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Case Report

Evaluation of a cardiac sarcoma with CT multislice contrast-enhanced and 18FDG-PET/TC

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ABSTRACT

We present the case of an adult male who arrived to our emergency room with progressive dyspnea that had been ongoing for 2 months. During the radiological investigation, we found a large intracardiac mass, which invaded the pericardium, pulmonary trunk, pulmonary arteries, and left ventricle. Studies done with the 18FDG-PET/CT scan helped us to determine the malignant nature of the mass and to suspect the diagnosis of rhabdomyosarcoma.

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Introduction

According to the Armed Forces Institute of Pathology, primary tumors of the heart and pericardium are rare, having an incidence of 0.001 % to 0.28% in autopsy series [1], while metastatic cardiac tumors are about 30 times more common than primary neoplasm [2,3].

About 75% of all tumors of the heart in adult age are benign with myxoma being the most common benign tumor (50%).

Malignant tumors account for the remaining 25%: mesenchymal tumors constitute 2/3 of the cases, with the most common pathologies being angiosarcoma and rhabdomyosarcoma followed by fibrosarcoma, leiomyosarcoma, liposarcoma, and other poorly differentiated sarcomas [4,5].

Mesothelioma and lymphoma account for the other 1/3 of malignant tumors.

Case presentation

A 74-year-old man visited the emergency department for progressive dyspnea that had begun in the previous 2 months. Upon arrival, the patient had a mild respiratory distress, with a respiratory rate of 22/min and a heart rate of 88 bpm. He had a history of hypertension and hypercholesterolemia, but no history of immunodepression or exposure to asbestos. After having ruled out myocardial infarction with an electrocardiogram, a contrast-enhanced chest computed tomography (CT)

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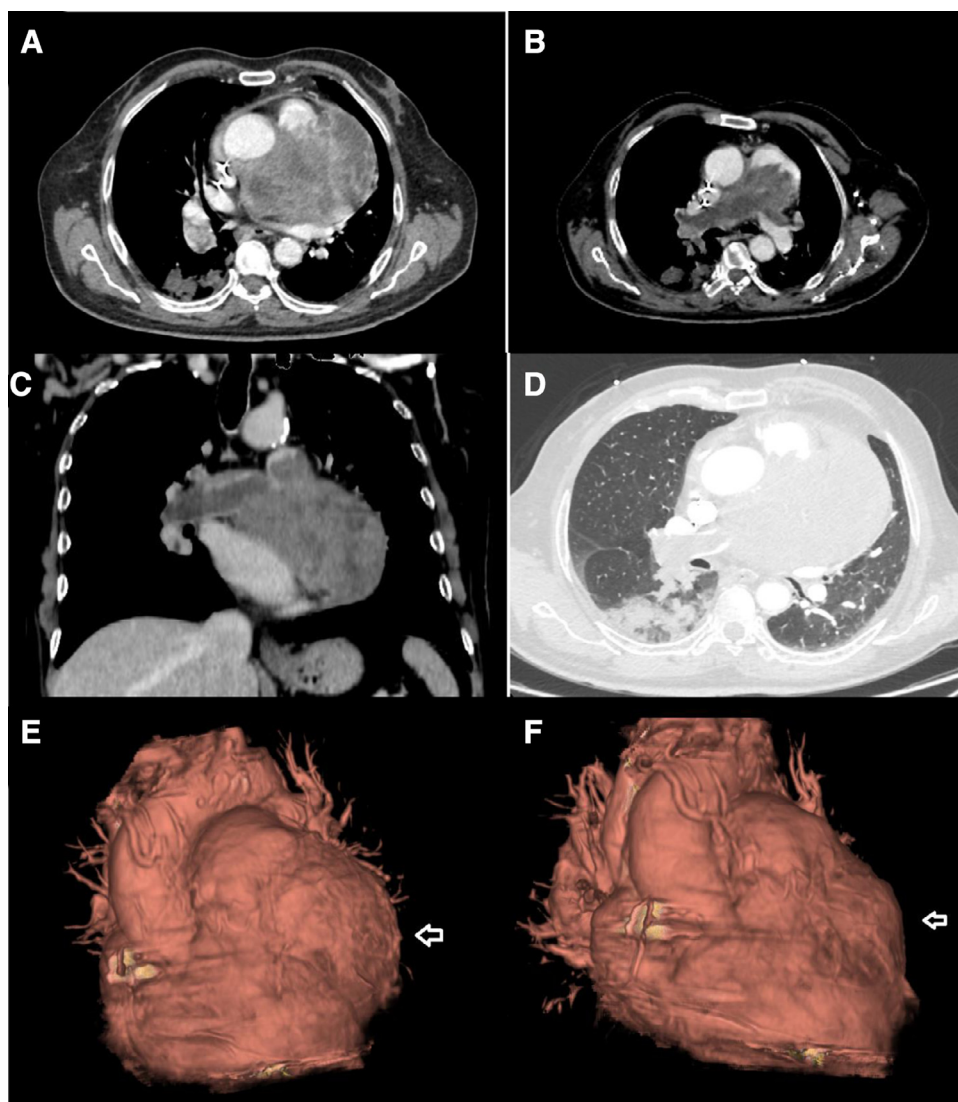


