

[CASE REPORT]

A Mass Filling the Right Atrium: Primary Cardiac Rhabdomyosarcoma

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Abstract:

A 43-year-old woman presented with worsening shortness of breath and lower-extremity edema. Echocardiography and computed tomography showed obstruction of blood flow due to a mass filling the right atrium. Emergency surgery was performed for circulatory failure. Primary cardiac rhabdomyosarcoma was diagnosed based on a histological examination. The patient died about two months after the diagnosis despite surgical excision and radiation therapy. The poor prognosis may have resulted from the grossly incomplete removal of the tumor and chemotherapy intolerance. We herein report a case of primary cardiac rhabdomyosarcoma filling the right atrium and offer possible reasons for the poor prognosis.

Key words: rhabdomyosarcoma, primary cardiac tumor, combined modality approach, prognosis

(Intern Med 57: 3575-3580, 2018)

(DOI: 10.2169/internalmedicine.0657-17)

Introduction

Rhabdomyosarcoma is a malignant solid tumor deriving from mesenchymal cells that differentiate into striated muscle. Primary cardiac tumors are rare, with an incidence of 0.001% to 0.28% in an autopsy series, and approximately 75% are benign while 25% are malignant (1). The Armed Forces Institute of Pathology reports that rhabdomyosarcoma is the second-most common type of malignant cardiac tumors, comprising 21% of cases, after angiosarcoma, which accounts for 31% of cases (1). Due to its rarity, only a few case reports on primary cardiac rhabdomyosarcoma are available.

We herein report a case of primary cardiac rhabdomyosarcoma originating in the right atrium that caused circulatory failure.

Case Report

A 43-year-old woman presented to a clinic in a bedridden

state with a 1-month history of dyspnea and lower-extremity edema and was referred to our hospital. A physical examination showed a respiratory rate of 16 breaths per minute, a heart rate of 100 beats per minute (regular), a blood pressure at 104/76 mmHg, a body temperature of 37.0°C, and a percutaneous oxygen saturation of 97% on ambient air. Jugular venous distension and pretibial edema were observed. The rest of the findings on examinations, including auscultation and palpation, were unremarkable. Echocardiography revealed a movable isoechoic mass filling the cavity of the right atrium (RA) to the intravascular lumen of the inferior vena cava (IVC) (Fig. 1). Contrast-enhanced computed tomography (CT) identified a mass extending from the RA to the common iliac veins and obstructing the blood flow (Fig. 2). The laboratory findings showed N-terminal pro-brain natriuretic peptide 3,142 pg/mL, aspartate aminotransferase 534 U/L, alanine aminotransferase 765 U/L, and D dimer 17 µg/mL.

Emergency surgery was performed in order to prevent sudden death from circulatory failure. A tumor occupied the entire RA cavity with its stalk firmly attached to the inferior

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Received: December 13, 2017; Accepted: May 29, 2018; Advance Publication by J-STAGE: August 10, 2018

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wall of the RA near the IVC (Fig. 3). The elastic, hard tumor was excised from the intima of the RA wall, which appeared coarse and infiltrated. The excised tumor had a smooth gray surface and was 70×50×30 mm in size (Fig. 3). Multiple thrombi filling the IVC to the common iliac veins were then removed. Microscopically, the tumor consisted of atypical spindle-shaped cells with focal hypercellularity around vessels in hematoxylin and eosin staining (Fig. 4). Immunostaining showed that the specimen was negative for cytokeratin AE1/AE3 (a protein contained in epithelial cells) and positive for vimentin (a protein contained in non-epithelial cells), desmin (a protein contained in muscle cells), and myogenin (a protein contained in striated muscle cells) (Fig. 4). Based on these histological findings, primary cardiac rhabdomyosarcoma was diagnosed.

