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CASE REPORT

Primary sarcoma of the right ventricle: surgical and adjuvant therapy

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Abstract We describe surgical and adjuvant therapeutic management of a right ventricular (RV) sarcoma and pulmonary artery occlusion. Echocardiographic evaluation of a 39-year-old man with exertional dyspnea revealed a tumor mass in the right ventricle, pulmonary trunk, and bilateral pulmonary arteries. The tumor was resected with concomitant pulmonary valvotomy, but the resection was incomplete. The RV outflow was reconstructed with an allograft patch, and a stentless valve was implanted for pulmonary valvular function. The pulmonary trunk and arteries were enlarged with allograft patches. The tumor was undifferentiated sarcoma and caused postoperative pulmonary artery restenosis. Radiotherapy improved pulmonary perfusion (reduction of RV pressure), but the patient died of brain metastasis. Undifferentiated cardiac sarcomas associated with pulmonary hypoperfusion should be resected even if incompletely, and radiation therapy could alleviate reduced pulmonary perfusion.

Key words Cardiac tumor · Right ventricular sarcoma · Stentless valve pulmonary valve replacement · Radiation therapy

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Introduction

Primary right ventricular (RV) sarcoma involving the pulmonary arteries is rare. The symptoms of this condition are related to low cardiac output due to reduced pulmonary blood flow resulting from obstruction of the RV outflow tract and pulmonary trunk. We describe a patient with primary RV sarcoma that almost totally obstructed the pulmonary arteries.

Case

A 39-year-old man who presented at a nearby hospital with exertional dyspnea, easy fatigability, and syncope was referred to our department with a diagnosis of an RV mass occupying the bilateral pulmonary arteries. Upon admission, the patient was asymptomatic at rest, and his medical history revealed no risk factors for thromboembolism. Chest radiography showed an enlarged cardiac shadow with clear lung fields. Electrocardiography showed a regular sinus rhythm at 90 beats/min. Arterial blood gas analysis revealed slight hypoxia (P_{O_2} 87.4 mmHg) with hypocapnia (P_{CO_2} 29.6 mmHg) under an oxygen mask (oxygen flow 8 l/min).

Transthoracic two-dimensional echocardiography and computed tomographic (CT) scanning (Fig. 1) revealed pericardial effusion (estimated volume > 500 ml) and a lobular mass in a markedly enlarged RV extending to the pulmonary trunk and bilateral branches and involving the pulmonary valve. The pressure gradient between the right ventricle and the right atrium (ΔPG), calculated from the tricuspid regurgitant jet velocity measured with Doppler echocardiography, was 91.4 mmHg (the estimated RV pressure >100 mmHg).

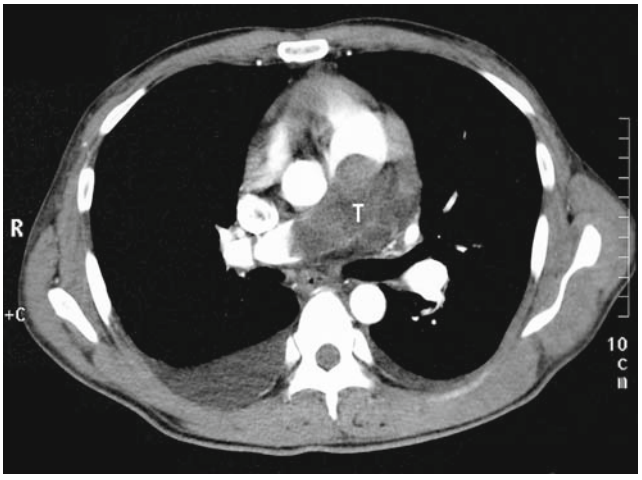

Fig. 1 Preoperative computed tomography scan. The tumor mass (*T*) is located in bilateral pulmonary arteries

