Case Report

Primary Cardiac Angiosarcoma Evaluated on PET/CT

Angiossarcoma Primário Cardíaco Avaliado em PET/TC

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Introduction

Cardiac angiosarcoma is a rare malignant endothelial tumor that is aggressive and has an adverse prognosis.[1,2] It features a high mortality rate because of the tendency for local recurrence and high incidence of systemic metastases.[1] Therefore, its early diagnosis is very important, as is the determination of its local and distant extension, to ensure its proper therapeutic management. The objective of this case report was to show the value of positron emission tomography associated with computed tomography with 18-fluorodeoxyglucose (^{18}F-FDG PET/CT) in the diagnosis and staging of cardiac angiosarcoma.

Caso Report

A 39-year-old man with dyspnea and palpitations at rest for approximately 15 days was admitted to the emergency room with signs of cardiac tamponade and underwent pericardiocentesis. During the hospitalization, the pericardial effusion recurred, which required a pericardial window and biopsy. The pericardial fluid showed a hematic aspect and high cellularity with a predominance of lymphomonocytic and low glucose, motivating empirical treatment with rifampicin, isoniazid, pyrazinamide, and ethambutol for possible tuberculosis.

Transthoracic echocardiography showed a fixed mass adherent to the lateral wall of the right ventricle (RV), right atrium (RA), and posterior tricuspid leaflet measuring approximately 30 mm × 34 mm. Transesophageal echocardiography showed a solid homogeneous mass adherent to the lateral wall of the RA projecting into the RV inlet and causing a flow obstruction with dimensions of 58 mm × 47 mm and without vascularization by microbubbles.

As the patient's condition failed to improve despite treatment, it was necessary to continue the investigation to better diagnose the cardiac mass. Magnetic resonance imaging (MRI) was not possible because of hemodynamic instability (lipothymia) at the time of the examination.

Keywords

Angiosarcoma; Positron-emission tomography; Diagnostic imaging.

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Therefore, the patient was referred for ^{18}F-FDG PET/CT imaging. This examination revealed a cardiac mass with an epicenter in the RA (Figures 1 and 2), heterogeneous density, a more inferior solid component, and a superior area of necrosis/liquefaction measuring approximately 75 mm × 60 mm × 86 mm with a maximum standardized uptake value (SUVmax) of ^{18}F-FDG of 19.6. Diffuse nodular thickening of the entire pericardium, including its recesses (SUVmax of 16.0) and hypermetabolic thoracic lymph nodes, was also observed with no evidence of distant metastases.

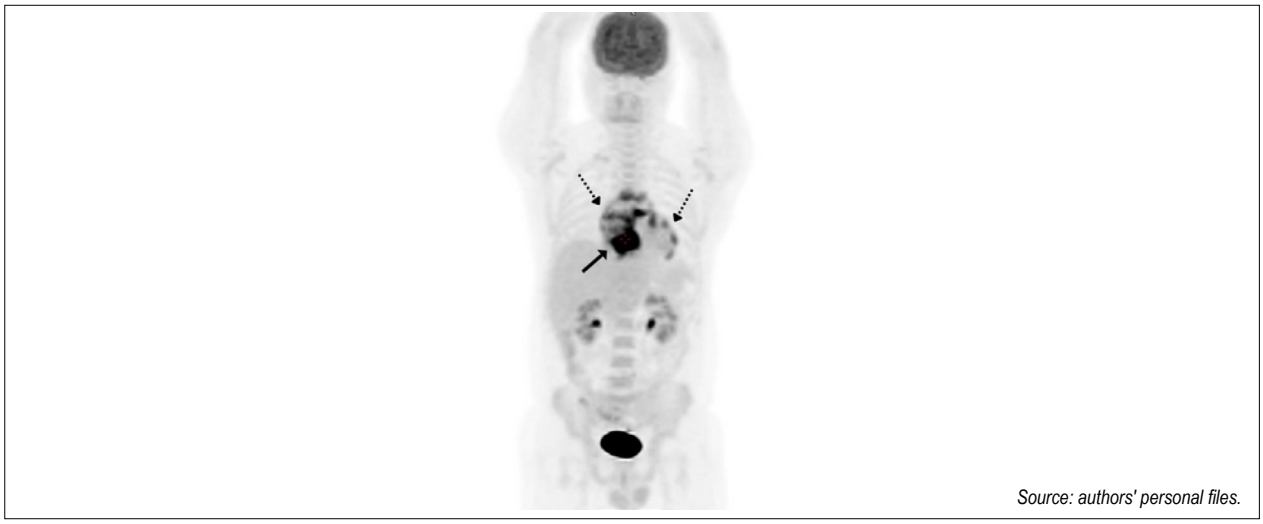
These findings were suggestive of a high-grade primary malignant cardiac tumor. An anatomopathological study revealed that it was a fusocellular neoplasm (Figure 3), and the immunohistochemical study findings were compatible with angiosarcoma, with positive Ki-67 staining in 50% of the neoplastic cells.

