'Double cortex' sign on FDG-PET/CT in diffuse band heterotopia

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ABSTRACT

F-18 Fluorodeoxyglucose (FDG) Positron emission tomography/Computed Tomography (PET/CT) has come to play an increasingly important role for the pre-surgical evaluation of drug resistant epilepsy and complements Magnetic Resonance Imaging (MRI) in the evaluation of grey matter heterotopias. This case illustrates the characteristic pattern of metabolic abnormality in diffuse band heterotopia (DBH) which is otherwise called double cortex syndrome. The presence of metabolic activity in the heterotopic inner cortical band and in the overlying true cortex gives rise to the 'double cortex' sign on FDG-PET, concurrent CT provides a good anato-metabolic coregistration.

Keywords: Band heterotopia, epilepsy, F-18 FDG, PET/CT

A 26-year-old female patient with history of seizure onset at 6 months of age was referred for F-18 Fluorodeoxyglucose (FDG) Positron emission tomography/Computed Tomography (PET/CT) study. She had complaints of sudden fall backwards or forwards with loss of responsiveness occurring multiple times a day. There was a history of delayed developmental milestones and poor scholastic performance. She was on optimal doses of four antiepileptic drugs. 296 MBq of F-18 FDG was injected intravenously and the patient was rested for one hour during which Electroencephalography (EEG) monitoring confirmed the interictal state. This was followed by the PET/CT acquisition on a Biograph mCT scanner (Siemens). The transaxial PET images [Figure 1a] revealed abnormal tracer accumulation in the subcortical region extending across frontal, parietal, temporal and occipital lobes. The fused transaxial PET/CT images [Figure 1b] clearly delineated this uptake to a bilateral symmetrical band located between the ventricular wall and cortical mantle involving the frontal, parietal, temporal and occipital lobes in both the hemispheres. FDG uptake in the subcortical band was ≥ overlying cortical FDG uptake with heterogeneity which was most marked in the frontal lobes [Figure 1b-arrow]. MRI images of the patient clearly demonstrated the thick isointense subcortical band extending across the frontal, parietal and temporal [Figure 1c-arrow] lobes suggestive of diffuse band heterotopia (DBH).

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Band heterotopia is a type of grey matter heterotopia that results from neuronal migration anomaly and falls under the broad category of malformations of cortical development.[1] It is commoner in females and the syndrome is usually associated with a mutation in the *doublecortin gene* (Xq22.3-q23) and less frequently in the *LIS1* (17p13.3) gene. It may be partial or complete[2] and it is typically the complete type which will give rise to the 'double cortex' sign on FDG-PET. The presence of glucose metabolism in the heterotopic band of grey matter basically indicates the presence of synaptic activity. Prior studies have reported normal to high metabolism in the heterotopic neurons in the interictal state.[3-6] Hypermetabolism in the subcortical band suggesting epileptogenic activity and correlating with the ictal EEG pattern has also been demonstrated in DBH.[7] This case of DBH demonstrated normal to mild heterogenously increased metabolism in the heterotopic subcortical band extending along the frontal, parietal and temporal cortices giving rise to the 'double cortex' sign. Volder *et al.*,[8] have suggested that the relatively high glucose utilization noted in DBH represents physiological activity. The hypothesis that has been proposed for the presence of high metabolic activity focally in the interictal state is a lack of synaptic revision, with persistence of exuberant synapses in the abnormally located neurons. This appears to be the most plausible explanation for the metabolic characteristics of the heterotopic band in DBH.

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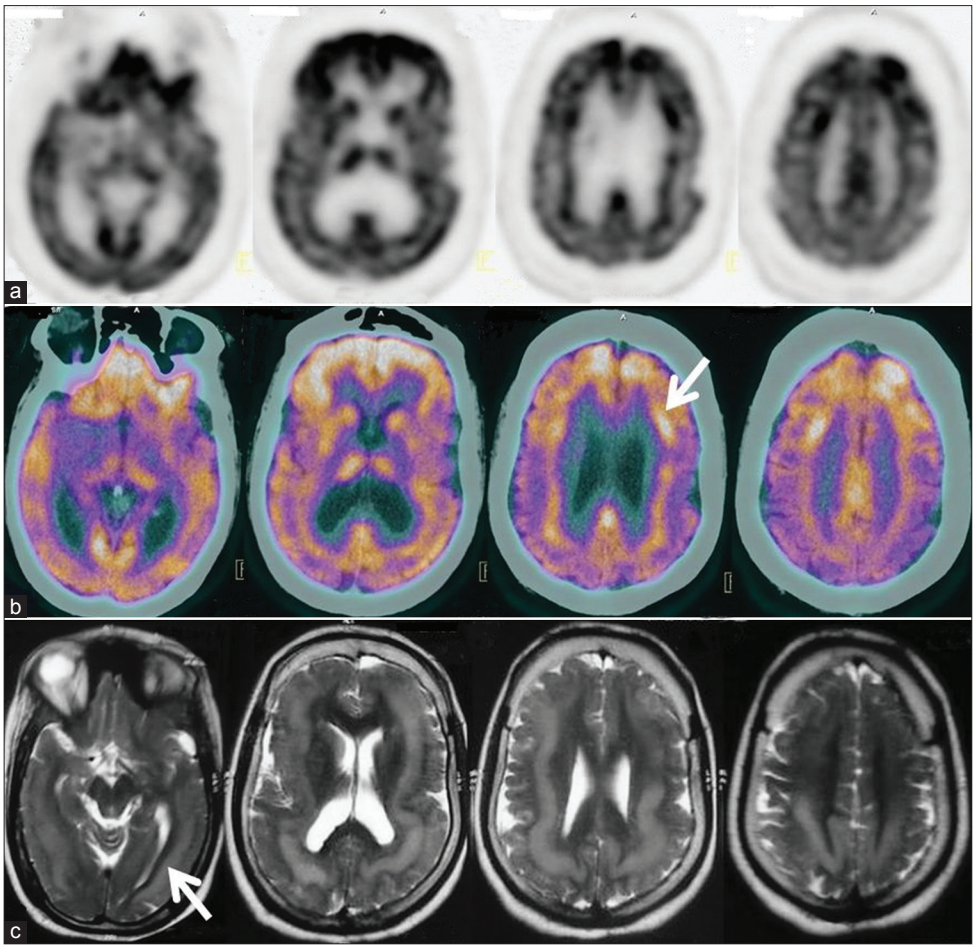

Figure 1: The transaxial PET images (a) revealed abnormal tracer accumulation in the subcortical region extending across the frontal, parietal, temporal and occipital lobes. The fused transaxial PET/CT images (b) clearly delineated this uptake to a bilateral symmetrical band located between the ventricular wall and cortical mantle involving the frontal, parietal, temporal and occipital lobes in both the hemispheres. (c) MRI images of the patient clearly demonstrated the thick isointense subcortical band (arrow) suggestive of DBH

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