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Primary Cardiac Angiosarcoma in a Patient with Syncope: A Case Report

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A 70-year-old woman presented at our institution's emergency room with syncope. She had a history of chest discomfort, dyspnea on exertion, and cough in the prior 3 months. Laboratory findings were as follows: white blood cell count, 7390/μL; hemoglobin, 10.7 g/dL; C-reactive protein, 1.7 (<0.5) mg/dL; aspartate aminotransferase/alanine aminotransferase, 649/455 U/L; and N-terminal pro–B-type natriuretic peptide (NT-proBNP), 1045.2 ρg/mL.

Chest radiography showed cardiomegaly, and chest computed tomography (CT) revealed a 6.8 cm-sized inhomogeneous mass enhancement in the right atrium, narrowing of the SVC, and hemopericardium effusion (Fig. 1A). And transthoracic echocardiography (TTE) showed an heterogenous echogenic mass measuring 5.22×3.02 cm in the right atrial (RA) cavity (Fig. 1B). To identify the primary origin of the RA mass, positron emission tomography (PET) was performed, and it revealed a hypermetabolic mass with central necrosis inside the right atrium. Besides this, no other abnormal fluorodeoxyglucose (FDG) uptake findings suggested lymph nodes or distant metastasis. Owing to the peculiarity of this condition, this case was discussed with our institutional multidisciplinary team to determine an appropriate biopsy method for histological confirmation, and a consensus was reached to perform an intracardiac biopsy via the femoral vein. Thus, an intracardiac echocardiography (ICE)-guided RA mass biopsy was performed. ICE showed a mass-like lesion with multi-lobular tissue density (Fig. 1D). During the biopsy, the patient experienced loss of consciousness, and her blood pressure dropped to 70/40 mmHg, confirming the diagnosis of car-

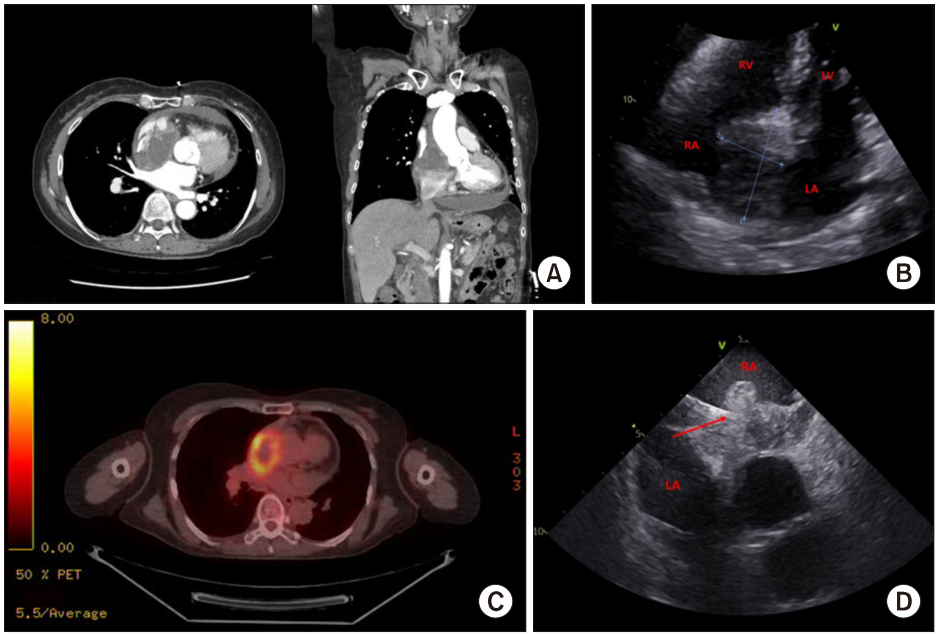

FIG. 1. (A) Chest computed tomography shows a 6.8-cm-sized mass in the right atrium. Narrowing of the superior vena cava (SVC). (B) Transthoracic echocardiography (TTE) shows a mass inside the right atrium. (C) Positron emission tomography is performed to detect the primary origin of the right atrial (RA) mass. (D) Intracardiac echocardiography (ICE) shows a right atrial (RA) mass. The biopsy forceps were positioned within the mass located in the right atrium (arrow).

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Article History:
Received August 10, 2023
Revised August 26, 2023
Accepted August 28, 2023

https://doi.org/10.4068/cmj.2023.59.3.211
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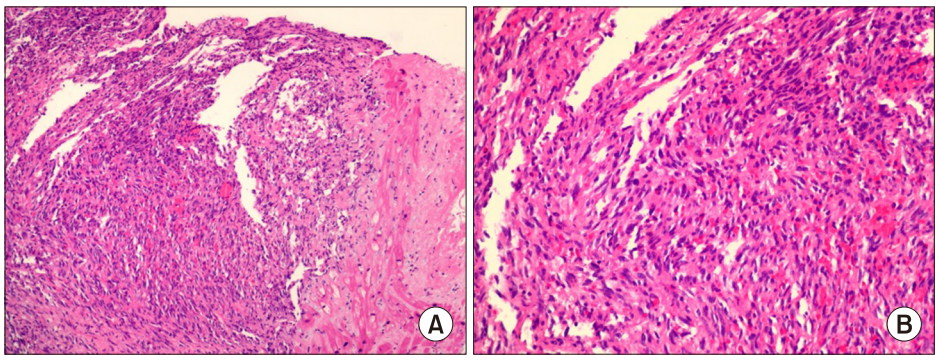

Fig. 2. Pathological findings using hematoxylin and eosin (H&E) stains. (A) Spindle cell proliferation at low magnification (×100). (B) The spindle cells show moderate pleomorphism and hyperchromatic nuclei at high magnification (×200).

diac tamponade. Consequently, emergency pericardiocentesis was performed, resulting in an improvement in the patient's consciousness.

Pathological findings revealed spindle cell proliferation with a small slit-like space in the hemorrhagic background when viewed at low magnification using hematoxylin and eosin staining. At higher magnification, the spindle cells showed moderate pleomorphism and hyperchromatic nuclei (Fig. 2). Immunohistochemistry revealed positive findings for CD31 and CD34 but negative findings for desmin, cytokeratin, S100, and Melan A. Therefore, based on these findings, we diagnosed the patient with angiosarcoma in the right atrium. However, given the invasive nature of the mass into the RA wall, complete surgical resection was not feasible. Owing to this, we planned on treating the patient using a combination of radiotherapy and chemotherapy under the supervision of the hemato-oncology team.

Primary cardiac angiosarcoma is a rare and highly aggressive tumor of endothelial origin. This is due to the rapid progression of the disease and its propensity for metastasis. Symptoms of primary cardiac angiosarcoma may vary depending on the affected area and may include chest discomfort, dyspnea, and cough. In some cases, syncope may occur, as in the current patient, because of the narrowing of the SVC. While studies have demonstrated that cardiac angiosarcoma can invade surrounding vascular structures,[1,2] many cases do not exhibit symptoms, even when such invasions occur. Therefore, histological confirmation through biopsy is important in conjunction with various imaging techniques. Although pathological confirmation based on biopsy findings is important, performing a biopsy in the atrium carries a substantial risk of acute complications. Therefore, it is very important to carefully select an appropriate biopsy method for patients with intracardiac mass.

CONFLICT OF INTEREST STATEMENT

None declared.

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