Primary Ewing Sarcoma in the Right Ventricle in an Adult Patient: A Case Report

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Abstract

Ewing sarcoma, a highly aggressive round-cell neoplasm of unknown origin, rarely occurs as a primary cardiac tumor. In this intriguing case, we describe an instance of primary Ewing sarcoma in an adult patient's right ventricle (RV).

The patient, a 27-year-old man, presented with symptoms including epigastric pain, fever, tachycardia, nausea, vomiting, shortness of breath, and a dry cough. Imaging studies, including a computed tomography scan and echocardiogram, revealed an anterior mediastinal mass arising from the free wall of the RV and extending into the pericardium. The mass exerted pressure on the RV outflow tract and the pulmonary artery.

Surgical intervention was deemed necessary based on the clinical presentation and paraclinical findings. The mass was meticulously dissected from the cardiac tissue, and the RV outflow tract was resected, along with the pulmonary valve and the main pulmonary artery. These structures were replaced with a composite Dacron graft, anastomosed to the bifurcation of the main pulmonary artery. Additionally, the mass near the superior vena cava and the pulmonary vein region was carefully excised.

The patient was successfully weaned off the cardiopulmonary bypass pump in stable condition and transferred to the ICU. Histopathological analysis confirmed the diagnosis of small round cell sarcoma, specifically Ewing sarcoma. Following recovery, the patient was discharged and scheduled for regular follow-up appointments.

Following discharge, the patient was referred to a radiotherapy center and underwent radiotherapy. During a follow-up examination 1 year later, no signs of disease recurrence or progression were observed, offering a promising outlook. This case underscores the importance of timely referral and treatment for cardiac Ewing sarcoma, which may significantly improve prognosis.

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Background

Ewing sarcoma is a highly aggressive round-cell neoplasm of unknown origin, comprising approximately 5% of bone tumors, primarily affecting long bones.[1] It is a relatively uncommon primary soft tissue tumor, representing 20% to 30% of all reported Ewing sarcoma cases.[2] Most cases occur in patients aged 10 to 30 years.[3] Among primary cardiac tumors, only 25% are malignant, with sarcomas accounting for 95% of these malignancies. Ewing sarcoma typically arises in the bones of children, making cardiac Ewing sarcoma exceedingly rare. Few cases of this occurrence have been documented in the literature.[4]

Metastases at the time of diagnosis are uncommon, observed in fewer than 25% of patients. Nonetheless, due to its high recurrence rate following local therapy and its propensity to metastasize, Ewing sarcoma is considered a systemic disease that often requires multimodality treatment.[5]

This study presents a case report of primary Ewing sarcoma in the right ventricle (RV) of an adult patient.

Case Report

The patient was a 27-year-old man who presented with symptoms including epigastric pain, fever, tachycardia, nausea, vomiting, shortness of breath, and a dry cough. He reported a 5 kg weight loss over the preceding 2 months. Transthoracic echocardiography revealed extensive pericardial effusion, physiological tamponade (without clinical signs of tamponade), and a pericardial mass, prompting hospitalization. A subsequent computed tomography (CT) scan identified an anterior mediastinal mass within the pericardial space (Figure1). Subsequent transesophageal echocardiography revealed an echogenic mass (4×9 cm) arising from the free wall of the RV, exerting pressure on the right ventricular outflow tract (RVOT) and pulmonary artery within the pericardial space (Figure 2). Surgical intervention was deemed necessary based on the clinical presentation and paraclinical findings. The patient underwent general anesthesia and median sternotomy with cardiopulmonary bypass. Upon opening the pericardium, the mass was identified in the RVOT region, where it was firmly attached (Figure 3). The mass was meticulously dissected and separated from the cardiac tissue. The RVOT, along with the pulmonary valve and main pulmonary artery, was resected and removed. This area was replaced with a composite Dacron graft, and the graft was anastomosed to the bifurcation of the main pulmonary artery (Figure 4). Moreover, the mass near the superior vena cava and the pulmonary vein region was successfully excised.

Following reperfusion, the patient's heart rate stabilized, marking a critical milestone in the treatment process. Hemostasis was achieved, and the patient was successfully weaned off the cardiopulmonary bypass pump. He was then transferred to the ICU for postoperative monitoring.

Six hours postoperatively, the patient was extubated and transferred to the general ward after 2 days. Histopathological analysis confirmed the presence of small round cell sarcoma, specifically Ewing sarcoma, confirming the initial diagnosis. Following an uneventful recovery, the patient was discharged after 5 days and referred to a radiotherapy center for adjuvant treatment.

The effectiveness of the radiotherapy was demonstrated during a follow-up examination 1 year later. A contrast-enhanced CT scan revealed no evidence of disease recurrence or metastasis, indicating a positive outcome.

