

Pericardial Synovial Sarcoma: Case Report and Literature Review

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Introduction

Pericardial synovial sarcoma is an extremely rare primary malignant tumor of the heart and has an unclear prognosis.

The purpose of this report is to describe the case of a patient with pericardial synovial sarcoma whose initial presentation was severe pericardial effusion and dramatic weight loss. Despite treatment, the patient died in one year. Then, we reviewed the medical literature for the epidemiology, clinical picture, relevance of complementary imaging tests, therapeutic conduct and prognosis of this neoplasm.

Case Report

Female patient, 29 years old, developed dyspnea on mild exertion that evolved into orthopnea in four days; associated with mild chest pain that improved with knee-chest position. The patient lost 11 kg in two months. She denied smoking or alcohol consumption. The patient's mother had presented breast neoplasm.

On examination, she was mildly tachypneic with a respiratory rate of 24 rpm and decreased breath sounds in the lung bases, no adventitious sounds. The patient had tachycardia, heart rate of 110 bpm, hypophonic heart sounds, paradoxical pulse characterized by a drop in blood pressure of 105 x 63 mmHg to 80 x 58 mmHg during deep inspiration and pathological jugular venous distention at 45°.

Electrocardiogram showed signs of low ventricular voltage. Echocardiography revealed significant pericardial effusion with signs of diastolic restriction and oval, hyperechoic image with a diameter of 2 cm in the pericardial sac adjacent to the Left Atrium (LA) wall, suggesting clot.

The patient underwent urgent pericardial drainage with removal of 1000 mL of serohemorrhagic fluid, which laboratory analysis showed the presence of exudate with large amount of red blood cells. Gram-positive bacteria, Acid-Alcohol Resistant Bacilli (AARB), neither neoplastic cells were found. Adenosine deaminase levels were normal.

Keywords

Sarcoma, Synovial / surgery; Heart Neoplasms / surgery; Pericardial Effusion; Magnetic Resonance Imaging.

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After ten days in hospital, during which the patient was clinically stable, she was discharged for outpatient investigation. With a medical appointment scheduled for the following week, the patient returned only after five months, complaining of fatigue on mild exertion.

On this occasion, the echocardiogram revealed a large hyperechoic mass in posterior topography, extending from the LA to the middle portion of the left ventricle (LV). Within the mass, there were oval hypoechoic images, some of which presented septa suggestive of central necrosis.

Magnetic Resonance imaging (MRI) of the heart was requested, which revealed a large heterogeneous mass within the pericardial sac, adjacent to the free wall of the LA and the mid-basal portion of the LV lateral wall, measuring 9.3 x 8.0 x 4.3 cm in diameter and multiple septa inside (Figure 1). The central portion of the mass was hypointense on T1-weighted spin-echo sequence, hyperintense on T2-weighted spin-echo sequence and showed no decrease in signal intensity in the fat saturation pre-pulse sequence. The first-pass perfusion imaging revealed good vascularization of the mass periphery and septa, and no significant central vasculature. On late enhancement sequence, there was intense gadolinium uptake by the mass periphery and septa and absence of gadolinium uptake by the central region. The appearance suggested a large malignant tumor of the pericardium with multiple areas of central necrosis, apparently without invasion of adjacent structures by the tumor.

It was decided to refer the patient to cardiac surgery in order to achieve complete excision of the mass for diagnosis and therapeutic purposes.

During the surgery, a capsule covering the mass was found. The capsule was opened and inside it, there was a large friable tumor with the appearance of "fish meat". It had multiple septa and a large amount of necrotic material inside. The tumor had ill-defined borders and was extremely attached to the heart, without cleavage plane, which prevented its complete excision (Figure 2). Therefore, only one partial resection of the tumor was performed.

Pathological examination showed spindle cells consistent with the diagnosis of pericardial synovial sarcoma.

Chemotherapy and radiotherapy were indicated, but the patient failed to follow treatment from the start. She only started chemotherapy nine months later. After the second cycle, the patient developed progressive dyspnea, fever, dry cough and significant decline in overall condition. Admitted on an emergency, she was diagnosed with severe community-acquired pneumonia associated with severe neutropenia. Despite the prescription of appropriate antibiotic therapy,

