Interesting Image
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^{18}F-FDG PET/CT Imaging for Treatment Response Assessment of Cardiac Primitive Neuroectodermal Tumor

Kardiyak Primitif Nöroektodermal Tümörde Tedavi Yanıtı Değerlendirmede ^{18}F-FDG PET/BT

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Abstract

Primitive neuroectodermal tumors (PNETs) are rare and aggressive members of the small round cell carcinoma family. Generally, PNETs are classified into two main groups: PNETs of the central nervous system and PNETs of the peripheral nervous system. Herein, we report the therapy response assessment of a rare case of isolated cardiac PNET using ^{18}F-fluorodeoxyglucose (^{18}F-FDG) positron emission tomography/computed tomography (PET/CT) imaging. Given that physiological cardiac FDG uptake is typically observed, assessing FDG avid lesions in the myocardium presents a challenge for FDG PET/CT. This case holds significance because of the rarity of the disease and the challenging nature of the site for FDG PET/CT imaging.

Keywords: Primitive, neuroectodermal, cardiac, ^{18}F-fluorodeoxyglucose, positron, emission, tomography, treatment, response

Öz

Primitif nöroektodermal tümörler (PNET) küçük yuvarlak hücreli tümörler ailesinin nadir görülen ve agresif bir üyesidir. PNET'ler genellikle santral sinir sisteminin PNET'leri ve periferik sinir sisteminin PNET'leri olmak üzere iki ana grupta değerlendirilmektedir. Burada, nadir görülen bir izole kardiyak PNET olgusunda tedavi yanıtının ^{18}F-FDG pozitron emisyon tomografisi/bilgisayarlı tomografi (PET/BT) ile değerlendirmesi sunulmuştur. Fizyolojik kardiyak FDG tutulumu göz önünde bulundurulduğunda, miyokarddaki FDG tutan lezyonların değerlendirilmesi FDG PET/BT için zorluk oluşturmaktadır. Bu olgu, hastalığın nadirliği ve FDG PET/BT ile değerlendirilmesi için zorlu doğası nedeniyle önem taşımaktadır.

Anahtar kelimeler: Primitif, nöroektodermal, kardiyak, ^{18}F-fluorodeoksiglikoz, pozitron, emisyon, tomografi, tedavi, yanıtı

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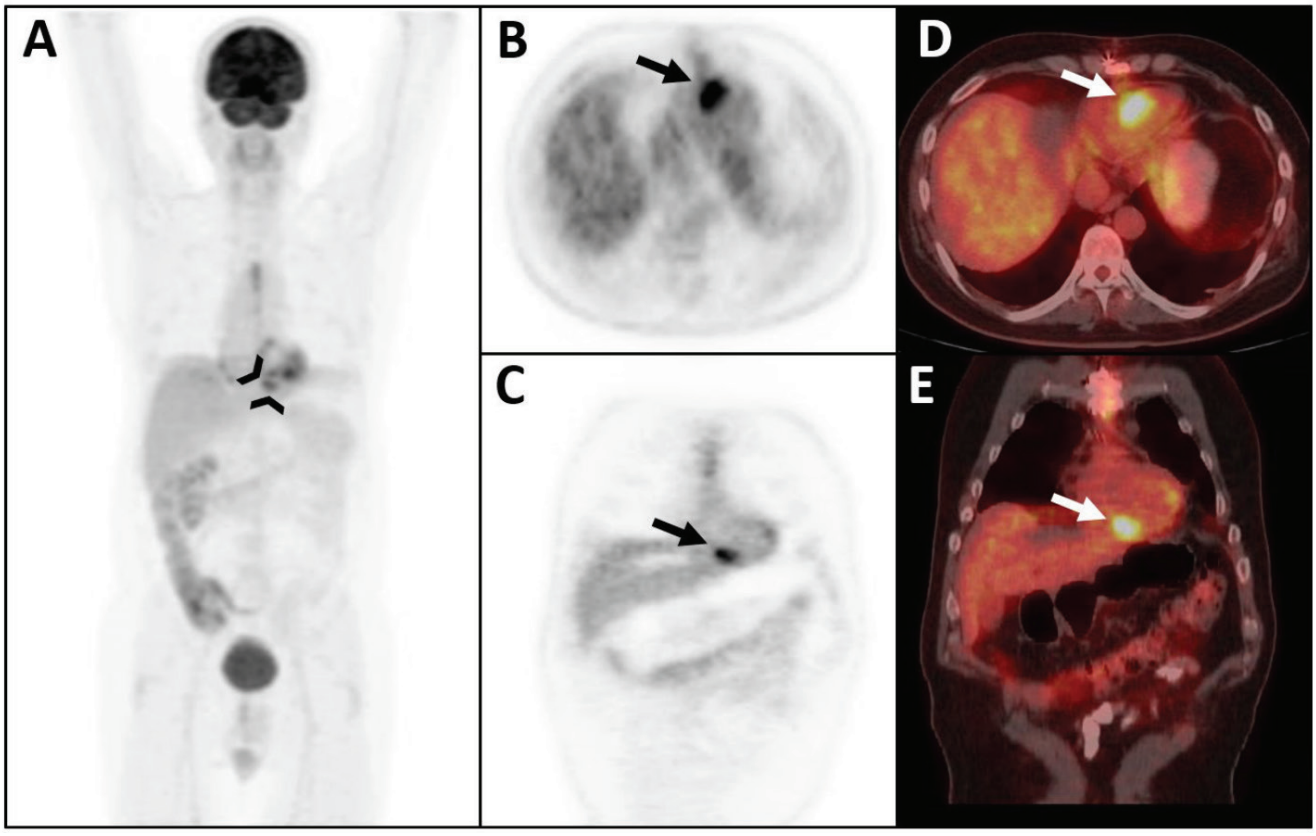