Discussion

Primary cardiac angiosarcoma is a clinically rare cardiac neoplasm, with approximately 200 cases described to date[1] and an incidence of approximately 0.017%.[2] It is highly invasive and has a poor prognosis.[1,2] This tumor can occur in any part of the heart, but it most frequently occurs on the right side, especially in the RA, and rarely in the epicardium, pericardium, and RV.[2] It usually causes chest pain, vomiting, cough, hemoptysis, shortness of breath, fatigue, and arrhythmia. It has a high mortality rate owing to the tendency for local recurrence and high incidence of systemic metastases.[1]

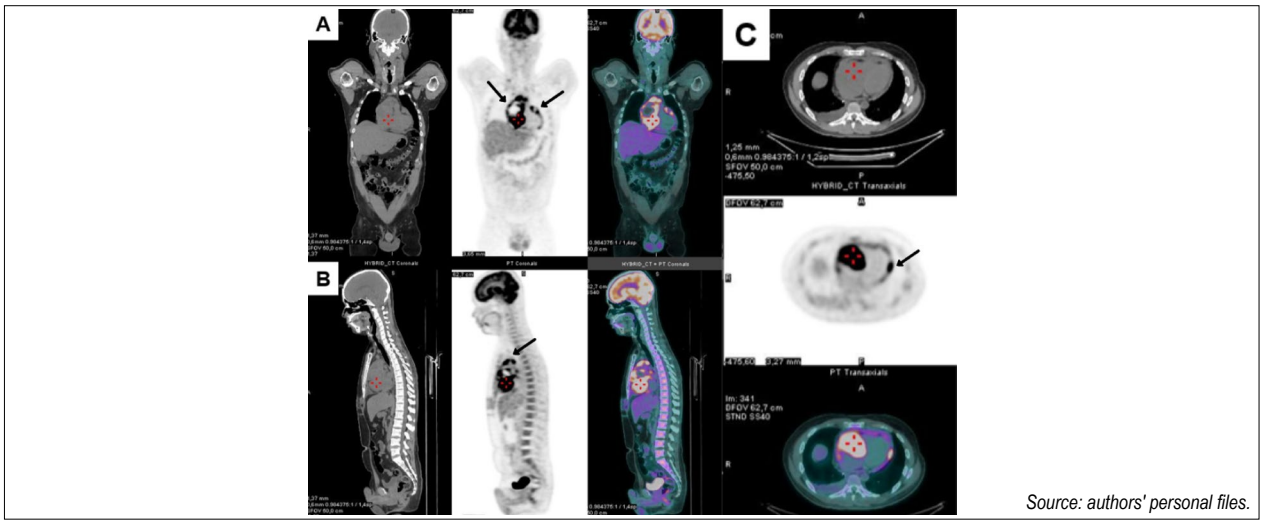
The late diagnosis and rarity of these tumors makes it difficult to define the best treatment and prognostic factors. In addition, they are resistant to radiation and chemotherapy,[2] therefore, surgical resection is currently considered the ideal treatment modality. In the present case, surgical excision was not possible because of the extent of cardiac involvement. One week after the biopsy, the patient experienced new cardiac tamponade, underwent another pericardial window, and on the 3rd day after the procedure died suddenly.