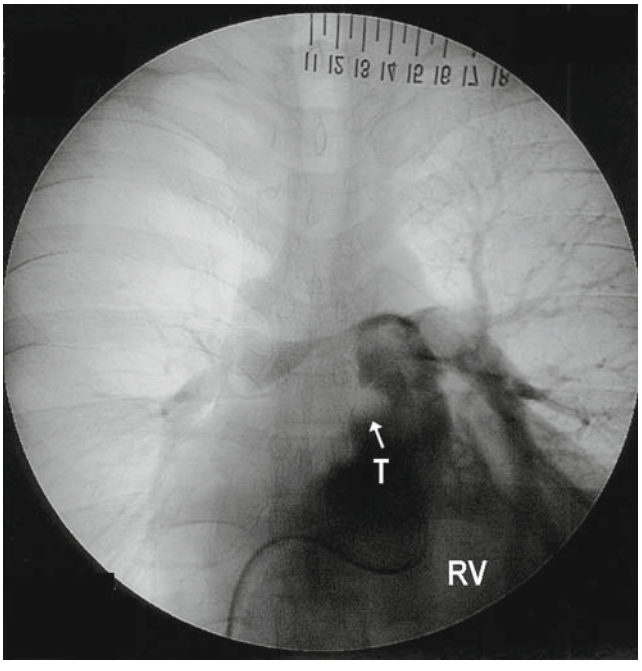

Fig. 2 Preoperative right ventricular angiography. The lobular mass (*T*) in the right ventricular outflow tract extends to the pulmonary trunk and bilateral pulmonary arteries. The right upper branch of the pulmonary artery is not opacified. *RV*, right ventricle

Cardiac catheterization demonstrated systolic RV pressure of 116 mmHg, mean right atrial pressure of 18 mmHg, and systolic aortic pressure of 120 mmHg. RV angiography revealed a lobular mass in the RV outflow tract extending to the pulmonary trunk and bilateral pulmonary branches, but the right upper branch of the pulmonary artery was not opacified (Fig. 2).

A cytological examination of drained pericardial effusion did not reveal any tumor cells. However, we performed emergency tumor resection because pulmonary hypoperfusion due to the tumor was life threatening. We exposed the heart through a median sternotomy. An elastic hard mass was palpable behind the pulmonary trunk and in the left side of the RV outflow tract. After heparinization, cardiopulmonary bypass (CPB) was established by cannulating the ascending aorta and then cannulating the superior vena cava directly, and the inferior vena cava through the right atrial wall for venous return. The ascending aorta was cross-clamped, and then the heart was arrested by antegrade and retrograde infusion of oxygenated tepid blood cardioplegia. A right atriotomy was performed to examine whether the tumor mass extended toward the RV inflow, but a tumor mass was not found in this location. Subsequently, we incised the pulmonary trunk longitudinally and extended the incision proximally toward the RV outflow tract and distally toward the left pulmonary artery. To expose the right pulmonary artery, the ascending aorta and superior vena cava were transected, and the right pulmonary artery was then incised distally from the incision on the pulmonary trunk down to just proximal to the upper lobar branch.

A whitish, lobular, elastic hard tumor was firmly attached to the RV outflow tract and pulmonary trunk, and the pulmonary valve annulus appeared to be involved. The mass firmly adhered to the intima of the left pulmonary artery and had partially invaded the wall of the left pulmonary artery, extending to both the upper and middle/lower lobar branches (Fig. 3A,B). The tumor in the pulmonary arteries was removed except for the part that had invaded the arterial wall of the left pulmonary artery. The infundibular septum and the posterior wall of the pulmonary trunk also appeared to be invaded by the tumor. In the RV outflow tract and pulmonary trunk, the tumor was resected piece by piece together with the pulmonary valve, but it could not be completely removed. The incisions of the pulmonary trunk and arteries were closed with an allograft patch to enlarge the lumen diameter and a stentless valve (Medtronic Freestyle stentless xenograft, 27 mm) was then implanted to minimize pulmonary regurgitation (Fig. 3C). The incision on the RV outflow tract was closed with an allograft patch (Fig. 3D).

The patient was weaned from CPB, and his postoperative recovery was satisfactory. Two-dimensional and Doppler echocardiography on postoperative day (POD) 6 revealed a smaller tumor with a concomitant decrease in the ΔPG to 30.0 mmHg (estimated RV pressure 40 mmHg). Postoperative symptoms such as exertional dyspnea and easy fatigability obviously diminished.

A pathology examination (light microscopy) showed that a large part of the tumor was necrotic, and its outer

