Case Report
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^{18}F-FDG PET-CT for Evaluation of Cardiac Angiosarcoma: A Case Report and Review of Literature

Kardiyak Anjiosarkomun İncelenmesi için ^{18}F-FDG PET-BT: Bir Olgu Sunumu ve Literatür Derlemesi

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Abstract

Cardiac angiosarcomas are rare neoplasms. We here present the case of a 24 year old male with a cardiac mass which was characterised as malignant on ^{18}F-Fluorodeoxyglucose (^{18}F-FDG) positron emission tomography-computed tomography (PET-CT). In addition ^{18}F-FDG PET-CT also demonstrated pericardial infiltration and bone metastases. The tumor was confirmed to be angiosarcoma on biopsy and palliative chemotherapy was started. Here we have highlighted the potential role of ^{18}F-FDG PET-CT in patients with cardiac angiosarcoma and presented a brief review.
Key Words: Hemangiosarcoma, heart, PET Scan, ^{18}F-FDG, bone, metastasis

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Özet

Kardiyak anjiyosarkomlar nadir neoplazilerdir. Bu makalede ^{18}F-Florodeoksiglukoz (^{18}F-FDG) pozitron emisyon tomografi-bilgisayarlı tomografi (PET-BT) ile malin olduğu saptanan kardiyak kitlesi olan 24 yaşında bir olguyu sunduk. ^{18}F-FDG PET-BT aynı zamanda perikardiyal infiltrasyon ve kemik metastazını da gösterdi. Tümör, biyopsi sonucunda anjiyosarkom olarak doğrulandı ve palyatif kemoterapiye başlandı. Bu makalede kardiyak anjiyosarkomlu hastalarda ^{18}F-FDG PET-BT'nin potansiyel rolünü vurguladık ve kısa bir derleme sunduk.
Anahtar Kelimeler: Kardiyak anjiyosarkom, PET,^{18}F-FDG, Kemik metastazı

Çıkar Çatışması: Yazarlar bu makale ile ilgili olarak herhangi bir çıkar çatışması bildirmemiştir.

Introduction

Primary cardiac tumors are rare, with an incidence ranging from 0.001% to 0.028% in autopsy reports. About one fourth of these tumors are malignant, with angiosarcoma being the most common malignant cardiac tumor (1,2). Cardiac angiosarcomas are neoplasms of mesenchymal cells. Only 200 cases have been described in the literature (3). These tumors are resistant to radiation and chemotherapeutic agents, therefore, surgical resection remains the treatment of choice. But the diagnosis is often delayed as the symptoms are usually non-specific. The tumor has a high mortality rate as it has a tendency for local relapse along with a high incidence of systemic

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metastases. Survival may range from a few days to years (4,5). As cardiac angiosarcomas are highly malignant tumors with poor prognosis, early diagnosis is mandatory. Therefore, it is important to determine the exact extent of the primary lesion, to detect local recurrence and distant metastases for appropriate therapy management (6). Here, we have aimed to report the potential benefits of ^{18}F-Fluorodeoxyglucose (^{18}F-FDG) positron emission tomography-computed tomography (PET-CT) imaging for examination of malignant potential and exploration of distant metastases in a young patient of cardiac angiosarcoma. In addition we have reviewed the published literature regarding the utility of ^{18}F-FDG PET-CT in cardiac angiosarcoma.