The surgery restored the blood flow and led to the improvement of congestive heart failure. On postoperative day (POD) 1, CT showed that the tumor had disappeared from the RA but that the thrombi remained in the IVC at the renal vein level, left iliac vein, and hepatic veins (Fig. 5). To prevent the formation of pulmonary embolism, an IVC filter was placed, and anticoagulation therapy was initiated. Despite the therapy, follow-up CT on POD 8 showed the intact

thrombi and a new lung-metastatic tumor (Fig. 5). Follow-up CT on POD 22 showed that the previously excised tumor had reemerged from the RA to the IVC and that the lung-metastatic tumor and thrombi had increased in size (Fig. 5). Fluorodeoxyglucose-positron emission tomography visualized the recurrent cardiac tumor from the RA to the IVC and the metastatic tumor in the right lung on POD 25 (Fig. 6). Radiation therapy 45 Gy was given from POD 39 to POD 55 to reduce the tumor but was ineffective. Chemotherapy was considered for the perioperative period but was not administered due to the patient's poor performance status at admission (Eastern Cooperative Oncology Group

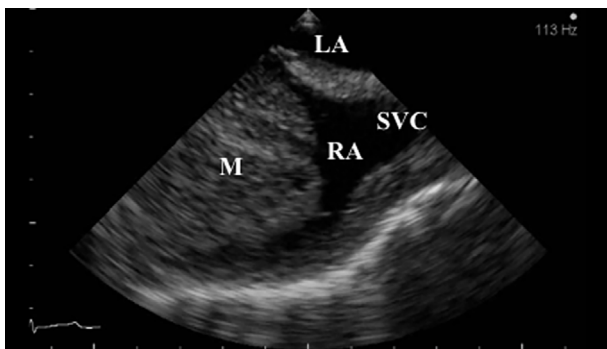


Figure 1. Transesophageal echocardiography (mid-esophageal view 90°) revealed a movable isoechoic mass filling the right atrium. LA: left atrium, M: mass, RA: right atrium, SVC: superior vena cava

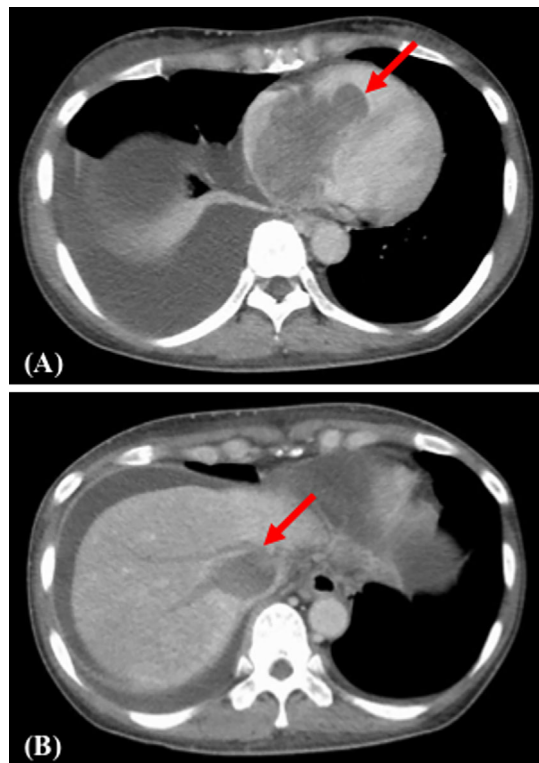


Figure 2. Contrast-enhanced computed tomography on admission showed a mass extending from the right atrium (arrow) (A) to the inferior vena cava (arrow) (B).

