

Primary Angiosarcoma in the Right Atrium Diagnosed by a Cardiac Tumor Biopsy Using Intracardiac Echocardiography

Genki Naruse¹, Masanori Kawasaki^{2*}, Komei Yanase³, Toshiki Tanaka¹

¹Department of Cardiology, Gifu University Graduate School of Medicine, Gifu, Japan, ²Department of Cardiovascular Medicine, Gifu Heart Center, Gifu, Japan,

³Department of Respiriology, Gifu University Graduate School of Medicine, Gifu, Japan

Abstract

A 47-year-old woman consulted a doctor due to a persistent cough. Computed tomography revealed a 30 mm × 60 mm intracardiac mass in the right atrium. Because of lung metastasis, her respiratory status did not allow a more invasive procedure, such as general anesthesia. Although intracardiac echocardiography (ICE) during percutaneous transcatheter biopsy (PTB) is not covered by medical insurance, we performed PTB under ICE guidance. Pathology and immunohistochemistry revealed primary cardiac angiosarcoma. Primary cardiac angiosarcoma is a rare tumor with a poor prognosis. After seven cycles of chemotherapy, the pulmonary metastasis was clearly improved. The patient is alive 18 months after the first consult, even though the mortality of angiosarcoma is high. ICE during PTB allowed us to choose appropriate chemotherapy and improve her pulmonary metastasis. ICE during PTB reduces the need for a diagnostic open-chest procedure that requires a more invasive approach.

Keywords: Biopsy, intracardiac echocardiography, primary angiosarcoma, right atrium

INTRODUCTION

Malignant tumors of the heart have a poor prognosis. The mean survival time is 5 months after the resection.^[1] Although advanced imaging modalities such as computed tomography and cardiac magnetic resonance provide important information on tumors,^[2] the pathological examination is still necessary for the diagnosis and selection of treatment. A biopsy is generally guided by transesophageal echocardiography (TEE). However, TEE is semi-invasive and frequently induces vomiting and discomfort, and patients are required to lie quietly, particularly during biopsy procedures; TEE should also be avoided during pulmonary vein ablation procedures for the same reason. Intracardiac echocardiography (ICE) is a promising imaging modality for detailed intracardiac atrial imaging and safer biopsy procedures.^[3,4]

CASE REPORT

A 47-year-old woman consulted a doctor due to a persistent cough. Multiple pulmonary nodules were identified in her chest X-ray, and she was transferred to our hospital,

where we diagnosed multiple pulmonary nodules and pericardial effusion. Blood examination showed no specific abnormalities that would indicate infection or collagen diseases. Transthoracic echocardiography confirmed a mass in the right atrium [Figure 1a], and contrast-enhanced computed tomography also revealed a 30 mm × 60 mm intracardiac mass in the right atrium [Figure 1b-d]. Fluorodeoxyglucose positron emission tomography showed abnormal uptake in the right atrial mass and multiple pulmonary nodules. These findings suggested a primary malignant tumor in the right atrium and multiple pulmonary metastases. The pathology of a transbronchial lung biopsy and cytology of pericardial fluid were negative. Therefore, we performed a percutaneous transcatheter biopsy (PTB) of the right atrial tumor under ICE guidance after obtaining consent from the ethical committee of our institution, because ICE during PTB is not covered by medical insurance

Address for correspondence: Dr. Masanori Kawasaki,
Department of Cardiovascular Medicine, Gifu Heart Center, 4-14-4
Yabuta-Minami, Gifu 500-8384, Japan.
E-mail: masanori@ya2.so-net.ne.jp

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in Japan. The patient gave written informed consent before the procedure.

A 5.5 French (Fr) cardiac biptome was advanced through a 6 Fr long sheath in the right jugular vein, and a 9-MHz Ultra ICE catheter (Boston Scientific Corporation, Natick, MA, USA) was advanced through a 9 Fr long sheath in the femoral vein [Figure 2a]. The right atrial tumor was clearly visualized [Figure 2b], and six biopsy specimens were taken. This image was same as the image that was previously obtained by TEE [Figure 2c]. There were no complications related to the procedure. Pathology and immunohistochemistry revealed primary cardiac angiosarcoma. After seven cycles of chemotherapy (paclitaxel 100 mg/m²), the pulmonary metastasis was clearly improved [before: Figure 3a and after: Figure 3b]. The patient was alive 18 months after the first consult, even though the mortality of angiosarcoma is high.

DISCUSSION

Primary cardiac tumors are rare, with an incidence ranging from 0.001% to 0.28% in an autopsy series.^[5] Approximately 75% of primary cardiac tumors are benign, with myxomas accounting for 50% of them, and the rest are malignant. For the diagnosis of primary cardiac tumors, cardiac biopsy is necessary. However, PTB requires high skill, particularly in the atrium, because the atrial wall is thin. In addition to a report of cardiac tumors that were diagnosed by a PTB in the right ventricle, primary cardiac lymphoma diagnosed by a PTB in the right atrium has been reported.^[6,7] We described a case of primary cardiac angiosarcoma that was diagnosed by PTB in the right atrium.