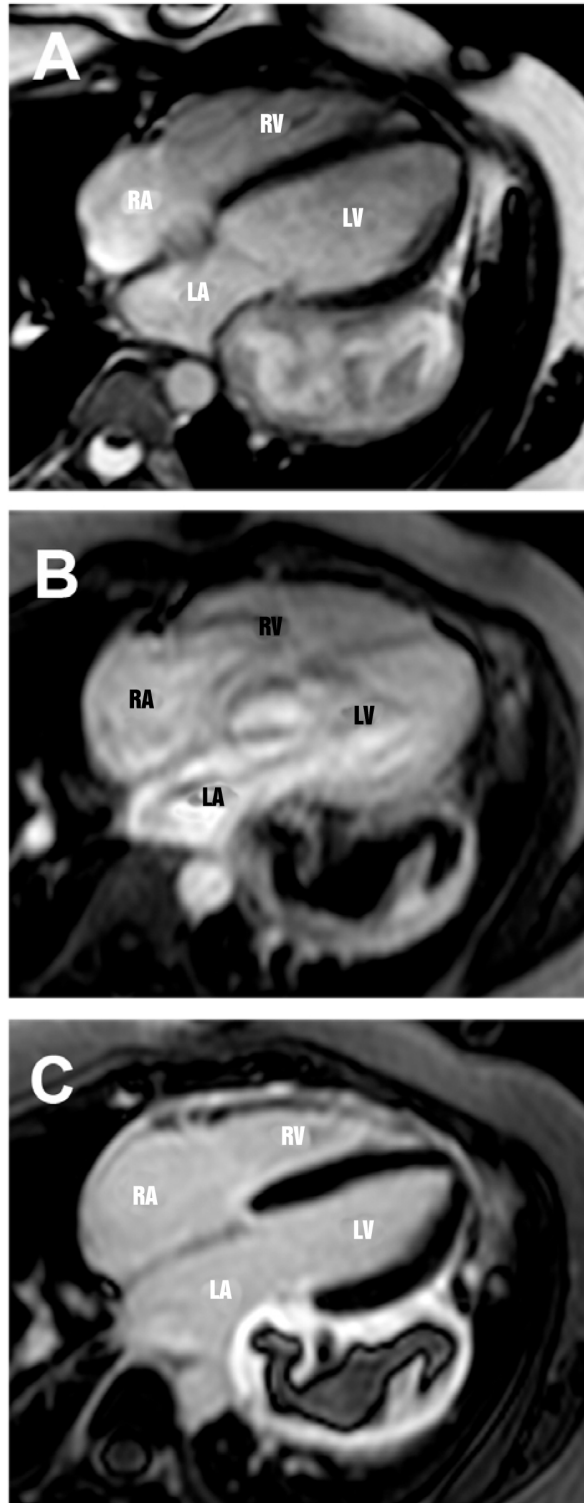


Figure 1 - Cardiac magnetic resonance imaging. A. Cine imaging sequence showing a large heterogeneous tumor with multiple septa inside, located within the pericardial sac, adjacent to the LA free wall and the mid-basal portion of the LV lateral wall. B. First-pass perfusion sequence. Note the tumor periphery and septa perfusion. There is no perfusion of the central region of the tumor (necrosis). C. Delayed enhancement sequence. Note the intense gadolinium uptake by the tumor periphery and the septa. There is no gadolinium uptake by the central region (necrosis). RA: right atrium; RV: right ventricle; LA: left atrium; LV: left ventricle.

