^{18}F-Fluorodeoxyglucose Positron Emission Tomography-Computed Tomography in Response Assessment of Perivascular Epithelioid Cell Tumor of the Pelvic Cavity to Irinotecan and Temozolomide

Abstract

Perivascular epithelioid cell tumors (PEComas) are a rare variety of mesenchymal tumors composed of distinctive cells that show a focal association with blood vessel walls and usually express melanocytic and smooth muscle markers. We present a case of 38-year-old male, diagnosed with PEComa of the pelvic cavity who underwent serial ^{18}F-fluorodeoxyglucose positron emission tomography-computed tomography scans for the assessment of response to the chemotherapeutic combination of irinotecan and temozolomide.

Keywords: *Fluorodeoxyglucose, perivascular epithelioid cell tumor, positron emission tomography-computed tomography*

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A 38-year-old male presented to urology outpatient department with chief complaints of increase in the frequency of urination and stools. He was referred for contrast enhanced computed tomography of the abdomen and pelvis that revealed a large heterogeneous mass lesion with areas of internal necrosis and heterogeneous contrast enhancement measuring ~ 10.3 cm × 9.5 cm × 11.8 cm in the pelvic cavity. For further evaluation, he was advised to undergo ^{18}F-fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT) scan that showed increased FDG uptake (maximum standardized uptake value [SUVmax] ~ 16.7) in the mass lesion of the pelvic cavity [Figure 1a-f]. Biopsy of the mass was done that showed cellular tumor composed of cells arranged in sheets and in vague nesting pattern. On immunohistochemistry, tumor cells were found to be positive for spinal muscular atrophy, HMB45 (focal), Melan A (diffuse), TFE3 (focal), S100 (focal), estrogen receptor, progesterone receptor, CD56, and calponin (focal) while negative for desmin, myogenin, synaptophysin, SOX10, WT1, chromogranin A, and pan CK. Immunostaining for INI 1, BRG1, and H3K27me3 showed retained nuclear expression in the tumor cells. MIB-1 labeling index was approximately 20%, and the histopathological diagnosis was made of aggressive perivascular epitheloid cell tumor (PEComa). Then, the patient underwent six cycles of combined chemotherapeutic regime comprising irinotecan and temozolomide. Follow-up ^{18}F-FDG PET-CT after 6 months showed a significant interval reduction in size and FDG uptake (SUVmax ~ 10.1) of the mass lesion measuring ~ 5.1 cm × 5.2 × cm × 4.4 cm [Figure 2a-f].

PEComa is driven by tuberous sclerosis complex gene mutation causing upregulation of mechanistic target of rapamycin pathway which controls multiple cellular processes, including GLUT1 function, so high FDG uptake in malignant PEComa could reflect over activation of mTOR pathway and is useful for staging/restaging and response assessment to chemotherapeutic agents in oncology practice.[1] Contrast-enhanced CT shows heterogeneous enhancement, whereas magnetic resonance imaging images reveal characteristic masses that are isointense on T1-weighted image and heterogeneously hyperintense on T2-weighted image.[2] ^{18}F-FDG PET-CT can be of useful value in differentiating between benign and aggressive variety of PEComas as aggressive varieties have traditionally

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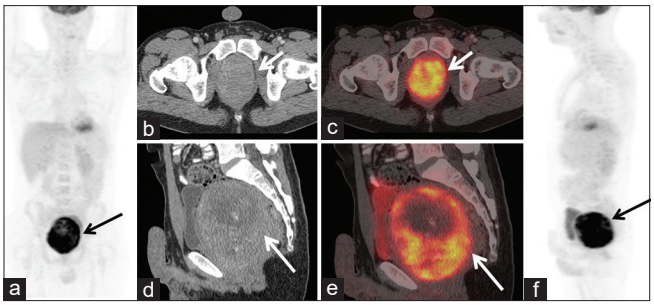

Figure 1: (a) Maximum intensity projection image of fluorodeoxyglucose positron emission tomography-computed tomography showing an enlarged area of fluorodeoxyglucose uptake in the pelvis also seen in the sagittal section maximum intensity projection (f). (b) Axial computed tomography section of the pelvic cavity showing heterogeneous mass lesion showing increased fluorodeoxyglucose uptake in the fused axial positron emission tomography-computed tomography (c). (d) Sagittal section computed tomography of the pelvis showing heterogeneous mass showing increased fluorodeoxyglucose uptake in the fused positron emission tomography-computed tomography image (e)

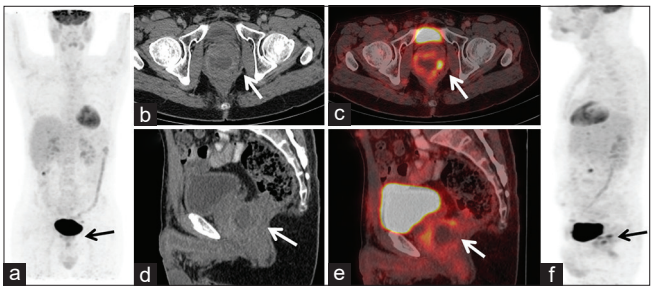

Figure 2: (a) Maximum intensity projection of follow-up fluorodeoxyglucose positron emission tomography-computed tomography scan after 6 months showing faint area of radiotracer concentration in the pelvis also seen in sagittal section maximum intensity projection (f, black arrow). (b) Axial section computed tomography of the pelvis showing heterogeneous mass lesion with predominant necrosis and fluorodeoxyglucose uptake in the periphery in fused positron emission tomography-computed tomography image (c). (d) Sagittal section computed tomography showing heterogeneous mass lesion with predominant necrosis and fluorodeoxyglucose uptake in the periphery in fused positron emission tomography-computed tomography image (e)

demonstrated high FDG uptake (SUVmax values ranging from 3.19 to 72.2) while benign ones demonstrating low or no FDG uptake.[3-10] Through this case, the authors want to underscore the importance of ^{18}F-FDG PET-CT in response assessment to chemotherapeutic agents in this rare group of mesenchymal tumors as very few reports have been published in the literature.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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