Diffusion and Perfusion MRI in Creutzfeld-Jakob Disease

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SYNOPSIS

Four patients with autopsy proven Creutzfeld-Jakob disease (CJD) were studied ante mortem using diffusion and perfusion MRI. All were shown to have multifocal cortical gyral hyperintensity and variable subcortical involvement on diffusion sequences. Diffusion changes were more pronounced at high b-values (2500 vs 1000). Regionally prolonged perfusion transit times were co-localized to areas of diffusion restriction in the cortex.

INTRODUCTION

Creutzfeld-Jakob disease is a rare, uniformly fatal neurodegenerative condition accompanied by prion protein accumulation in the brain. The typical clinical presentation is of a rapidly progressive dementia, gait instability, and myoclonus. Ante mortem confirmation is imperfect, but electroencephalography, spinal fluid assays for the neuron-specific enolase 14-3-3, and biopsy of the pharyngeal tonsil can be supportive. Recently, several groups have shown a characteristic pattern of high signal on diffusion weighted images (DWI) in the cortex and basal nuclei of affected patients. In this study we investigated the utility of adding high b-value DWI and perfusion weighted imaging (PWI) in the MR investigations of patients with suspected CJD.

MATERIALS AND METHODS

Four patients with a rapidly progressive dementia suspicious for CJD were referred for diagnostic MRI. There were 3 men and 1 woman, ranging in age from 43-85 years. MRI exams were done at 1.5T (GE Medical Systems) and included diffusion (b=1000) and gadolinium-enhanced dynamic susceptibility perfusion exams in addition to routine sequences. Three patients also had b=2500 DWI sequences. Perfusion was analyzed with both in-house and commercial software (GE) to yield parameter maps.

RESULTS AND DISCUSSION

All patients showed abnormalities in the cortex on b=1000 DWI, and all had slightly prolonged mean transit times on PWI in the affected regions. Changes were more conspicuous on high b-value diffusion images. Autopsy confirmed typical spongiform changes and positive testing for PrPsc (prion protein) in each case.

CONCLUSIONS

Gyral signal abnormalities can be detected on DWI in patients with CJD, and these changes are more easily seen on high b-value studies. Perfusion parameter maps show slightly longer transit times in the regions which appear abnormal by DWI. The addition of these sequences may assist in the diagnostic evaluation of patients with rapidly progressive dementia.

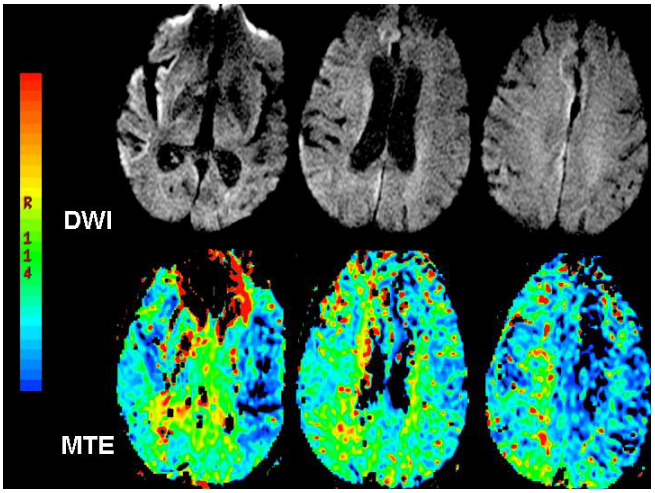

Figure 1. 85 year old man with CJD. DWI (B=1000) shows high signal in the right sylvian cortex, caudate body, and cingulate gyrus. A slightly prolonged transit time is also seen in the right hemisphere.

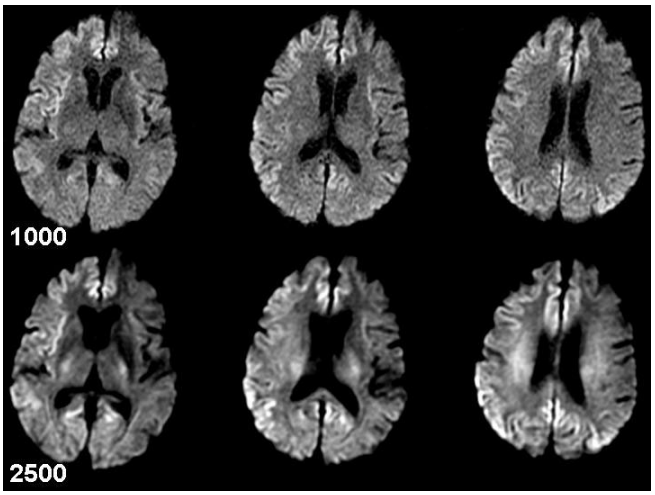

Figure 2. 43 year old woman with CJD. Gyral signal abnormalities are seen in both hemispheres, right greater than left. Changes are much more conspicuous on high b-value (2500) images shown on the lower panel.