Case Report

A 24 year old male patient was admitted to the emergency department with progressive dyspnoea. Patient had tachycardia (PR: 100/minute) and low blood pressure (100/60 mmHg) at presentation. The intensity of the heart sounds was reduced. All other physical examinations were normal. On chest X-ray a massive cardiomegaly was seen. Transthoracic echocardiography showed a large hypoechoic ill-defined cardiac mass. Subsequently patient underwent contrast enhanced CT that showed a large mass draping the cardiac root and ascending aorta, left ventricular outflow tract, right ventricular outflow tract, infundibulum and the basal interventricular septum (Figure 1a, 1b). Patient was then referred to our department for ^{18}F-FDG PET-CT for further characterisation of the cardiac mass as well as for metastatic work up. The patient fasted overnight. Blood glucose level was 96 mg/dl. A dose of 370 MBq of ^{18}F-FDG was injected intravenously. PET-CT acquisition was done after a 45 minute uptake period. 50 ml of non ionic iodinated intravenous contrast (Visipaque, 320 mg I/ml, GE) was administered and scan was acquired after a delay of 50 seconds. Contrast enhanced 8F-FDG PET-CT revealed a large ill defined non enhancing soft tissue density mass (single largest dimension 8.1 cm) in the interventricular septum (proximal two-third) extending into the right ventricle upto right ventricle outflow tract, the aortic root and the right A-V valve with a small extension along the left ventricular wall with increased ^{18}F-FDG uptake (SUVmax-8.3) (Figure 1c-1f). Pericardial and minimal right pleural effusion were noted. Pericardial effusion was loculated mainly on the right side and the inferior surface of the heart (Figure 2a). Also, increased ^{18}F-FDG uptake was seen in the pericardium, likely due to pericardial infiltration by the tumor (SUVmax-5.1) (Figure 2b, 2c). The SUVmax of the normal myocardium was 1.5. Apart from the cardiac findings, there was a sclerotic lesion in left iliac bone with increased ^{18}F-FDG uptake (SUVmax-4.5) (Figure 2d, 2e). Another focus of increased ^{18}F-FDG uptake was noted in left ala of sacrum (SUVmax-3.7) with minimal sclerosis on CT (Figure 2f, 2g). ^{18}F-FDG PET-CT findings were suspicious for a cardiac tumor with skeletal metastases. Biopsy from the cardiac lesion was performed which showed spindle cell neoplasm with CD-31 positivity, suggesting angiosarcoma.

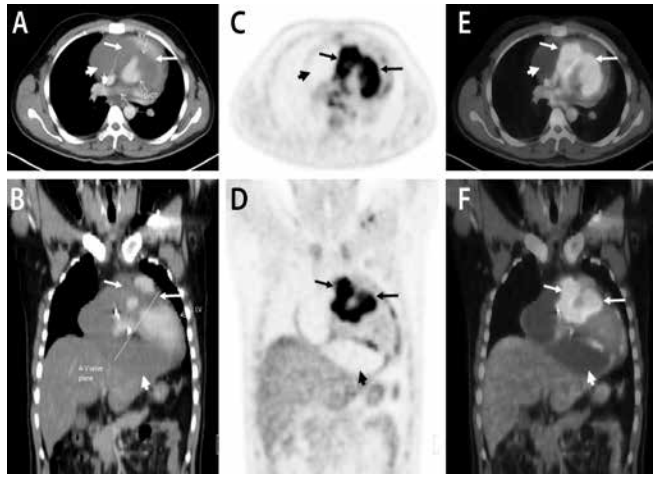

Figure 1. Contrast enhanced CT (Transaxial A, Coronal B) revealed a large ill defined non enhancing soft tissue density mass (bold arrows) in the interventricular septum (proximal two-third) extending into the right ventricle upto right ventricle outflow tract, the aortic root and the right A-V valve with a small extension along the left ventricular wall. ^{18}F-FDG PET (Transaxial C, Coronal D) and contrast enhanced ^{18}F-FDG PET-CT (Transaxial E, Coronal F) shows increased ^{18}F-FDG uptake in the soft tissue mass seen on contrast enhanced CT (arrows, SUVmax-8.3). Also noted is loculated pericardial effusion without significant ^{18}F-FDG uptake on ^{18}F-FDG PET (arrowheads). RV-right ventricle; LV-left ventricle; RVOT-RV outflow tract; LVOT-LV outflow tract; A-V-atrio-ventricular.

