Clinical note

Findings of the $^{18}$F-FDG PET-CT in a cardiac angiosarcoma complicated by a cardiac rupture

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Abstract

Primary malignant tumors of the heart are a rare condition. The most common type is the cardiac angiosarcoma. The symptoms of this disease are very nonspecific and can be very difficult to diagnose by conventional imaging techniques. We report the case of a male patient with cardiac angiosarcoma who also had a rare complication, this being cardiac rupture, which required the use of $^{18}$F-FDG PET-CT to demonstrate the mass malignancy and to reach a definitive diagnosis.

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Hallazgos de la PET-TC con $^{18}$F-FDG en un angiosarcoma cardíaco complicado con una rotura cardíaca

Resumen

Los tumores malignos primarios del corazón son una patología muy poco frecuente y de ellos, el tipo más frecuente es el angiosarcoma cardíaco. Esta patología tiene una clínica muy inespecífica y puede ser muy difícil de diagnosticar por técnicas convencionales de imagen. Presentamos el caso de un paciente con un angiosarcoma cardíaco que además presentaba una complicación muy poco frecuente, una rotura cardíaca, lo que hizo necesario el uso de la $^{18}$F-FDG PET-TC para demostrar la malignidad de la masa y poder llegar a un diagnóstico definitivo.

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Introduction

Although extremely infrequent the angiosarcoma is the most common cardiac malignant primary tumor. Conventional imaging techniques are usually enough for its diagnosis, but sometimes studies with advanced imaging methods are necessary to reach it. We present one patient with cardiac angiosarcoma who debuted with a cardiac wall rupture that led to misdiagnosis by conventional imaging.

Case report

A 34-year-old sportsman with no relevant familiar history, no toxic habits, and personal history of multiple injuries from falls of bike, biking and climbing was admitted at emergency department with increased heart rate in relation to the exercise. He noticed similar episodes since about six months ago, one of them accompanied by chest pain radiating to the left arm. Also he noticed that he did not have the same physical performance and pain in the left anterior ribs. An initial EKG was consistent with atrial flutter, so the patient was restored to sinus rhythm, after which he remained asymptomatic and hemodynamically stable. After cardioversion, a transthoracic ultrasonography identified a heterogeneous pericardial mass which was communicated with the right atrium (RA), presenting bidirectional blood flow from the RA to the mass. A contrast enhanced thoracic and abdominal CT was performed to characterize this mass. It showed a poorly defined cardiac mass within the pericardium, isodense to myocardium, which involved the RA, right ventricle (RV) and the whole diaphragmatic surface of the heart. After contrast injection the mass showed a heterogeneous enhancement. Due to history of several posttraumatic
Fig. 1. (A) Axial cardiac-MRI FIESTA sequence, double inversion pulse T2-weighted and double inversion pulse T1-weighted images. Anterolateral cardiac wall rupture (black arrow) along with a pericardial mass hyperintense on both T1-weighted and T2-weighted images (white arrow). Signal void within the mass (red arrow) on T1-weighted and T2-weighted images consistent with flow disturbances between the mass and the RA, reported as pericardial hematoma. (B) Axial contrast enhanced cardiac-CT acquired on arterial, venous and delayed phases. Cardiac mass within the pericardium, isodense to myocardium, involving the RA and inferolateral and anterior wall of the RV (white and black arrows). The RA is deformed and displaced by the mass. After contrast injection the mass present a significant global and progressive enhancement. It was reported as a cardiac tumor.

Injuries the first diagnosis was a posttraumatic pericardial hematoma secondary to RA rupture and, less likely, with a pericardial neoplasm infiltrating cardiac structures. A cardiac-MRI was compatible with RA rupture (Fig. 1A). However coronary arteries were not studied by MRI and a coronary-CT was recommended. Despite the previous diagnosis of RA rupture, coronary-CT findings were consistent with a solid cardiac malignancy, infiltrating right side chambers, involving the right coronary artery, and with high and progressive enhancement after contrast administration (Fig. 1B). Because of disagreement between different imaging techniques and in order to plan a proper treatment, an $^{18}$F-FDG PET/CT was requested to establish whether or not the mass had a malignant etiology. After a fasting period of 18 h, $^{18}$F-FDG PET/CT scan was acquired at 45 and 90 min. It showed a very heterogeneous area of enhancement within the cardiac muscle in right sided chambers and multiple hypermetabolic foci with an irregular distribution in the RV and RA (Fig. 2), which were also persistent on delayed phases (Fig. 3). This is a very unusual finding for right sided chambers on...