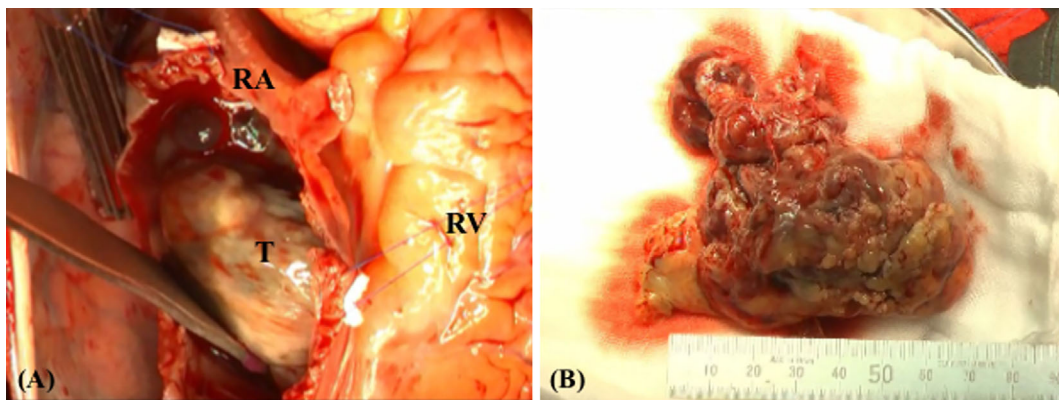


Figure 3. The tumor was attached to the inferior wall of the right atrium near the inferior vena cava (A). The excised tumor had a smooth gray surface and a size of 70×50×30 mm (B). RA: right atrium, RV: right ventricle, T: tumor