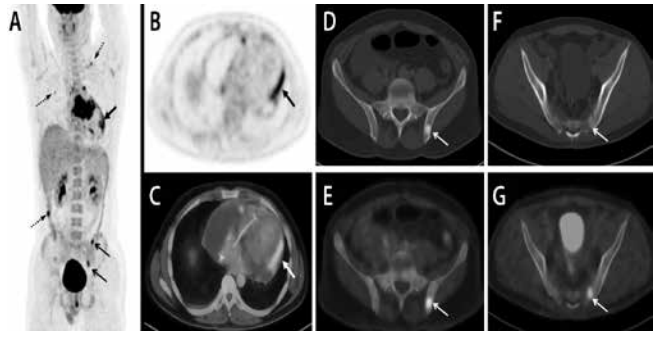

Figure 2. Apart from the primary tumor, PET Maximum Intensity Projection (MIP) image shows increased ^{18}F-FDG uptake along the left lateral border of the heart (A, bold arrow) along with two foci of increased ^{18}F-FDG uptake in left hemipelvis (arrows). Other foci of benign FDG uptake (broken arrows) on MIP image were seen in left supraclavicular region (central venous line), right hemithorax (right lung infection) and ascending colon (normal colonoscopy).Transaxial ^{18}F-FDG PET (B) and contrast enhanced ^{18}F-FDG PET-CT (C) show moderate pericardial effusion with increased ^{18}F-FDG uptake in pericardium (arrow) likely infiltration (SUVmax-5.1). Minimal right pleural effusion is also noted. Transaxial CT (D) and ^{18}F-FDG PET-CT (E) show a sclerotic lesion in left iliac bone (arrow) with increased ^{18}F-FDG uptake (SUVmax-4.5). Transaxial CT (F) and ^{18}F-FDG PET-CT (G) show another focus of increased ^{18}F-FDG uptake in left ala of sacrum (SUVmax-3.7) with minimal sclerosis on CT (arrow).

Table 1. Review of literature-^{18}F-FDG PET/PET-CT for primary cardiac angiosarcoma reported in the English literature

SL	Author	Age/ Sex	Symptoms	Conventional Imaging	^{18}F-FDG PET/PET-CT findings		Treatment	Follow up
					Primary tumor	Metastasis		
1.	Freudenberg et al. (2002)	17/M	Queasiness, dyspnoea	MRI & CT-Mass in the RA & ? lung metastasis	RA mass (SUVmax-7.5)	None	Sx+Ct	_
2.	Hoffmeier et al. (2005)	8/M	Dyspnoea	MRI-Mass almost totally obstructing the left ventricular cavum with infiltration of the myocardium.	LV mass (SUVmax-5.4)	None	Cardiac transplantation	Symptomatic improvement and no recurrence till 6 months
3.	Hori et al. (2007)		Dyspnoea, facial swelling	CT-RA mass	RA mass (SUVmax-9.9)	None	Sx+Ct	No recurrence
4.	Juergens et al. (2007)	62 /F	_	MRI- LA mass	LA mass, that infiltrated LV, mitral valve annulus, and the great coronary (SUVmax-5.4)	None	Sx +Ct	20 months; F/U PET-CT showed local recurrence with lung metastasis
5.	Nakamura-Horigome et al. (2008)	49/M	Cardiac tamponade	CT- RA mass with thickening of the RV wall	Increased ^{18}F-FDG uptake in the mass	None	Ct+RT	12 months; No progression or metastasis
6.	Higashiyama et al. (2009)	60/F	Dyspnoea	CT- Inhomogeneously enhancing RA mass with RV infiltration	RA mass (SUVmax-5.6)	Mediastinal node	Sx	6 months; Died
7.	Büyükşirin et al. (2009)	61/M	Cough, hemoptysis, chest pain	CT-RA mass with b/l lung parenchymal ground glassing	Increased ^{18}F-FDG uptake in the mass; Uptake in lung parenchyma not high	None	Sx	Died of respiratory failure
8.	Kadota et al. (2010)	49/F	Epigastric discomfort, Cardiac tamponade	CT & MRI-Two masses in the anterior RA & pericardial space	RA mass (SUVmax-9.9)	None	Sx	Day 34-admitted with cerebral haemorrhage-suspected metastasis; Day 79-Died
9.	Ak et al. (2011)	55/F	_	MRI-Preoperative; RA mass with extension to adjacent pericardium; Pericardial effusion	Postoperative; Recurrent RA mass (SUVmax-16.4)	None	Ct	_
10.	Bouma et al. (2011)	50/F	Dyspnoea, chest and shoulder pain	CT & MRI-Large RA mass extending into the wall of the SVC	RA mass with intense ^{18}F-FDG uptake	None	Sx	5 months; bone and liver metastasis; started on Ct-RT
11.	Shao et al. (2011)	-	-	CT- RA mass; misdiagnosed as myxoma	RA mass (SUVmax-5.5)	None	Sx	_

