^{18}F-FDG PET/CT in detection of sarcomatous degeneration of renal angiomyolipoma in setting of tuberous sclerosis

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ABSTRACT

Angiomyolipomas (AMLs) of kidneys are one of the common extracranial manifestations of tuberous sclerosis (TSC). AMLs when large may cause life-threatening hemorrhage, but seldom undergo malignant degeneration. We describe the appearance of renal AML degenerated to angiosarcoma on ^{18}F-flruorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) and contrast-enhanced CT (CECT).

Keywords: Angiosarcoma, contrast-enhanced computed tomography, flruorodeoxyglucose, positron emission tomography/computed tomography, renal angiomyolipomas, tuberous sclerosis

A 36-year-old female patient of tuberous sclerosis (TSC) presented with pain abdomen. A contrast-enhanced computed tomography (CECT) scan was advised which showed gross distortion of renal architecture along with multiple well-encapsulated lesions with predominantly fatty component intermixed with areas of increased tissue density and arising from the both the kidneys consistent with the diagnosis of the renal angiomyolipoma (AML). Another large lobulated soft tissue density mass was found to be arising from mid and lower pole of the left kidney, which demonstrated heterogenous contrast enhancement on arterial phase and delayed washout on venous phase. For further evaluation of the nature of the mass the patient was referred to our department for ^{18}F-fluorodeoxyglucose positron emission tomography/CT (^{18}F-FDG PET/CT). PET/CT images revealed gross distortion of renal architecture along with multiple, well-encapsulated lesions with predominantly fatty component intermixed with areas of increased tissue density and arising from both the kidneys with no significant radiotracer uptake consistent with renal AML [Figure 1] (bold arrow). Another large lobulated soft tissue density mass with nonuniform radiotracer uptake (maximum standardized uptake value (SUV_{max}) - 11.6) was found to be arising from mid and lower pole of the left kidney. Surprisingly, when the images of CECT and ^{18}F-FDG PET/CT were correlated, the relative hypoenhancing areas of the lesion with delayed venous filling in CECT demonstrated highest ^{18}F-FDG uptake indirectly reflecting perfusion-metabolism mismatch [Figure 1, line arrow]. Since FDG uptake is unusual in lesions of TSC, the treating physician advised a biopsy which yielded the diagnosis of renal angiosarcoma.

TSC complex is a neurocutaneous syndrome involving multiple organ systems.[1] The two genes involved, TSC-1 and TSC-2, are located on chromosomes 9 and 16, respectively.[2] Skin represent the most frequent site of involvement and renal changes are the main cause of morbidity and mortality in TSC, the most common being renal AMLs.[3,4] Other important features include pulmonary lymphangiomyomatosis, retinal astrocytoma, hepatic AML, and cardiac rhabdomyoma.[5] Although the lesions of the TSC are usually silent on ^{18}F-FDG PET/CT study, ^{18}F-FDG PET/CT can be useful for identifying other neoplasms like malignant perivascular epitheliod cell tumors (PEComas), primary lung cancers, and lymphomas, on a background of diffuse infiltrative and cystic lung disease, thoracic and abdominal lymphadenopathy, cystic lymphangiomyomas, and renal AML in patients with TSC.[6] Though there are case reports describing malignant transformation of renal AML in patients with TSC,[7,8] to our best of knowledge no such report has been described with ^{18}F-FDG PET/CT. Our case describes the appearance of sarcomatous degeneration of AML detected on ^{18}F-FDG PET/CT in a patient with TSC.

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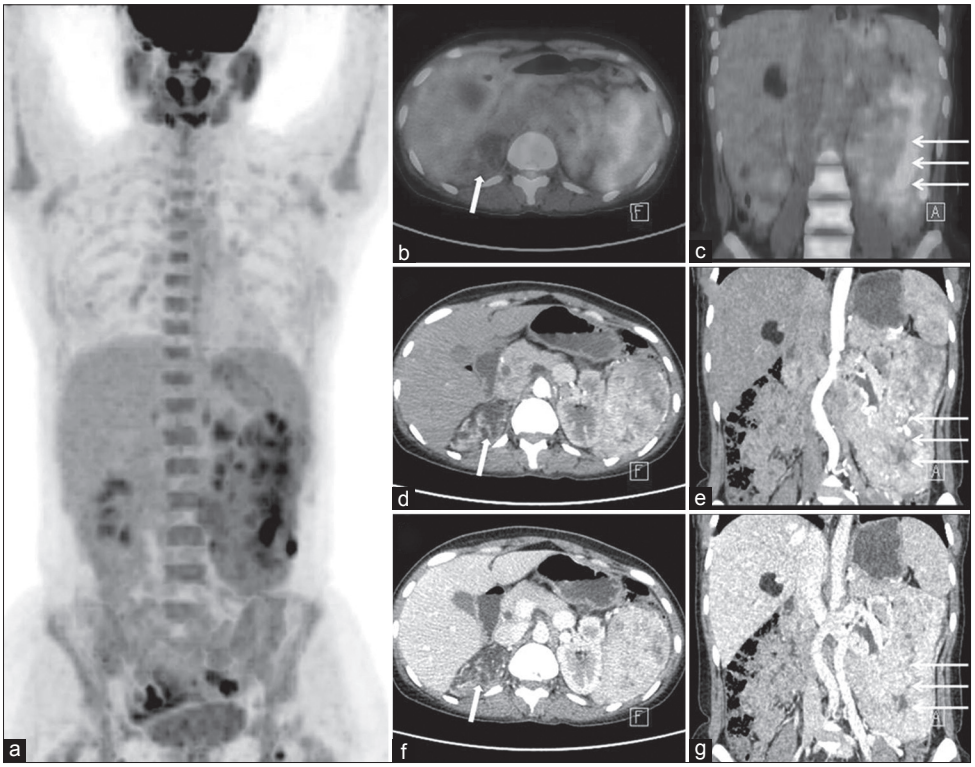

Figure 1: ^{18}F-flruorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) revealing well-encapsulated lesions arising from both the kidneys with no significant radiotracer uptake consistent with renal angiomyolipomas (a and b; bold arrow). Another large lobulated mass with nonuniform radiotracer uptake was noted in mid and lower pole of the left kidney (b and c) which demonstrated heterogenous contrast enhancement on contrast-enhanced CT (CECT) in arterial phase (d and e) and delayed washout on venous phase (f and g). The hypoenhancing areas of the lesion on delayed venous filling demonstrated highest radiotracer uptake reflecting perfusion-metabolism mismatch (c, e, and g; line arrow)

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