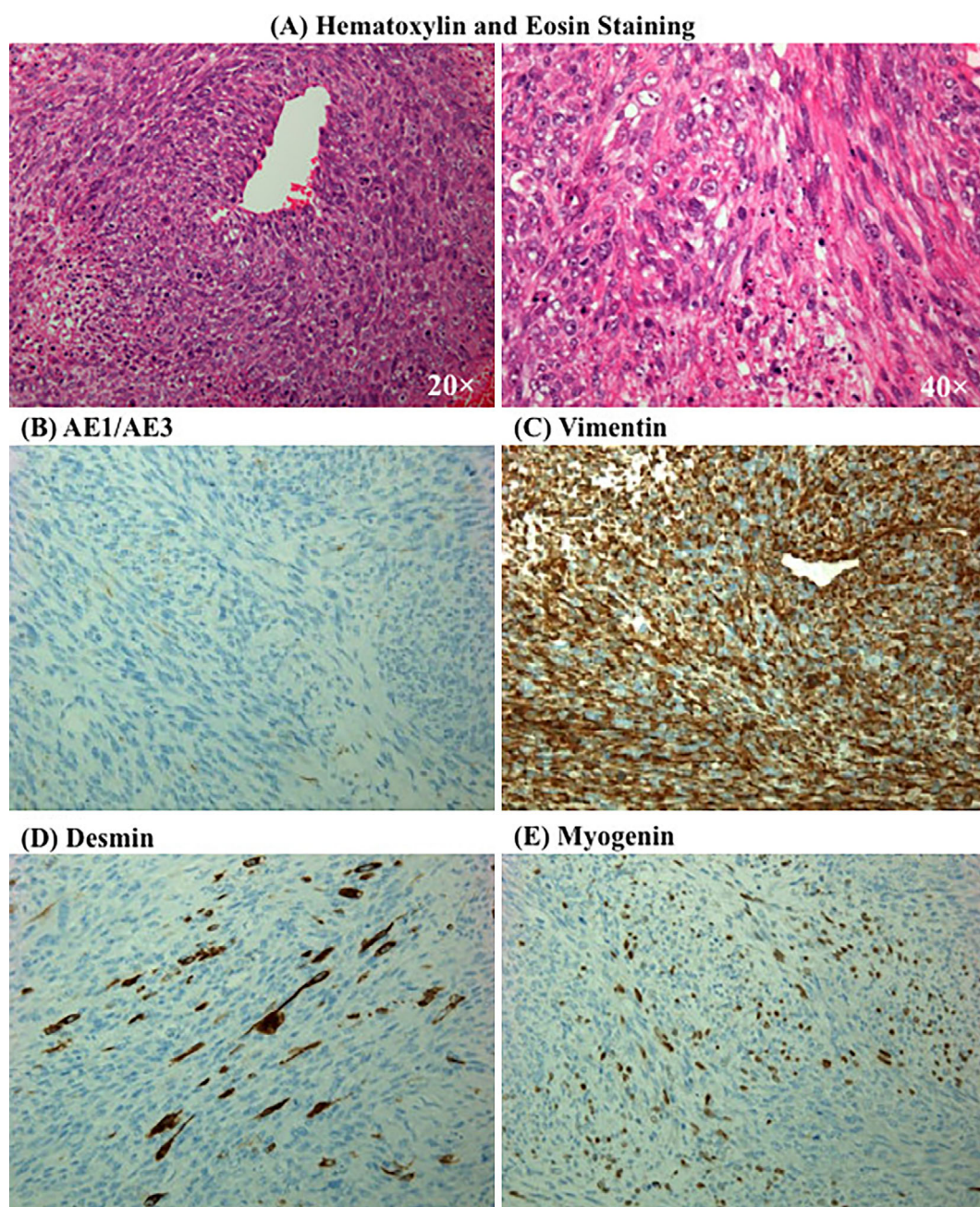


Figure 4. Atypical spindle-shaped cells were dense around vessels on Hematoxylin and Eosin staining (A). The tumor was negative for cytokeratin AE1/AE3 (B) and positive for vimentin, desmin, and myogenin, as is characteristic of rhabdomyosarcoma (C-E).

Performance Status 3). Gradually, the patient suffered multiple organ failure, lost consciousness, and died on POD 68.

Discussion

Symptoms in patients with cardiac tumors, including primary cardiac rhabdomyosarcoma, are caused by the obstruction of blood flow, the severity of which in turn depends on the sites and sizes of the tumors (2, 3). Patients also often experience embolic complications caused by detached tumor tissues and tumor-induced thrombi, such as cerebral, pulmonary, and peripheral embolisms (3). Furthermore, tumor infiltration into the myocardia causes restrictive cardiomyopa-

thy, cardiac tamponade, and arrhythmia, especially atrioventricular block (3). Our patient had right cardiac failure caused by the obstruction of the blood flow due to a tumor filling the RA but none of the other complications at the time of admission.

As in our case, cardiac tumors are diagnosed by a combination of echocardiography and CT or magnetic resonance imaging (MRI), which generally provides enough information for preoperative planning (4). Initial transthoracic echocardiography can often identify a cardiac mass. Transesophageal echocardiography can give a more vivid picture of the mass and allow the tumor to be distinguished from a thrombus, its size to be determined, and its location and po-

