scans show an uneven irregular hypermetabolic huge mass (arrows) (max standardized uptake value; 8.4 g/mL).

F. Specimen from the cardiac mass shows elongated spindle cells with long, blunt-end nuclei (arrow) that was compatible with leiomyosarcoma ($\times 200$; hematoxylin and eosin stain).

cardium. On the other hand, sarcoma uses the hematogenous spread route by disseminated tumor cells in the blood stream. In this path, common sites of disease are the myocardium and endocardium.

Patients with metastatic cardiac tumors have new cardiac symptoms such as heart failure, valvular disease, conduction defect, arrhythmias, or syncope. The most common symptom is dyspnea, but it doesn't suggest cardiac metastasis obviously because the lung metastasis may be seen in many of the patients (8). When evaluating the cardiac metastasis, pericardium should be closely observed as pericardial involvement has been reported in up to 40% of the patients with cardiac metastasis and 15% of cytological examination of pericardial effusion are malignant (7). In previous reports, an intracardiac or pericardial mass was the most specific indicator of cardiac metastasis. The cardiac metastasis is common in the right chamber of the heart, but occurs on the left side when accompanied by lung metastasis.

Our case of cardiac metastatic nonvisceral LMS showed general features mentioned above. On the other hand, pericardial effusion was inconsistent with the reported general features. Obviously, CT is a helpful tool for evaluation of an intracardiac mass, such as the location, morphology, and the extent of cardiac masses. Takenaka et al. (8) claimed that echocardiography should be performed on the patients with suspicious manifestations of cardiac metastasis because the chest CT may have failed to find cardiac metastases even if tumors existed. Although radiologic multimodalities might be useful for the diagnosis, it cannot be confirmed only by radiologic study.

The prognosis of most cardiac metastases is extremely poor. Previous literatures describe perioperative mortality rate to be 40% and average survival time to be 5 months (7, 8). Radiation therapy is recommended and surgical resection is limited. There are a few differential diagnostic considerations to diagnose the cardiac metastasis. The important alternate diagnosis is intracardiac thrombus. Clues to this diagnosis are the location in the left side of the heart, including the left atrium or appendage, a lack of internal enhancement, and a predisposing clinical status including a previous infarction or dysrhythmia. Despite much less common incidence, primary tumors should be listed on the differential diagnosis. The most common intracardiac neoplasm is myxoma, which is a benign neoplasm, typically located in the left atrium, and has an ovoid or spherical shape. Primary malignancies such as sarcomas and lymphoma are rare (9). In previous reports, they have been frequently located in the RV and both atria, which is the same anatomical region with cardiac metastasis. For this reason, the distinction between primary and secondary cardiac tumors is usually done by clinical conditions. It is challenging even for experienced radiologists to clearly distinguish a secondary tumor from a primary tumor only using imaging findings. However, radiologists need to consider the possibility of the metastatic cardiac mass and look for diagnostic clues such as clinical grounds, mass location, and pericardial effusion.

Author Contributions

Conceptualization, all authors; data curation, P.S.; supervision, K.H.; writing—original draft, P.S.; and writing—review & editing, all authors.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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우심실로 전이된 내장 외 연부조직 평활근육종: 증례 보고 및 문헌 고찰

박상민* · 김희경

평활근육종은 평활근육세포에서 기원하는 연부조직 육종이다. 평활근육종은 자궁에 흔하게 발생하지만 후복막강, 복강 그리고 혈관조직에도 발생할 수 있다. 자궁 또는 혈관에서 기원하는 평활근육종이 심장에 전이된 증례는 많이 보고되었으나, 내장 외 연부조직에서 생긴 평활근육종은 매우 드물게 발생하며 심장으로의 전이는 더욱 드문 것으로 알려져 있다. 이에 좌측 측복부에서 우심실로 전이된 평활근육종의 심초음파 소견과 조영증강 컴퓨터단층촬영 소견을 경험하여 보고하고자 한다.

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