Comparison MRI findings and positron emission tomography of Creutzfeldt-Jakob disease with V180I mutation

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Abstract

We report MRI and positron emission tomography (PET) findings of a 78-year-old man of Creutzfeldt-Jakob disease with codon 180 mutation who was difficult to be differentiated from mitochondrial encephalo- myopathy, lactic acidosis and stroke-like episodes (MELAS). The patient showed slow progression of dementia including memory disturbance, aphasia, apraxia, and left hemispatial agnosia during several months. He was suspected of MELAS because of the high-intensity lesion in the occipital lobe by diffusion-weighted MRI (DWI), and elevation of lactic acid in cerebrospinal fluid and serum, while mitochondrial abnormality was not found in muscle biopsy and rCBF was decreased at the lesion by PET. Finally, genotype revealed familial CJD with V180I mutation. The cortical edema was progressed after 6 months and then reduced after 14 months, while high intensity area on DWI was expanding for entire cortex after 14 months. PET showed marked decrease in rCBF in earlier stage and the area was larger than DWI lesion by MRI. The result revealed that the decrease of rOEF and acid increase of lactic acid in the right occipital area anaerobic metabolism in the area suggesting that PET findings in conjunction with MRI are useful for differentiating patient with CJD.