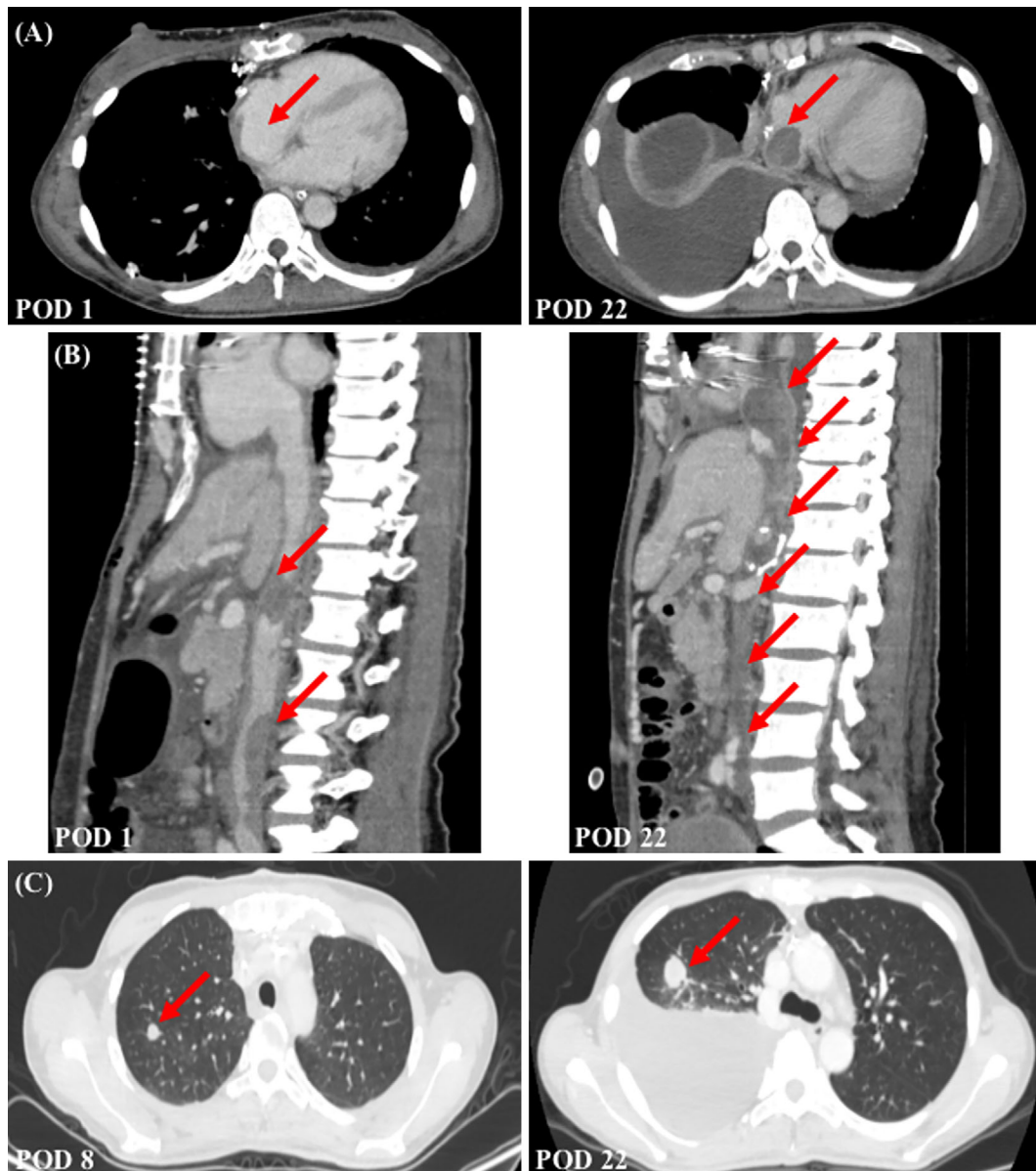


Figure 5. Contrast-enhanced computed tomography showed the reemergence of the tumor in the right atrium to the inferior vena cava (arrow) (A), the enlargement of thrombi in the inferior vena cava (arrow) (B), and the emergence and growth of a lung-metastatic tumor (arrow) (C).

sition in relation to the valves to be assessed (5). Some studies have reported that primary cardiac rhabdomyosarcoma has no predominant localization within any area of heart (1-3), but the World Health Organization has recently reported that the most common sites of primary cardiac rhabdomyosarcoma are the ventricles (6). Based on this update, the involvement of the RA as the originating site of primary cardiac rhabdomyosarcoma in our case appears to be rare. Both CT and MRI can provide precise images of the tumor's anatomical structure and the extent of its invasion and metastasis (7). Ultimately, surgical specimens are necessary for a correct diagnosis and staging, based on which appropriate therapy can be initiated.

The standard therapy for rhabdomyosarcoma is a combination of surgery, chemotherapy, and radiation therapy, an approach that has been developed over the past several dec-

ades through a series of large-scale trials carried out by the Intergroup Rhabdomyosarcoma Study Group now known as the Soft Tissue Sarcoma Committee of the Children's Oncology Group (7-11). Although complete surgical resection is still an important prognostic factor, the most important component of the combined modality approach is systemic chemotherapy, as metastatic lesions are usually diffuse by the time symptoms appear. Vincristine, dactinomycin, and cyclophosphamide (VAC) are the standard regimen for rhabdomyosarcoma. The specific treatment regimens have been discussed elsewhere (12). In addition, radiation therapy is administered to achieve local control in patients with residual microscopic or gross disease following surgery and chemotherapy (12). In our case, primary cardiac rhabdomyosarcoma was incompletely removed with gross residual disease, for which radiation therapy was later administered. However,

