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Primary Cardiac Lymphoma A Rare Case of Pulmonary Tumor Embolism

Gabriel Pérez Baztarrica, MD; Norma Nieva, MD; Luis Gariglio, MD;
Flavio Salvaggio, MD; Rafael Porcile, MD

Human immunodeficiency virus can cause pericarditis, myocarditis, cardiomyopathies, endocarditis, other valvular compromises, cardiac involvement by neoplasm, pulmonary hypertension, arrhythmia, and thromboembolic disease. Myocardial involvement by tumors has been reported in rare cases, with Kaposi sarcoma and non-Hodgkin lymphomas being the most frequent tumor types. In this case, we introduce a 51-year-old man who presented with a febrile syndrome lasting for 30 days. He had just been on a 7-day course of azithromycin prescribed for a respiratory infection. On day 10 after he completed the antibiotic course, fever recurred without evidence of an identifiable infectious focus. Because of the aforementioned clinical presentation and to rule out infectious endocarditis, an echocardiogram was performed on the patient, which showed an image in the right atrium that could correspond to a vegetation. On physical examination, heart rate was 100 bpm, blood pressure 120/75 mm Hg, and jugular distention 3/3 without inspiratory change. Laboratory data were as follows: white blood cells $12\,300/\text{mm}^3$, C-reactive protein $+++$, and erythrocyte sed-

imentation rate 67 mm/h. Serology for human immunodeficiency virus was positive. Findings on transesophageal echocardiography were as follows: a round mass located at the posterior portion of the interatrial septum protruding toward the left atrium and the right atrium and infiltrating into the posterior wall of the right atrium. The mass was 7 cm in diameter, multilobulated, seemed to have a firm consistency, and showed occlusive behavior at the level of the confluence with the inferior vena cava (Figure 1A). A second mass situated on the free wall of the right atrium along the base of implantation of the tricuspid valve and the posterior wall of the right ventricle was observed. It appeared ≈ 6 cm in diameter and less echogenic. Severe pericardial effusion with collapse of the right chambers was present as well. Pericardiocentesis was performed, and samples of pericardial fluid were sent for cytological analysis. The clinical course of the patient was favorable. For further evaluation of the intracardiac mass, cardiac magnetic resonance imaging with eventual biopsy was considered. Coronary magnetic resonance angiography reported a heterogeneous, multilobulated, T1 hypoin-

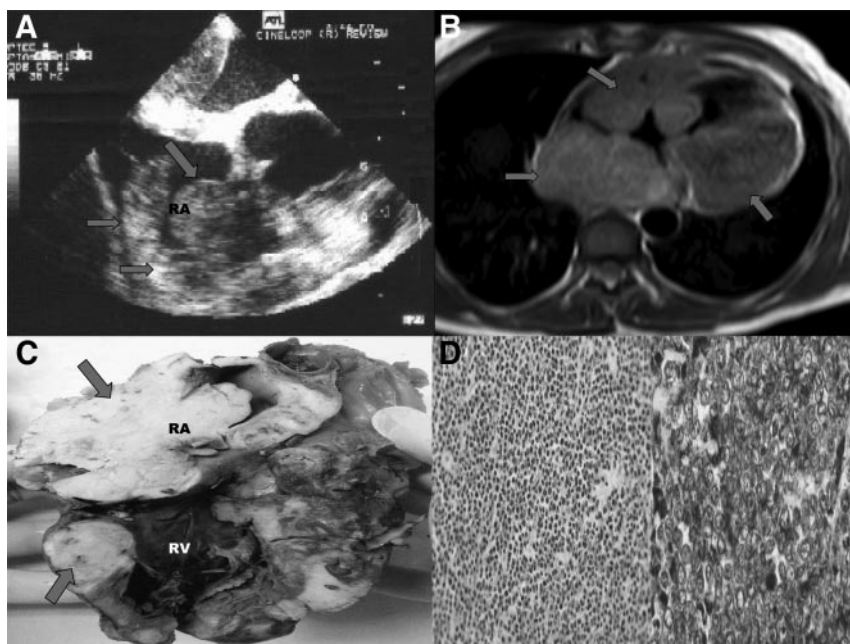


Figure 1. A, Transesophageal echocardiography showing a round mass located at the right atrium and occlusive behavior at the level of the confluence with the inferior vena cava. B, Cardiac magnetic resonance image showing a nodular mass (arrows). C, An autopsy revealed a firm cardiac mass with multiple nodules (arrows). D, Histological section from the cardiac mass and pulmonary emboli revealed diffuse proliferation of large B-cell lymphoma (hematoxylin-eosin and $\text{CD}20^+$). RA indicates right atrium; RV, right ventricle.