Fig. 2. Maximum intensity projection PET image (A), coronal PET/CT fused images (B), PET and cardiac-MRI FIESTA fused images, 45 min after $^{18}$F-FDG administration (C). Highly significant $^{18}$F-FDG uptake in the LV reported as physiologic. Heterogeneous $^{18}$F-FDG enhancement in right sided chambers, mainly in RA and inferolateral wall of RV (white arrows). No extracardiac findings were detected.
Among the other Cardiac angiosarcoma...Less frequently...Debut with cardiac wall rupture is an extremely rare...when almost...Transthoracic ultrasound...Focusing on the imaging detection, chest X-ray is...Signs and symptoms are very unspecific...For this reason it is important to know...Dyspnea is the most...Uptake may range from 25% of cases, and more than 2/3 are benign.1–4,6 Among the other malignant primary cardiac tumors, angiosarcoma is the most frequent with a prevalence of 0.001–0.03%.4,8 They typically occur between 30 and 50 years of life, with a male to female ratio of 2–3:1. Although they may arise in every heart chamber, they are mainly located in the RA (80% of cases).4,5 Cardiac angiosarcomas have an aggressive behavior with local infiltration and fast growth, most of them having metastatic dissemination at diagnosis, mainly to the lung.2,4,6 Signs and symptoms are very unspecific and present late in the disease process, when it is on an advanced stage with decreased cardiac function.1–6 Dyspnea is the most frequent symptom, but right-heart failure and pericardial disease are also common due to RA and pericardial involvement.2,4,5,6 Other clinical findings are arrhythmias, pleuritic pain, valvular dysfunction, pericardial effusion or symptoms related to metastatic spread.1,4,5,7 Debut with cardiac wall rupture is an extremely rare event and very few cases has been reported on the literature.1–4,6–8 Cardiac angiosarcoma has a poor prognosis on account of the advanced stage of the disease at diagnosis,1,2,4–8 when almost all tumors (80%) are unresectable with metastatic extension.3–6 Median survival after diagnosis range between 6 and 12 months despite multidisciplinary treatment, which includes chemotherapy, radiotherapy, surgery or even heart transplantation.2,4–8 Imaging studies have a pivotal not only for its early diagnosis but also to establish tumor’s location, size and extent. Nevertheless in most of the cases only histological findings will lead to final benign or malignant characterization. From a radiological point of view, cardiac angiosarcoma is usually found as a mass in the RA. Differential diagnosis includes benign tumors such as atrial myxoma, malignant tumors, and non-tumoral cardiac masses such as thrombus.2,6 Focusing on the imaging detection, chest X-ray is often the initial diagnostic tool. It may show cardiomegaly, which otherwise is an extremely unspecific finding.4,8 Transthoracic ultrasonography sensitivity for cardiac masses is 97%, although it is operator-dependent and is limited for the assessment of tumor infiltration.1–4,6,7 Subsequently CT and MRI are the best imaging methods to evaluate cardiac masses. They reveal anatomic details beyond ultrasonography capability, demonstrate infiltration of myocardium, pericardium and adjacent tissues and allow detection of metastatic spread. However, there are cases as the one we present whose diagnosis of angiosarcoma is not well established by the methods above described, requiring further studies before any invasive procedure. Few cases have been reported about the role of 18F-FDG PET/CT in patients with cardiac angiosarcoma, although it seems to be useful in cases with unclear diagnosis by other imaging techniques.8 For this reason it is important to know the differences in glucose uptake between physiologic and pathologic myocardium. In normal conditions glucose is the main energy supply for the myocardium; however, after a fasting period of 18 h, its metabolism is modified and glucose is changed for free fatty acids. Despite of this fact there is still a 30–40% of the myocardium function supported by glucose that explain why even in resting conditions it shows 18F-FDG uptake.5,9,10 Uptake may range from almost non-existent to a highly significant one that can lead us to misdiagnosis of tumoral activity.5,9,10 Normally glucose enhancement is only homogenously identified in the LV.5,10 Less frequently it can be detected on the RV, mainly in patients suffering COPD or RV insufficiency, but it is extremely rare in the atriums.9 Therefore on account that 80% of cardiac angiosarcomas arise in the atriums, atrial 18F-FDG uptake is a strong predictor for malignancy, more even when it presents as an irregular enhancement. In addition when malignancy has been proved, 18F-FDG PET/CT scan also allows the staging workup within the same imaging acquisition.