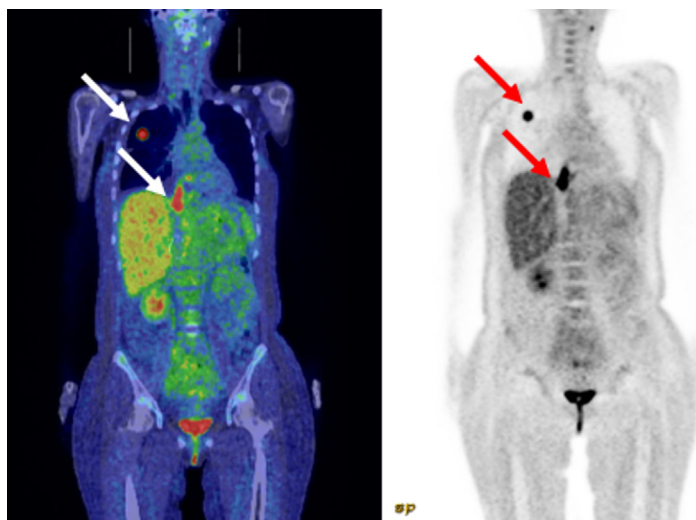


Figure 6. Fluorodeoxyglucose-positron emission tomography revealed the recurrent cardiac tumor extending from the right atrium to the inferior vena cava (arrow) and the metastatic tumor in the right lung (arrow).

chemotherapy was not given due to the patient's poor performance status even though it is the mainstay for the patient suspected of having microscopic metastasis on admission. Our patient died about two months after the diagnosis despite surgical excision and radiation therapy.

Although the combined modality approach has reportedly improved the prognosis of childhood rhabdomyosarcomas at common sites, including the orbits, head and neck, and genitourinary and biliary tracts, patients with primary cardiac rhabdomyosarcoma are expected to survive less than a year in most cases (1, 2, 5, 13). This relatively poor prognosis may be attributed to the significant risk of lethal cardiac functional loss resulting from complete resection of primary cardiac rhabdomyosarcoma with pathologically clear margins; only partial resection is therefore possible, although rhabdomyosarcomas at common sites can be completely excised without lethal complications. Alternatively, the status of some patients with primary cardiac rhabdomyosarcoma may be too critical to support chemotherapy at the time of the diagnosis due to its rapid progression, metastasis, and complications, including blood flow obstruction, cerebral and pulmonary embolisms, restrictive cardiomyopathy, cardiac tamponade, and arrhythmia. Radiation therapy may also be difficult, as the exposure of the ventricular wall to a radical radiation dose causes severe cardiomyopathy and pericarditis. Some reports have shown that heart transplantation is the last resort in cases for which these treatments are ineffective and metastasis can be ruled out (3, 4). However, this option seems unrealistic, as metastatic lesions usually become diffuse by the time cardiac symptoms appear, as mentioned above. In fact, the surgical option does not confer any significant benefit on the prognosis. Uberfuhr et al. reported a mean length of survival of 18 months for 4 patients with cardiac tumors who had been treated by heart transplantation with postoperative chemotherapy (14). Several other studies have suggested that adults have worse out-

comes than children, possibly due in part to the inadequacy of the primary treatment. These studies insist that adult patients be treated in the same manner as children in general, although rhabdomyosarcomas are more common in children than in adults, and the treatment is therefore mainly based on pediatric data (15, 16). This emphasis on the combined modality approach to primary cardiac rhabdomyosarcoma is worth considering, as it may result in a better prognosis, although its therapeutic effect might be limited.

Conclusion

Our patient died approximately two months after the diagnosis despite surgical resection and radiation therapy. This poor prognosis may be not only because the primary cardiac rhabdomyosarcoma quickly infiltrated all layers of the heart and metastasized to the extracardiac organs but also because a combined modality approach of surgery, chemotherapy, and radiation therapy could not be deployed.

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

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