With the development of new imaging techniques, an increasing number of cases are being diagnosed earlier.[3] CT, MRI, and ^{18}F-FDG PET/CT can help determine the extent of infiltration and presence of potential metastases. On contrast-enhanced CT, cardiac angiosarcomas commonly show inhomogeneous centripetal enhancement. Cardiac MRI allows for better characterization of the soft tissues and tumor in addition to being superior to CT in the assessment of myocardial and pericardial infiltration.[2]



Source: authors' personal files.

Figure 1 – *Maximum intensity projection image of positron emission tomography associated with computed tomography with 18-fluorodeoxyglucose showing a marked increase in glycolytic metabolism in the cardiac mass involving the right atrium and ventricle (solid black arrow) with extension to the pericardium (dotted black arrows).*



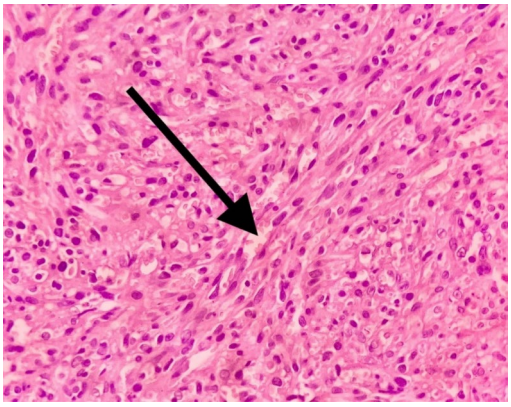
Source: authors' personal files.

Figure 2 – *Positron emission tomography (PET) associated with computed tomography (CT) with 18-fluorodeoxyglucose (18F-FDG) images (A, coronal CT, PET, and PET/CT sections; B, CT, PET, and PET/CT sagittal sections; C, CT, PET, and PET/CT axial sections) showing marked accumulation of 18F-FDG in the tumor (red cross marker) and sparse hypermetabolic foci in the pericardium (black arrows).*

Moreover, ^{18}F-FDG PET/CT has been used to better characterize and stage these tumors, as it aids the accurate determination of the tumor's extent, metabolic activity, and potential metastases, as it is a whole-body examination.[1,4] In addition, it makes it possible to characterize a malignant tumor in a non-invasive way,[4] which is very important for sparing the heart muscle.

The tumor metabolic characterization on ^{18}F-FDG PET/CT is based on the calculation of the SUVmax of FDG on the tumor mass versus the glucose uptake of the blood pool and the normal myocardium, permitting differentiation between hypermetabolic tumors and myocardial physiological uptake. In such a way that uptake is low in the blood pool and normal myocardium, it is higher in benign tumors and significantly higher in malignant primary tumors. Using a SUVmax cutoff of 3.5, the sensitivity in determining malignancy can reach 100%, while at a cutoff of slightly higher than 4.6, the specificity increased to 100%, although sensitivity decreased to 94%. Therefore, the use of an SUVmax cutoff of 3.5–4.6 has good accuracy for the diagnosis of malignant cardiac tumors.[5]

In the present case, ^{18}F-FDG PET/CT clearly demonstrated the extent of the primary tumor and the high uptake of ^{18}F-FDG in the cardiac lesion (SUVmax of 19.6), indicating the malignant nature of the tumor. This diagnostic approach makes it possible to characterize a malignant tumor noninvasively. Here we highlight the potential of ^{18}F-FDG PET/CT for diagnosing and staging primary cardiac angiosarcoma.



Source: authors' personal files.

Figure 3 – *Fusocellular neoplasm composed of cells with moderate atypia that outlines the vascular spaces (black arrow).*

Authors' contributions

Data collection and analysis: Silva PHR, Montenegro CEL, Flamini MEDM, Pontual MVNO and Brandão SCS; Manuscript writing: Silva PHR; Critical review of the article: Montenegro CEL and Brandão SCS, Consent to be responsible for all aspects of the work: Silva PHR, Montenegro CEL, Flamini MEDM, Pontual MVNO and Brandão SCS.

Conflict of interest

The authors have declared that they have no conflict of interest.

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