Discussion

Malignant heart diseases are rare, with a prevalence estimated at 0.3%, and the majority of them are secondary to metastatic neoplasms involving the heart. Primary cardiac tumors represent only 25% of cases, and more than 2/3 are benign.1–4 Among the other malignant primary cardiac tumors, angiosarcoma is the most frequent with a prevalence of 0.001–0.03%.4,8 They typically occur between 30 and 50 years of life, with a male to female ratio of 2–3:1. Although they may arise in every heart chamber, they are mainly located in the RA (80% of cases).4,5 Cardiac angiosarcomas have an aggressive behavior with local infiltration and fast growth, most of them having metastatic dissemination at diagnosis, mainly to the lung.2,4,6 Signs and symptoms are very unspecific and present late in the disease process, when it is on an advanced stage with decreased cardiac function.1–6 Dyspnea is the most frequent symptom, but right-heart failure and pericardial disease are also common due to RA and pericardial involvement.2,4,5,6 Other clinical findings are arrhythmias, pleuritic pain, valvular dysfunction, pericardial effusion or symptoms related to metastatic spread.1,4,5,7 Debut with cardiac wall rupture is an extremely rare event and very few cases has been reported on the literature.1–4,6–8 Cardiac angiosarcoma has a poor prognosis on account of the advanced stage of the disease at diagnosis,1,2,4–8 when almost all tumors (80%) are unresectable with metastatic extension.3–6 Median survival after diagnosis range between 6 and 12 months despite multidisciplinary treatment, which includes chemotherapy, radiotherapy, surgery or even heart transplantation.2,4–8 Imaging studies have a pivotal not only for its early diagnosis but also to establish tumor’s location, size and extent. Nevertheless in most of the cases only histological findings will lead to final benign or malignant characterization. From a radiological point of view, cardiac angiosarcoma is usually found as a mass in the RA. Differential diagnosis includes benign tumors such as atrial myxoma, malignant tumors, and non-tumoral cardiac masses such as thrombus.2,6 Focusing on the imaging detection, chest X-ray is often the initial diagnostic tool. It may show cardiomegaly, which otherwise is an extremely unspecific finding.4,8 Transthoracic ultrasonography sensitivity for cardiac masses is 97%, although it is operator-dependent and is limited for the assessment of tumor infiltration.1–4,6,7 Subsequently CT and MRI are the best imaging methods to evaluate cardiac masses. They reveal anatomic details beyond ultrasonography capability, demonstrate infiltration of myocardium, pericardium and adjacent tissues and allow detection of metastatic spread. However, there are cases as the one we present whose diagnosis of angiosarcoma is not well established by the methods above described, requiring further studies before any invasive procedure. Few cases have been reported about the role of 18F-FDG PET/CT in patients with cardiac angiosarcoma, although it seems to be useful in cases with unclear diagnosis by other imaging techniques.8 For this reason it is important to know the differences in glucose uptake between physiologic and pathologic myocardium. In normal conditions glucose is the main energy supply for the myocardium; however, after a fasting period of 18 h, its metabolism is modified and glucose is changed for free fatty acids. Despite of this fact there is still a 30–40% of the myocardium function supported by glucose that explain why even in resting conditions it shows 18F-FDG uptake.5,9,10 Uptake may range from almost non-existent to a highly significant one that can lead us to misdiagnosis of tumoral activity.5,9,10 Normally glucose enhancement is only homogenously identified in the LV.5,10 Less frequently it can be detected on the RV, mainly in patients suffering COPD or RV insufficiency, but it is extremely rare in the atriums.9 Therefore on account that 80% of cardiac angiosarcomas arise in the atriums, atrial 18F-FDG uptake is a strong predictor for malignancy, more even when it presents as an irregular enhancement. In addition when malignancy has been proved, 18F-FDG PET/CT scan also allows the staging workup within the same imaging acquisition.

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