FDG-PET/CT findings of sarcomatous transformation in neurofibromatosis: a case report

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We herein report FDG-PET/CT findings of sarcomatous transformation in a patient with neurofibromatosis type 1 (NF-1). About 5% of patients with NF-1 develop sarcomatous transformation of a malignant peripheral nerve sheath tumor which arises from plexiform neurofibromas and is often associated with a poor prognosis. Morphologic imaging techniques such as Magnetic Resonance Imaging (MRI) and Computed Tomography (CT) are the standard methods to define the anatomic extent of the tumor, although tumor heterogeneity prevents reliable differentiation between benign and malignant lesions. The degree of fluoro-deoxyglucose (FDG) uptake correlates with histologic grade in neurogenic tumors in NF-1 patients. Our patient had a huge mass in the left gluteus area with a large nearly circular focus of increased FDG uptake in the tumor. The mass had a photopenic center. The maximum Standard Uptake Value (SUV_{max}) of this mass was 6.6. There was CT evidence of invasion of the left iliac wing, left acetabulum, and left superior pubic ramus; however there was no increased FDG uptake in these areas on the PET study. We surmised that the high FDG uptake indicated a high grade sarcoma, which was confirmed histologically. There was also a focal region of increased uptake in the L5 vertebral body, correlating with the CT hypodense lesion, with 2.9 SUV_{max}. FDG-PET/CT can identify sarcomatous change from benign neurogenic tumor with minimal misregistration, and can also detect metastatic disease. This case illustrates the importance of evaluating both metabolic and morphologic abnormalities to be able to formulate a proper treatment plan. This information can be obtained in a single session, using PET/CT.

Key words: FDG-PET/CT, neurofibromatosis, sarcomatous transformation, malignant peripheral nerve sheath tumor