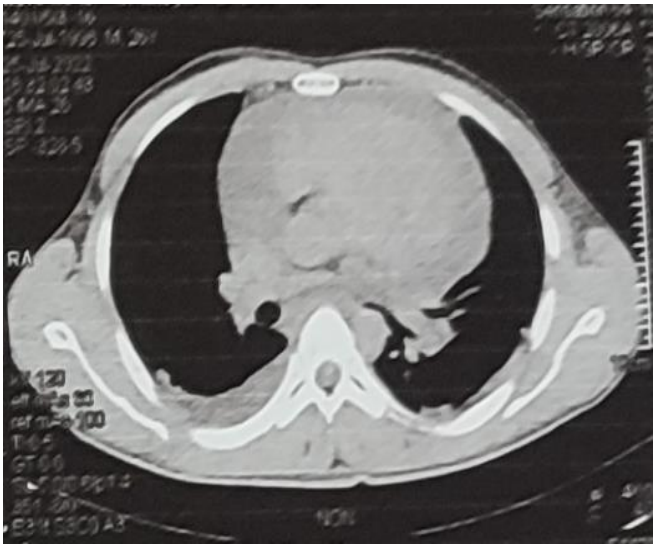


Figure 1. A computed tomography scan showed a mass in the pericardial space located in the anterior mediastinum



Figure 2. Transesophageal echocardiography showed an echogenic mass measuring 4×9 cm, originating from the free wall of the right ventricle. It exerted pressure on the right ventricular outflow tract and pulmonary artery within the pericardial space.

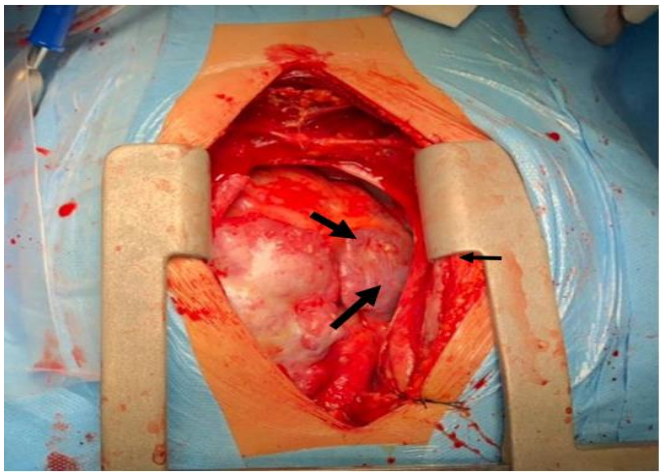


Figure 3. A portion of the mass (indicated by the black arrow) was identified as protruding through the right atrium, the free wall of the right ventricle, and the main pulmonary artery.

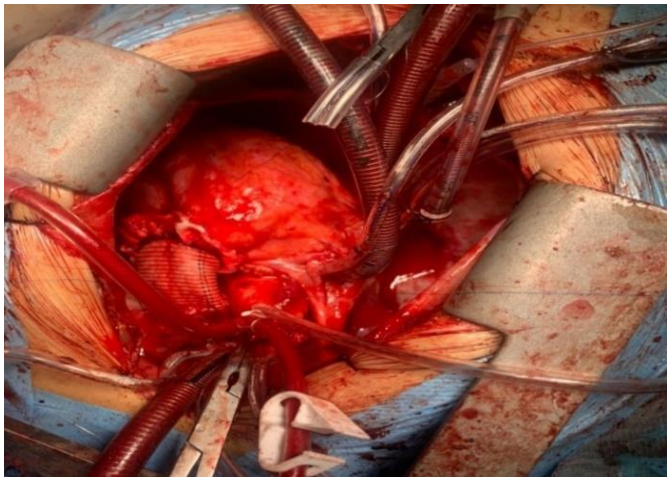


Figure 4. The right ventricular outflow tract, along with the pulmonary valve and main pulmonary artery, was surgically excised and reconstructed using a Dacron graft.

Discussion

Ewing sarcoma is a highly aggressive cellular neoplasm, accounting for approximately 5% of bone tumors, with a predilection for long bones.[1] According to previous reports, the incidence of extraosseous Ewing sarcoma is 0.4 cases per million individuals, which is tenfold lower than that of skeletal Ewing sarcoma.[6]

Malignancies that commonly metastasize to the heart include bronchogenic carcinoma, renal cell carcinoma, Wilms tumor, chondrosarcoma, and osteosarcoma.[7] At the time of diagnosis, approximately 25% of patients with Ewing sarcoma present with metastases to the lungs or other bones.[7]

The differential diagnosis of cardiac masses can be categorized as neoplastic or non-neoplastic. Neoplastic cardiac masses encompass primary benign tumors, primary malignant tumors, and secondary (metastatic) tumors. Secondary tumors are reported to be 20 to 40 times more common than primary cardiac tumors. Non-neoplastic cardiac masses include thrombi of various origins, pericardial cysts, and vegetations.[8]