Case Report

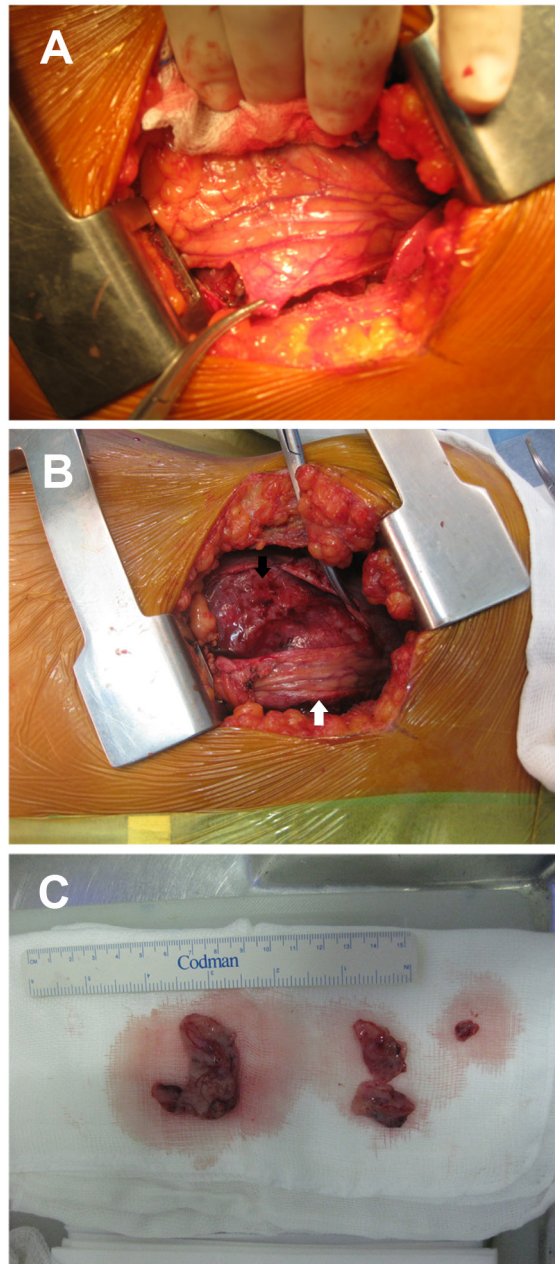


Figure 2 - Photos of the surgery. A: Capsule covering the tumor. B: The capsule (white arrow) was open and inside there is a large friable pericardial tumor (black arrow) with the appearance of "fish meat", imprecise borders and extremely attached to the heart. C: Tumor fragments with multiple septa and a large amount of necrotic material inside.

recombinant human granulocyte colony-stimulating factor and the support of intensive care, the patient developed respiratory failure and septic shock in two days and died.

Discussion

Primary cardiac tumors are rare, with a reported incidence of 0.05% in a study of 12,485 autopsies. Secondary involvement

of the heart and pericardium by extracardiac tumors is twenty times more common than primary cardiac tumors. The neoplasms that most often secondarily affect the cardiac structures are the tumors of lung and breast, melanoma and lymphoma, either by distant metastasis or by direct invasion¹.

Around 80% of primary cardiac tumors are benign. The atrial myxoma is the most common one. About 20% of

primary cardiac tumors are malignant. The sarcoma is the one most frequently found, followed by mesothelioma and lymphoma². Among the cardiac sarcomas, the pericardial synovial sarcoma is extremely rare. It originates from mesenchymal pluripotent cells and gets its name from the histologic similarity to synovial tissue in formation³.

The signs and symptoms caused by the primary cardiac sarcoma are extremely variable and nonspecific. Clinical manifestations can be divided into four categories: systemic; embolic; cardiac; and secondary to metastasis.

The most common systemic manifestations are fever, chills, fatigue and weight loss, and this patient showed significant weight loss of 11 kg in two months. Embolic manifestations occur mainly in sarcomas invading the cardiac cavities, causing pulmonary or systemic embolism. Cardiac manifestations depend on the location of the tumor: predominantly pericardial, intramural or intracavitary. In the case of pericardial sarcoma, the most common initial clinical picture results from massive hemorrhagic pericardial effusion and cardiac tamponade^{3,4}. The organs most often affected by primary cardiac sarcoma metastases are the lungs, brain and bones.

Imaging methods are extremely important for the diagnosis of cardiac tumors, which are often an incidental finding in tests performed for other reasons. In the case of pericardial tumors, echocardiography can identify the mass and it is important for quantifying pericardial effusion and evaluating the presence of signs of diastolic restriction, as in the case of this patient.

MRI and computer-aided tomography help determining the location and extent of the tumor and assessing local invasion of adjacent organs. They are also essential for surgical planning. MRI, by using multiple techniques, is important to characterize the tumor tissue in a detailed manner, enabling the evaluation of characteristics suggestive of malignancy⁵. Due to the rapid growth of malignant tumors, they may present central necrotic areas characterized for being hypointense on T1-weighted spin-echo sequence, hyperintense on T2-weighted spin-echo sequence and for not presenting gadolinium uptake in the sequences of perfusion and delayed enhancement, as demonstrated in the present pericardial sarcoma. Other characteristics suggestive of malignancy, well demonstrated by MRI, include: involvement of more than one heart chamber, if the tumor is attached to the heart through a large base, extracardiac extension and presence of pericardial effusion⁵.

The pericardial sarcoma is a highly aggressive neoplasm. Therefore, in most cases it is not possible to perform a complete surgical resection. In a study with 24 patients, only 22.7% of resected cardiac sarcomas had free lesion edges⁶. Complete surgical resection is the main impact factor in patient survival. Adjuvant chemotherapy and radiotherapy are recommended when the surgery is incomplete or for recurrent tumors⁷. However, despite treatment, the prognosis of patients with cardiac sarcoma is reserved, with median survival of only 25 months⁶.

This report described the case of a pericardial synovial sarcoma, an extremely rare primary cardiac tumor which, despite proper treatment, has an unclear prognosis.

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