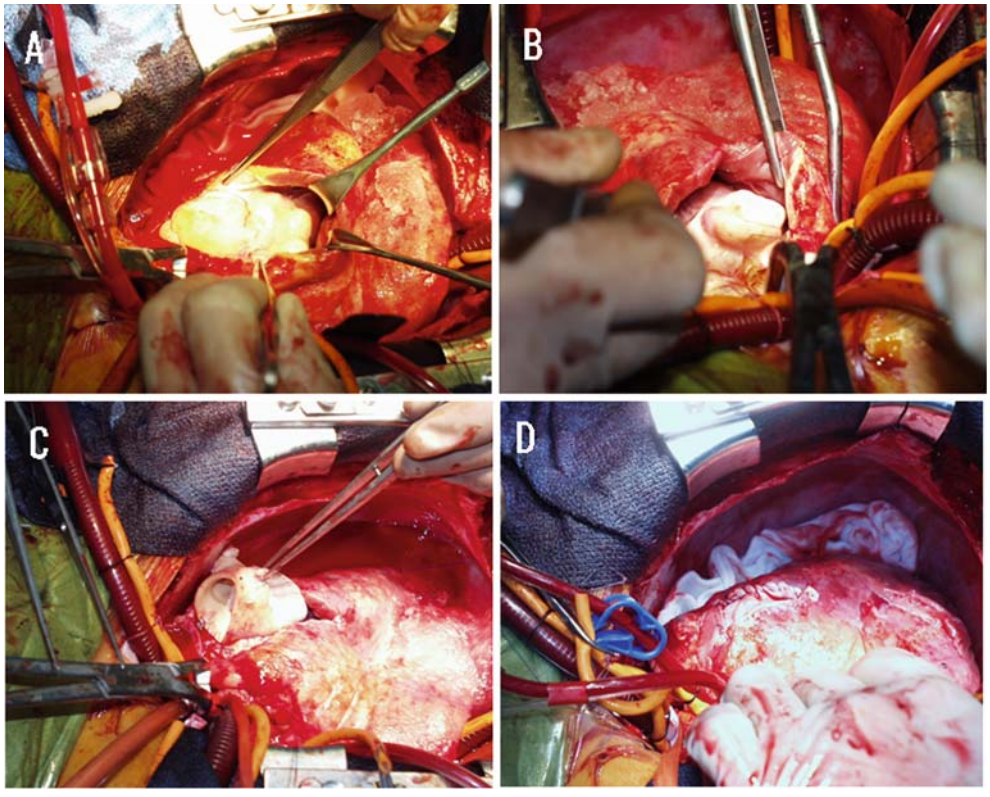

Fig. 3 Intraoperative view. A whitish, lobular, elastic hard mass is firmly attached to the right ventricular outflow tract (**A**) and pulmonary trunk (**B**) and appeared to involve the pulmonary valve annulus. A stentless valve (Medtronic Freestyle stentless xenograft, 27 mm) was implanted (**C**). The incision of the right ventricular outflow tract was closed with an allograft patch (**D**)

layer contained tumor cells with a high nuclear/cytoplasmic ratio in (Fig. 4). Immunocytochemistry revealed that the tumor was positive for vimentin and neuron-specific enolase but negative for desmin, smooth muscle actin, muscle-actin-specific monoclonal antibody, and myoglobin, indicating that the tumor was an undifferentiated sarcoma.

The tumor gradually proliferated again and caused restenosis of the pulmonary trunk and symptoms that progressively worsened over a few weeks. On POD 33, we performed adjuvant chemotherapy including cisplatin (CDDP, *cis*-diaminedichloroplatinum [II]) and adriamycin (80 and 20 mg/m^2 body surface area, respectively) based on a regimen of chemotherapy for soft tissue sarcomas (undifferentiated sarcoma).[1–3] After chemotherapy, however, two-dimensional echocardiography revealed no evidence of reduction of the tumor size. The ΔPG did not change from before chemotherapy and after it (50.7 and 52.1 mmHg, respectively), but it increased to 78.4 mmHg on POD 58. External irradiation of the right ventricle and pulmonary arteries started on POD 49, and the patient received a total of 54 Gy (30 doses of 1.8 Gy) for 6 weeks. After radiation therapy, the ΔPG decreased to 45.8 mmHg, although the size of the tumor did not change. The general state of the patient was temporarily improved presumably because the pulmonary restenosis was relieved. However, he died of brain metastasis 6 months later.

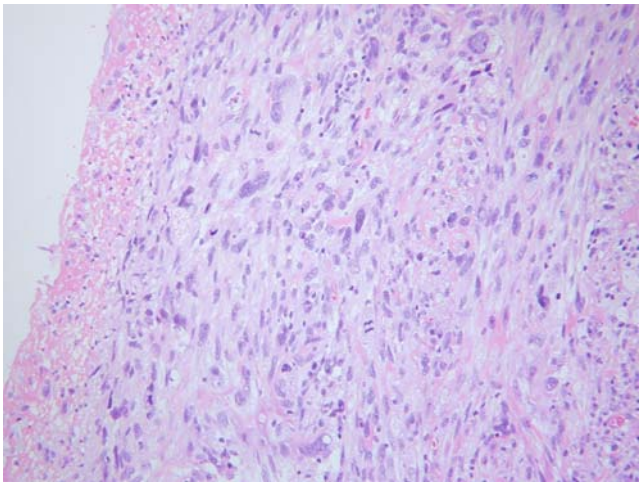

Fig. 4 Photomicrograph of the tumor, which is largely necrotic. Tumor cells in the outer layer have a high nuclear/cytoplasmic ratio. (H&E)

Discussion

The reported incidence of primary cardiac tumors ranges from 0.0017% to 0.03%.[4] The computer search of primary cardiac tumors (75 patients) by Burke et al.[5] found that cardiac sarcomas account for <25% of primary cardiac tumors, and undifferentiated sarcomas account

for 24% of primary cardiac sarcomas (18/75 patients). They also noted that undifferentiated RV sarcomas were identified in only 2 of 18 undifferentiated cardiac sarcomas.