Although rare, the incidence of extracardiac Ewing sarcoma exhibits a bimodal distribution, with peak occurrences in children (<5 y) and adults (>35 y).[9]

Cardiac tumors can present with symptoms such as shortness of breath, arrhythmias, or syncope, often due to obstruction of left ventricular inflow or outflow.[10] Metastases to the RV may also occur, potentially leading to right heart failure caused by obstruction of the RV's inflow or outflow.[10] In addition, tumors or thrombi can embolize, resulting in syncope or even fatal outcomes. Myocardial ischemia may develop secondary to external compression of the coronary arteries or embolic events within the coronary circulation.[11]

A definitive diagnosis is confirmed through the pathological examination of a biopsy sample, though this is not always feasible. CT and magnetic resonance imaging are valuable tools in aiding the diagnostic process.[12] Surgical intervention is recommended for patients exhibiting signs of obstruction or cardiac dysfunction.[13] In certain cases, emergency surgery may be warranted due to the risk of embolism.[14] However, in some instances, surgical removal of the cardiac mass may be delayed or avoided, particularly in the presence of concurrent pulmonary metastasis and a poor prognosis.[12] Following complete resection of the sarcoma, chemotherapy and long-term follow-up are recommended for patients with primary cardiac sarcoma, given the high rates of metastasis and recurrence.[4]

This case highlights the importance of timely referral and treatment of cardiac Ewing sarcoma, which may significantly improve prognosis.

References

1. Mirra JM. Bone tumors. clinical, radiologic, and pathologic correlations 1989; 226-48.
2. Abboud A, Masrouha K, Saliba M, Haidar R, Saab R, Khoury N, et al. Extraskeletal Ewing sarcoma: diagnosis, management and prognosis. Oncol Lett. 2021 ;21(5):354.
3. Iwamoto Y. Diagnosis and treatment of Ewing's sarcoma. Japanese journal of clinical oncology 2007; 37: 79-89.
4. Ushigusa J, Mukae Y, Takamatsu M, Nogami E, Furutachi A, Itoh M, et al. Adult-onset primary Ewing's sarcoma of the right atrium: a case report. Surg Case Rep. 2019;5(1):171.
5. Angervall L, Enzinger F. Extraskeletal neoplasm resembling Ewing's sarcoma. Cancer 1975; 36: 240-51.
6. van den Berg H, Heinen RC, van der Pal HJ, Merks JH. Extra-osseous Ewing sarcoma. Pediatric Hematology and Oncology 2009; 26: 175-85.
7. Cotterill SJ, Ahrens S, Paulussen M, Jürgens HF, Voûte PA, Gadner H, *et al.* Prognostic factors in Ewing's tumor of bone: analysis of 975 patients from the European Intergroup Cooperative Ewing's Sarcoma Study Group. J Clin Oncol 2000; 18: 3108-14. doi: 10.1200/jco.2000.18.17.3108
8. Huh J, Noh CI, Kim YW, Choi JY, Yun YS, Shin HY, *et al.* Secondary cardiac tumor in children. Pediatr Cardiol 1999; 20: 400-3. doi: 10.1007/s002469900500
9. Applebaum MA, Worch J, Matthay KK, Goldsby R, Neuhaus J, West DC, *et al.* Clinical features and outcomes in patients with extraskeletal Ewing sarcoma. Cancer 2011; 117: 3027-32.
10. Petrovic M, Zhao B, Thangam M, Loyalka P, Buja LM, Kar B, *et al.* Ewing sarcoma in the right ventricle. Texas Heart Institute Journal 2016; 43: 458-60.
11. Dotson J, Urella M, Shenouda M, Abu-Hashyeh A, Lebowicz Y. Extraosseous Ewing sarcoma presenting with inferior ST-elevation myocardial infarction and systemic emboli due to

tumor thrombus and invasion of the left atrium. Case Rep Orthop. 2020; 2020:3861927.
12. Kulkarni SK, Moorthy N, Somanna S, Cholenahally Nanjappa M. A rare case of Ewing's sarcoma/primitive neuroectodermal tumor metastasizing to left atrium through left inferior pulmonary vein. Echocardiography 2018; 35: 885-8. doi: 10.1111/echo.13897
13. Faim D, Francisco A, Pires A. Cardiac tumours in children: a single-centre experience and literature review. Cardiol Young 2022; 1-10. doi: 10.1017/s1047951122000531
14. Murad R, Bakkar R, Sukkar R, Mahmoud F, Al-Dairy A. A Case of Ewing Sarcoma Presenting With Cardiac Metastasis. World J Pediatr Congenit Heart Surg 2019; 10: 632-4. doi: 10.1177/2150135119846047