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SL	Author	Age/ Sex	Symptoms	Conventional Imaging	^{18}F-FDG PET/PET-CT findings		Treatment	Follow up
					Primary tumor	Metastasis		
12.	Rahbar et al. (2012) (n=6)	61/F	_	CT-Mass involving LA & LV; Malignant	Mass in LA & LV (SUVmax-5.3)	None	_	_
		48/F	_	CT-RA mass; Malignant	RA mass (SUVmax-7.6)	Lung, Bone	_	_
		64/F	_	CT-mass involving RA & RV; Malignant	Mass involving RA & RV (SUVmax-9.8)	None	_	_
		51 /F		CT-RV mass; Likely malignant	RV mass (SUVmax-10.7)	None	_	_
		74 /F	_	CT-RA mass involving SVC & IVC; Malignant	RA mass involving SVC & IVC (SUVmax-10.2)	Liver, lung, bone	_	_
		41/F	_	CT-RA mass; Malignant	RA mass (SUV max-6.8)	None	_	_
13.	Bilski et al. (2012)	30 /M	Precordial pain	CT-Preoperative; RA mass	Postoperative; RA mass (SUVmax-10.3); Concomitant pericardial effusion	Bone	Sx+Ct	_
14	Present case (2013)	24/M	Dyspnoea	CT-proximal IVS mass extending into the RV upto RVOT and the right A-V valve with a small extension along the LV wall	Mass in the IVS (proximal two-third) extending into the RV upto RVOT and right A-V valve (SUVmax-8.3); pericardial effusion	Bone	Ct	5 months; significant symptomatic improvement; continuing Ct

AA-Ascending Aorta; b/l-Bilateral; Ct-Chemotherapy; F/U-Follow up; IVC-Inferior Vena Cava; IVS-Inter ventricular septum; LA-Left atria; LV-Left ventricle; LVOT-Left ventricle outflow tract; RA-Right atria; RT-Radiotherapy; RCA-Right coronary artery; RV-Right ventricle; RVOT-Right ventricle outflow tract; SVC-Superior vena cava; Sx-Surgery

Hence, the diagnosis of cardiac angiosarcoma with skeletal metastases was reached. Patient is undergoing multiagent chemotherapy and has shown significant clinical improvement at 5 months of follow up.

Literature Review and Discussion

Early diagnosis and prompt management is crucial for the survival of the patients with cardiac angiosarcoma. With the availability of various non-invasive and advanced diagnostic tools, an early diagnosis of this rare lesion is possible (7). Although a positive correlation has been found between ^{18}F-FDG accumulation and the degree of malignancy for many tumors, high ^{18}F-FDG uptake in myocardium does not necessarily mean a malignant lesion. The degree and extent of ^{18}F-FDG myocardial activity may be heterogeneous and variable. Patients with myocardial ischemia, coronary artery disease, atherosclerotic plaques, etc may have a focal increased ^{18}F-FDG uptake (8,9). Moreover, respiratory motion can sometimes lead to inhomogeneties in myocardial FDG uptake, more so in the lateral and anterior regions and to a lesser extent in the septal region (10).

^{18}F-FDG PET-CT has been evaluated for characterisation, staging and restaging of cardiac angiosarcomas. A brief review of literature in this regard is presented in Table (1 1,12,13,14,15,16,17,18,19,20,21,22,23). Rahbar et al. in their study on 24 patients with cardiac tumors (including 6 angiosarcomas) concluded that with a cut-off SUVmax of 3.5, ^{18}F-FDG PET-CT could be used to noninvasively determine malignant tumors with a sensitivity of 100% (22). In the present case too, the ^{18}F-FDG uptake was high in the cardiac lesion (SUVmax-8.3), which pointed towards the malignant nature which was confirmed on biopsy.

^{18}F-FDG PET-CT is widely employed for staging of various tumors. Being a whole body imaging modality, ^{18}F-FDG PET-CT is useful for the detection of distant metastases which may be missed on routine conventional imaging modalities. ^{18}F-FDG PET-CT has been sparsely used for staging of cardiac angiosarcoma, given the rarity of such tumors (Table 1). In such patients ^{18}F-FDG PET-CT can accurately determine the extent of the primary tumour as well as demonstrate distant metastases, if present. In the present case too, ^{18}F-FDG PET-CT clearly demonstrated the extent of the primary tumor. In addition the patient had skeletal metastases which were detected on ^{18}F-FDG PET-CT. In early stages without distant metastases, surgery followed by postoperative chemotherapy remains the treatment of choice while palliative chemotherapy with/without cytoreductive surgery is the pathway of management in patients with metastatic disease (11,24). As our patient had metastatic disease on PET-CT, he is undergoing chemotherapy and showed symptomatic improvement. It is to be remembered that the prognosis of metastatic cardiac angiosarcoma remains poor even with chemotherapy and thus ^{18}F-FDG PET-CT can categorise such patients into prognostic groups.

Thus, the present case and the published literature show the potential of ^{18}F-FDG PET-CT in management of patients with cardiac angiosarcoma starting from helping in reaching a diagnosis to staging to restaging and possibly prognostication. However, false positive causes should be kept in mind.

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