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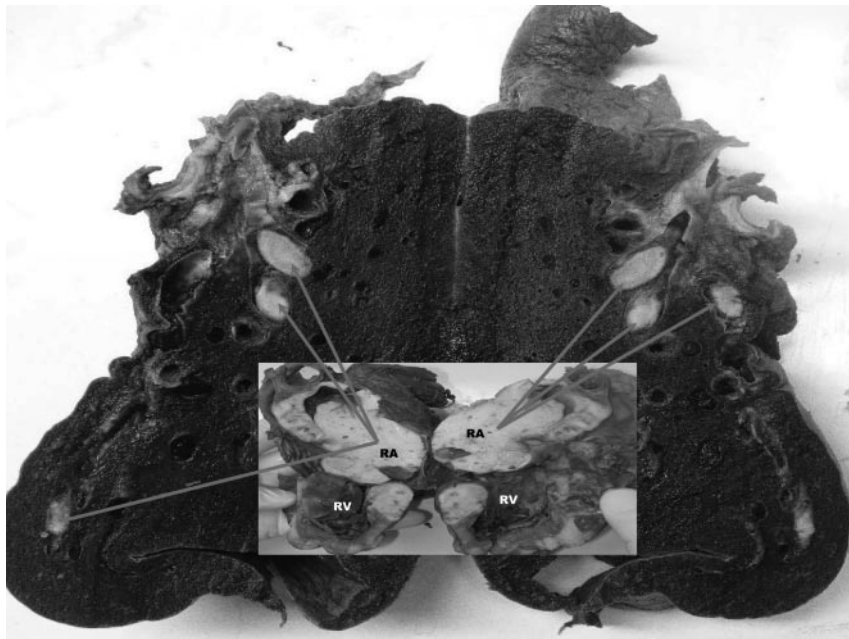


Figure 2. Both lungs showed multiple, grossly visible tumor emboli (arrows). RA indicates right atrium; RV, right ventricle.

tense, T2 hyperintense, vascularized mass showing areas of hemorrhage and necrosis, occupying the right atrium almost entirely and protruding toward the atrial septum and posterior wall of the left atrium and infiltrating the pericardium. The mass spread into the right ventricle and involved both inflow and outflow tracts as well as the free wall. The diameter of the mass was 8 cm, and, according to the radiological features, it could correspond to a sarcoma (Figure 1B). During workup, the patient presented with cough, hemoptysic expectoration, and hemodynamic decompensation followed by death. Necropsy examination revealed primary cardiac diffuse large B-cell lymphoma associated with tumoral pulmonary embolism (Figures 1C, 1D, and 2).

Of all primary cardiac tumors, lymphomas are rare, especially among immunocompetent individuals (1.3%).¹ B-cell lymphomas are the most common type. They usually involve the right chambers and pericardium.² B-cell lymphoma progresses quickly, and delay in diagnosis is associated with poor prognosis. Patients can be asymptomatic or present with symptoms related to tumor extension and/or embolus (obstruction, arrhythmia, pulmonary or systemic embolism, conduction disturbances, fever, weight loss, and cardiac tamponade).^{2,3} Some patients present with chest pain. Reported cases of tumoral pulmonary embolism, as in the present case, are extremely rare.^{4,5} Among the diagnostic tools, transthoracic echocardiogram should be considered as a first approach. Its sensitivity depends on the size of the masses and the acoustic (sonic) window of each patient. It is also used for assessing associated pericardial effusion. Transesophageal echocardiography represents a useful tool, as demonstrated in this case.⁶ Computed tomography and magnetic resonance imaging allow not only the diagnosis of polypoid, ill-defined,

infiltrating masses but also provide information about the type of tumor (eg, vascularization, necrosis).¹⁻³ Although imaging techniques are of great value, the best tool for a diagnosis with certainty remains anatomopathological (cytology of pericardial fluid or transvenous biopsy of the mass). Transvenous biopsy is a controversial procedure because of the risk of pulmonary embolism.²⁻⁴ Treatment options include chemotherapy and radiology. Surgical treatment is palliative (for relieving obstruction). In terms of prognosis, mean survival of patients with sarcoma is 9 to 11 months depending on the stage of the disease at the time of diagnosis.⁶

Disclosures

None.

References

1. Sato Y, Matsumoto N, Kinukawa N, Matsuo S, Komatsu S, Kunimasa T, Yoda S, Tani S, Takayama T, Kasamaki Y, Kunimoto S, Furuhashi S, Takahashi M, Saito S. Successful treatment of primary cardiac B-cell lymphoma: depiction at multislice computed tomography and magnetic resonance imaging. *Int J Cardiol*. 2006;113:e26–e29.
2. Burling F, Devlin G, Heald S. Primary cardiac lymphoma diagnosed with transesophageal endocardiography-guided endomyocardial biopsy. *Circulation*. 2000;101:e179–e181.
3. Nakata A, Hirota S, Takazakura E. Primary cardiac lymphoma diagnosed by percutaneous needle biopsy. *Int J Cardiol*. 1998;65:201–203.
4. Skalidis E, Parthenakis F, Zacharis E, Datsis G, Vardas P. Pulmonary tumor embolism from primary cardiac B-cell lymphoma. *Chest*. 1999;116:1489–1490.
5. Bestetti R, Soares F, Soares E, Oliveira S. Primary lymphoma of the right atrium with fatal neoplastic pulmonary embolism. *Am Heart J*. 1992;124:1088–1090.
6. Antoniadis L, Eftychiou C, Petrou P, Bagatzounis A, Minas M. Primary cardiac lymphoma: case report and brief review of the literature. *Echocardiography*. 2009;26:214–219.