Figure 1. A 45-year-old man with a history of orchiectomy 21 years ago due to yolk sac tumor underwent thoracoabdominal computed tomography (CT) for routine follow-up. A left adrenal mass and a cardiac soft tissue lesion in the right ventricle were detected. After that, the adrenal mass and cardiac lesion were excised surgically. Pathology report of the adrenal mass was consistent with mature teratoma, but the cardiac lesion was diagnosed as primitive neuroectodermal tumor (PNET). One month after surgery, ^{18}F-fluorodeoxyglucose (^{18}F-FDG) positron emission tomography (PET)/CT was performed to evaluate disease status. Maximum intensity projection (MIP) image (A, black arrowheads), axial and coronal PET (B, C, black arrows), and PET/CT fusion (D, E, white arrows) images of PET/CT scan revealed increased FDG uptake in the right ventricle area, previous surgery site, without any marked pathology on CT component of the study, which was interpreted as probable residual malignant disease, indicating that surgery with R0 margin safety cannot be guaranteed in such surgical excision of cardiac tumoral masses. Moreover, it is not usual to observe physiologic FDG uptake in the right ventricle in particular where no physiologic FDG uptake is not seen in the left ventricle or elsewhere in the heart, unless there is an underlying clinical situation of other etiologies such as right ventricular hyperthrophia and cardiac failure, which are not present in this patient. There was no other abnormal FDG uptake in the rest of the body in favor of metastasis.