Angiosarcoma is one of the various forms of malignant tumors, accounting for approximately 8% of primary cardiac

tumors.^[8] The prognosis of primary cardiac angiosarcoma is very poor, with a mean survival of 3.8 ± 2.5 months without surgical resection.^[9] Survival times range from 12 to 30 months, even in patients with various combinations of surgery, chemotherapy, radiation, and/or transplantation.^[10] Prognostic factors such as tumor grade, age, gender, growth pattern, and margin status have not been clearly elucidated. For this reason, an immediate diagnosis is needed when an angiosarcoma is suspected. In the present case, because of lung metastasis, her respiratory status did not allow general anesthesia. We chose ICE-guided PTB that made it possible to obtain tumor tissue safely during cardiac catheterization. This allowed us to choose appropriate chemotherapy and improve her pulmonary metastasis.

CONCLUSION

Primary cardiac angiosarcoma is a rare tumor with an extremely fatal prognosis due to its invasive behavior. Therefore, a prompt and safe procedure for diagnosis is imperative for a better prognosis. We performed ICE during PTB at our own expense, because the ICE procedure during PTB is not covered by medical insurance in Japan. Based on the usefulness of ICE in the present case of primary cardiac angiosarcoma, ICE during PTB should be covered by Japanese medical insurance. ICE during PTB reduces the need for a diagnostic open-chest procedure that requires a more invasive approach.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and

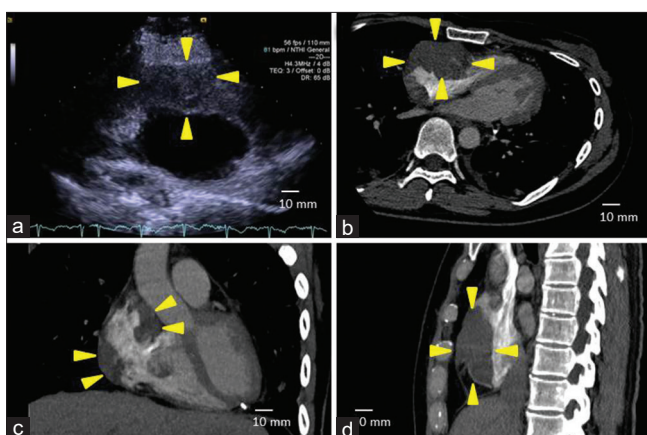


Figure 1: Images of intracardiac mass at the time of initial consultation. (a) Transthoracic echocardiography shows a hypodense mass attached to the anterior atrial free wall (arrowhead), (b) contrast-enhanced computed tomography (axial view). Arrowhead: intracardiac mass in the right atrium, (c) contrast-enhanced computed tomography (coronal view). Arrowhead: intracardiac mass in the right atrium, (d) contrast-enhanced computed tomography (sagittal view). Arrowhead: intracardiac mass in the right atrium

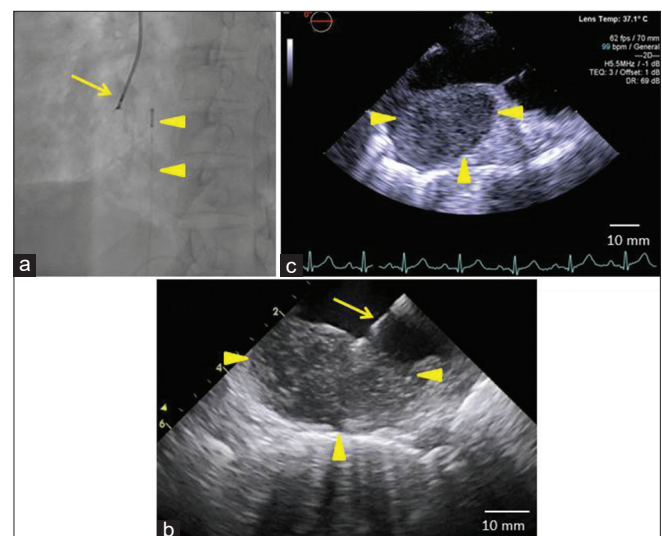


Figure 2: Images of intracardiac mass. (a) Catheter for intracardiac echocardiography (arrowhead) and cardiac biptome (arrow), (c) transesophageal echocardiography shows a hypodense mass attached to the anterior atrial free wall (arrowhead), (b) biptome (arrow) contacting the tumor (arrowhead) visualized on intracardiac echocardiography

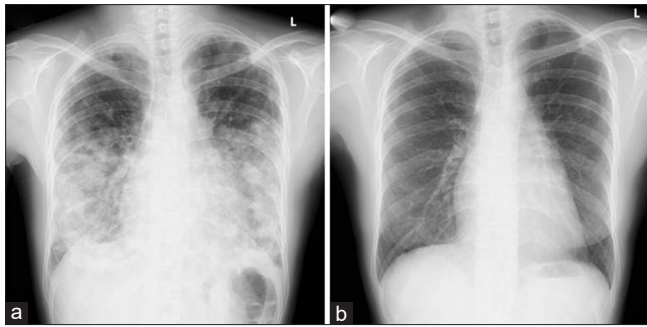


Figure 3: Chest X-ray before and after chemotherapy. (a) Before chemotherapy, (b) after chemotherapy

initials will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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