Diffusion-Weighted Imaging and FLAIR in Creutzfeldt-Jakob Disease

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Purpose
Creutzfeldt-Jakob disease (CJD) presents with rapidly progressive dementia and movement disorder. Creutzfeldt-Jakob disease is secondary to either inherited or acquired prion disease of the brain. In addition to the “classical” CJD (sporadic, iatrogenic, and familial), a new variant (vCJD) has been identified, which is related to ingestion of contaminated beef. MR imaging plays a major role both in the triage of patients for brain biopsy, and possibly in predicting diagnosis. The described MR findings in CJD include flair and diffusion changes within gray matter, sparing white matter. This predilection for gray matter and constellation of involved locations rarely is seen in other conditions, and in the context of a rapidly progressive dementia may be quite specific for CJD. We undertook a prospective review of the neuroradiologic manifestations of 10 CJD patients referred for evaluation for a new treatment protocol using quinacrine, assessing their MR findings.

Materials & Methods
Ten patients underwent MR, (echo-planar diffusion tensor, axial and coronal flair, spin-echo T2-weighted imaging). ADC maps were created for all scans. Quinacrine therapy was initiated in four patients; two stopped after a brief decline. Follow-up imaging was performed on one patient after 10 weeks of therapy.

Results
Tissue diagnosis by biopsy or autopsy was obtained on eight patients and is planned on the other two. One patient (not a US resident) had vCJD and 9 had classic CJD; 6 definite sporadic, 2 probable sporadic, 1 definite familial. The vCJD patient’s scans demonstrated T2 prolongation and reduced diffusion involving both medial thalamus, but sparing the neocortex. Follow-up after 10 weeks of quinacrine therapy showed no change on imaging and only slight decrease in ADC. Clinical status had declined slightly. MR imaging of the 9 patients with classic CJD typical demonstrated T2 prolongation and reduced diffusion in the basal ganglia, cortical regions of the temporal lobe, cingulated gyrus, and/or hippocampus, as well as generalized atrophy (see Tables). Coronal flair and diffusion best delineated these subtle changes within the cortex.

<table>
<thead>
<tr>
<th>Classic CJD</th>
<th>FLAIR</th>
<th>Neocortex</th>
<th>Striatum</th>
<th>Thalamus</th>
<th>Cingulate</th>
<th>Hippo/M</th>
<th>Inf Frontal</th>
<th>Insula</th>
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<tbody>
<tr>
<td>DWI</td>
<td>Neocortex</td>
<td>Striatum</td>
<td>Thalamus</td>
<td>Cingulate</td>
<td>Hippo/M</td>
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Conclusion
Creutzfeldt-Jakob disease is a prion disorder with predominant involvement of multiple cortical and subcortical locations. There is a different pattern of predominant involvement in the classical and variant forms, but the previously reported “pulvinar sign” is not 100% specific for vCJD. Classic CJD is typified by abnormal signal on FLAIR and diffusion in scattered areas of neocortex, medial and inferior temporal lobes, cingulate gyrus, and corpus striatum. Prominent abnormality in the medial thalamus with sparing of neocortex is typical of vCJD, but also has been seen in at least one “classic” patient. Treatment regimen with quinacrine did no reversal of findings in two patients, however the progress may have stabilized.