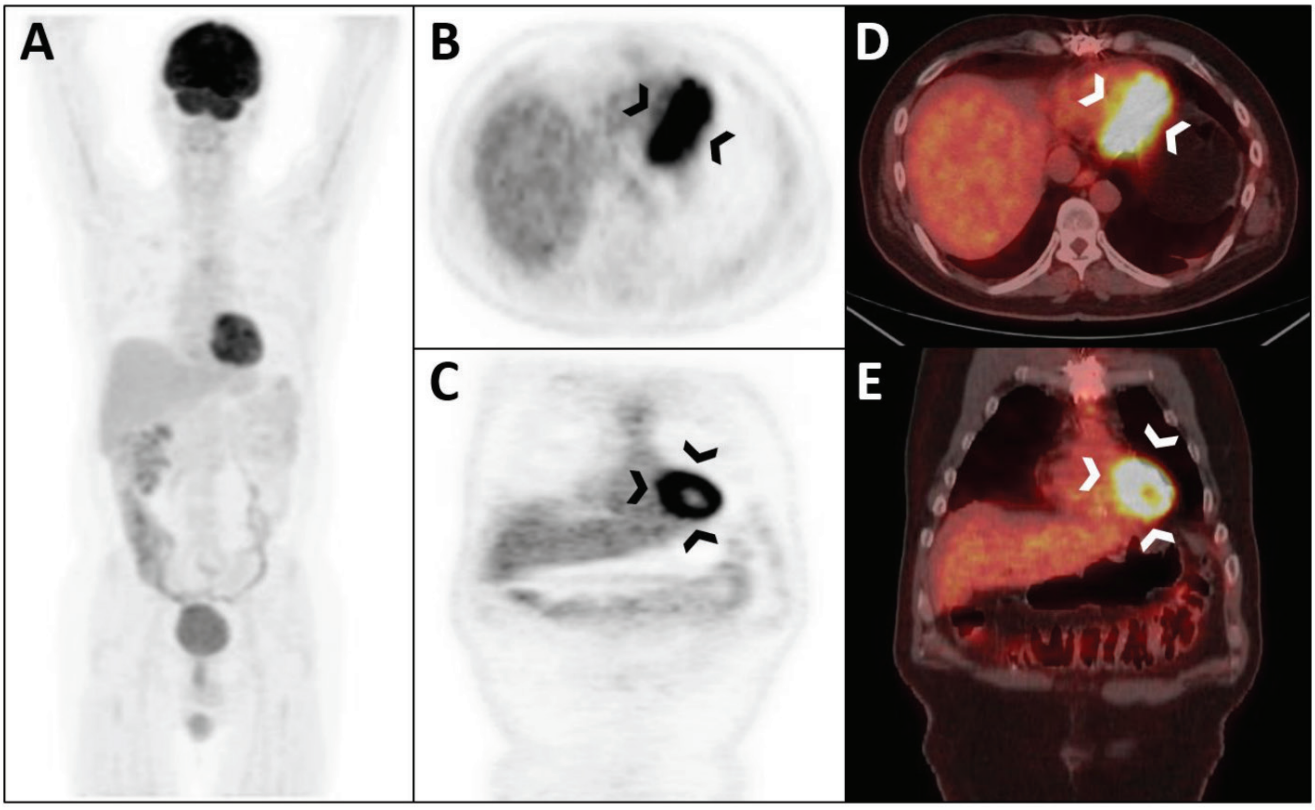

Figure 2. The patient was treated with chemotherapy after detection of residual malignant disease. Following chemotherapy, another [18]F-FDG PET/CT scan was performed to evaluate the therapy response. MIP image (A), axial and coronal PET (B, C), and PET/CT fusion (D, E) images of the second scan revealed no abnormal FDG activity in the right ventricle. However, unlike the first scan, there was physiological FDG uptake in the left ventricle (arrowheads). PNETs are rare and aggressive members of the small round cell carcinoma family and primarily stem from neural crest cells (1). When mentioning PNETs, the distinction between PNETs of the central nervous system (cPNETs) and PNETs of the peripheral nervous system (pPNETs) must be considered first, since they are fairly different from each other both clinically and pathologically (2). The term cPNETs no longer exists in the World Health Organization classification of tumors of the central nervous system as it was removed from the 4th edition which was publihed in 2016 (3). pPNETs and Ewing's sarcoma are considered to be on two edges of the neuroectodermal differentiation spectrum, with pPNETs on the most differentiated side and Ewing's sarcoma on the least differentiated side (4). A limited number of cases of demonstration of pPNETs occurring at different sites of the body by FDG PET/CT, generally with high FDG avidity, have been reported in the literature (5,6,7,8,9). Considering the rarity of pPNETs, cardiac pPNETs, especially isolated cardiac pPNETs, are even rarer. Only a few cases of isolated cardiac pPNETs have been reported in the literature (10,11,12); among them, only one case demonstrated the disease with FDG PET/CT (10). In this case, both the detection of probable residual disease and the therapy response of an isolated cardiac pPNET was demonstrated by FDG PET/CT imaging. Because physiological cardiac FDG uptake is usually observed, evaluation of the FDG avid lesions of the myocardium is challenging for FDG PET/CT (13). However, although FDG uptake is usually seen in the left ventricle, owing to relatively high muscle content and contractile activity, it is not common to observe physiological cardiac FDG uptake solely in the right ventricle, unless there is an underlying disease status affecting the right ventricle, such as in our case (14). Therefore, this case is quite important for such a rare disease in a challenging site for FDG PET/CT imaging.

Ethics

Informed Consent: Since the information provided is anonymous, obtaining informed consent from the patients was deemed not required.

Authorship Contributions

Surgical and Medical Practices: M.E.M., M.F.B., Concept: M.E.M., M.F.B., Design: M.E.M., M.F.B., Data Collection or Processing: M.E.M., M.F.B., Analysis or Interpretation: M.E.M., M.F.B., Literature Search: M.E.M., M.F.B., Writing: M.E.M., M.F.B.

Conflict of Interest: No conflicts of interest were declared by the authors.

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