Fig. 1 – Computed tomography; (A) Axial section, intracardiac mass that infiltrate left ventricle and pericardium; (B) axial section, invasion of pulmonary trunk and both pulmonary arteries; (C) coronal section, both pulmonary arteries, pericardium and left ventricle invaded; (D) axial section, lung window, pulmonary neoplastic embolism phenomena on lower lobe of right lung; (E, F) 3D reconstructions of the mass (white arrow) invading pericardium.

(Iobitridolo; volume: 100 mL; flow rate: 3.5 mL/s, slice thickness: 2.5 mm) was performed to exclude pulmonary embolism. The exam documented a large intracardiac mass which showed nonhomogeneous contrast-enhancement during contrast CT acquisition with some necrotic areas, that infiltrated left ventricle, pericardium, pulmonary trunk, and both pulmonary arteries, mainly involving the right branch, which extended into the hilum and to the arterial branches tributary for the ipsilateral lower lobe, where neoplastic embolism phenomena were documented (Fig. 1).

A FDG-PET/CT (298 Mbq-120 mL Iobitridol 300 mg/mL) was then performed and that excluded any primary noncardiac tumor or any other distant metastasis. The cardiac mass had a nonhomogeneous uptake of FDG with a Maximum standardized uptake value (SUVmax) of 32.3. Increased uptake of FDG was also documented for mediastinic lymphnodes (Fig. 2).

No biopsy was performed because the patient died soon after his hospitalization due to cardiorespiratory failure.

No autopsy was performed because the family declined consent.

Discussion

Cardiac tumors are rare and can be divided into primary cardiac tumors (PCT) and metastatic tumors. Most frequent metastatic lesions come from breast, lung, kidney, skin and hematopoietic tumors [6]. Almost 75% of PCT are benign tumors, with atrial mixoma being the most frequent one [4,7]. Malignant PCT accounts for 25% of the cases, of which about 2/3 are sarcomas, in particular angiosarcoma and

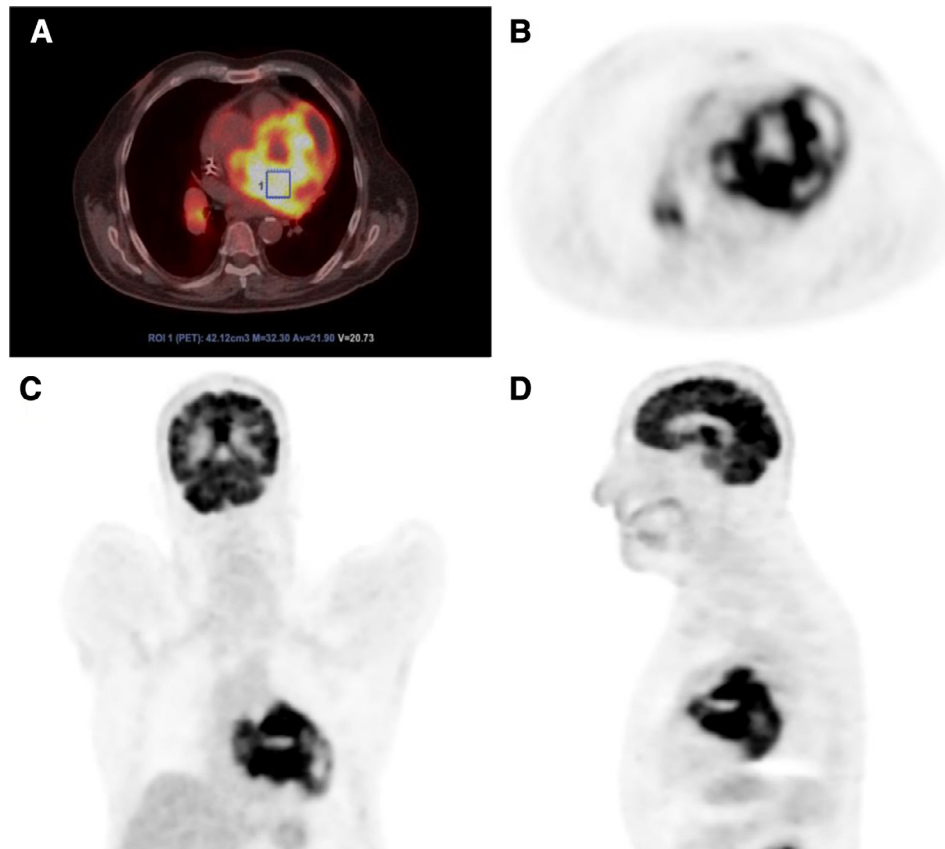


Fig. 2 – The ^{18}F -fluorodeoxyglucose positron emission tomography/computed tomography images. (A) PET/CT fused images showing intense FDG uptake (SUVmax = 32.3); (B–D) axial, coronal, and sagittal reconstruction.