A tumor in the RV outflow tract can remain asymptomatic unless pulmonary flow critically decreases. Diagnosis is difficult because the symptoms are nonspecific, mostly comprising progressive exertional dyspnea, easy fatigability, cough, and syncope of variable duration.[6,7] Physical examination and chest radiography are considered ineffective for early diagnosis of cardiac tumors.[6] On auscultation, a systolic murmur would be audible at the left sternal border if stenosis exists between the RV outflow and pulmonary artery tract. If RV dilatation or pericardial effusion is present, chest radiography might reveal cardiac enlargement. Our patient had exertional dyspnea, systolic heart murmur on auscultation, and cardiomegaly on chest radiography. Two-dimensional echocardiography was useful for this patient as it revealed a shadow obstructing the pulmonary trunk and a dilated RV cavity. Doppler echocardiography and cardiac catheterization also helped to reveal almost systemic RV pressure. Angiography might provide useful information on the patency of the pulmonary arterial branches. Pulmonary thromboembolism is considered difficult to differentiate from a tumor in the RV outflow tract and pulmonary trunk.[7]

Pulmonary hypoperfusion seems to be a key factor in the decision on whether to proceed with emergency surgery in terms of the survival of patients with tumors obstructing the pulmonary artery. According to the results of treatment for pulmonary artery sarcomas,[6,8] the median survival without surgical treatment is thought to be 1.5 months regardless of chemotherapy or irradiation. Resection has lengthened the median survival to 10 months, and resection associated with adjuvant therapy has lengthened median survival but has not prolonged life beyond 3 or 4 years.[9] However, most patients with pulmonary trunk sarcoma have undergone only incomplete resection.[6,10]

Although the tumor in our patient might not have been a candidate for complete resection at the time of the preoperative diagnosis, we decided on surgical resection to avoid sudden death resulting from a reduction in pulmonary blood flow. The surgical technique included (1) tumor resection associated with resection of the involved pulmonary valve, (2) reconstruction of pulmonary valvular function, and (3) enlargement of the pulmonary arteries, pulmonary trunk, and RV outflow. These procedures were aimed at delaying reobstruction of the RV outflow-to-pulmonary artery tract and maintaining stable peripheral pulmonary perfusion. Pulmonary endoarterectomy was performed for the part of the tumor that invaded the intima of the pulmonary arteries. The prevention of the pulmonary hypoperfusion and the resultant pulmonary hypertension caused by peripheral pulmonary microembolism soon after surgery might be an important early determinant of mortality. Pulmonary valvular function has been reconstructed using a bioprosthetic valve.[11,12] We implanted a stentless valve instead of a mono-cusped biomaterial patch to obtain competent pulmonary valvular function because the patient needed sufficient valvular function to maintain pulmonary blood flow against pulmonary hypertension resulting from distal tumor microembolization. Also, patch enlargement of the pulmonary arteries, which would delay pulmonary artery reobstruction by the tumor growth, seemed effective in lengthening the survival of our patient.

Whether adjuvant therapy for residual disease after tumor resection is related to the prognosis of patients with a sarcoma in the RV outflow tract and pulmonary arteries is unknown. Chemotherapy, including CDDP and adriamycin (doxorubicin hydrochloride), failed to suppress tumor growth in our patient, whereas irradiation partially relieved pulmonary artery restenosis, which alleviated his symptoms. Chemoradiotherapy for pulmonary artery sarcoma has been shown to have a beneficial effect on short-term morbidity (respiratory symptoms caused by pulmonary artery obstruction)[13] and long-term survival.[14] Because we had no evidence-based regimen of radiation therapy for cardiac or pulmonary artery sarcoma, we calculated the total dose and duration of irradiation based on several case reports.[13,14] Chemotherapy was thought to be more effective on tumor growth than radiotherapy because we had not seen any study suggesting that radiotherapy was preferable to chemotherapy based on beneficial effects of radiotherapy on cardiac or pulmonary artery sarcomas. That was the reason why chemotherapy was given priority to radiotherapy in our patient. Adjuvant therapy for residual sarcomas after noncurative resection should be applied on an individual basis because effectiveness might depend on tumor characteristics.

Conclusion

Undifferentiated cardiac sarcoma associated with pulmonary hypoperfusion should be resected even if incompletely, and pulmonary valve replacement may be effective for maintaining forward pulmonary blood flow. Irradiation as adjuvant therapy for the residual sarcoma could improve a prognosis after surgery.

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