rhabdomyosarcoma, followed by fibrosarcoma, leiomyosarcoma, liposarcoma, and other poorly differentiated sarcomas [4,5] while the last 1/3 is composed of cardiac lymphomas and mesotheliomas.

The majority of primary cardiac sarcomas are located in the atrial cavities. According to the literature, about half of right atrium tumors are malignant, while the majority of those arising in the left atrium are benign, most of which being myxomas [8].

Echocardiography is the first line exam. CT or magnetic resonance imaging can confirm the suspicion of malignancy [9,10] and can be employed to determine the possible presence of a primary noncardiac tumor, thus helping us to rule out the possibility of the mass being a metastasis. FDG-PET/CT may help in the diagnosis as it can differentiate a benign from a malignant lesion, with a 100% sensitivity and about 86% specificity, using a cutoff of SUVmax of 3.5 [11]. Biopsy may also be performed if there is suspicion of a lymphoma (this kind of tumor must be suspected in immunodepressed patients) and must be followed by chemotherapy and/or radiotherapy [12]. Immunohistochemistry is the final step to identify the histotype of the tumor [7].

Angiomyosarcoma is a very rare cardiac neoplasia with only 200 cases described in the literature [13]. Typical localizations of cardiac angiomyosarcoma are the right atrium, the pericardium, the tricuspidal valve, and the vena cava. The

pathology is more frequently diagnosed in more advanced states due to the aspecificity of its symptoms; therefore, metastatic finding at the time of diagnosis is typical, making the prognosis severe [14]. The advanced disease state upon diagnosis makes the treatment controversial: the surgical excision is the first line treatment, but chemotherapy and radiotherapy have well-established postoperative roles because of the high probability of metastasis, particularly pulmonary; targeted therapy may also be considered [7].

Rhabdomyosarcoma is the second most common malignant tumor of heart, with an incidence of about 20%. It is rare in the pediatric age and more common in adulthood, but it has no well-defined age prevalence. It originates from the cardiac striated muscle, with higher incidence in men compared to women [1]. Unlike angiomyosarcoma, it can involve any of the heart chambers and it can grow invasively in either a single or in multiple locations. Invasion of the cardiac valves and spread to neighboring organs, pericardium, pleura, and mediastinum has been reported [4]. Similarly to angiomyosarcoma, symptoms usually manifest in late stage disease and may be not specific for cardiac disease, but more indicative of malignancy (fever, anorexia, weight loss), or pericardial disease (dyspnea, chest pain, pleural effusion, and embolic phenomena). Valvular infiltrations may restrict blood flow mimicking a stenosis of the mitral or tricuspid valve. Heart wall infiltration may result in hypertrophic or restrictive cardiomyopathy [15].

Cardiac tumors are often diagnosed because of thrombotic or neoplastic embolism that may cause stroke, pulmonary artery embolism, or peripheral limb vasculature [16].

In this report, we present a primary cardiac malignancy evaluated by contrast-enhancement chest CT and by 18FDG-PET/CT scan. Due to the lack of specific symptoms, the patient presented to the emergency department with only dyspnea and, after excluding a myocardial infarction with electrocardiogram, was evaluated with a contrast-enhancement chest CT which showed a large intracardiac mass that infiltrated left ventricle, pericardium, pulmonary trunk, and both pulmonary arteries, with a major extension on the right branch, involving the hilum and the arterial branches tributaries of the ipsilateral lower lobe where neoplastic embolism phenomena were documented.

FDG-PET/CT helped us to determine, noninvasively, the malignancy of the tumor as well and excluded the presence of possible secondary lesions as no further neoplastic disease was detected.

The history of the patient, the localization of the tumor, and the absence of pleural lesions helped us to exclude the diagnosis of mesothelioma, while origin, extension, and local infiltration of mediastinal pleura and pericardium oriented towards the diagnosis of rhabdomyosarcoma instead of angiosarcoma.

Despite advances in imaging techniques and their increasing clinical availability, most patients are diagnosed at an advanced stage because the symptoms lack and the prognosis of the patients does not survive more than 12 months from the time of diagnosis.

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